Mayo Clinic Images in Internal Medicine

Self-Assessment for Board Exam Review

Editor
Furman S. McDonald, M.D.

MAYO CLINIC SCIENTIFIC PRESS

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DEDICATED TO

Four very special people:

- Charles H. Rohren, MD—the best physical diagnostician I know. A great physician, teacher, mentor, and friend. You are the reason I am an internist today. I will always see you when I think of the physician I want to be.
- Darryl S. Chutka, MD—a great physician and teacher. When you told me to "make the book as large as you like," I don't think you expected what would come of it. Without your support and allowance for creative freedom, this book would never have been written.
- Henry J. Schultz, MD—the greatest single influence on my professional life. Program director, adviser, friend. I hope that this book will be useful to the residents you care so much about, of whom I know I am one.
- Elizabeth S. McDonald, MD, PhD—my devoted wife. Thanks for putting up with me while I put so much time into this book.

One very special program:

Proceeds from the book will support the Mayo International Health Program, which defrays the cost of elective experiences for Mayo Clinic residents and fellows caring for underserved patients in international settings.

Furman S. McDonald, MD

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PREFACE

Mayo Clinic Images in Internal Medicine: Self-Assessment for Board Exam Review was begun in 1998 as an adjunct to a presentation for the Mayo Internal Medicine Board Review Course. Images were presented in random order with brief case scenarios. The reader was encouraged to consider the cases before turning the page to read a few learning points about each image. That first book contained 38 cases and 57 images from 6 contributors.

Year by year the work was expanded until now the current book has 173 cases with 287 images from 28 contributors. As with the first book, all images are from patients seen at Mayo Clinic, and the cases have been designed to illustrate important teaching points. American Board of Internal Medicine–type board review questions accompany the cases. The references from which the content for each case scenario was built are provided so the reader can obtain further information. Cases are presented in random order to be a more effective review tool that simulates examination (and clinical practice) conditions. However, with the increase in size, a detailed index was developed to allow the reader to find cases more efficiently. In addition, this edition introduces chain referencing, an innovative learning technique that allows the reader to move from case to case based on specialty designation. With access to the information in this book through random case review, the detailed reference index, and specialty chain referencing, *Mayo Clinic Images in Internal Medicine: Self-Assessment for Board Exam Review* is really three effective learning texts bound in one cover.

It is hoped that *Mayo Clinic Images in Internal Medicine: Self-Assessment for Board Exam Review* will be an aid for preparation for board examination certification or recertification and medical clerkship review and also a useful update for practicing clinicians.

Furman S. McDonald, MD
Editor

ACKNOWLEDGMENTS

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ABBREVIATIONS

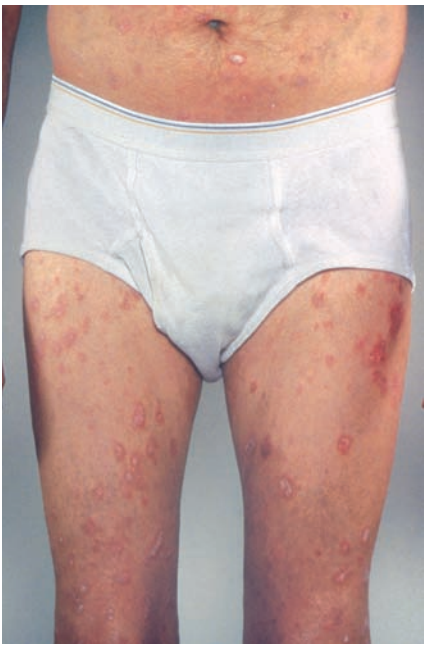
The chain references listed at the bottom of the content pages use the following abbreviations for specialties:

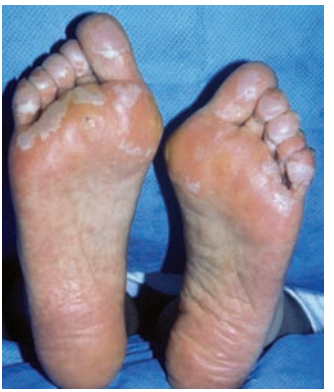
Allergy/Immunol	Allergy and Immunology
CAM	Complementary and Alternative Medicine
Crit	Critical Care
CV	Cardiovascular Diseases
Derm	Dermatology
Endo	Endocrinology
ENT	Ear, Nose, and Throat
Genet	Genetics
GI	Gastroenterology
Hem	Hematology
ID	Infectious Diseases
Musc	Musculoskeletal Diseases
Neph	Nephrology
Neuro	Neurology
Ob/Gyn	Obstetrics and Gynecology
Oncol	Oncology
Ophth	Ophthalmology
Pulm	Pulmonary Diseases
Rheum	Rheumatology
Toxicol	Toxicology
Vasc	Vascular Diseases

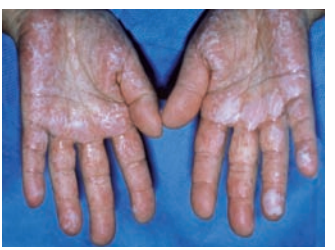
How to Use This Book

Most atlases of internal medicine and review texts group images and content by specialty, an order that works well for reference volumes. However, it does not resemble the random order of cases in general clinical practice or on examinations. Furthermore, learning styles differ. The editors have endeavored to make *Mayo Clinic Images in Internal Medicine: Self-Assessment for Board Exam Review* useful for examination study, practice, and reference, realizing that users of the book may have different needs and different styles of learning. With this understanding, we suggest the following ways to use this book:

1. **Random case review:** The order of the cases presented in this book was determined with a random number generator; thus, a case does not lend clues to the diagnosis or questions related to the subsequent case. This most closely simulates a general practice or examination setting. People who used the prototypes of this book found this order useful for examination review.
2. **Specialty-specific review with use of chain references:** Some readers may want to review cases in a single specialty. The bottom of the content page for each case includes abbreviations for the specialty classifications of the case followed by a page number directing the reader to the next case of that specialty. After the last case in any specialty, the page numbers refer to the first case in the series. Thus, the reader can open the book at any page and review specifically the cases of any specialty by following the chain references.
3. **Cases by specialty:** To find all the cases of any specialty, a list of cases categorized by specialty and page number is provided in the Appendix. Because some cases can be classified with more than one specialty, these lists may overlap.
4. **Reference by index:** For readers who want to find a particular image or content related to a particular subject, an extensive index is provided.







A 37-year-old man received a bone marrow transplant 2 weeks previously for acute myelogenous leukemia. Two days ago, itching, diarrhea, pain and numbness in his palms and soles, and the skin lesions shown here developed. Each of the following statements about this condition is true *except*:

a. Use of HLA-identical grafts eliminates the risk of this condition
b. This condition typically occurs 7 to 21 days after transplantation
c. Increased age is a risk factor for this condition
d. Sex mismatch (female donor, male recipient) is a risk factor for this condition
e. Values on liver function tests are often increased in this condition

Acute Graft-Versus-Host Disease

Answer: a

- Occurs 7 to 21 days after transplantation
- Donor T cells attack host HLA antigens
- Mean frequency in adults with HLA-identical grafts is 35%
- Risk factors include the following:

  Increased age
  HLA mismatch
  Sex mismatch (female donor, male recipient)
  Irradiation
  Suboptimal immunosuppression
- Affects the skin, gastrointestinal tract, and liver
- Pruritus and pain may be the first sensations, commonly followed by an erythematous measles-like maculopapular exanthem. Acral erythematous lesions may develop on palms, soles, and ears
- Blistering and exfoliation are common
- Digestive tract involvement may present (from less severe to more) as nausea, vomiting, anorexia, diarrhea, malabsorption, abdominal pain, ileus, and ascites
- Values on liver function tests are often increased; jaundice and hepatomegaly may develop
- Treatment involves intensifying immunosuppression, such as high-dose corticosteroids, cyclosporine, and cyclophosphamide
- Severe cases (grade IV) have a high mortality rate (>80%)

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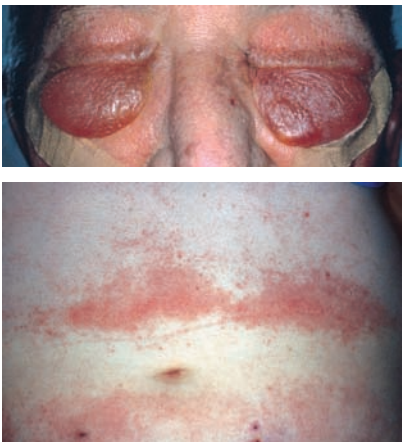

An 82-year-old man presents with 8 weeks of periorbital edema, 6 weeks of rash, and 2 weeks of bilateral shoulder and arm aches and weakness. What is the diagnosis?

a. Polymyalgia rheumatica
b. Inclusion body myositis
c. Hypothyroidism
d. Polymyositis
e. Dermatomyositis

Dermatomyositis

Answer: e

- Three causes of idiopathic inflammatory myopathy:

  Dermatomyositis (DM)
  Polymyositis
  Inclusion body myositis

- Dermatomyositis has a bimodal age distribution
- Skin findings associated with DM:

  Heliotrope hue of the eyelids
  Rash of the metacarpophalangeal and proximal interphalangeal joints (Gottron's papules)
  Photosensitivity dermatitis of the face

- Proximal myositis with associated pain and weakness is characteristic of DM
- The serum creatine kinase and aldolase values are usually increased in DM
- The anti-Jo1 antibody is present in 25% of patients with DM
- The electromyogram is characteristic, but not diagnostic, of the inflammatory myopathies
- A muscle biopsy is suggested for all patients with inflammatory myopathy
- Patients with DM have an increased risk for cancer
- DM is treated with immunosuppressive drugs

Reference
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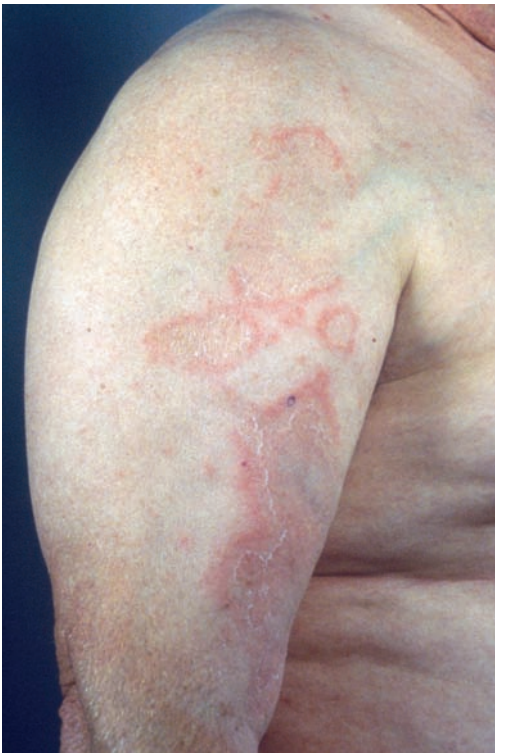
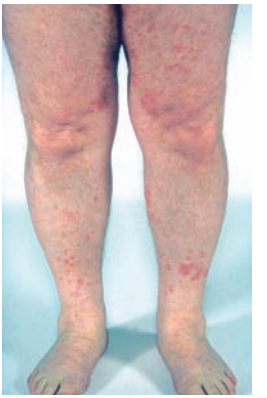
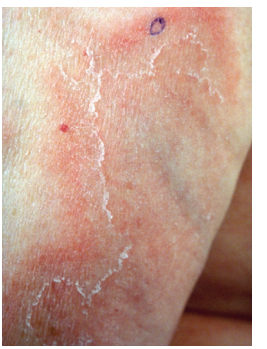






The lesions in this patient developed 2 weeks after hiking in the woods of the Upper Midwest in April. If left untreated, this patient is at risk for development of all of the following *except*:

a. Fever, lymphadenopathy, meningismus
b. Carditis
c. Ischemic bowel
d. Cranial nerve palsies
e. Arthritis

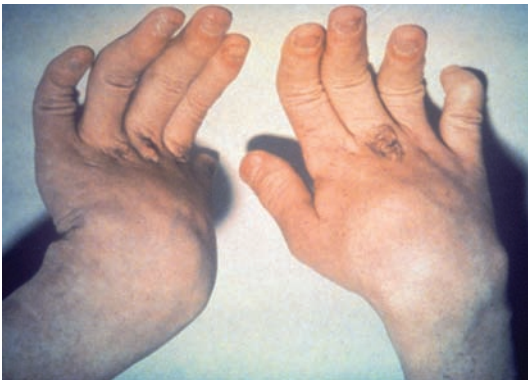
Lyme Disease

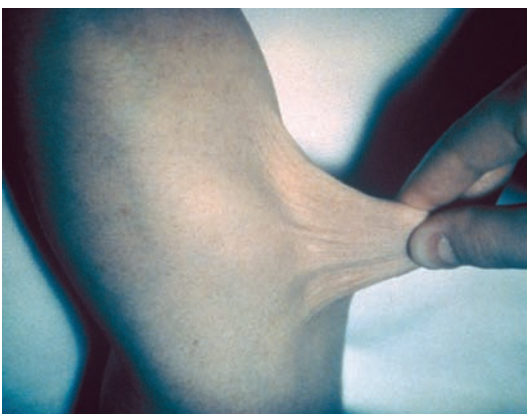
Answer: c

- Causative agent, *Borrelia burgdorferi*, is transmitted by *Ixodes* ticks
- Highest incidence is during spring and summer
- Tick must be attached more than 24 hours for transmission to occur
- Stage 1: Within 30 days of infection, erythema chronicum migrans occurs in 80% of patients and may be associated with fever, lymphadenopathy, and meningismus. The rash resolves within 4 weeks
- Stage 2: Onset is weeks to months after stage 1. Neurologic abnormalities occur in 10% to 15% of patients. Carditis occurs in 10% of patients
- Stage 3: Begins months to years after infection. Arthritis occurs in 50% of untreated patients, becoming chronic in 10% to 20%
- Enzyme-linked immunosorbent assay is positive for diagnosis within 2 to 6 weeks
- Early-stage disease may be treated with doxycycline, amoxicillin, or cefuroxime
- Neurologic or cardiac involvement requires high-dose ceftriaxone or penicillin G

Reference

Habermann TM. *Mayo Clinic Internal Medicine Board Review 2004-2005*. Philadelphia: Lippincott Williams & Wilkins; 2004:59, 180, 564-565, 755, 1000-1001.





These are the hand and skin findings of a 26-year-old woman who complains of early satiety. Her past medical history is notable for upper gastrointestinal bleeding and rectal prolapse. What is the diagnosis?

a. Ehlers-Danlos syndrome
b. Cutis laxa
c. Osteogenesis imperfecta
d. Congenital contractural arachnodactyly
e. Marfan syndrome

Ehlers-Danlos Syndrome

Answer: a

- Ehlers-Danlos syndrome is characterized by highly elastic connective tissue
- Many forms (up to 15) of Ehlers-Danlos syndrome exist
- The autosomal-dominant forms of the disease account for 90% of reported cases
- Patients have hyperextensible and lax joints that are prone to dislocation
- Patients with skin manifestations have hyperextensible, fragile skin that heals poorly, characteristically forming wide, thin, "fish-mouth" scars. The skin may have a velvety texture
- Patients are predisposed to the following:

  Gastrointestinal motility disorders
  Visceral diverticulosis
  Mitral valve prolapse (up to 50% of patients)
  Dilatation of the aortic root
  Pes planus
  Scoliosis
  Degenerative arthritis
  Pneumothorax
  Dilatation of the pulmonary artery
  Angina

Reference
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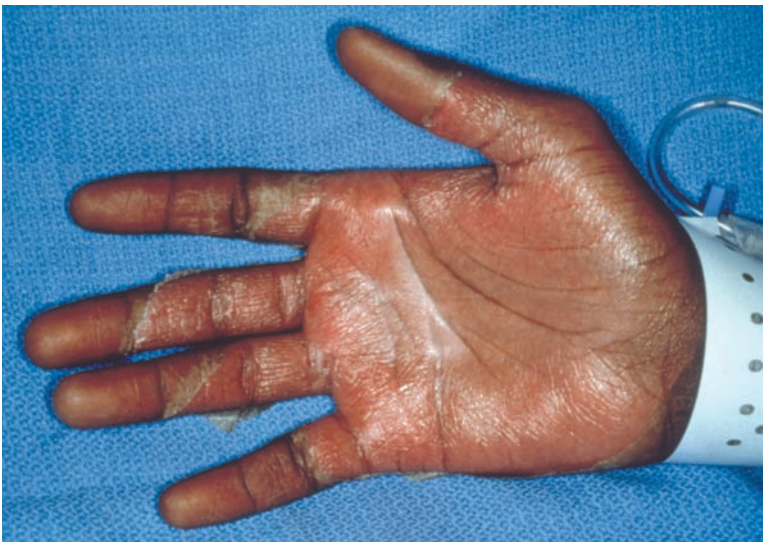

Clinical criteria for toxic shock syndrome include all of the following *except*:

a. Fever
b. Positive blood cultures
c. Hypotension
d. Involvement of 3 or more organ systems
e. Erythroderma

Toxic Shock Syndrome

Answer: b

- Acute life-threatening illness caused by production of staphylococcal exotoxin (toxic shock syndrome toxin-1, TSST-1)
- Centers for Disease Control and Prevention criteria: 6 required for *confirmation*; 5 of 6 indicate a *probable* case

1. Fever	Temperature of more than 38.9°C
2. Rash	Diffuse, macular, and erythematous
3. Desquamation	Especially of palms and soles
4. Hypotension	In adults, systolic blood pressure less than 90 mm Hg
5. Multisystem involvement	Severe myalgias Diarrhea, vomiting Liver dysfunction Thrombocytopenia Renal insufficiency Mental status changes
6. Exclusion of other causes	Negative results of blood, throat, cerebrospinal fluid cultures (usually *not* positive for *Staphylococcus aureus*)

- Associations: menstruation with prolonged use of tampons, surgery (even if wound is not worrisome), barrier contraceptives
- With early and aggressive management and judicious use of antibiotics and supportive care, the mortality rate for toxic shock syndrome has been estimated at 3%. The use of corticosteroids (to lessen the overwhelming inflammatory response) and immunoglobulins remains controversial

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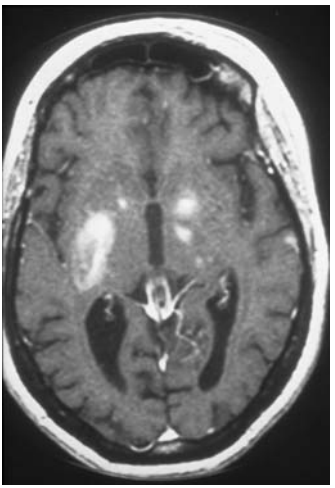

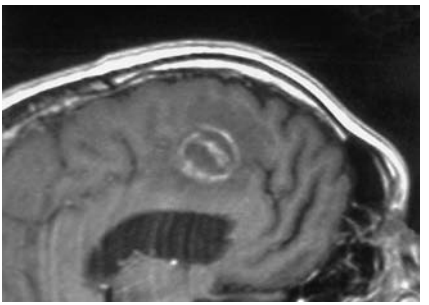

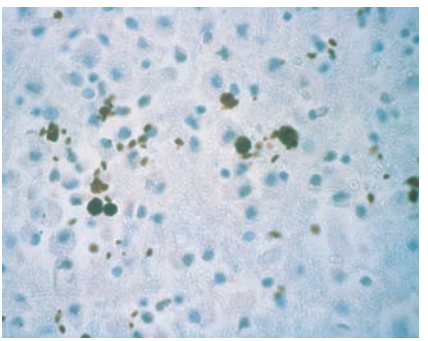

A 37-year-old man with acquired immunodeficiency syndrome (AIDS) presents with fever, seizures, and altered mental status. His last known CD4 cell count was less than 50 cells/mm^3. Which one of the following is the best prophylactic agent for this condition?

a. Rifabutin
b. Trimethoprim-sulfamethoxazole
c. Pyrimethamine
d. Fluconazole
e. Acyclovir

Answer: b

- The preferred prophylactic agent for *Pneumocystis carinii* and *Toxoplasma gondii* is trimethoprim-sulfamethoxazole
- If the patient is allergic to sulfonamides, pyrimethamine *and* dapsone are efficacious
- Without prophylaxis, central nervous system toxoplasmosis will develop in 30% to 50% of patients with AIDS who are seropositive for *Toxoplasma* IgG
- Magnetic resonance imaging is more sensitive than computed tomography for detecting central nervous system lesions of toxoplasmosis
- Typical appearance is of multiple ring-enhancing lesions within the brain parenchyma
- Central nervous system lymphoma can mimic toxoplasma encephalitis
- Patients with a single lesion, who are seronegative, or who do not respond to antitoxoplasmosis therapy should be referred for brain biopsy
- On biopsy, immunoperoxidase stain shows cysts and tachyzoites of *T. gondii*

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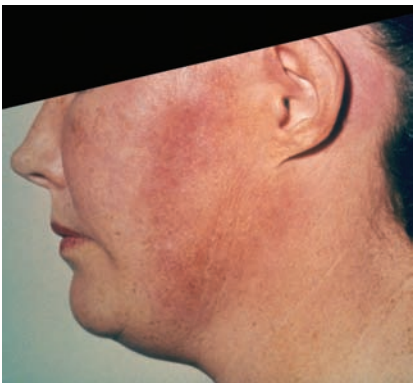
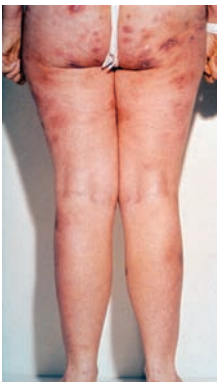
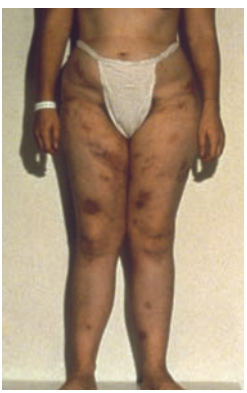
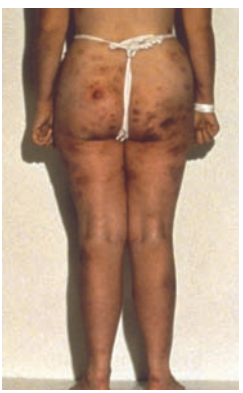

In this 26-year-old woman, these tender, subcutaneous, nodular lesions developed recurrently with fever. Biopsy reveals fat necrosis. She also has increased values on liver function tests, and peripheral smear demonstrates a leukemoid reaction. What is this syndrome called?

a. Kikuchi-Fugimoto disease
b. Weber-Christian disease
c. Letterer-Siwe disease
d. Kuhnt-Junius disease
e. Smith-Strang disease

Weber-Christian Disease, or Relapsing Febrile Nonsuppurative Nodular Panniculitis

Answer: b

- Idiopathic, relapsing, remitting, inflammatory, infiltrative, nodular, nonsuppurative, tender panniculitis that often presents with fever
- The hallmark is tender subcutaneous nodules
- Lesions occasionally drain an oily substance
- Most commonly affects white women after the second decade of life
- Also may involve deep intravisceral fat, resulting in constitutional symptoms, bleeding tendencies, thrombosis, hepatic failure, nodular lung disease, congestive heart failure, and pancreatic dysfunction
- Visceral involvement portends a poor prognosis
- Diagnosis is by exclusion of other causes and skin biopsy showing inflammatory changes in the subcutaneous fat layers
- No completely effective therapy is known. Corticosteroids and nonsteroidal anti-inflammatory drugs are usually helpful for acute exacerbations. Hydroxychloroquine has been used with some success
- Associated diseases include systemic lupus erythematosus, pancreatitis, α_1-antitrypsin disease, lymphoproliferative diseases, infections, and trauma

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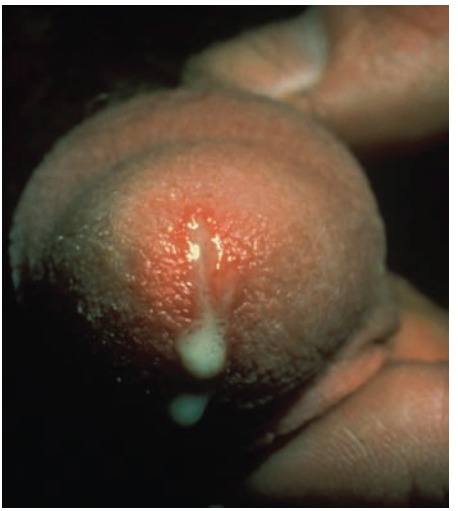

In this case, in addition to diagnostic testing, which empiric therapy is most appropriate?

a. Imipenem
b. Oxacillin and gentamicin
c. Vancomycin and gentamicin
d. Ceftriaxone and doxycycline
e. Amphotericin B

Answer: d

- Urethral discharge with burning on urination and an itchy urethra are typical symptoms

Neisseria gonorrhoeae:	Gram-negative intracellular diplococcus Diagnosis by molecular gene probe, Gram stain, or culture Injectable ceftriaxone is first-line therapy Add doxycycline or azithromycin for treatment of *Chlamydia trachomatis* because 15% to 25% of patients with gonorrhea also have *Chlamydia*
Nongonoccocal urethritis:	Symptomatically indistinguishable from gonococcal variety, but much more frequent Most common causes are *C. trachomatis* (25%-40%) and *Ureaplasma urealyticum* (40%-50%) Other causes include *Mycoplasma genitalium*, *Trichomonas vaginalis*, and herpes simplex virus

- Sexual partners also should be treated and intercourse avoided until treatment is completed

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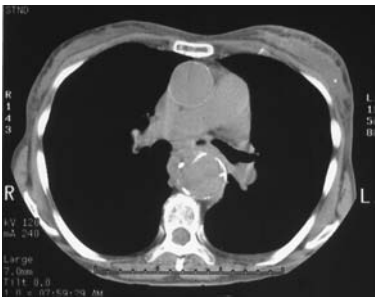
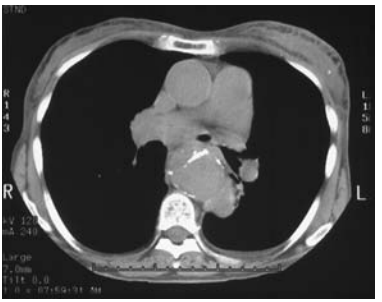
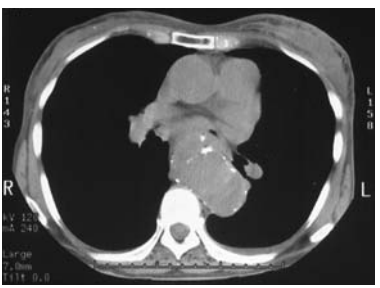

A 54-year-old woman presents with nonreproducible upper back discomfort and dysphagia. She has a history of coronary artery disease and abdominal aortic aneurysm repair 6 years ago. Thoracic computed tomography scans are shown. In the United States, which one of the following risk factors is most commonly associated with this condition?

a. Syphilis
b. Cystic medial necrosis
c. Rheumatoid arthritis
d. Hypertension
e. Atherosclerosis

Descending Thoracic Aortic Aneurysm

Answer: e

- Aortic aneurysms are classified by location (abdominal or thoracic) and shape (fusiform or saccular, as in this case)
- Thoracic aneurysms are further classified as ascending or descending, based on their location proximal or distal to the aortic arch
- Most often aortic aneurysms are asymptomatic
- Compression of adjacent structures (the esophagus in this case) may result in symptoms. Leakage of blood from the aneurysm may cause acute pain and may be a sign of impending rupture
- Acute rupture without warning is the most common cause of symptoms. Therefore, clinical suspicion for the diagnosis should be high
- When one aneurysm is detected, the patient should be screened for the presence of other occult aneurysms
- Size correlates with risk of rupture, but not as exactly as for abdominal aortic aneurysms. Nonetheless, most authors favor surgical management for thoracic aortic aneurysms that are more than 6 cm in diameter
- Atherosclerosis is the most commonly associated condition. Others include hypertension, giant cell arteritis, syphilis, Ehlers-Danlos syndrome, rheumatoid arthritis, trauma, cystic medial necrosis, and Marfan syndrome

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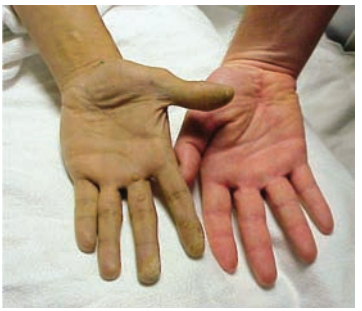

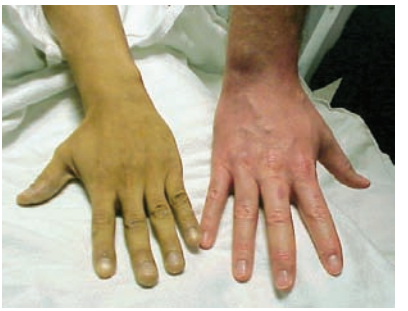

A patient with human immunodeficiency virus had a rash in association with trimethoprim-sulfamethoxazole used as prophylaxis for *Pneumocystis carinii* pneumonia. An alternative medication was used, but the patient took 3 times the normal dose by mistake. He became dyspneic, and headache, nausea and vomiting, and the skin discoloration shown here developed (a normal hand is shown for contrast on the right).

1. What was the alternative medication?
   1a. Pentamidine
   1b. Pyrimethamine-sulfonamide
   1c. Atovaquone
   1d. Dapsone
   1e. Clindamycin-primaquine

2. In addition to supplemental oxygen, which one of the following would be the most appropriate antidote?
   2a. *N*-Acetylcysteine
   2b. Amyl nitrite
   2c. Methylene blue
   2d. Deferoxamine
   2e. 4-Methylpyrazole

Methemoglobinemia Due to Dapsone Overdose

Answer 1: d

Answer 2: c

- Methemoglobin is the ferric form of hemoglobin ($HbFe^{3+}$) that cannot bind oxygen
- Although sometimes inherited, it is more often formed by oxidative stress
- Among drugs, nitrites are commonly associated with this condition
- Chocolate-brown or slate-blue cyanosis unrelieved with oxygen suggests the diagnosis
- Arterial blood gas studies will show normal arterial oxygen pressure but low oxygen saturation. Pulse oximetry results will be inappropriately increased (the pulse oximeter result will be higher than the actual oxygen saturation, although the absolute value may still be below normal)
- Methylene blue is the antidote for patients with severe hypoxia, unless they have glucose-6-phosphate dehydrogenase deficiency, in which case it may worsen the clinical condition. If the patient has this deficiency, ascorbic acid can be used to reduce the methemoglobin
- *N*-Acetylcysteine is the antidote for acetaminophen overdose
- Amyl nitrite is an antidote for cyanide overdose because nitrites precipitate the formation of methemoglobin, which is an alternative binder for cyanide. Thus, it would be contraindicated in methemoglobinemia
- Deferoxamine is the antidote for iron overdose
- 4-Methylpyrazole is an alternative to ethanol for the treatment of methanol toxicity

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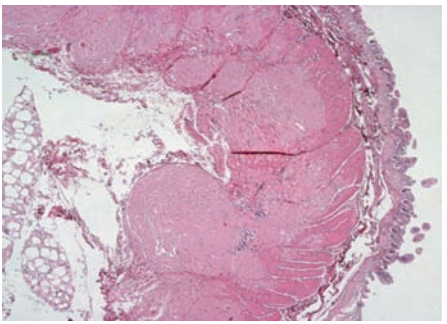

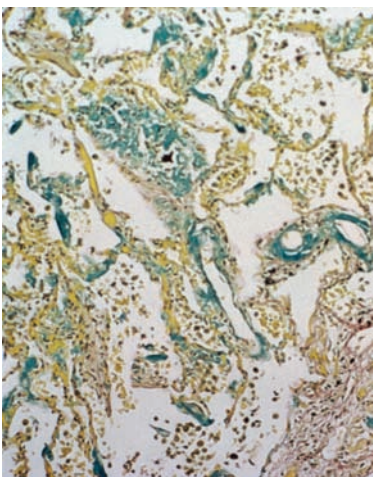

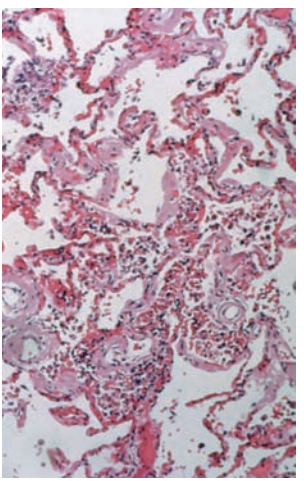

A 57-year-old man with known lymphoproliferative disorder presents with gastrointestinal bleeding and diarrhea. Periorbital purpura may occur in this patient and is associated with which one of the following?

a. Exposure to sunlight
b. Diffuse muscular pain
c. Proctoscopic examination
d. Fat aspirate biopsy
e. Hypertension

Amyloidosis With Pulmonary and Gastrointestinal Involvement

Answer: c

- Amyloidosis may have multiorgan involvement, including liver, kidney, gastrointestinal tract, and heart. Up to 90% of patients with primary amyloidosis have cardiac dysfunction
- The liver may become infiltrated in up to 25% of cases and indicates extensive involvement and a poor prognosis
- Manifestations result from deposition of an amorphous, insoluble protein-polysaccharide complex in the tissue
- There are several types of amyloid, and their classification is based on the type of protein fibrillar deposition:

  Type AA is associated with reactive systemic amyloidosis and is found in hereditary or acquired chronic inflammatory disease

  Type AL is associated with systemic amyloidosis and is found in multiple myeloma and monoclonal gammopathies
- Gastrointestinal manifestations include diarrhea, megacolon, and fecal incontinence
- Periorbital purpura may occur after proctoscopic examination
- Fat aspiration is confirmatory of the diagnosis in up to 80% of patients. Rectal biopsy confirms the diagnosis in up to 75% of patients
- Congo red stain of histologic specimens may reveal a characteristic apple-green birefringence under crossed polarized light. This remains the standard test for diagnosis

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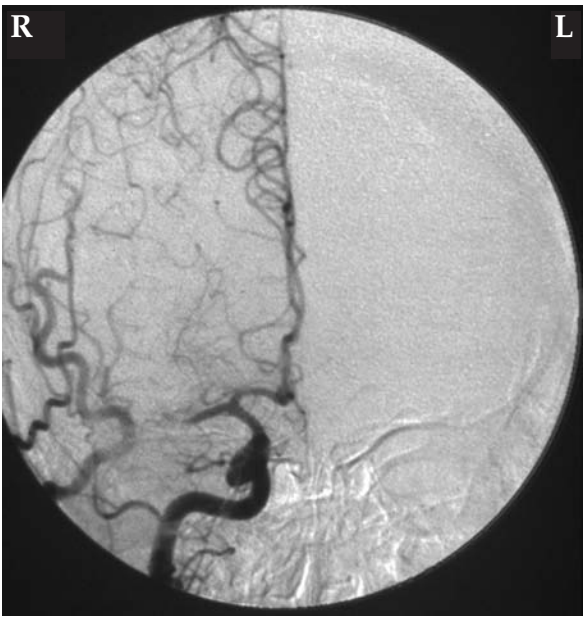

A 31-year-old man with *Streptococcus viridans* endocarditis has acute right hemiparesis and dysarthria. Which one of the following is the most likely cause?

a. Conversion disorder
b. Left brain abscess
c. Seizure with Todd paralysis
d. Left middle cerebral artery occlusion
e. Atypical migraine

Left Middle Cerebral Artery Occlusion Due to Embolus in Infective Endocarditis

Answer: d

- Extracardiac manifestations of infective endocarditis include the following:

  Embolic events
  Suppurative complications
  Immunologic reactions
  Osler's nodes (which are painful)
  Roth's spots
  Increased rheumatoid factor
  Glomerulonephritis

- Systemic embolization occurs in 20% to 40% of patients with left-sided endocarditis
- Embolization may occur at any time, but risk decreases with duration of treatment
- Specifically vascular phenomena associated with infective endocarditis include the following:

  Arterial emboli
  Mycotic aneurysms
  Intracranial hemorrhages
  Conjunctival hemorrhages
  Pulmonary infarcts (septic)
  Janeway lesions (hemorrhagic lesions on the palms or soles, usually painless)
  Splinter hemorrhages

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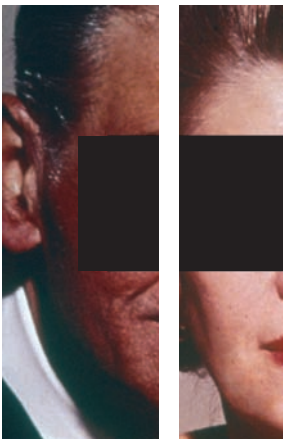

A 40-year-old man presents with hyperpigmentation, malabsorptive diarrhea, weight loss, recurrent arthritis, and adenopathy. His wife is shown for contrast. Which one of the following tests is likely to confirm the diagnosis?

a. Magnetic resonance imaging of the head
b. Computed tomography with thin cuts of the adrenal glands
c. Small bowel biopsy with periodic acid-Schiff staining
d. Dexamethasone suppression test
e. Cosyntropin stimulation test

Whipple's Disease

Answer: c

- Whipple's disease is a chronic relapsing, systemic infectious disease involving the central nervous system, heart, kidneys, and small bowel
- It occurs primarily in white middle-aged men
- The causative agent is a gram-positive bacillus, *Tropheryma whippelii*
- The name of the bacillus was chosen from the Greek word *trophe*, which means "nourishment," thus highlighting the primary clinical feature of malabsorption
- From 40% to 50% of patients have hyperpigmentation in sun-exposed areas and scars
- Other signs and symptoms include weight loss (80%), diarrhea (75%), arthralgia (70%), lymphadenopathy (55%), abdominal tenderness (50%), fever (40%), edema (25%), glossitis (20%), splenomegaly (10%), ascites (5%)
- 90% of patients have steatorrhea on 72-hour stool studies
- Anemia of chronic disease and thrombocytosis are common
- Joint symptoms may precede intestinal manifestations by years
- Diagnosis is established with small bowel biopsy showing periodic acid-Schiff–positive granules in macrophages with gram-positive acid-fast bacillus–negative bacilli
- Treatment is with trimethoprim-sulfamethoxazole for 1 year
- Response to antibiotics is dramatic; most symptoms completely resolve within 2 to 4 weeks
- Relapses are common (up to 40%)

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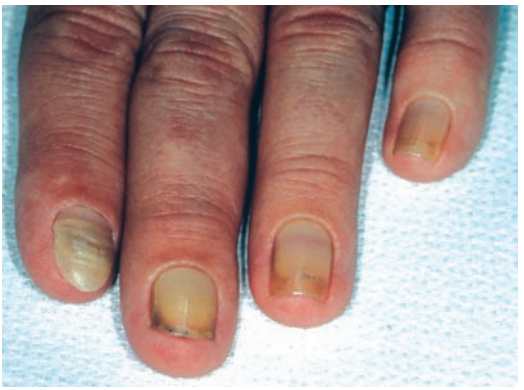

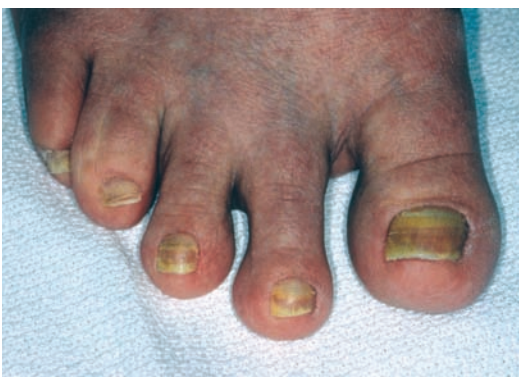

The nails shown here are those of a 56-year-old ill-appearing woman. Which one of the following organ systems has the highest likelihood of being diseased?

a. Liver
b. Kidneys
c. Heart
d. Lungs
e. Pancreas

Yellow Nail Syndrome

Answer: d

- Classic triad:

  Yellow nails
  Extremity lymphedema
  Pleural effusions

- Nail findings:

  Discoloration: yellow to yellow-green or brownish yellow
  Slow growth
  Onycholysis

- Pulmonary associations (not in 100%):

  Pleural effusion (35%-40%; lymphocyte predominant, often bilateral, one-third are recurrent)
  Restrictive and obstructive lung defects
  Bronchiectasis (20%)

- Systemic associations (not in 100%):

  Rhinosinusitis
  Chronic edema of lower extremities
  Breast edema
  Raynaud's phenomenon
  Pericardial effusion

- Lymphatic insufficiency is common and results in the edema
- Nail changes may be reversible and do not necessarily correlate with other manifestations of the syndrome (such as pulmonary disease, edema)
- No treatment has been proved effective

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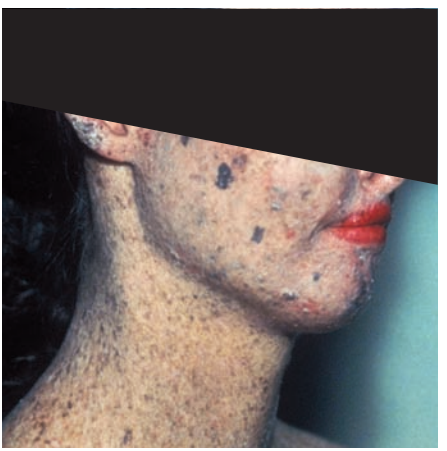

In this 15-year-old girl, these lesions developed in sun-exposed areas. She is exquisitely sensitive to sunburn. What is the cause of this rare disorder?

a. Systemic lupus erythematosus
b. Defective porphyrin metabolism
c. Autoimmune reaction to self-antigens in the skin
d. Autosomal recessive defect in DNA excision repair mechanism
e. A vertically transmitted hepatitis virus

Xeroderma Pigmentosum

Answer: d

- Autosomal recessive disorder found in all racial groups
- Multiple mutations have been identified, but all involve defects in the excision repair mechanism of damaged DNA in skin exposed to ultraviolet light (nucleotide excision repair)
- Skin findings:

  Actinic keratoses
  Cutaneous melanoma
  Progressive atrophy
  Irregular pigmentation
  Telangiectases
  Basal cell carcinoma
  Squamous cell carcinoma

- Patients are very sensitive to sun exposure, as evidenced by reports of tongue tumors
- Skin cancers develop at an early age (median, 8 years)
- Most patients die of these malignancies by the third decade of life
- Risk for skin cancer is 2,000- to 10,000-fold greater in patients with xeroderma pigmentosum than in age-matched controls
- Ophthalmic and neurologic disorders also occur
- Diagnosis is established with the fibroblast survival test after exposure to ultraviolet light

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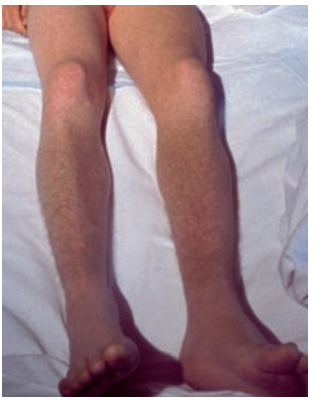

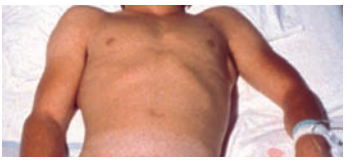

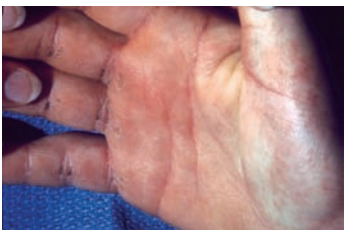

This 25-year-old man was hiking in North Carolina 1 week before development of rash and prostrating illness, including hypotension, meningoencephalitis, anemia, and bleeding. What is the most appropriate course of action?

a. Blood cultures and empiric therapy with vancomycin, gentamicin, and metronidazole
b. Blood cultures, supportive care, lumbar puncture, and empiric doxycycline
c. Supportive care until serologic diagnosis can be made to direct therapy
d. Discussion with family about withdrawal of support and palliative care
e. Transesophageal echocardiography

Rocky Mountain Spotted Fever (RMSF)

Answer: b

- Although first described in Idaho in 1896, RMSF is most common in the mid-Atlantic states and Oklahoma, not the Rocky Mountains
- *Rickettsia rickettsii*, a gram-negative intracellular bacterium, is inoculated into humans by ticks (*Dermacentor variabilis* and *D. andersoni*, the dog and wood tick, respectively) after at least 6 hours of feeding
- Most cases (>90%) occur between April and September
- Incubation time is 2 to 14 days (mean, 7 days)
- Rash begins on the extremities and moves centrally
- Major symptoms and signs: fever (88%-100%), headache (79%-93%), myalgia (72%-92%), rash (74%-90%), known tick bite (54%-66%), nausea and vomiting (55%-60%), classic triad of fever, headache, and rash (45%-60%), classic tetrad of fever, headache, rash, and history of tick bite (3%-18%)
- Serologic test result is not positive until 7 to 10 days after exposure; therefore treatment should not be delayed for diagnosis
- Mortality rate is 20% to 25% in 8 to 15 days without appropriate treatment and 5% despite appropriate treatment
- Treatment of choice is doxycycline. Other tetracyclines and chloramphenicol are also effective

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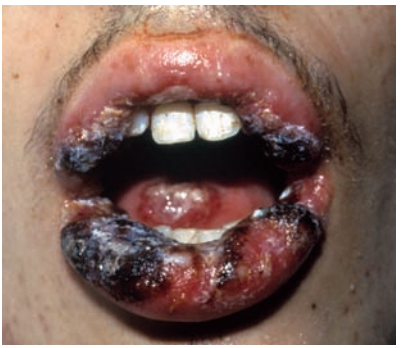

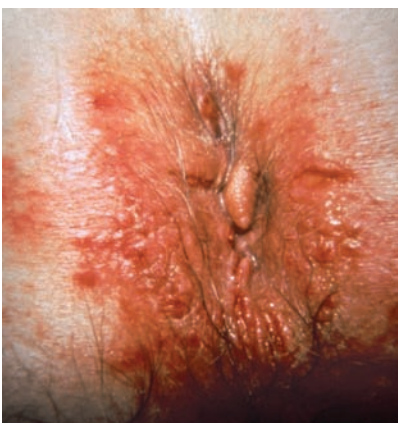

A 17-year-old boy with these lesions complains of abdominal pain and back pain. All of the following are true of this disease *except*:

a. From 10% to 20% of patients have arthritis that mirrors disease activity
b. It may be associated with ankylosing spondylitis
c. Renal oxalate stones occur in 5% to 10% of patients
d. Smoking has been associated with a preventive effect
e. Pathologic findings are marked by segmental bowel inflammation and skip lesions

Answer: d

- Lesions can develop anywhere from the mouth to the anus
- Pathologic findings are marked by segmental inflammation of involved bowel with skip lesions
- Associations include intestinal fistulas, strictures, and perianal disease
- 10% to 20% of patients have associated arthritis that mirrors bowel disease activity
- Crohn's disease may be associated with ankylosing spondylitis (positive for HLA-B27)
- Skin lesions are present in 10% of patients and include pyoderma gangrenosum, aphthous ulcers, and erythema nodosum
- Renal oxalate stones occur in 5% to 15% of patients and are due to malabsorption of calcium (oxalate is then preferentially absorbed)
- Unlike ulcerative colitis, in which smoking appears to have some preventive effect, smoking is a risk factor for Crohn's disease. Smokers also have a higher incidence of recurrent Crohn's disease
- Smoking cessation should be strongly encouraged for patients with Crohn's disease
- There is no effective prophylaxis for Crohn's disease
- Treatment depends on the site and severity of the disease and may involve sulfasalazine, corticosteroids, 6-mercaptopurine, metronidazole, infliximab, surgery, and nutritional support

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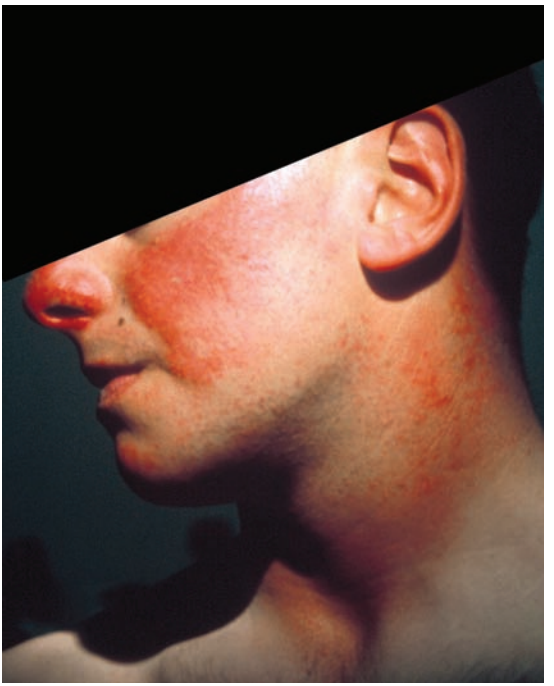

After treatment for urethritis, an 18-year-old man presents with the skin condition shown here. Other appropriate uses for the agent implicated in this case include all of the following *except*:

a. A 45-year-old woman with pelvic inflammatory disease
b. A 62-year-old man with early-stage Lyme disease
c. A 29-year-old pregnant woman with *Vibrio cholerae*
d. A 55-year-old woman with *Borrelia recurrentis*
e. A 30-year-old man with community-acquired pneumonia

Tetracycline-Induced Phototoxic Dermatitis

Answer: c

- The major toxic effects of the tetracycline class of agents (tetracycline, minocycline, and doxycycline) include the following:

  Rash
  Urticaria
  Angioedema
  Photosensitivity
  Dyspepsia

- More unusual side effects include the following:

  Exacerbation of underlying uremia
  Acute fatty liver disease
  Pancreatitis
  Pseudotumor cerebri

- These agents must be avoided in pregnancy because they can cause retardation of bone growth and discoloration of teeth in children
- These agents are the drugs of choice for treatment of rickettsial and chlamydial infections and for community-acquired pneumonia in adults younger than 40 years
- Serious adverse reactions to one of the tetracycline class of agents should preclude future prescribing of other tetracyclines because of cross-reactivity

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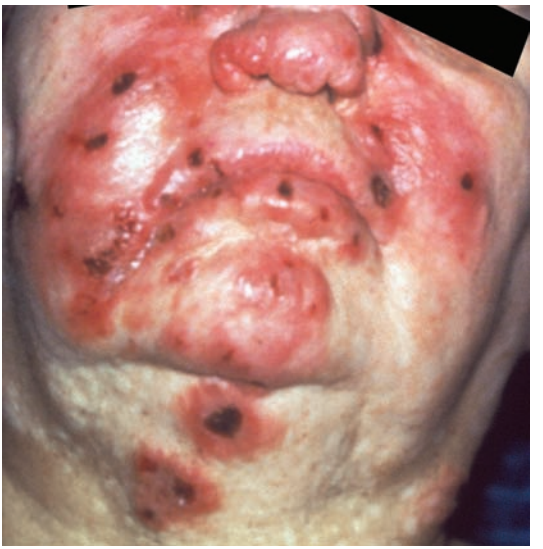

A 42-year-old woman presents with pink-red macules of the face, palms, and soles. She has had multiple sexual partners. After she is treated with an antibiotic, fever, hypotension, and worsening of the rash develop. Which one of the following occurred?

a. Drug allergy
b. Sepsis syndrome
c. Jarisch-Herxheimer reaction
d. Angioedema
e. Stevens-Johnson reaction

Jarisch-Herxheimer Reaction

Answer: c

- The Jarisch-Herxheimer reaction occurs within 1 to 2 hours of treatment of syphilis with antibiotics, especially penicillin
- The reaction is caused by release of pyrogen from the spirochetes
- It is most common during the treatment of secondary syphilis (70%-90% of cases)
- The reaction is characterized by the following:

  Fevers
  Chills
  Sweats
  Headache
  Hypotension
  Worsening of the skin lesions

- The reaction usually resolves within 24 hours of treatment
- Treatment of the reaction is supportive only
- Similar reactions have been reported with treatment of the following:

  Lyme disease
  Borreliosis
  Brucellosis
  Typhoid fever
  Trichinellosis

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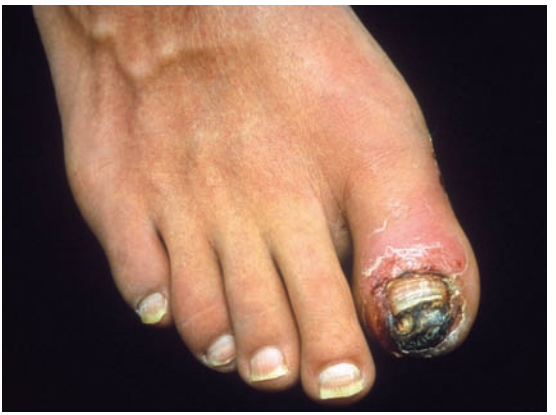

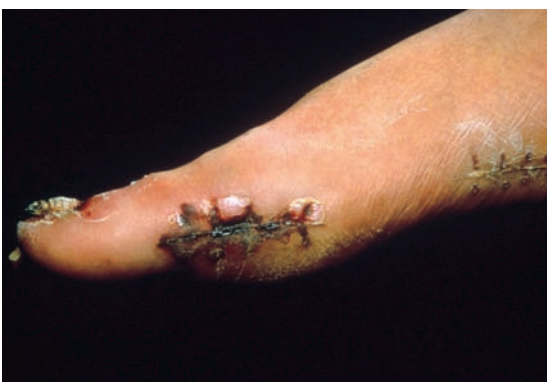

A 34-year-old male smoker reports having rest pain and lower extremity digital ulcers for 2 months. He also reports a 1-year history of claudication. Which one of the following is the best means of preventing amputation?

a. Smoking cessation
b. Calcium channel blockers
c. Peripheral artery bypass
d. Pentoxifylline
e. Warfarin anticoagulation

Buerger's Disease (Thromboangiitis Obliterans)

Answer: a

- The cause of Buerger's disease is unknown, but it is closely linked to smoking
- Few, if any, cases occur in the absence of tobacco use
- The disease affects the small and medium-sized arteries and veins of the extremities
- Affected vessels are thrombosed and infiltrated with inflammatory cells, resulting in ischemia
- Intraluminal thrombus contains microabscesses
- Markers that are common in other vasculitides are usually negative or normal in Buerger's disease, including sedimentation rate, C-reactive protein, complement, cryoglobulins, antinuclear antibodies, and rheumatoid factor
- In the Mayo Clinic experience, the incidence of the disease declined from 104 per 100,000 in 1947 to 12.6 per 100,000 in 1986
- Most patients (75%) are men
- 40% of persistent smokers with Buerger's disease face amputation
- The disease arrests with smoking cessation, and patients who quit smoking usually avoid amputation

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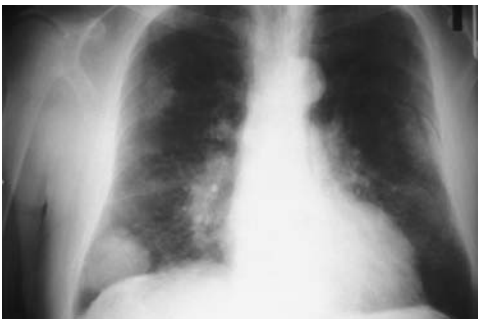

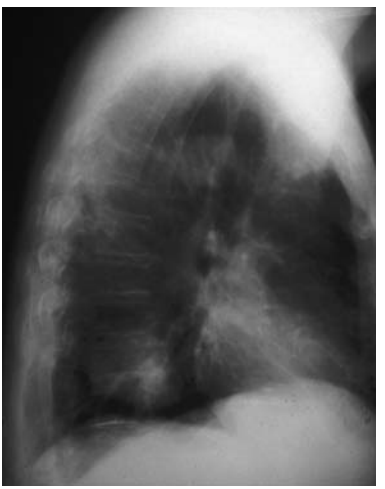

A 78-year-old male smoker presents with profound weight loss and a chronic cough. During World War II he was employed at the local navy shipyard. Which one of the following is the most common pulmonary finding in this disorder?

a. Pulmonary embolism
b. Pleural effusion
c. Pleural plaques
d. Pulmonary fibrosis
e. Malignant mesothelioma

Asbestos Exposure

Answer: c

- Exposure to asbestos fibers has several clinical associations, including pleural effusions, pleural plaques (most common), and malignant mesothelioma. Lower lobe abnormalities predominate in asbestos exposure
- Cumulative dose and time since first exposure are key determinants to the toxicity of asbestos illnesses. Clinical manifestations may occur 15 to 40 years after initial exposure
- Two classes of asbestos fibers:

  Serpentine–curly. Chrysotile is a subtype and accounts for 95% of asbestos used worldwide

  Amphibole–straight and rodlike, greater persistence in lung tissue, and generally considered more pathogenic than serpentine asbestos fibers, which more readily dissolve in vivo
- Asbestosis refers to diffuse interstitial lung disease (pulmonary fibrosis), which in severe cases may lead to cor pulmonale
- Evidence suggests that release of free radicals has a pivotal role in inducing lung injury after asbestos exposure
- Smoking does not increase the death rate in patients with malignant mesothelioma. It does, however, greatly increase the death rate in patients with asbestosis (up to 3 times)
- Smoking in combination with asbestos exposure substantially increases the risk for development of bronchogenic carcinoma (shown in this case)

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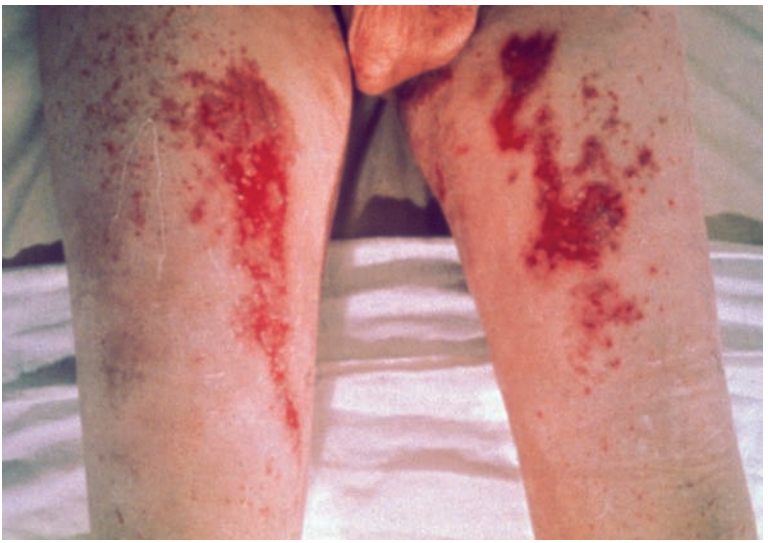

This patient presents with the skin lesions shown and with persistent hyperglycemia. Additional features of this syndrome include all of the following *except*:

a. Fever
b. Glossitis
c. Anemia
d. Diarrhea
e. Weight loss

Glucagonoma Syndrome With Necrolytic Migratory Erythema

Answer: a

- Syndrome is rare; occurs at 40 to 60 years
- Diagnosis is made with a serum glucagon test
- About 70% of patients have necrolytic migratory erythema: peeling, crusting, erosions of perineum and perioral areas
- Other features may include:

  Normochromic normocytic anemia
  Diarrhea
  Weight loss
  Glossitis, stomatitis, and angular cheilitis
  Diabetes mellitus
  Hypoaminoacidemia (glucagon enhances degradation of amino acids)

- Associations include pancreatic duct and islet cell tumors of the pancreas and, in 20% of cases, multiple endocrine neoplasia type I. Remainder of cases are sporadic
- Malignancy of glucagonomas is proportional to size. Generally, most malignant tumors are more than 5 cm
- The liver is the most common site of distant metastases
- Management in setting of glucagonoma may include resection, somatostatin, intravenous amino acid infusion, hepatic artery embolization, and zinc replacement
- Pseudoglucagonoma syndrome occurs with liver cirrhosis, Crohn's disease, celiac sprue, and malabsorption syndromes

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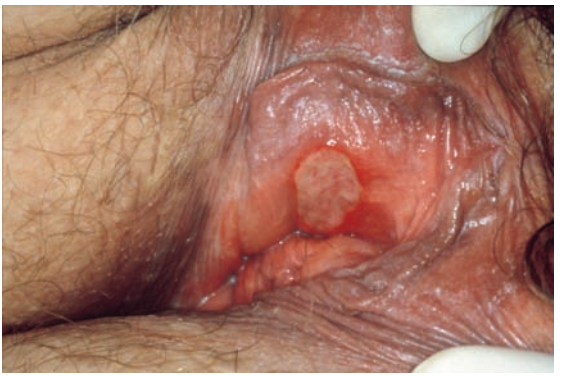

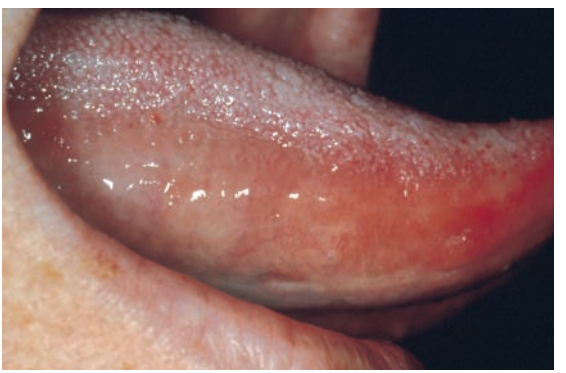

A 60-year-old woman presents with arthritis and a long history of recurrent painful ulcers of the mouth and genitalia. Past medical history is notable for venous thromboembolism and uveitis. What is the diagnosis?

a. Primary syphilis
b. Herpes simplex
c. Sarcoidosis
d. Behçet's syndrome
e. Niacin deficiency

Behçet's Syndrome

Answer: d

- Behçet's syndrome is a chronic, relapsing, multisystem, inflammatory disorder characterized by recurrent oral aphthous ulcers and any 2 of the following:

  Genital aphthous ulcers
  Uveitis
  Cutaneous nodules or pustules
  Phlebitis
  Arteritis
  Arthritis
  Meningoencephalitis

- Behçet's syndrome is more common in patients of Japanese and Middle Eastern descent
- Patients are at high risk for venous thromboembolism (deep venous thrombosis and pulmonary embolism occur in 7%-37% of patients)
- Pulmonary vascular involvement may result in serious hemoptysis; indeed, hemoptysis is the cause of death in 39% of patients
- Chest radiographs may show the following:

  Infiltrates
  Effusion
  Pulmonary artery aneurysms
  Prominent pulmonary arteries

- Treatment usually includes corticosteroids or other immunosuppressive agents

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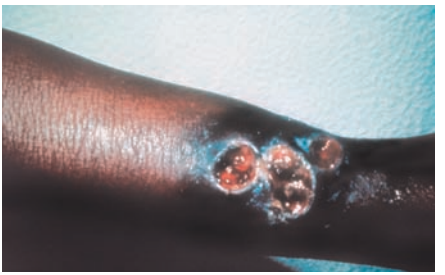

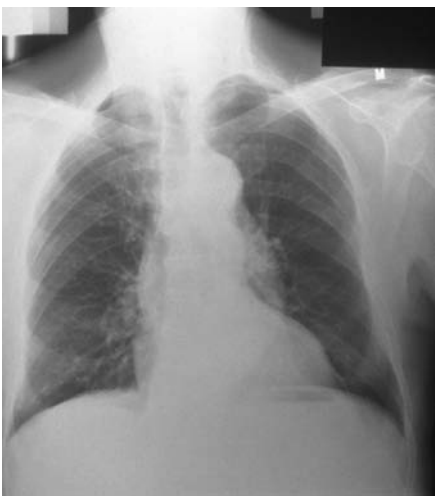

A 42-year-old woman with a past history of multiple sexually transmitted diseases and progressive dementia has the skin lesions shown here and shortness of breath. Of the procedures listed below, which one is most important for directing definitive therapy for the unifying diagnosis?

a. Lumbar puncture
b. Echocardiography
c. Magnetic resonance imaging
d. Triple-phase bone scanning
e. Skin biopsy

Tertiary Syphilis

Answer: a

- Manifestations include the following:

  Gummatous osteomyelitis
  Aortitis (with ascending aortic aneurysm and aortic regurgitation)
  Neurosyphilis
  Tabes dorsalis

- Neurosyphilis is the most common form of tertiary syphilis in the United States
- Fluorescent treponemal antibody absorption test is positive in 98%, and VDRL test is positive in 70%
- Seropositivity mandates cerebrospinal fluid examination because neurosyphilis is often asymptomatic
- Symptomatic neurosyphilis usually involves 1 of 2 syndromes:

  Meningovascular syphilis
    Occurs 4-7 years after initial infection
    Focal deficits (such as cerebrovascular accidents, cranial nerve deficits)
  Parenchymatous neurosyphilis
    Occurs decades after initial infection
    Varied deficits, including:
      General paresis
      Chronic progressive dementia
      Tabes dorsalis

- Treatment of choice for neurosyphilis is penicillin G 12 to 24 million units intravenously for 10 to 14 days
- Serologic titer should decline more than 4-fold and cerebrospinal fluid should normalize within 2 years of treatment

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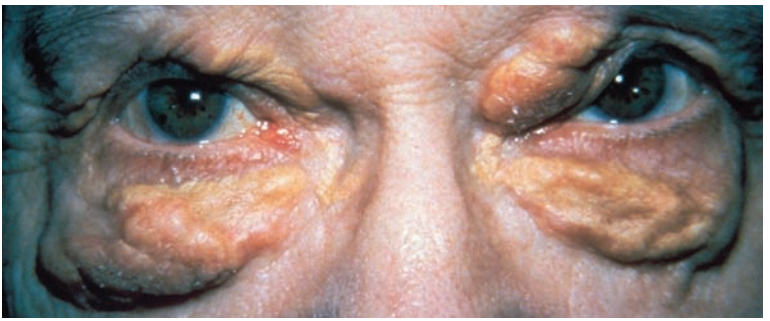

A 50-year-old woman presents with the lesions shown here, which are yellow on cut section. All of the following are true about this condition *except*:

a. These lesions develop in 10% of patients with primary biliary cirrhosis
b. These lesions consistently correlate with increased cholesterol levels
c. These lesions are benign
d. Incidence increases with age
e. Women are more likely to have this condition than men

Xanthelasma (Xanthoma Palpebrarum)

Answer: b

- Xanthelasma is the most common cutaneous xanthoma
- Soft, semisolid, or calcific yellow plaques on eyelids or inner canthi are characteristic
- Diagnosis is purely clinical
- Microscopic examination reveals lipid-filled foamy histiocytes
- There is no consistent association with hyperlipidemia
- Serum lipid values are normal in many series (25%-70%; average, 50%)
- Xanthelasma is more common in women than in men
- Incidence increases with age
- Lesions tend to be symmetric, permanent, and progressive
- Xanthelasma may be associated with increased β-lipoprotein values in young adults
- Xanthelasma is often noted in descriptions of primary biliary cirrhosis, but it is a late manifestation and occurs in only 10% of patients with this type of cirrhosis
- The lesions alone are benign
- Unless the lesions obstruct vision, treatment is for cosmetic reasons only
- If pursued, treatment may involve excision, laser ablation, and topical trichloroacetic acid
- Recurrence after all forms of treatment is common

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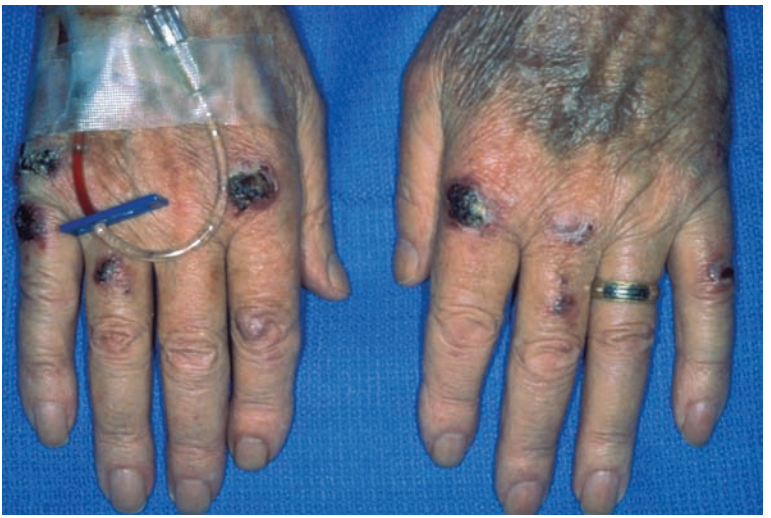

A 75-year-old man presents with weight loss, painful plaques on his hands, and a low-grade fever. This syndrome is most often associated with patients who have which one of the following?

a. Celiac sprue
b. Acquired immunodeficiency syndrome (AIDS)
c. Acute leukemia
d. Multiple myeloma
e. Endocarditis

Sweet's Syndrome

Answer: c

- Sweet's syndrome is also known as acute febrile neutrophilic dermatosis
- Originally described by Robert Sweet in 1964 as having 4 features:

  Fever
  Blood neutrophilic leukocytosis
  Raised painful plaques on the limbs, face, and neck
  Histologic findings of dense infiltration of the dermis by mature neutrophils
- The syndrome may be idiopathic, drug-related, or associated with malignancy
- In up to 20% of cases, the syndrome is associated with hematologic illness; of these, 50% are acute myelogenous leukemia
- Extracutaneous manifestations include diffuse arthralgias, proteinuria, and episcleritis
- Appropriate management includes treatment of the underlying disorder and use of systemic corticosteroids
- Potassium iodide and colchicine may be used in patients who should not take corticosteroids

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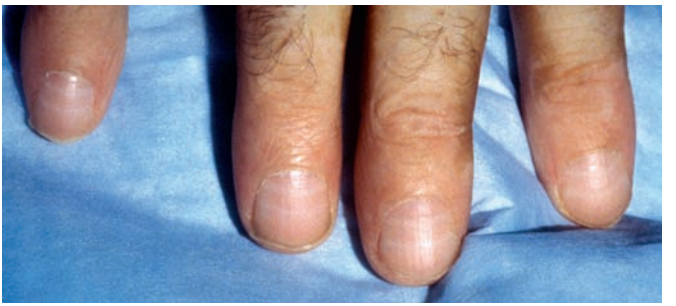

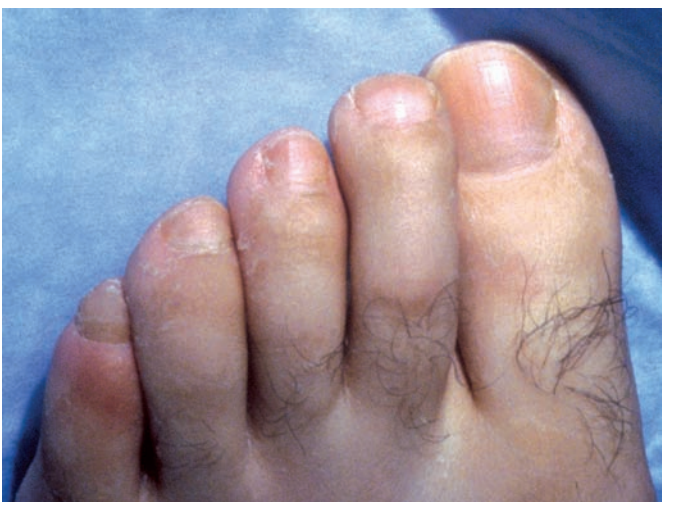

A 37-year-old microelectronics worker presents with "stocking-glove" peripheral sensory neuropathy, distal weakness, and the nail changes shown here. Which one of the following is the most effective treatment?

a. Plasma exchange
b. Systemic antifungal medication
c. Intravenous immunoglobulin
d. Avoidance of further exposure to the causative agent
e. Chelation therapy

Mees' Lines Caused by Arsenic Toxicity

Answer: d

- Arsenic exposure can occur in the microelectronics industry (gallium arsenide), from water in some wells, in the smelting industry, from pesticides, and from intentional poisoning
- Manifestations of arsenic toxicity include hair changes, exfoliative dermatitis, sensorimotor polyneuropathy, nephropathy, abdominal pain, nausea, vomiting, diarrhea, and mucosal erosions
- Urinary arsenic levels show recent exposures
- Analysis of hair or nails reveals arsenic months after exposure
- Avoidance of further exposure is the only proven treatment for chronic toxicity. Neuropathic changes may improve, but often not completely
- Mees' lines:

  Transverse striate leukonychia (white lines in transverse direction on nails)
  Result from disruption of nail plate keratinization
  Occur 2 to 3 weeks after acute arsenic ingestion. Also occur in chronic poisonings
  Distance from Mees' lines to the nail fold can be used to estimate the time since exposure
  Also may occur from poisoning with thallium, lead, sulfonamides, pilocarpine, and chemotherapy and from stress of severe systemic illness

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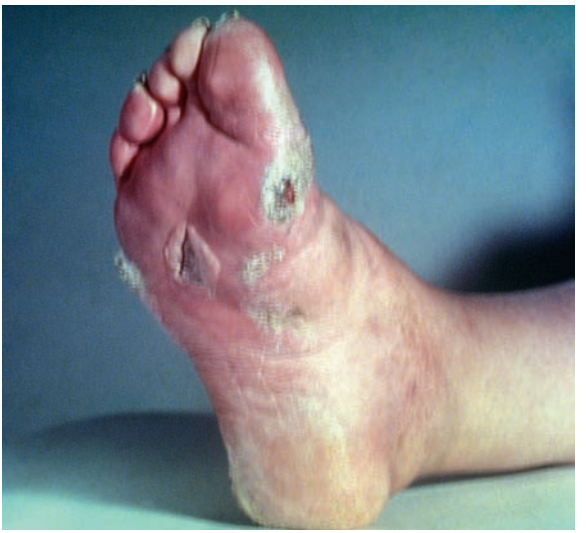

The lesions shown developed in a 41-year-old nonsmoking man after 5 days of dry cough and fever. About 2 weeks previously, he had been hiking in Arizona. What is the most likely diagnosis?

a. Atherosclerosis obliterans
b. Endocarditis
c. Histoplasmosis
d. Blastomycosis
e. Coccidioidomycosis

Coccidioidomycosis

Answer: e

- *Coccidioides immitis* is a dimorphic fungus endemic to the southwestern United States
- Valley fever is named after the San Joaquin Valley in California, where the first patients with the clinical syndrome (including fever, cough, fatigue, and weight loss) were identified in 1896
- Travel history is very important to recognizing this condition
- Impaired cellular immunity (such as from steroids, transplantation, human immunodeficiency virus) increases the risk of infection and reactivation of infection, which may have been acquired years previously
- Extrapulmonary manifestations are usually focal. Investigation for them should be prompted by history and physical examination: headache → lumbar puncture, nonhealing ulcers → biopsy, joint effusion → aspiration, skeletal pain → tests for osteomyelitis
- Erythema nodosum, if present, is a good prognostic sign
- Culture, biopsy with silver stains, and serologic testing in appropriate clinical settings are used for diagnosis
- If the patient is immunocompetent, infection is usually self-limited, resolving in weeks to months without treatment
- Reasons for treatment include weight loss of more than 10%, night sweats for longer than 3 weeks, infiltrates in both lungs or more than half of one lung, persistent adenopathy, titers of more than 1:16, lack of development of hypersensitivity antigens, symptoms for longer than 2 months, and persistent absence from work

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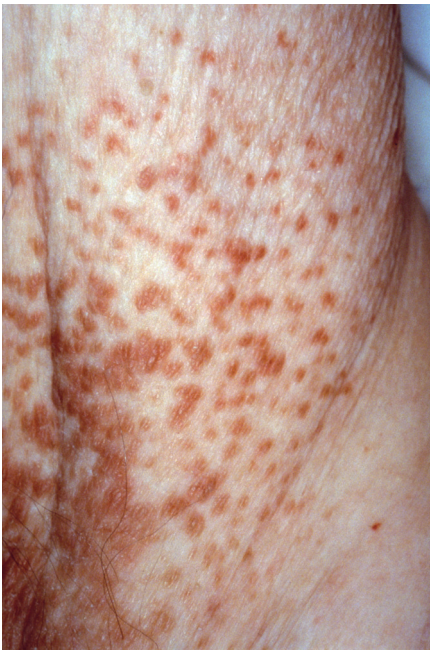

A 50-year-old woman presents with chronic pruritus. On examination, you notice red-brown papular lesions, as shown here. Her pulse is 110 beats per minute, and she appears flushed. The systemic manifestations of this illness are primarily from endogenous release of which one of the following?

a. Interleukin-1
b. Histamine
c. Norepinephrine
d. Acetylcholine
e. Dopamine

Urticaria Pigmentosa

Answer: b

- Mastocytosis (mast cell disease) may be broadly subclassified as follows:

  Indolent mastocytosis
    Cutaneous (urticaria pigmentosa, mastocytoma)
    Systemic
  Mastocytosis with associated hematologic disorder
    Myeloproliferative
    Malignant lymphoma
  Lymphadenopathic mastocytosis with eosinophilia
  Mast cell leukemia

- The skin lesions typically urticate with minimal trauma (Darier's sign)
- The systemic findings of tachycardia, flushing, and diarrhea are due to mast cell degranulation or release of histamine
- Diagnosis is based on increased urinary levels of the histamine metabolites (*N*-methylimidazole acetic acid and methylhistamine). These are more sensitive and specific than total histamine levels
- Disease activity also may be correlated with plasma tryptase levels
- Management includes judicious use of histamine$_1$ and histamine$_2$ antagonists
- Cutaneous mastocytosis may be especially responsive to psoralen with ultraviolet light of A wavelength (PUVA) photochemotherapy

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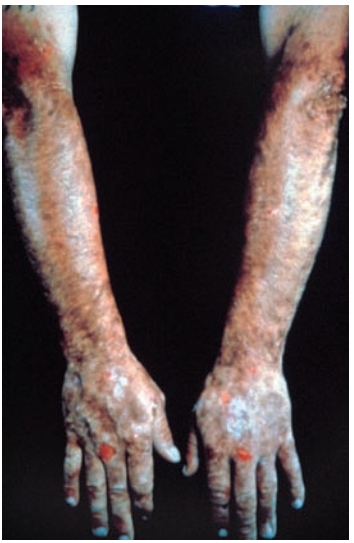
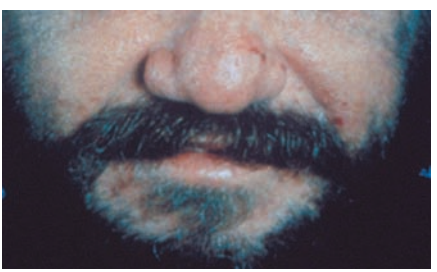




A 45-year-old laborer presents with vesicles and bullae in light-exposed areas. These lesions leave scars. He is an alcoholic. He reports no abdominal pain or loose stools. His mental status is normal. What is the diagnosis?

a. Porphyria variegata
b. Acute intermittent porphyria
c. Porphyria cutanea tarda
d. Coproporphyria
e. None of the above

Porphyria Cutanea Tarda

Answer: c

- Porphyria cutanea tarda (PCT) is the most common porphyria
- Patients with PCT have reduced activity of hepatic uroporphyrinogen decarboxylase
- If inherited, the defect is autosomal dominant
- Skin findings include the following:

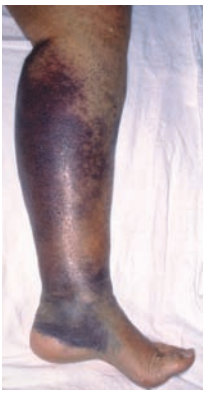
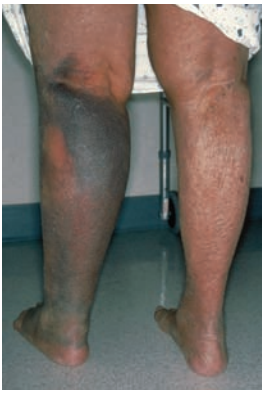
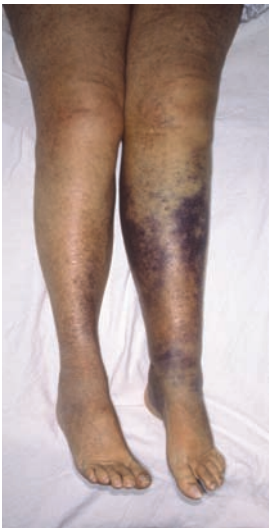
  Photosensitivity
  Dorsal erosions and bullae
  Hyperpigmentation
  Facial reddening

- Patients with PCT do not experience gastrointestinal or neurologic symptoms
- Associations include the following:

  Alcohol
  Hepatitis C
  Estrogens
  Iron
  Other toxins (such as hexachlorobenzene)

- Diagnosis is made by assessing urinary porphyrins
- The mainstay of treatment is phlebotomy, although chloroquine has been used

Reference
Habermann TM. *Mayo Clinic Internal Medicine Board Review 2004-2005.* Philadelphia: Lippincott Williams & Wilkins; 2004:183, 461.

A 71-year-old woman presented with swelling of the left calf. Deep venous thrombosis was diagnosed, and therapy with intravenous heparin was started. Subsequently, a large hematoma developed on the left lower extremity. What should be done now?

a. Stop heparin therapy
b. Perform ultrasonography of the right popliteal fossa
c. Give intravenous antibiotics
d. a + b + c
e. a + b

Ruptured Baker's Cyst

Answer: e

- Condition is named for William M. Baker (1839-1896), an English surgeon
- The signs and symptoms of a ruptured Baker cyst can mimic those of acute deep venous thrombosis (DVT)
- Baker's cyst is also known as "pseudothrombophlebitis" and popliteal cyst
- The frequency of a ruptured Baker cyst among patients with suspected acute DVT is less than 4%
- The predictive values of the history and physical examination for DVT are poor
- In a series of more than 3,000 patients evaluated for DVT, 95 (3.1%) had a Baker cyst (Langsfeld et al.)
- Ultrasonography is the best test for both DVT and Baker's cyst

References

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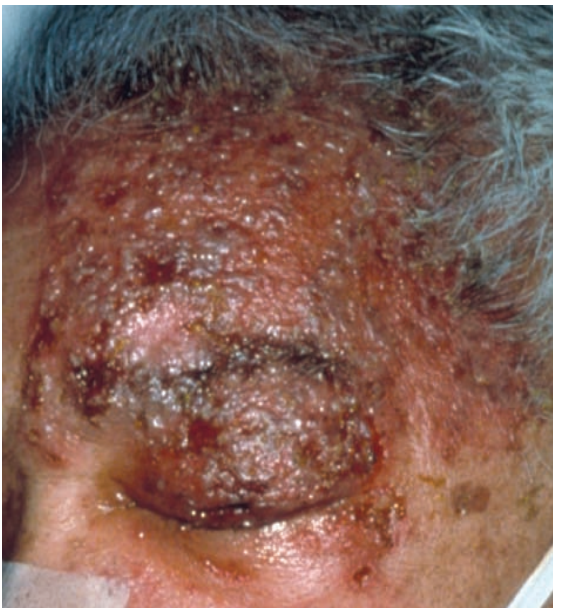
Volteas SK, Labropoulos N, Leon M, et al. Incidence of ruptured Baker's cyst among patients with symptoms of deep vein thrombosis. *Br J Surg*. 1997;84:342.



A 75-year-old man has profound obtundation, pneumonia, and a vesicular rash. All of the following are appropriate therapies *except*:

a. Acyclovir
b. Famciclovir
c. Foscarnet
d. Corticosteroids
e. Valacyclovir

Answer: d

- Primary infection with varicella-zoster virus (VZV) commonly occurs in childhood and manifests as chickenpox. Infectivity is highest just before the onset of rash
- The virus then may enter a latent stage and remain dormant in dorsal root and cranial nerve ganglia
- Illnesses associated with VZV include the following:

  Varicella pneumonia
  Encephalomyelitis
  Zoster ophthalmicus (cranial nerve V [ophthalmic branch] involvement) (as in this case)
  Shingles (painful vesicular rash in a dermatomal distribution which does not cross the midline)
  Cerebellar ataxia
  Acute retinal necrosis

- Postherpetic neuralgia is one of the most debilitating and common complications of VZV
- VZV infection may be resistant to acyclovir, especially in patients with acquired immunodeficiency syndrome
- The use of corticosteroids remains controversial. Some studies have shown a reduction in the duration of postherpetic neuralgia, and other studies have had equivocal results
- Varicella-zoster immune globulin may be administered (within 96 hours) for postexposure prophylaxis of varicella

References

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Cohen JI, Brunell PA, Straus SE, et al. Recent advances in varicella-zoster virus infection. *Ann Intern Med*. 1999;130:922-932.
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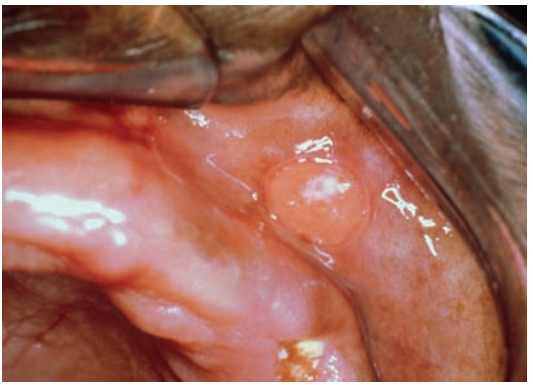

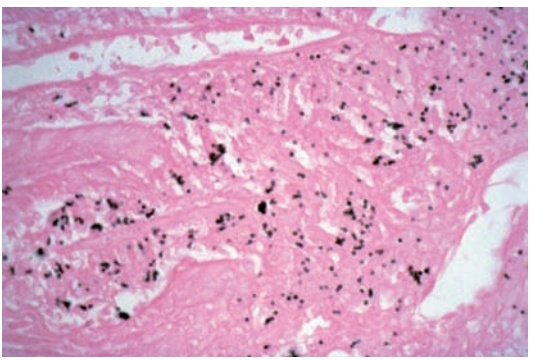

A 28-year-old patient with newly diagnosed human immunodeficiency virus (HIV) presents to your office with cough, fever, and fatigue. He remarks to you that he, nonetheless, has been enjoying the great outdoors at his log cabin along the banks of the Ohio River. This patient most likely has which one of the following?

a. *Cryptococcus neoformans*
b. *Histoplasma capsulatum*
c. *Coccidioides immitis*
d. *Blastomyces dermatitidis*
e. *Candida albicans*

Answer: b

- *Histoplasma capsulatum* is especially prevalent in the Ohio and Mississippi river valleys
- It exists as a dimorphic fungus and is associated with excavation projects, building sites, and bird droppings
- Mode of transmission is aerosol and is the most common cause for mediastinal granuloma. Clinically active disease develops in less than 5% of exposed persons
- Symptoms and clinical findings in the immunocompetent host include a flu-like pulmonary illness, pericarditis, arthritis or arthralgia, or erythema nodosum
- Early manifestations include a localized or patchy pneumonitis. Hematogenous dissemination to other organ systems occurs within the first 2 weeks
- Mucosal oral lesions, splenomegaly, weight loss, and fever are not uncommon findings. Adrenal insufficiency may occur from adrenal gland infiltration
- The disseminated form of histoplasmosis is more common in infants, elderly, and immunocompromised individuals
- Serologic test results are positive in up to 90% of patients with symptomatic illness. Host antibody responses reach high levels within 6 weeks of exposure. *H. capsulatum* antigens also may be rapidly detected in serum and urine
- Itraconazole is reasonable therapy for non–life-threatening forms of the illness. Amphotericin B is advised for life-threatening, severe infection

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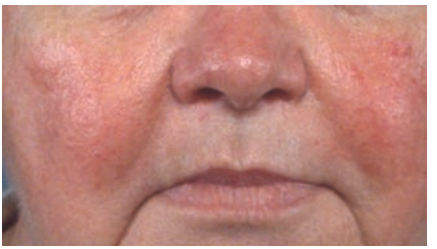

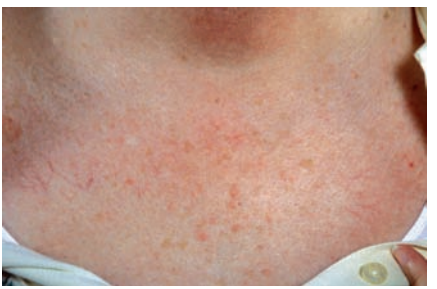

A 52-year-old woman has diarrhea, wheezing, and the flushing reactions shown here, which seemed to be associated with emotion and alcohol ingestion.

1. What is the best screening test?

1a. Urinary metanephrines
1b. Antimitochondrial antibodies
1c. Antinuclear antibodies
1d. Skin biopsy
1e. Urinary 5-hydroxyindoleacetic acid (5-HIAA)

2. Which one of the following likely will be most effective for alleviating the symptoms?

2a. β-Adrenergic blockade
2b. Psychiatry consultation
2c. Bulk fiber supplementation
2d. Somatostatin analogues
2e. Systemic corticosteroids

Carcinoid Syndrome

Answer 1: e

Answer 2: d

- Carcinoid tumors metastatic to the liver cause flushing, wheezing, and diarrhea
- Most common sites of origin are the following:

  Terminal ileum (90%)
  Lungs and bronchi
  Appendix
  Rectum
  Stomach

- The heart is involved in about 50% of patients who have pulmonary or liver metastases
- Cardiac involvement usually includes the right-sided valves (tricuspid and pulmonary regurgitation or stenosis)
- Serotonin-like substances are responsible for the symptoms
- Measurement of urinary 5-hydroxyindoleacetic acid (5-HIAA) is the diagnostic test of choice
- Computed tomography or magnetic resonance imaging is best for demonstrating liver metastases
- Somatostatin scintigraphy may help find lesions that are otherwise difficult to localize
- Somatostatin analogues are effective for relieving symptoms, but they do not cause the tumors to regress

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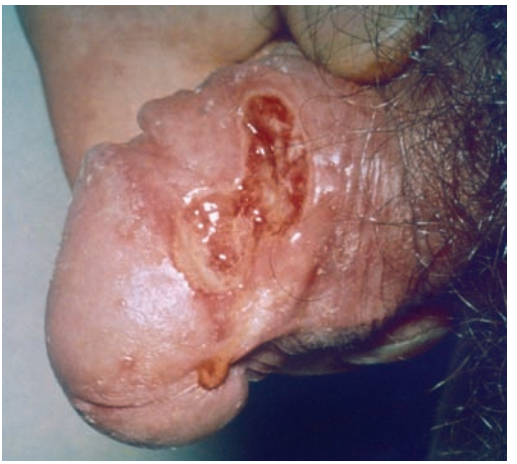
Kulke MH, Mayer RJ. Carcinoid tumors. *N Engl J Med*. 1999;340:858-868.



A 33-year-old sexually active man presents with this painless ulcer. Approximately how long ago did the inoculation occur?

a. 12 to 48 hours
b. 3 to 90 days
c. 3 to 6 months
d. 6 to 12 months
e. >1 year

Primary Syphilis

Answer: b

- A painless ulcer (chancre) appears at the site of inoculation within 3 to 90 days after infection
- The chancre will resolve spontaneously without treatment, but the patient remains infected
- Differential diagnosis includes herpes simplex and *Haemophilus ducreyi* (chancroid), but these ulcers are usually painful
- Diagnosis is with dark-field microscopy demonstrating spirochetes (*Treponema pallidum*)
- Fluorescent treponemal antibody absorption test is positive in 85% and VDRL test is positive in 70% of cases of primary syphilis
- Treatment of choice is benzathine penicillin 2.4 million units intramuscularly
- Follow-up serologic testing is recommended 6 to 12 months after treatment
- After successful treatment, VDRL test result should decrease at least fourfold

Reference

Habermann TM. *Mayo Clinic Internal Medicine Board Review 2004-2005*. Philadelphia: Lippincott Williams & Wilkins; 2004:592-593.

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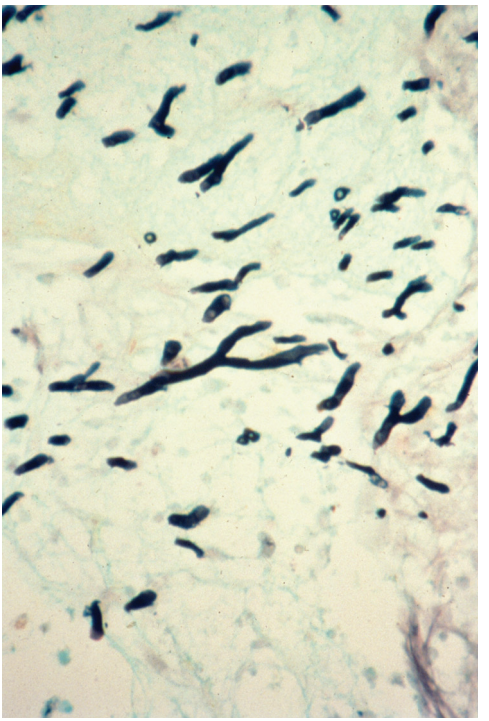

A 45-year-old woman is receiving her second cycle of CHOP (cyclophosphamide, hydroxydaunomycin, Oncovin [vincristine], prednisone) chemotherapy for diffuse large cell lymphoma. Dyspnea and cough develop, and the result of bronchoscopy with bronchoalveolar lavage is shown here. On chest radiography, a "fungus ball" is evident. Which one of the following is true regarding this finding?

a. It occurs in pathologically normal lung tissue
b. It commonly is implicated in aspiration pneumonia
c. Hemoptysis is a major complication
d. It may be responsible for underlying small bowel obstruction
e. Blood cultures are usually positive for fungemia

Aspergillosis

Answer: c

- Angioinvasive aspergillosis is the most severe manifestation of this infection and can result in massive hemoptysis. *Aspergillus fumigatus* is most commonly transmitted as an airborne pathogen
- Invasive aspergillosis most commonly occurs in immunocompromised hosts. Patients who have had bone marrow transplantation, are receiving chemotherapy, or have acquired immunodeficiency syndrome are especially predisposed
- *Aspergillus* organisms frequently colonize the respiratory tract; to document active infection, tissue infiltration should be demonstrated
- Bronchoalveolar lavage is positive for *Aspergillus* organisms in only 45% to 62% of patients with invasive aspergillosis
- Amphotericin B and itraconazole are acceptable therapies. Surgical resection (wedge resection or lobectomy) has been successful for isolated pulmonary infection
- The use of granulocyte colony-stimulating factor increases polymorphonuclear counts in neutropenic patients to better counteract *Aspergillus* infections
- "Fungus balls" are collections of fungal hyphae in preexisting lung bullae (such as caused by emphysema or tuberculosis)
- Computed tomography may show a layer of air surrounding the fungus ball (Monod sign)

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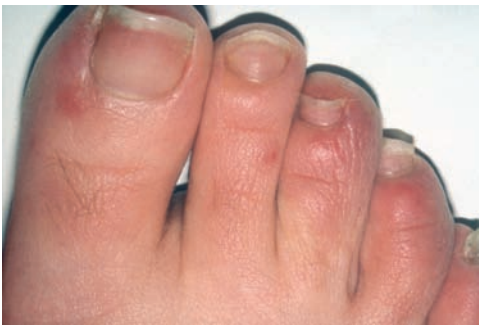
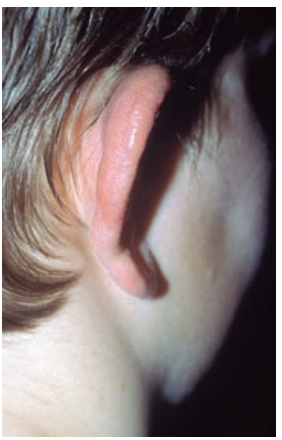
Paterson DL, Singh N. Invasive aspergillosis in transplant recipients. *Medicine (Baltimore)*. 1999;78:123-138.





A 16-year-old boy was playing broomball in southeastern Minnesota in January. The lesions shown here, which were painful, developed. All of the following may be appropriate in the treatment of this condition *except*:

a. Rapid rewarming of the affected areas in a bath at 40°C to 42°C for 15 to 30 minutes
b. Debridement of devitalized tissue after rewarming
c. Oral ibuprofen
d. Topical aloe vera to the affected areas
e. Massage of the affected areas

Answer: e

- Originally described in military populations, frostbite is common among urban and rural civilian populations as a result of the increase of outdoor winter activities and homelessness
- Alcohol consumption and psychiatric illness are important risk factors because they impair judgment with regard to appropriate duration of exposure
- Vasoconstriction and ischemia, as well as the direct cytodestruction of freezing, are the likely mechanisms of injury
- Hands and feet are involved in 90% of frostbite injuries. Other common sites are cheeks, nose, ears, and penis
- Treatment involves protection of the area until definitive rewarming can occur (freeze-thaw cycles are deleterious). Rapid rewarming in a bath at 40°C to 42°C for 15 to 30 minutes (or until completely thawed) with range-of-motion exercises is the preferred treatment
- Treatment after thawing includes debridement of devitalized tissue, elevation (reduces edema), tetanus prophylaxis, ibuprofen (inhibits thromboxane A_2, a mediator of rewarming inflammatory injury), aloe vera (a topical thromboxane A_2 inhibitor), and use of a whirlpool for the affected area
- Prophylactic antibiotics are controversial
- Massage and extreme heat are contraindicated
- Rewarming may be painful, necessitating analgesics
- The affected body part usually becomes extremely cold-intolerant

References

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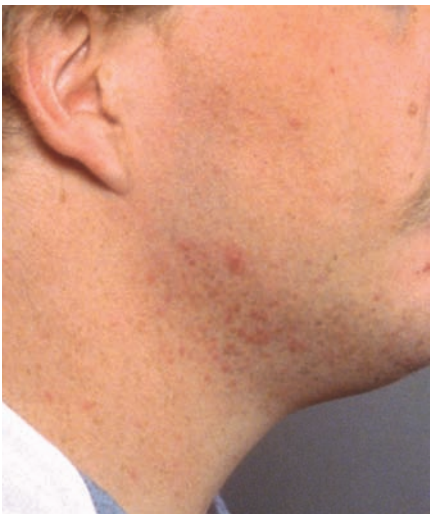
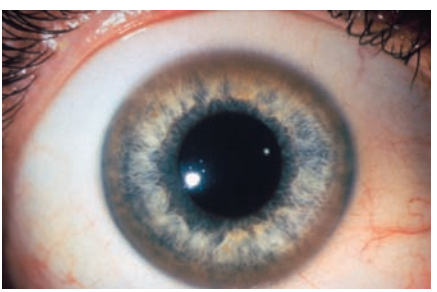
Murphy JV, Banwell PE, Roberts AH, et al. Frostbite: pathogenesis and treatment. *J Trauma*. 2000;48:171-178.





A 30-year-old man presents with personality changes, jaundice, joint pain, and this eye finding. The disease associated with these findings is caused by defects in metabolism of which element?

a. Zinc
b. Magnesium
c. Sodium
d. Selenium
e. Copper

Wilson's Disease (Hepatolenticular Degeneration)

Answer: e

- First described by Westphal, not Wilson, in 1883
- An autosomal recessive defect in biliary excretion of copper results in deposition in the liver, brain, and cornea, among other tissues
- Kayser-Fleischer rings are more frequent in patients with neurologic complications
- Neurologic findings include the following:

  Personality changes
  Rigidity
  Tremor
  Spasticity
  Dysarthria
  Dysphagia

- Liver manifestations include the following:

  Cirrhosis
  Cholestasis
  Hepatitis

- Arthropathy occurs in 50% of patients
- Wilson's disease was universally fatal until 1951, when it was discovered that British antilewisite, a chelator used for arsenic poisoning, increased urinary copper excretion and resulted in reversal of the tremor and rigidity of Wilson's disease
- Wilson's disease is now treated with penicillamine (increases urinary excretion of copper) with or without zinc (inhibits copper absorption in the gastrointestinal tract)

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Habermann TM. *Mayo Clinic Internal Medicine Board Review 2004-2005*. Philadelphia: Lippincott Williams & Wilkins; 2004:304-305, 955-956.
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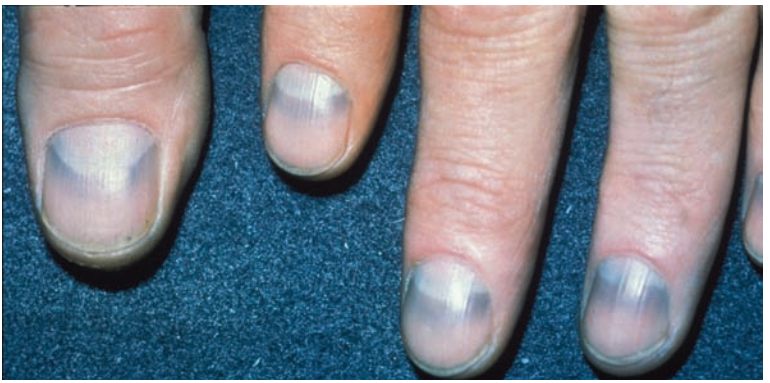

The man with the findings shown here had used a very old silver-containing sinus preparation for years. What is the diagnosis?

a. Muehrcke's nails
b. Terry's nails
c. Yellow nail syndrome
d. Green nail syndrome
e. Argyria

Argyria

Answer: e

- Findings include slate-gray or bluish discoloration of nails, skin, gingivae, sclerae, and some deep tissues as a result of long-term use of soluble silver salts
- Dark-field microscopic examination of sweat glands, hair follicles, and blood vessels shows refractile white granules
- Discoloration is not related to quantity of silver in skin
- Photoactivation reduces silver and results in more discoloration in areas exposed to sunlight
- Proprietary formulations for the nose, sinuses, and general cold remedies used to contain silver salts (now rare in the United States)
- More recently, retailers of health food supplements have promoted products containing colloidal silver protein for prevention of various ailments
- Discoloration is a result of prolonged use of silver-containing products (usually for more than 3 years)
- There is no effective treatment. Argyria is not reversible
- Differential diagnosis for blue nails includes the following:

  Wilson's disease
  Prolonged use of quinacrine or phenolphthalein
  Topical mercury
  Bacterial infection under the fingernail (such as *Pseudomonas aeruginosa* paronychia, but this is usually greener and rarely affects all the nails)

References
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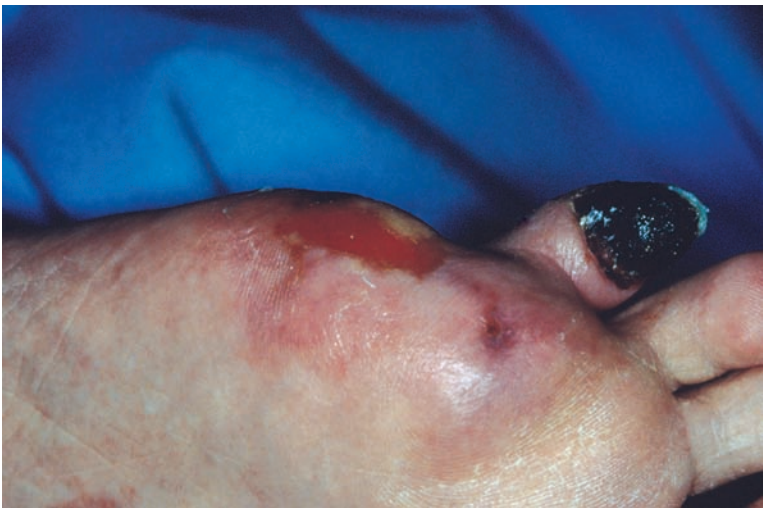
Koplon BS. Azure lunulae due to argyria. *Arch Dermatol.* 1966;94:333-334.



A 36-year-old woman with a history of multiple miscarriages has the lesions shown here. Her activated partial thromboplastin time is prolonged. Treatment should include which one of the following?

a. High-dose oral corticosteroids
b. Warfarin with a goal international normalized ratio of 3 to 4
c. Fresh frozen plasma
d. Plasma exchange and high-dose cyclophosphamide
e. Factor VIII transfusions for bleeding episodes

Answer: b

- APS may include recurrent arterial or venous thromboembolic disease, recurrent fetal loss, sterile endocarditis, livedo reticularis, and thrombocytopenia
- May be associated with rheumatic diseases (particularly systemic lupus erythematosus), certain infections, lymphoproliferative diseases, some drugs, and intravenous drug use
- Antibody associations include the following:

  Lupus anticoagulant, a coagulation inhibitor in vitro which increases clotting time in phospholipid-dependent tests (such as aPTT) with incomplete correction on mixing studies

  Antiphospholipid antibodies such as directed against cardiolipin, phosphatidyl serine, and β_2-glycoprotein I
- Neither antibody type is specific for the clinical syndrome. A causal connection between the antibodies and thrombosis is not fully delineated
- In patients who have had thrombosis, treatment is with warfarin anticoagulation (goal international normalized ratio 2.5-3.5 or higher, although higher-goal INRs have been questioned). Treatment is not recommended for incidental finding of the antibodies
- Catastrophic APS is simultaneous multiorgan involvement over a short time. Mortality is high (>50%) despite aggressive treatment

References

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Crowther MA, Ginsberg JS, Julian J, et al. A comparison of two intensities of warfarin for the prevention of recurrent thrombosis in patients with the antiphospholipid antibody syndrome. *N Engl J Med*. 2003;349:1133-1138.

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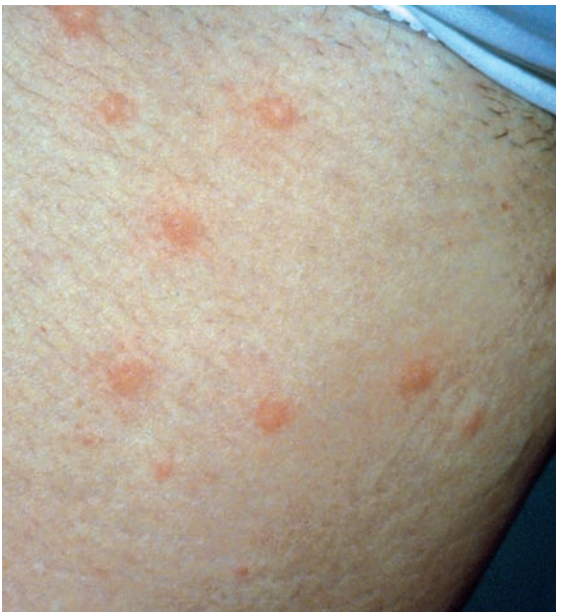

These pruritic papules developed in a real estate agent after inspecting a home in which there were several pets. Organisms similar to the ones that caused these lesions act as vectors for each of the following disease-causing microbes *except*:

a. *Yersinia pestis*
b. *Bartonella henselae*
c. *Rickettsia typhi*
d. *Rickettsia rickettsii*
e. *Rickettsia felis*

Flea Bites

Answer: d

- Dog and cat fleas (*Ctenocephalides canis* and *felis*) and rat fleas (*Xenopsylla cheopis*) commonly feed on the blood of humans and other warm-blooded animals
- Fleas are only 2- to 3-mm long but can jump up to 8 inches (20.3 cm) vertically and up to 16 inches (40.6 cm) horizontally
- After the blood meal, the female flea lays her eggs, and she may lay up to 2,000 eggs in a lifetime
- For every flea seen, there are 10 to 100 fleas in the area
- Body odor and sex hormones may make individuals more likely to be bitten; it may be that only one member of a family is bitten
- Flea bites present as pruritic papules on exposed skin surfaces, especially the lower extremities and ankles, usually sparing the genital, anal, and axillary areas. Itching is a result of the anticoagulant flea saliva that is injected before blood is eaten
- Affected areas should be gently cleaned. Wet compresses and mild topical corticosteroids may help the itching. Antibiotics are warranted only with superinfection, which occurs mostly due to intense scratching
- The environment should be cleaned thoroughly, vacuumed, and fumigated, especially areas where pets are located
- Fleas are the vectors for many disease-causing organisms, including *Yersinia pestis* (plague), *Rickettsia typhi* (murine typhus), *Rickettsia felis*, and *Bartonella henselae* (cat-scratch disease)

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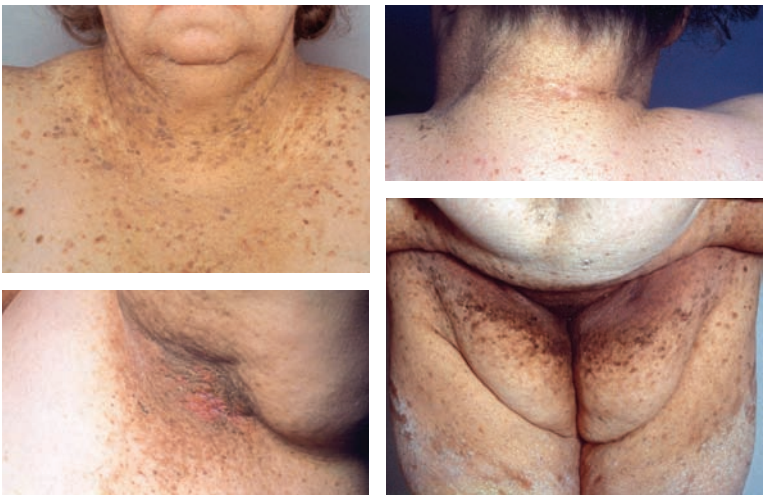

On physical examination, a 56-year-old obese woman with type 2 diabetes mellitus has soft, velvety, verrucous, brown pigmentation of the intertriginous regions. A previous physician correctly told her this skin finding was related to her obesity and diabetes. What disease, other than diabetes, also is associated with this skin finding?

a. Adenocarcinoma of the gastrointestinal tract
b. Cushing's disease
c. Polycystic ovaries
d. Thyroid disease
e. All of the above

Acanthosis Nigricans

Answer: e

- Acanthosis nigricans involves the intertriginous regions, especially the axillae and groin
- Acanthosis nigricans is associated with the following:

  Obesity
  Type 2 diabetes mellitus
  Cushing's disease
  Polycystic ovaries
  Thyroid disease
  Adenocarcinoma of the gastrointestinal tract
  Acromegaly
  Medications (e.g., prednisone, nicotinic acid)

- Acanthosis nigricans can be familial (usually autosomal dominant)
- Acanthosis nigricans is most common in persons younger than 40 years
- In older persons, especially if *thin*, acanthosis nigricans often is associated with an underlying malignancy

References
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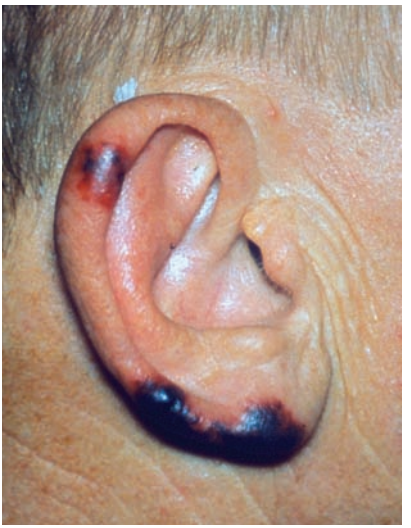

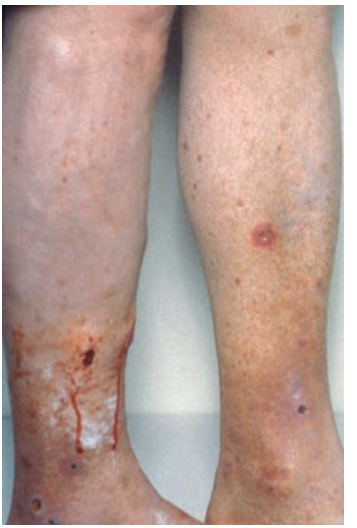

A 47-year-old man has hepatitis C, joint pains, weakness, polyneuropathy, and the skin lesions shown here. What is the most likely cause?

a. Churg-Strauss vasculitis
b. Buerger's disease
c. Porphyria cutanea tarda
d. Mixed cryoglobulinemic vasculitis
e. Polyarteritis nodosa

Type II (Mixed) Cryoglobulinemic Vasculitis

Answer: d

- Cryoglobulins are immunoglobulins that reversibly precipitate in cold temperatures and are classified by their immunoelectrophoresis and immunofixation patterns into types I, II, and III, depending on monoclonal and polyclonal distributions
- More than 70% of patients have one or more of the following: palpable purpura, arthralgia or arthritis, and weakness. The combination of all three major symptoms, known as Meltzer's triad, is found in less than 40% of patients
- Acral ulcerations and necrosis may occur where peripheral temperature is less than core temperature
- Polyneuropathy occurs in 40% to 70% of patients
- The vast majority of cases previously known as "essential mixed cryoglobulinemia" are now thought to be due to hepatitis C virus (HCV). Anti-HCV antibodies and HCV RNA are present in 85% to 90% of patients
- Mixed cryoglobulinemia may be present in more than 50% of HCV-infected patients. However, cryoglobulinemic vasculitis develops in only a minority of these patients
- If HCV antibodies or HCV RNA is absent, look for other associated disorders, such as infective endocarditis, lymphoma, myeloproliferative disorders, autoimmune diseases, and connective tissue disorders
- For non-HCV–related cases, treat the underlying disorder
- Sample collection is important. Venous blood must be kept at 37°C for 2 hours before serum is removed and placed at 4°C for up to 4 days to allow cryoproteins to precipitate

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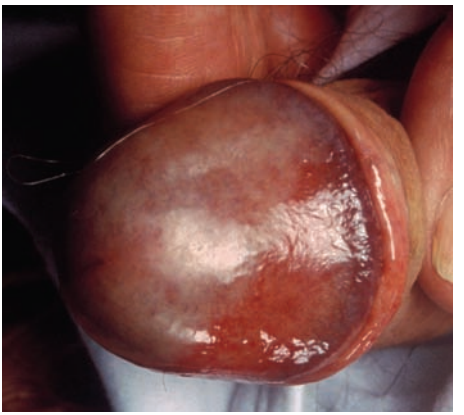

A 55-year-old man had the painless lesions shown here. Biopsy reveals carcinoma in situ.

1. What is this condition?
   - **1a.** Primary syphilis
   - **1b.** Erythroplasia of Queyrat
   - **1c.** Lichen planus
   - **1d.** Circinate balanitis of Reiter's syndrome
   - **1e.** Plasma cell balanitis of Zoon

2. What is the treatment?
   - **2a.** Triamcinolone ointment 0.1%
   - **2b.** One dose each of ceftriaxone 125 mg intramuscularly and azithromycin 1 g orally
   - **2c.** Melphalan (0.15 mg/kg per day) and prednisone (20 mg 3 times a day) for 7-day cycle, repeated every 6 weeks
   - **2d.** Benzathine penicillin G 2.4 million units intramuscularly
   - **2e.** Surgical excision

Erythroplasia of Queyrat

Answer 1: b

Answer 2: e

- Penile squamous cell carcinoma in situ was first described by Auguste Queyrat in 1911
- Lesions are slightly raised, velvety, shiny, erythematous to violaceous plaques that are dry, may be scaling, and are usually painless
- Lesions occur on the glans penis, prepuce, or urethral meatus
- Surgical removal of the involved areas with 5-mm margins virtually eliminates the risk of invasive squamous cell carcinoma. Topical laser treatments and 5-fluorouracil have been used for extensive involvement
- Without treatment, progression to squamous cell carcinoma occurs in up to 30% of cases
- Although the exact pathogenesis is unknown, the presence of human papillomavirus (HPV) DNA in several series suggests a viral oncogenesis
- Differential diagnosis for papulosquamous penile lesions includes psoriasis, lichen planus, lichen sclerosis, cicatricial pemphigoid, secondary syphilis, drug reactions, erythroplasia of Queyrat, bowenoid papulosis, circinate balanitis of Reiter's syndrome, and plasma cell balanitis of Zoon

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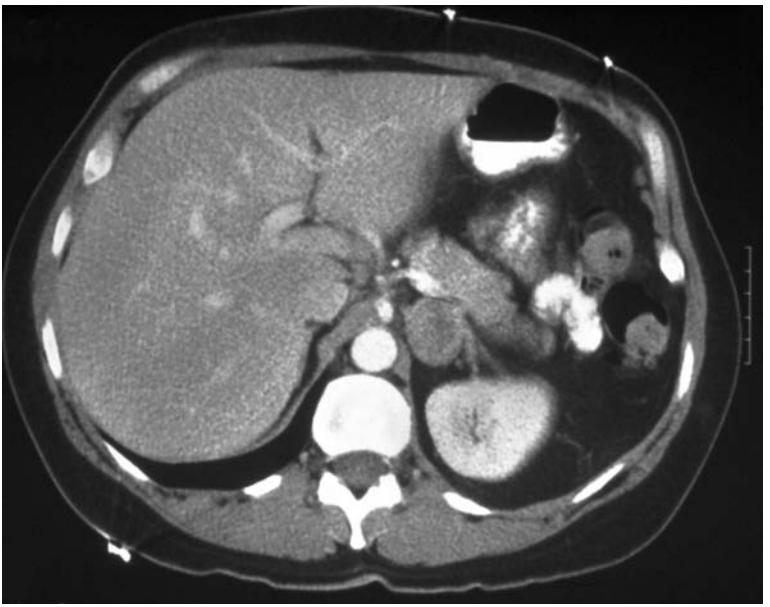


A 43-year-old woman with a 3-year history of poorly controlled hypertension, headaches, sweats, and palpitations is admitted to the hospital with acute myocardial infarction and congestive heart failure. Echocardiography shows global hypokinesis and a left ventricular ejection fraction of 10%. Coronary angiography is normal. Finding on computed tomography of the abdomen is shown here. What is the diagnosis?

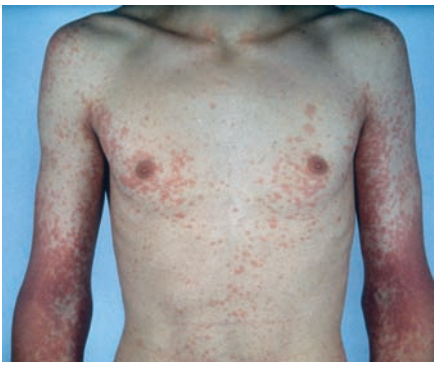
a. Carcinoid of the appendix
b. Pheochromocytoma
c. Hypernephroma
d. Renal artery stenosis
e. Aortic dissection

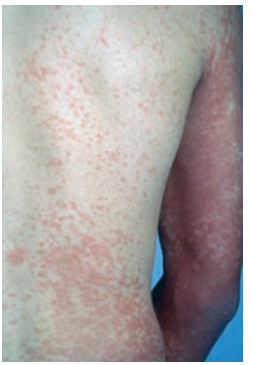
Pheochromocytoma

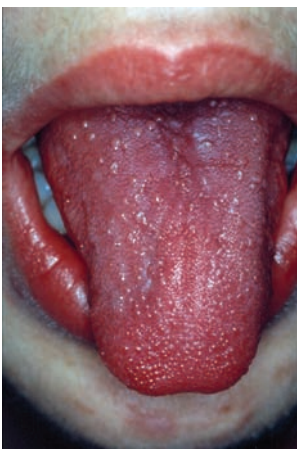
Answer: b

- The most common symptoms of pheochromocytoma are headaches, sweats, and palpitations
- Sustained hypertension is present in 50% of patients, and paroxysmal hypertension in 45%
- Other symptoms may include anxiety, pallor, heat intolerance, and weight loss
- The best screening test for pheochromocytoma is the 24-hour urine collection for metanephrines
- 80% to 90% of pheochromocytomas are in the adrenal medulla; 10% are malignant; 10% are familial; 10% are multiple or bilateral; 10% occur in children
- Diseases associated with pheochromocytoma include multiple endocrine neoplasia types IIA and IIB, von Hippel-Lindau disease, and neurofibromatosis
- This patient had catecholamine-induced cardiomyopathy, a complication of pheochromocytoma, which resolved after resection of the tumor
- Computed tomography of the abdomen is the usual imaging test to localize the tumor
- Treatment is surgical with preoperative α-adrenergic blockade before β-adrenergic blockade (to avoid unopposed α-activity with increased hypertension)

Reference
Habermann TM. *Mayo Clinic Internal Medicine Board Review 2004-2005*. Philadelphia: Lippincott Williams & Wilkins; 2004:223-224, 530-532.







A 16-year-old high school boy has a sore throat and the rash shown here, which started on the trunk and spread to the arms and legs but not the palms and soles. The appearance of his tongue is also shown. What is the most likely diagnosis?

a. Measles
b. Kawasaki disease
c. Toxic shock syndrome
d. Scarlet fever
e. Ehrlichiosis

Answer: d

- The causative agent is group A streptococcus (*Streptococcus pyogenes*)
- Strawberry tongue is caused by enlarged papillae
- The characteristic "sandpaper" rash spreads from trunk to extremities, beginning 1 to 2 days after initial fever, sore throat, and vomiting. Circumoral pallor also is thought to be characteristic
- After about a week, the rash desquamates, as do the palms and soles
- Hyperpigmentation in the bend of the elbow (Pastia's sign) is thought by some to be pathognomonic of scarlet fever
- Differential diagnosis includes measles, toxic shock syndrome, staphylococcal scalded skin syndrome, erythema infectiosum (fifth disease), Kawasaki disease, and drug reactions
- Before the advent of antibiotics and antipyretics, severe cases resulted in extremely high fevers (107°-108°F), painful lymphadenopathy, delerium, convulsions, and death
- Scarlet fever is much less common now than in the past. It is estimated that less than 5% of streptococcal infections in the United States present as scarlet fever
- Scarlet fever predisposes to rheumatic fever, the incidence of which also has declined with the advent of effective treatment
- Penicillin remains the treatment of choice

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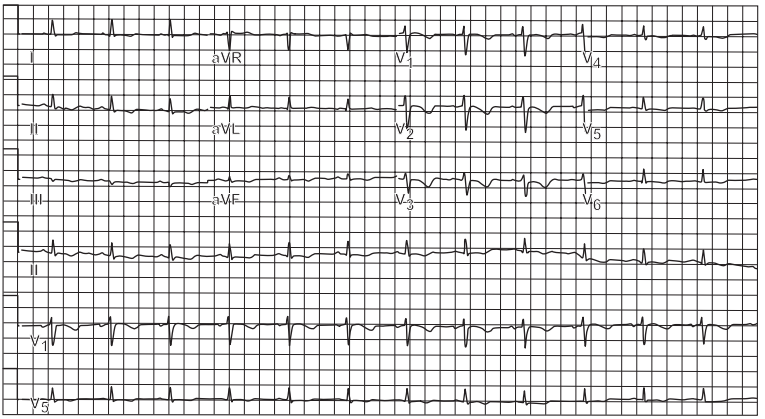

A 44-year-old woman presents with a 6-week history of exertional dyspnea, abdominal distention, and lower extremity swelling. Physical examination reveals bilateral lung rales, increased jugular venous pressure, ascites, and lower extremity edema. Electrocardiography shows low-voltage QRS complexes, yet echocardiography shows thick left ventricular walls that have a "granular" appearance. Troponin-T value has been persistently mildly increased. What is the median survival of patients with this condition?

a. 1 month
b. 7 months
c. 1 year
d. 3 years
e. 7 years

Primary Systemic Amyloidosis

Answer: b

- 90% of patients with primary systemic amyloidosis have cardiac dysfunction
- Other systems typically involved include the following:

  Liver
  Kidney
  Gastrointestinal tract
  Peripheral nerves

- Other clinical findings may include the following:

Macroglossia	Vocal hoarseness
Intestinal dysmotility	Carpal tunnel syndrome
Hepatomegaly	Fatigue
Peripheral neuropathy	Weight loss
Proteinuria	Orthostatic hypotension

- Heart failure is the most common cause of death of patients with cardiac amyloidosis
- The median survival of patients with primary systemic amyloidosis and overt congestive heart failure is 7 months
- Patients with cardiac amyloidosis have low-voltage QRS complexes on electrocardiography yet thick walls on echocardiography
- Increases of cardiac troponins (troponin T and troponin I) are associated with decreased survival in primary systemic amyloidosis

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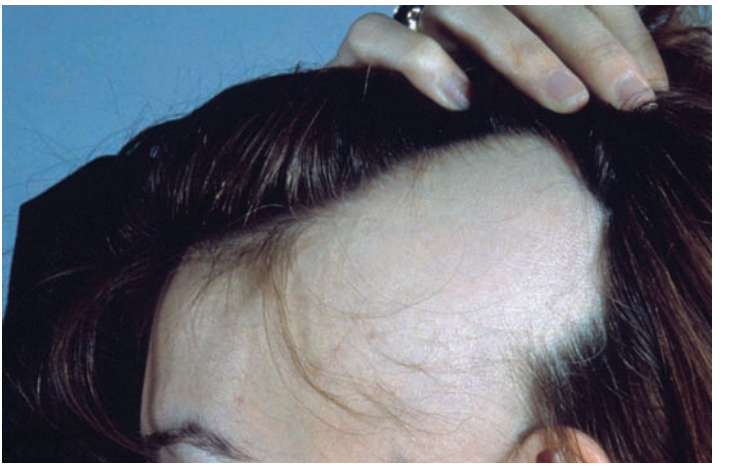

A 22-year-old woman with hypothyroidism has the patch of hair loss shown here. Biopsy of the affected area shows a T-cell–predominant infiltrate. What is the most likely cause?

a. Androgenic alopecia
b. Telogen effluvium
c. Traumatic alopecia
d. Alopecia areata
e. Tinea capitis

Alopecia Areata

Answer: d

- Almost 2% of the U.S. population is affected by alopecia areata
- The condition is characterized by well-circumscribed, circular areas of patchy hair loss
- Severe variants cause loss of all hair on the scalp (alopecia totalis) and body (alopecia universalis)
- Activated CD4 and CD8 T-lymphocyte infiltrates surround the follicles on histologic examinations
- The condition may be associated with hypothyroidism, pernicious anemia, hyperthyroidism, vitiligo, atopy, and Down syndrome
- Hair loss is often chronic and recurrent
- Treatment with intralesional corticosteroids, topical anthralin, or minoxidil may be attempted. Phototherapy, topical sensitizers, cyclosporin A, thymopentin, inosiplex, and topical nitrogen mustard have all been attempted, with variable success
- Hair follicles are not destroyed, so regrowth is always possible
- If healing occurs, there will not be scarring
- Although this condition is otherwise benign, patients may need psychological counseling because it can be emotionally devastating

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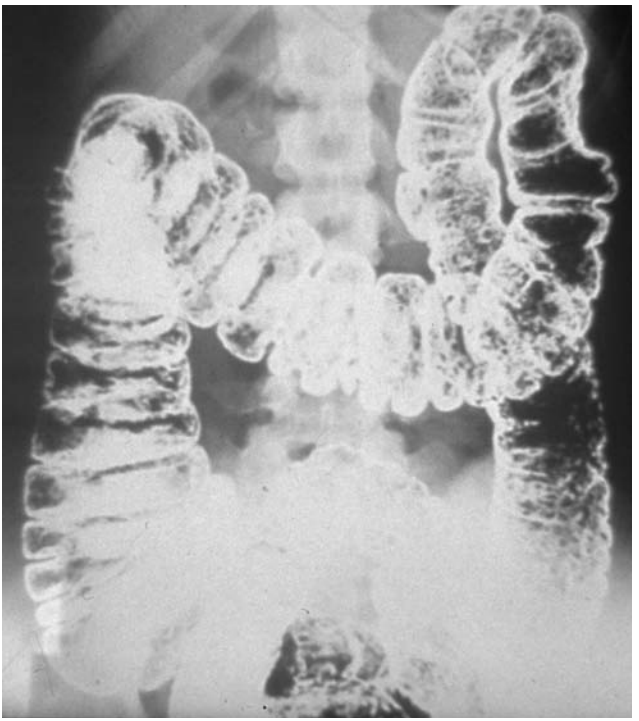

A 40-year-old woman presents for a physical examination. She has undergone excision of several bony prominences from her mandible. An ophthalmologist has been following pigmented lesions of the right eye. Her father died of colon cancer at age 42 years. Her colon radiograph is shown. What is the risk of colon cancer by age 40 years for patients with this syndrome?

a. Same as that in the general population
b. 20%
c. 40%
d. 60%
e. 80%

Gardner's Syndrome

Answer: d

- Gardner's syndrome is an autosomal dominant form of hereditary colon polyposis
- The syndrome is distinguished by extraintestinal lesions:

  Desmoid tumors
  Sebaceous cysts
  Lipomas
  Fibromas
  Supernumerary teeth
  Osteomas of the skull
  Scoliosis
  Retinal pigmented lesions
  Malignancies of nonintestinal origin (e.g., thyroid, adrenal, genitourinary)

- Colon cancer develops in 60% of patients by age 40 years and 95% of patients overall; hence, prophylactic total colectomy is recommended

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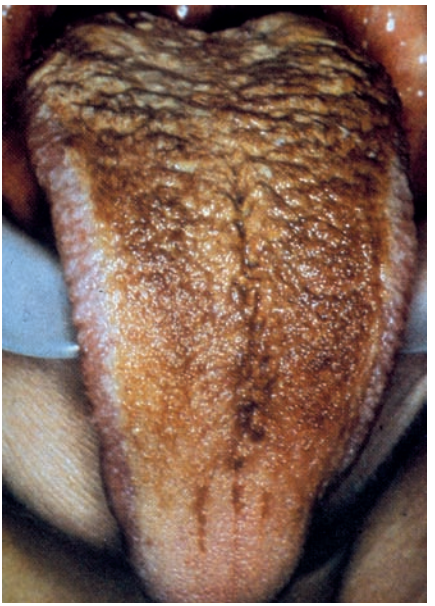

A 63-year-old male smoker presents with concern about the appearance of his tongue. He reports no other symptoms, but he was recently treated for bronchitis with amoxicillin-clavulanate. What is the diagnosis?

a. Geographic tongue
b. Black hairy tongue
c. Glossitis
d. Amyloidosis
e. Squamous cell carcinoma of the tongue

Black Hairy Tongue (Lingua Nigra)

Answer: b

- Black hairy tongue is due to hyperplasia and elongation of the filiform papillae and overgrowth of chromogenic bacteria and fungi
- Risk factors for black hairy tongue include the following:

  Poor oral hygiene
  Smoking
  Antibiotic use
  Limitation of tongue movements (such as caused by stroke)
- Treatment consists of frequent (twice daily) brushing of the tongue
- Black hairy tongue is a benign condition

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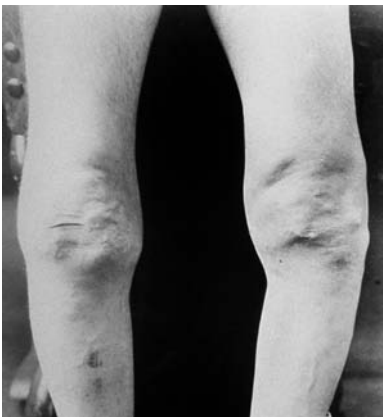
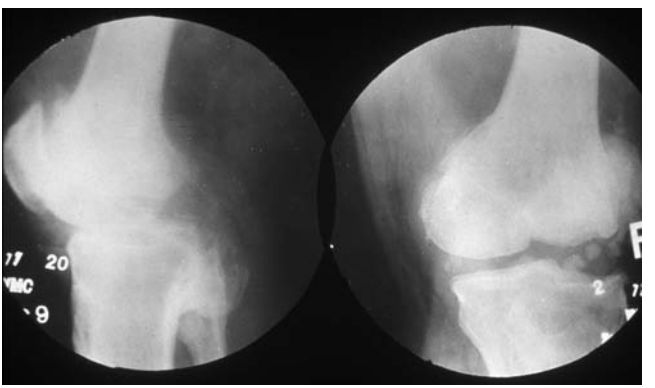




A 45-year-old man presents with an acutely swollen knee. He reports no pain. His past medical history is notable for type 1 diabetes mellitus, which has been poorly controlled. What is the diagnosis?

a. Septic arthritis
b. Tibial plateau fracture
c. Torn anterior cruciate ligament
d. Lyme arthritis
e. Charcot's arthropathy

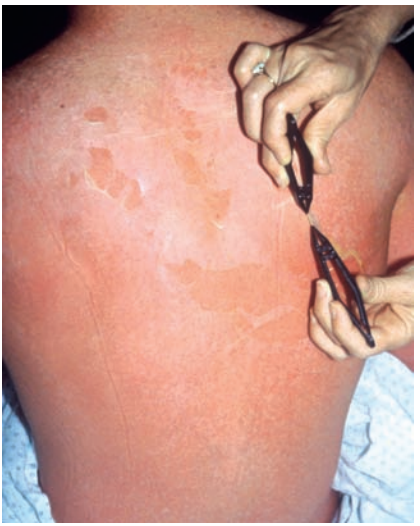
Charcot's Joint (Neuroarthropathy)

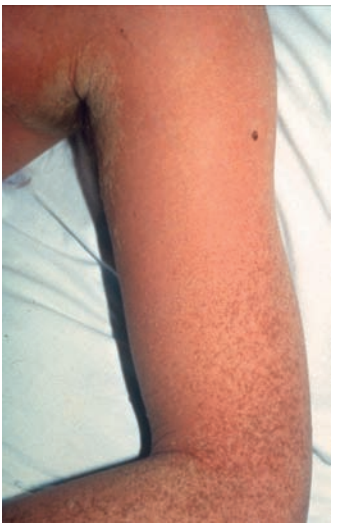
Answer: e

- Charcot's arthropathy is also known as neuropathic arthropathy
- Patients with Charcot's arthropathy have impaired pain and position sense; neuropathy and repeated injury lead to the arthropathy
- The most common cause of Charcot's arthropathy is diabetes mellitus
- Other causes include the following:

  Tabes dorsalis (classically involving the spine and hip)
  Leprosy
  Amyloidosis
  Syringomyelia (affects the shoulder)
- Charcot's arthropathy is a rapidly destructive process
- Radiographs reveal disorganized joint architecture
- Patients with diabetes who have Charcot's arthropathy have had diabetes for more than 16 years on average
- Although the patient described here had severe knee involvement, the most commonly affected area in patients with diabetes is the foot
- The suspicion for osteomyelitis should be high in patients with Charcot's arthropathy

Reference
Habermann TM. *Mayo Clinic Internal Medicine Board Review 2004-2005*. Philadelphia: Lippincott Williams & Wilkins; 2004:956.





A rash developed in a 14-year-old boy, first around his eyes and then on his trunk and extremities. The involved skin is tender.

1. What is the likely cause of this condition?

1a. Reactivation of varicella-zoster virus
1b. Henoch-Schönlein purpura
1c. Cutaneous vasculitis
1d. Staphylococcal exfoliative toxins
1e. Disseminated intravascular coagulation due to rickettsial infection

2. Which physical sign is shown on the patient's back?

2a. Mueller's sign
2b. Nikolsky's sign
2c. Osler's sign
2d. Litten's sign
2e. Schultze's sign
2f. Plummer's sign

Staphylococcal Scalded Skin Syndrome

Answer 1: d

Answer 2: b

- Staphylococcal scalded skin syndrome is caused by exfoliative toxins produced by strains of *Staphylococcus aureus*
- The syndrome is characterized by sudden onset of reddened skin that later loosens in large areas
- The syndrome begins periorbitally and spreads to the trunk and centrifugally to the limbs
- The involved skin is often tender
- The syndrome is more common in children than adults
- Risk factors in adults include immunocompromise, malignancy, rheumatic heart disease, intravenous drug use, renal disease, and diabetes
- Therapy includes fluid and electrolyte replacement, local care of exfoliated areas, and avoidance of secondary infections. Antistaphylococcal antibiotics are used, but their usefulness is uncertain given that preformed toxin is the cause
- The syndrome usually resolves within 10 days. Mortality increases with the age of the patient (3% in children, >50% in adults)
- Nikolsky's sign is bullae formation or sloughing caused by gentle lateral stroking

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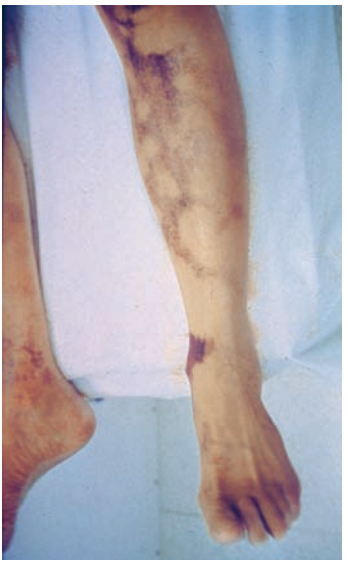

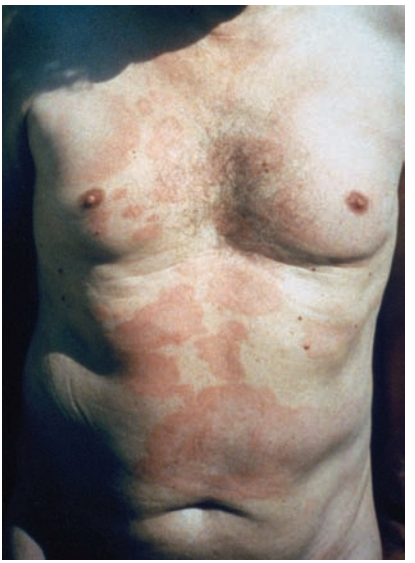

A 47-year-old missionary has lived in India for 15 years. The lesions shown here are painless and numb and developed over about 6 months' time. Which of the following is part of a first-line treatment regimen for this disease?

a. Glucose control
b. Dapsone
c. Gabapentin
d. Topical corticosteroids
e. Itraconazole

Leprosy (Hansen's Disease)

Answer: b

- The disease is caused by *Mycobacterium leprae*, which has never been cultured but can grow in some animal models
- Armadillos in the southwestern United States can be a reservoir, but most transmission is from human to human (close contacts)
- Most cases in the United States occur in immigrants
- *M. leprae* grows best at temperatures less than 30°C, including skin, peripheral nerves, anterior parts of eyes, upper respiratory structures, testes, hands, and feet
- *Tuberculoid leprosy* is marked early on by hypopigmented, anesthetic, sharply demarcated macules. Nerve involvement may lead to muscle atrophy (especially in the hands), ulcerations, resorption of digits, exposure keratitis, and corneal ulceration (including blindness)
- *Lepromatous leprosy*, as in the case described here, has extensive cutaneous involvement with diffuse, granulomatous macules, papules, or nodules. Common sites are face, ears, elbows, buttocks, and knees. There can be a loss of lateral aspect of the eyebrows and later development of thickened, corrugated facies (leonine facies). Clawhand, footdrop, claw toes, and plantar insensitivity are caused by peripheral nerve involvement (ulnar or median, common peroneal, and posterior tibial, respectively). Nasal septal perforation and hoarseness are advanced complications
- *Borderline leprosy* has a spectrum between tuberculoid and lepromatous leprosy
- First-line treatment includes dapsone with rifampin. Depending on the type of disease, clofazimine may be added. Treatment durations are long, 1 to 2 years

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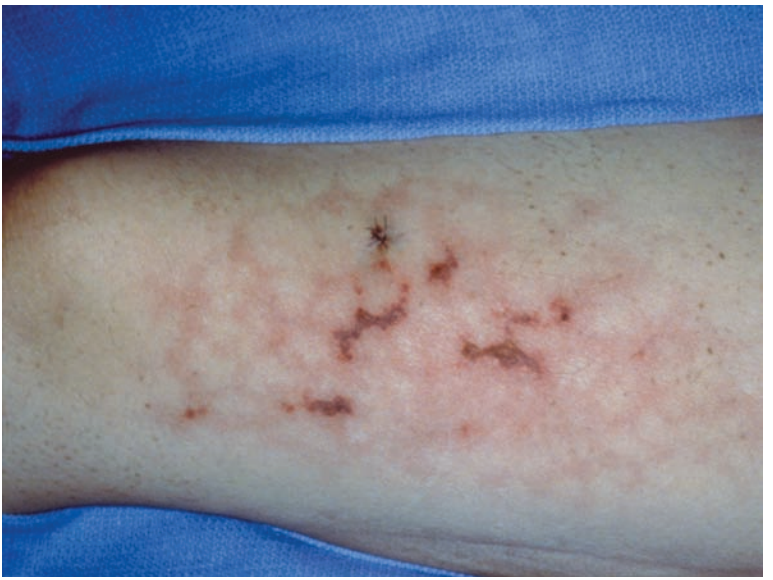

The cutaneous finding shown is most commonly associated with which one of the following medications?

a. Ciprofloxacin
b. Amantadine
c. Hydrocortisone
d. Verapamil
e. Ibuprofen

Livedo Reticularis

Answer: b

- Livedo reticularis manifests in a netlike, mottled pattern with a bluish discoloration. It may be deep blue in cold environments
- It is more common on the lower extremities
- Livedo reticularis may be broadly categorized as follows:

  Physiologic
    Also known as cutis marmorata
    Occurs in children and women
  Primary (idiopathic)
  Secondary
    Intravascular obstruction
      Cholesterol embolization
      Anticardiolipin antibody syndrome
      Cryoglobulinemia
      Polycythemia vera
    Arteritis
      Polyarteritis nodosa
      Systemic lupus erythematosus
    Medications
      Amantadine
      Quinidine
      Quinine

- Almost half of all cases involving cholesterol embolism (such as after cardiac catheterization) have cutaneous features of livedo reticularis
- Treatment includes management of the underlying cause and avoidance of cold exposure

References

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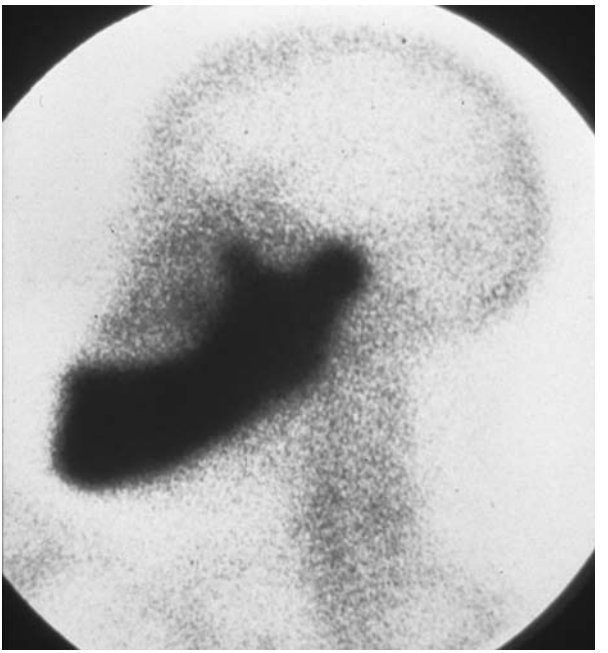

A patient presents with tibial bowing and complains of hearing loss. What is the most common complication of the disease affecting this patient?

a. Pulmonary fat embolism
b. Congestive heart failure
c. Hypercalcemia
d. Traumatic bone fractures
e. Aortic dissection

Paget's Disease

Answer: d

- Most elderly patients present with an isolated increase in the alkaline phosphatase value. Paget's disease will develop in up to 4% of Americans by the age of 60 years
- Paget's disease is characterized by disorganized bone remodeling and bone pain
- Bones involved by Paget's disease typically have three stages of evolution: lytic phase, transition to a mosaic pattern, and sclerotic phase
- The most common symptom is focal bone pain. The most common complications are traumatic and pathologic bone fractures
- In 0.7% to 5% of cases, osteosarcoma may develop in pagetic bone. In some series, almost half of patients with osteosarcoma who are older than 50 years have Paget's disease as a predisposing condition
- Congestive heart failure may be precipitated when more than 20% of the skeleton is involved
- First-line therapy is use of antiresorptive medication. Intravenous bisphosphonate is preferred because of its efficacy, minimal side effect profile, and logistics of dosing
- Hearing loss may be due to involvement of inner ear ossicles or to bony impingement of cranial nerve VIII in the auditory foramen

References

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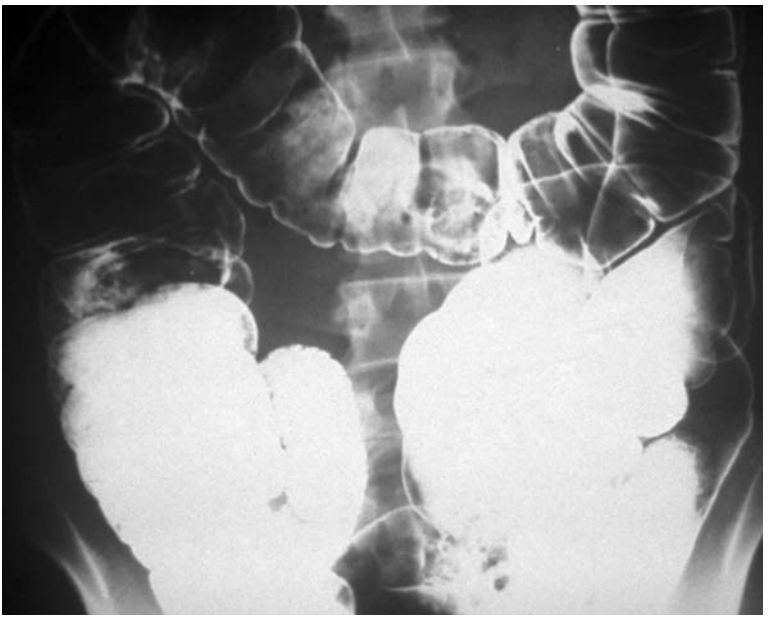

An 18-year-old man is distressed about the intermittent abdominal pain he has been experiencing for the past several months. Results of barium enema examination are shown here. This syndrome of intestinal polyposis is also associated with which one of the following?

a. Café au lait spots
b. Adenoma sebaceum
c. Macular pigmentation of oral mucosa
d. Dermatitis herpetiformis
e. Erythema marginatum

Peutz-Jeghers Syndrome

Answer: c

- Peutz-Jeghers syndrome is an autosomal dominant disorder characterized by hamartomatous polyps in the gastrointestinal tract
- Genetic penetrance in families is reported to be as high as 90%
- Polyps are usually large and pedunculated
- Frequency of involvement is as follows:

Small bowel	70% to 90%
Large bowel	50%
Stomach	25%

- Associated cutaneous lesions include macular pigmentation of the lips and periungual skin. This freckling may appear during the first decade of life and gradually fade away by the third decade
- Recurrent abdominal pain may be due to partial obstruction from polyps
- Patients also may present with catastrophic abdominal emergencies such as volvulus and intussusception
- Less commonly, polyps may be found at extraintestinal sites such as the nose, uterus, respiratory tract, urinary tract, and gallbladder

References
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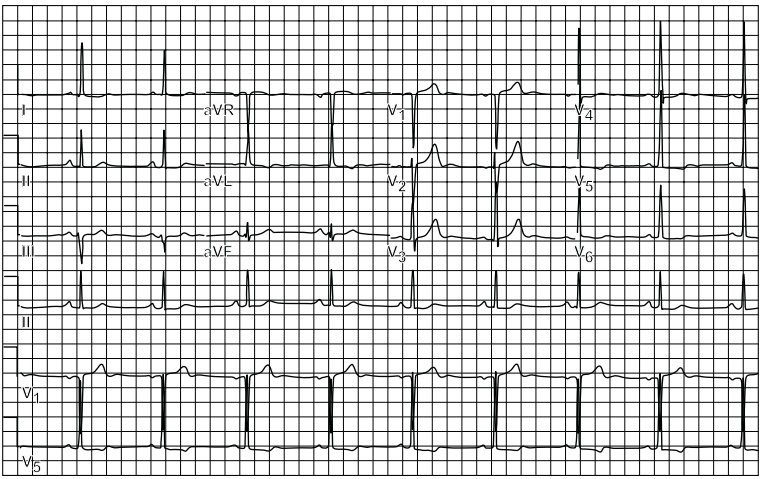
Tomlinson IP, Houlston RS. Peutz-Jeghers syndrome. *J Med Genet*. 1997;34:1007-1011.



A 67-year-old woman with a long-standing history of essential hypertension presents with the electrocardiogram shown here. Which one of the following electrocardiographic criteria may assist you in deducing her resultant cardiac anatomy?

a. $S(V_1) + R(V_5) > 3.5$ mV
b. $S(V_4) + R(V_2) > 3.5$ mV
c. $R(V_1) + S(V_5) > 3.5$ mV
d. $R(V_5) + S(V_1) > 3.5$ mV
e. S(III) + R(aVF) > 3.5 mV

Left Ventricular Hypertrophy

Answer: a

- Left ventricular hypertrophy (LVH) reflects an increase in cell mass of preexisting cardiac myocytes. Hyperplasia does not occur because myocytes are terminally differentiated after birth
- Cell hypertrophy occurs as a physiologic response to offset an increase in pressure load on the myocardium (LaPlace's law), as occurs in aortic stenosis or long-standing systemic hypertension
- Application of standardized LVH criteria to the Framingham study cohort yielded a prevalence of LVH of 16% in women and 19% in men
- Echocardiography is considerably more sensitive for detecting LVH than electrocardiography. However, infiltrative processes such as cardiac sarcoidosis may mimic LVH on echocardiography
- In cases of hypertrophic cardiomyopathy, the electrocardiogram typically shows LVH
- Prognostically, echocardiographically detected LVH is an independent risk factor for increased cardiovascular morbidity and mortality
- Substantial regression of LVH has been reported in studies of aortic valve replacement. Pharmacologic trials for hypertension have not yet matched these levels of regression, although this may be possible with better and more consistent blood pressure control
- Numerous electrocardiographic criteria exist for LVH. One of the most commonly used is the Sokolow-Lyon index: $S(V_1) + (R[V_5]$ or $R[V_6]) > 3.5$ mV or $R(aVL) > 1.1$ mV

References

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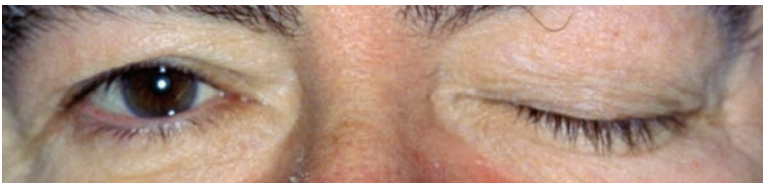
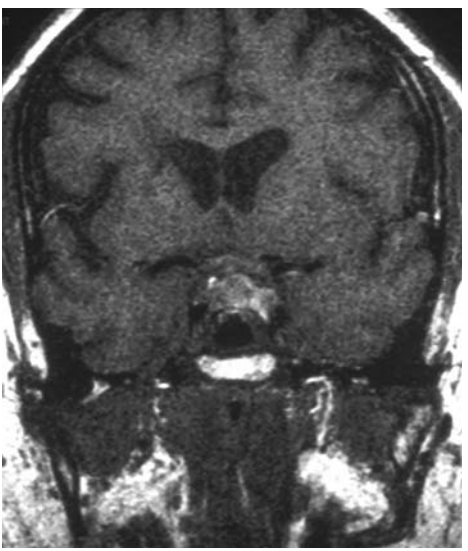
Lorell BH, Carabello BA. Left ventricular hypertrophy: pathogenesis, detection, and prognosis. *Circulation*. 2000;102:470-479.





A 69-year-old man presents with an acute severe headache. Physical examination reveals an isolated left third cranial nerve palsy. Laboratory studies show hyponatremia and abnormally low levels of luteinizing hormone, follicle-stimulating hormone, thyroid-stimulating hormone, adrenocorticotropic hormone, testosterone, thyroxine, and cortisol. What is the diagnosis?

a. Hypothalamic hemorrhage
b. Neurosarcoidosis
c. Pituitary apoplexy
d. Posterior pharyngeal abscess
e. Lymphocytic hypophysitis

Pituitary Apoplexy

Answer: c

- Pituitary apoplexy refers to hemorrhagic infarction of the pituitary gland
- Predisposing conditions include the following:

  Pituitary tumor
  Irradiated pituitary tumor
  Pregnancy
  Anticoagulation therapy
  Increased intracranial pressure
  Vascular disease
  Vasculitis

- Patients may present with the following:

  Headache
  Ocular palsies
  Visual field defects
  Nausea
  Altered mental state
  Acute adrenocortical crisis

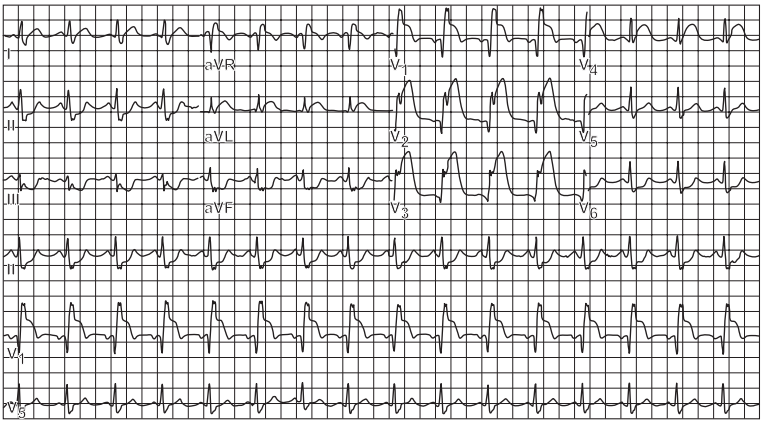
- Infarction of a pituitary adenoma may result in the following:

  Subarachnoid hemorrhage
  Compression of the optic chiasm
  Occlusion of the cavernous sinus

- The greatest immediate danger to patients with pituitary apoplexy is cortisol deficiency
- Long-term treatment often begins with neurosurgery (for decompression) and multiple hormone replacements

Reference

Habermann TM. *Mayo Clinic Internal Medicine Board Review 2004-2005*. Philadelphia: Lippincott Williams & Wilkins; 2004:204.



A 60-year-old man, a 20-pack-year smoker, arrives in the urgent care center complaining of lethargy and difficulty breathing. You note that he has a history of stable angina well managed with β-adrenergic blockers and nitrates. Emergency electrocardiography is done, and you conclude he is experiencing which one of the following?

a. Diffuse esophageal spasm
b. Acute inferior myocardial infarction
c. Acute posterior myocardial infarction
d. Acute pericarditis
e. Acute anteroseptal myocardial infarction

Answer: e

- Acute anteroseptal myocardial infarction is suggested by ST elevation (>0.1 mV) in precordial leads V_1-V_4
- Patients with anterior myocardial infarction have an increased cumulative mortality and duration of hospitalization and a more complicated follow-up course after hospitalization than patients with acute inferior myocardial infarction
- The poor prognosis associated with acute anterior compared with acute inferior myocardial infarction is independent of infarct size and type of infarction (that is, Q wave or non-Q wave)
- Poor prognosis may be attributable to the impact of anterior infarction on left ventricular ejection function and consequent dysfunction
- More aggressive management for acute anterior myocardial infarction (such as use of thrombolytics) results in better outcomes than similar strategies in patients with inferior infarctions
- The natural evolution of an anteroseptal infarction during 2 months is shown in the electrocardiograms on the following pages

References

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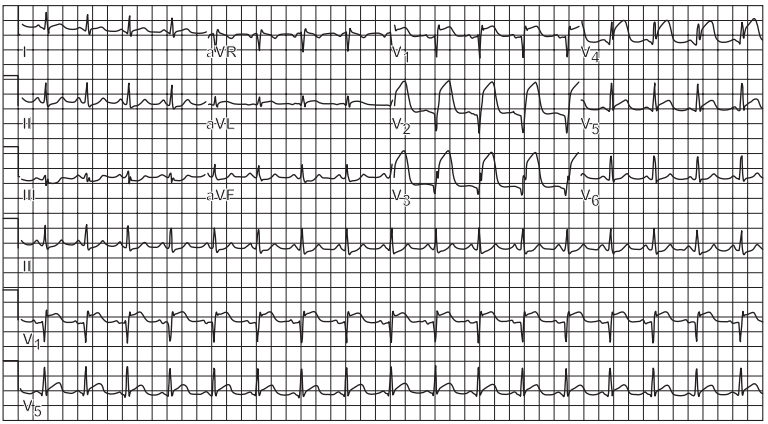
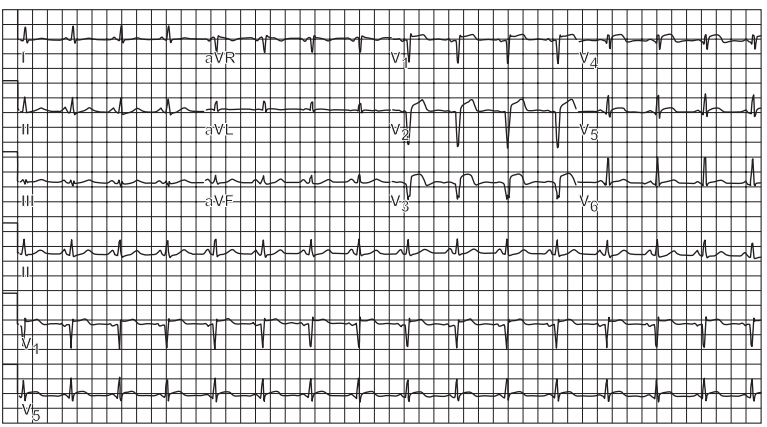
Stone PH, Raabe DS, Jaffe AS, et al. Prognostic significance of location and type of myocardial infarction: independent adverse outcome associated with anterior location. *J Am Coll Cardiol*. 1988;11:453-463.

Case 59 (continued): Evolution of Anteroseptal Myocardial Infarction

3 Hours after initial electrocardiogram

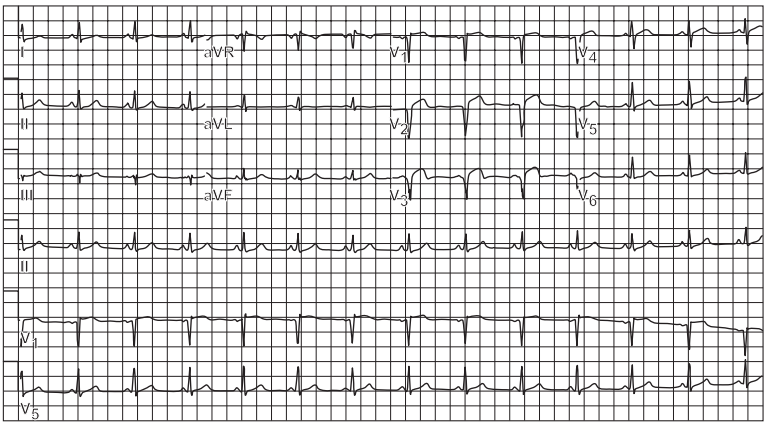


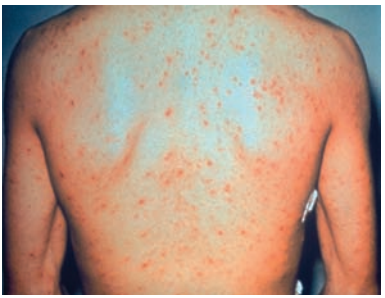
24 Hours after initial electrocardiogram

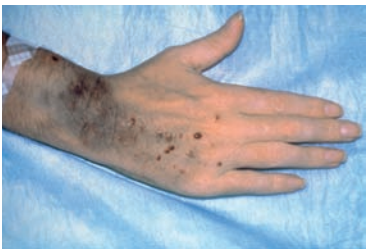


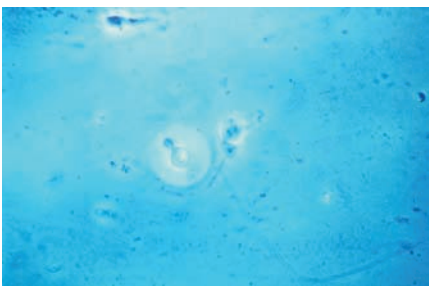
Case 59 (continued): Evolution of Anteroseptal Myocardial Infarction

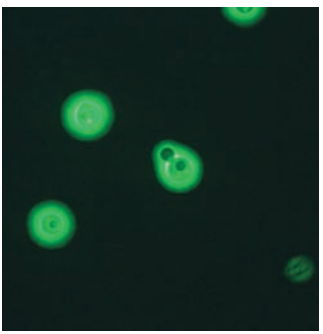
2 Months after initial electrocardiogram











The findings shown here are those for a 52-year-old man 6 months after liver transplantation. Which statement is true about this infection?

a. It develops in patients with humoral immunodeficiencies
b. It may be definitively diagnosed with serologic IgM assays
c. It may be effectively treated with itraconazole
d. It is transmitted by aerosolization
e. It is characterized by broad-based buds

Cryptococcosis

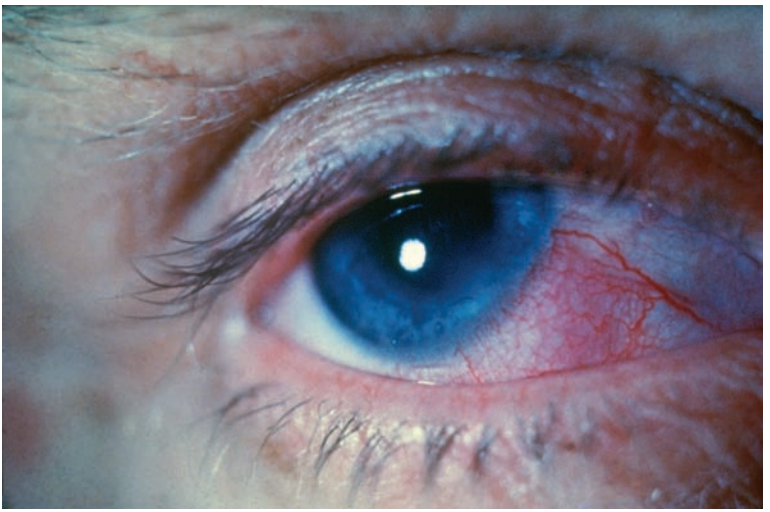
Answer: d

- *Cryptococcus neoformans* exists as a free living fungus in soil and avian excreta
- Inhalation of aerosolized organisms is the mode of transmission. Sweeping of bird cages or being in the presence of birds flapping their wings may easily predispose to this
- *Cryptococcus neoformans* is the only cryptococcal species that is pathogenic in humans
- Cryptococcosis is an opportunistic infection in patients with cell-mediated (T-cell) deficiency or dysfunction (such as patients receiving chemotherapy, with long-standing corticosteroid use, or with acquired immunodeficiency syndrome)
- Clinical manifestations include pneumonia, meningitis, and cranial nerve infiltration
- Definitive serologic test of choice is cryptococcal antigen test
- India ink stain may show yeastlike organisms with narrow-based buds
- Central nervous system (CNS) disease should be ruled out in patients with neurologic symptoms and in immunocompromised patients
- CNS disease is diagnosed with culture and cryptococcal antigen test of cerebrospinal fluid. If present, serial lumbar punctures to reduce intracerebral pressures may be needed
- Fluconazole may be used for non-CNS cryptococcosis. Amphotericin B with flucytosine is an accepted treatment for CNS disease
- Patients with acquired immunodeficiency syndrome require lifelong therapy with fluconazole after initial treatment to prevent relapses

References

Habermann TM. *Mayo Clinic Internal Medicine Board Review 2004-2005*. Philadelphia: Lippincott Williams & Wilkins; 2004:570.

Nosanchuk JD, Shoham S, Fries BC, et al. Evidence of zoonotic transmission of *Cryptococcus neoformans* from a pet cockatoo to an immunocompromised patient. *Ann Intern Med*. 2000;132:205-208.



A 54-year-old man with no significant past medical history except for hypertension (controlled with medication) presents with acute mild pain and redness of the eye. The redness involves only a portion of the eye. He otherwise feels well and reports no other symptoms. How should this condition be treated?

a. Observation only
b. Topical corticosteroids
c. Topical antibiotics
d. Topical β-adrenergic receptor antagonists
e. Corneal transplant

Episcleritis

Answer: a

- First described by Read in 1702
- Episcleritis is due to acute inflammation immediately underlying the conjunctiva
- Episcleritis is self-limited, although topical nonsteroidal anti-inflammatory drugs may hasten resolution
- Most patients with episcleritis do not have an associated underlying systemic disease
- Many patients have an associated ocular disorder (such as ocular rosacea, keratoconjunctivitis sicca, atopic keratoconjunctivitis)
- Unlike episcleritis, scleritis is very painful, destructive, and more often associated with underlying systemic disease, particularly the following:

  Rheumatoid arthritis
  Wegener's granulomatosis
  Tuberculosis
  Syphilis
  Herpes zoster

- If in doubt about the differentiation of episcleritis and scleritis, ophthalmologic consultation is recommended

References

Akpek EK, Uy HS, Christen W, et al. Severity of episcleritis and systemic disease association. *Ophthalmology*. 1999;106:729-731.

Pavesio CE, Meier FM. Systemic disorders associated with episcleritis and scleritis. *Curr Opin Ophthalmol*. 2001;12:471-478.

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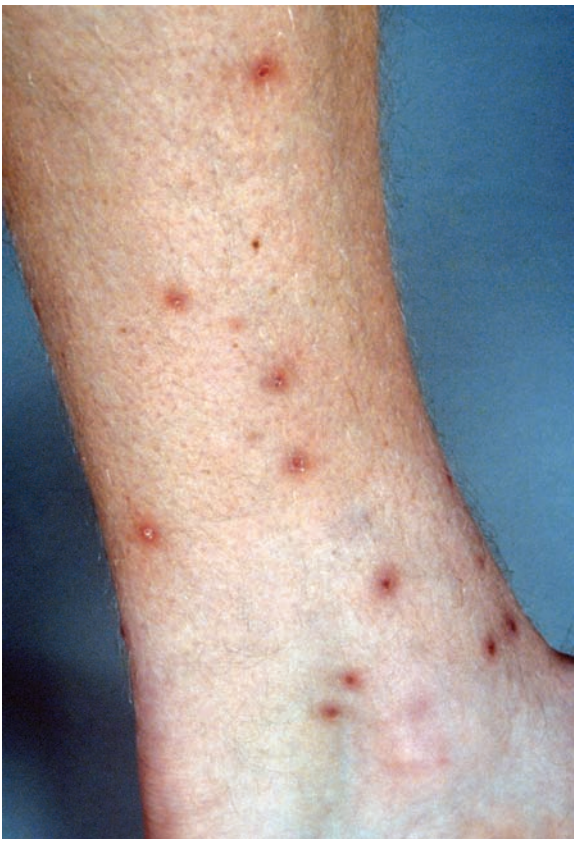


The itchy papules shown here developed in a 27-year-old man after picking blackberries on a hot summer mid-afternoon. What probably caused them?

a. Fleas
b. Harvest mites
c. Ticks
d. Bees
e. Mosquitoes

Harvest Mite (Chigger) Bites

Answer: b

- Lesions are caused by bites of the larvae of trombiculid (harvest) mites, *Eutrombicula alfreddugèsi* and others
- Known by many names:

Australia	scrub itch, ti tree itch
Ireland	orange tawny
Japan	kedani, akamushi
Mexico	Tlalzuatl, coloradillo
United Kingdom	harvest mite, harvest bug, red bug
United States	mower's mite, chigger, red bug

- Warm climates favored—Southern and midwestern United States during spring, summer, and fall. Normally feed on small mammals. Humans are incidental hosts. Mites wait on vegetation and attach themselves to hosts as they travel through the vegetation
- Similar mites are vectors of scrub typhus in tropical and subtropical Asia
- Bites are often located on legs and around the beltline, with mites stopping at areas of clothing constriction. Mites pierce the skin of hosts and place the stylostome (feeding tube) into the upper dermis for a blood, tissue, and lymph meal. The mite then falls off
- Initial contact results in little, if any, reaction. Subsequent exposure results in sensitivity and a severely pruritic reaction. The mite is gone by the time the reaction occurs. Small papules or wheals progress to pustules. Excoriations are the rule. Occasionally vesicles and bullae occur
- Treatment is with antipruritic therapies, topical corticosteroids, and warm baking soda baths
- Prevention with insect repellents along exposed skin areas is the best remedy

References

Millikan LE. Mite infestations other than scabies. *Semin Dermatol*. 1993;12:46-52.
Stawiski MA. Insect bites and stings. *Emerg Med Clin North Am*. 1985;3:785-808.

This pleural fluid was drained from a patient after a motor vehicle accident. Similar pleural fluid may be associated with all of the following *except*:

a. Pleural fluid triglyceride level of more than 100 mg/dL
b. Moderate congestive heart failure
c. Lymphangiomyomatosis
d. Thoracic duct obstruction
e. Normal serum triglyceride level

Chylous Effusion

Answer: b

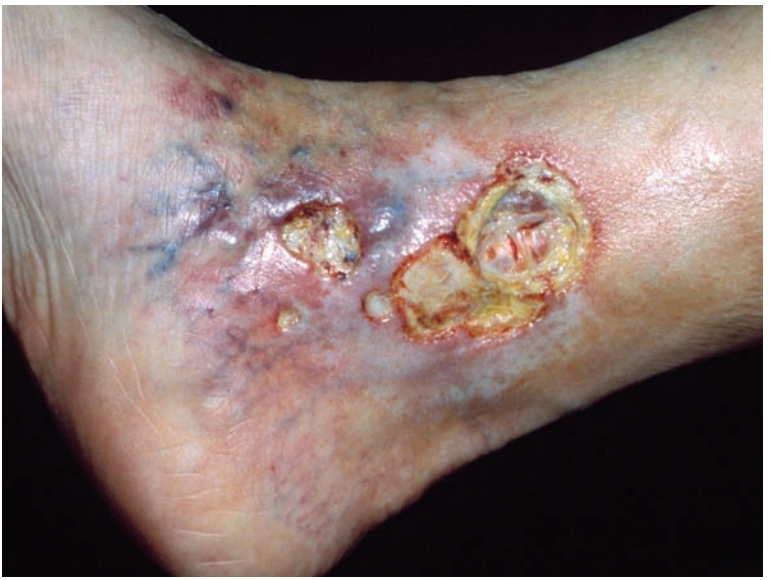
- Almost 70% of dietary fat absorbed through the lacteal system is circulated through the lymphatic channels
- Chyle can be differentiated from other fluids by its high pleural triglyceride level (more than 100 mg/dL) in the setting of normal serum triglyceride levels
- High pleural fluid cholesterol level is not by itself indicative of a true chylous effusion
- Numerous causes for chylous effusions may be broadly categorized as:

  Congenital (e.g., thoracic duct atresia)
  Traumatic (e.g., central line placement associated with subclavian vein thrombosis, after thoracic or esophageal surgery, penetrating injury)
  Intrinsic (e.g., neoplasm, pulmonary lymphangiomatosis)
  Extrinsic (e.g., infections, such as tuberculosis)

- Lymphangiography may elucidate the thoracic duct anatomy and localize the site and extent of lymphatic obstruction
- Conservative management includes pleural fluid drainage and restriction of fat intake (medium-chain triglycerides are acceptable)
- Chylous effusions resulting from surgical complications are generally best managed surgically

References

Habermann TM. *Mayo Clinic Internal Medicine Board Review 2004-2005*. Philadelphia: Lippincott Williams & Wilkins; 2004:883.
Merrigan BA, Winter DC, O'Sullivan GC. Chylothorax. *Br J Surg*. 1997;84:15-20.



The lesion shown here developed in a 57-year-old woman after she bumped her leg against a coffee table. She had had chronic discoloration in the area for many years before the ulcer formed. What type of ulcer is this?

a. Ischemic ulcer
b. Arteriolar (hypertensive) ulcer
c. Neurotrophic ulcer
d. Venous insufficiency ulcer
e. Pressure ulcer

Venous Insufficiency Ulcer

Answer: d

- Deep venous insufficiency (DVI) is one of the most common causes of leg ulcer in the United States, affecting up to 1% of the population
- The medial malleolar region is a common site of ulceration, which may be preceded by years of edema, aching pain, itching, brawny reddish tan discoloration, and lipodermatosclerosis
- Ulcers are often painful, but usually less so than ischemic ulcers
- Deep venous thrombosis is commonly associated with DVI, with edema occurring in about 67% of patients, skin pigmentation in 33%, and ulceration in 4%
- Compression stockings and elevation are usually effective treatments of chronic DVI
- Although general cleansing and wound care are warranted, topical antibiotics and antiseptics have not been shown to be helpful for healing venous insufficiency ulcers
- Compliance with conservative care can result in 5-year ulcer-free rates of 70%
- Surgical therapy is generally not needed, but for selected patients surgical ablation of superficial and perforating veins has had good results. Deep venous reconstruction is unusual but has been useful in some refractory cases

References

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Angle N, Bergan JJ. Chronic venous ulcer. *BMJ*. 1997;314:1019-1023.

Goodfield M. Optimal management of chronic leg ulcers in the elderly. *Drugs Aging*. 1997;10:341-348.

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Mayer W, Partsch H. Classification of chronic venous insufficiency. *Curr Probl Dermatol*. 1999;27:81-88.

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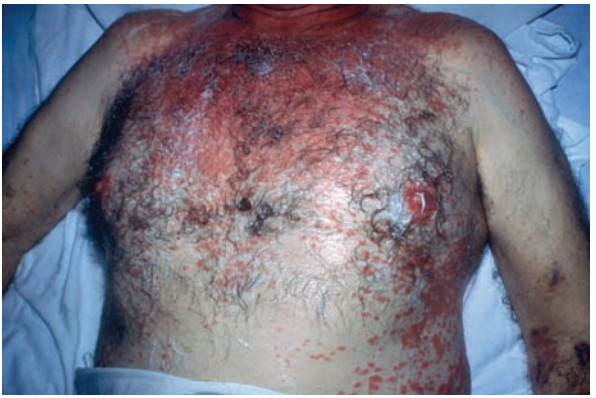

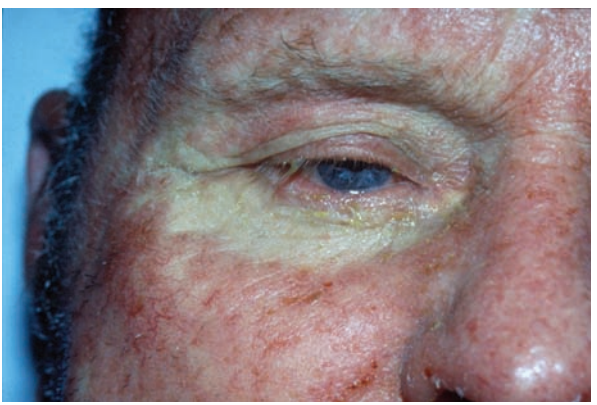

The blistering rash shown here and erosions of mucous membranes developed in a 54-year-old man 1 day after he started therapy with trimethoprim-sulfamethoxazole for an upper respiratory infection. What is this syndrome called?

a. Toxic shock syndrome
b. Stevens-Johnson syndrome
c. Red man syndrome
d. Scalded skin syndrome
e. Jarisch-Herxheimer reaction

Stevens-Johnson Syndrome (SJS) and Toxic Epidermal Necrolysis (TEN)

Answer: b

- Often starts as a morbilliform rash and fever. Risk factors for more severe reactions include blistering mucous membrane involvement, epidermal detachment, and increased age
- In SJS, less than 10% of total body surface area is affected
- In overlap, or transitional, SJS-TEN, 10% to 30% of total body surface area is affected
- In TEN, more than 30% of total body surface area is affected
- Amount of involvement has prognostic implications. TEN has the worst prognosis—30% of patients die of infection or pulmonary complications
- Any drug may cause the reaction, but most cases are associated with sulfonamides, penicillins, nonsteroidal anti-inflammatory drugs, and allopurinol
- Withdrawing the offending drug can reduce the risk of death by 30% per day
- Offending drugs with longer half-lives are more likely to be associated with poorer outcomes
- Treatment involves exquisite skin and eye care, fluids, nutrition, and aggressive treatment of superinfections. Care in a burn unit is appropriate. Ophthalmology and dermatology consultations are warranted
- Use of corticosteroids and other immunosuppressants, plasmapheresis, and intravenous immunoglobulin is controversial and not standard care

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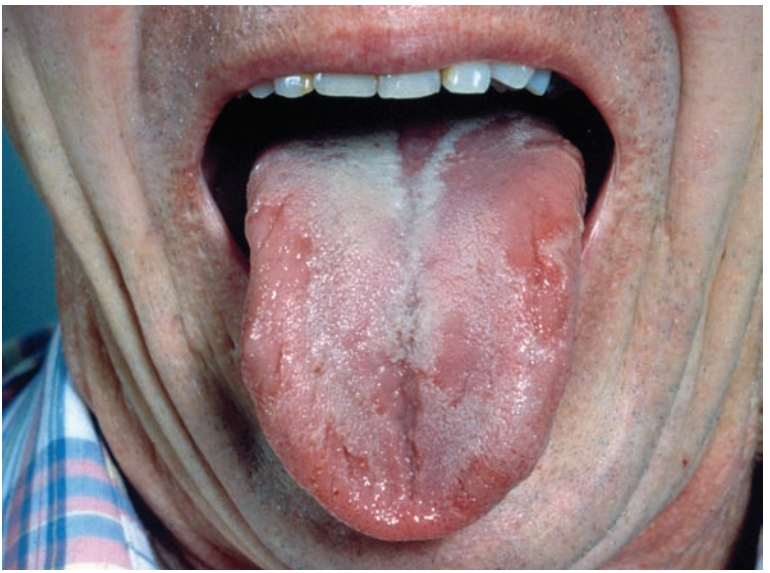


A 25-year-old man comes to you concerned about the appearance of his tongue, which he states has developed changing patterns over the past few months. He has no pain or associated symptoms. What is the most appropriate next step?

a. Reassure the patient that although there is no effective treatment, the condition is benign
b. Prescribe nystatin swish and swallow 4 times per day
c. Treat with topical corticosteroids
d. Biopsy the lesions for a definitive diagnosis
e. Refer the patient to a dermatologist

Geographic Tongue, or Benign Migratory Glossitis

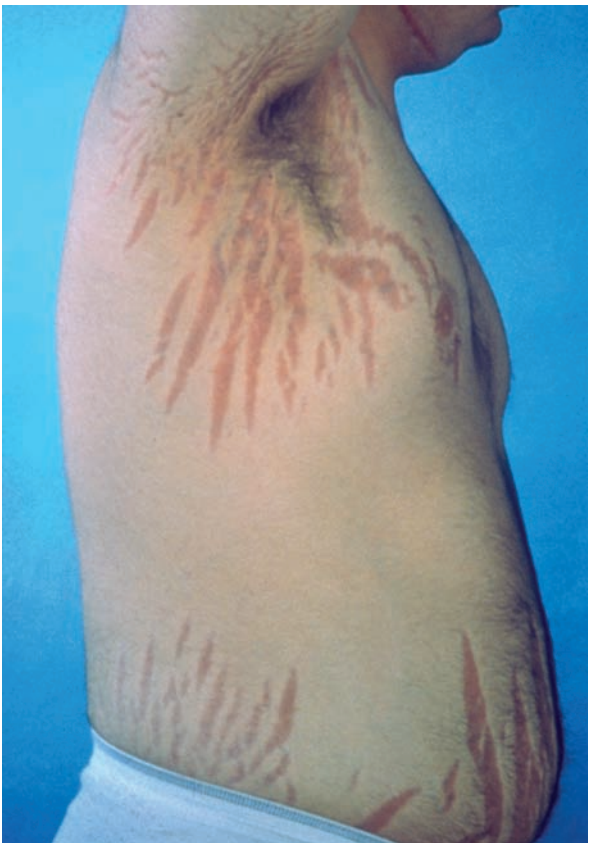
Answer: a

- Rapid destruction and regrowth of the filiform papillae cause changing patterns on tongue
- Most often asymptomatic, although spicy food, alcohol, and smoking may be irritants in the condition
- Cause unknown
- Differential diagnosis includes the following:

  Lichen planus
  Leukoplakia
  Reiter's syndrome
  Pustular psoriasis
  Drug eruptions

- The diagnosis is established by continued observation, revealing the changing patterns of geographic tongue
- The patient should avoid smoking and ingesting irritating foods
- The patient should be reassured that the condition is benign
- No treatment is needed, and none has been proved to help

Reference
Powell FC. Glossodynia and other disorders of the tongue. *Dermatol Clin*. 1987;5:687-693.



The 42-year-old man shown here presents with fatigue and muscle weakness. This patient also may have all of the following features *except*:

a. Thin, friable skin
b. Hypertension
c. Weight loss
d. Acne
e. Centripetal fat distribution

Cushing's Syndrome

Answer: c

- Cushing's syndrome may be either exogenous (more common) or endogenous. It occurs most commonly in women between 30 and 50 years of age
- Features of cortisol excess include the following:

  Truncal obesity
  Moon facies
  Acne
  Weight gain (the most common finding)
  Violaceous striae
  Muscle weakness
  Osteoporosis
  Thin, friable skin with easy bruisability
  Hyperglycemia
  Psychiatric disturbances

- In women, additional problems include breast atrophy, amenorrhea, and hirsutism
- The most common form of endogenous Cushing's syndrome is Cushing's disease, in which hypersecretion of corticotropin from the pituitary is the dominant finding. Transsphenoidal microadenectomy or hypophysectomy may be appropriate treatment
- Other endogenous forms include ectopic corticotropin-producing tumors (plasma corticotropin generally is not suppressed with the dexamethasone suppression test) and, rarely, corticotropin-releasing hormone–producing tumors
- The most sensitive test for Cushing's syndrome is 24-hour urine free cortisol test

References
Clayton LH, Dilley KB. Cushing's syndrome. *Am J Nurs.* 1998;98:40-41.
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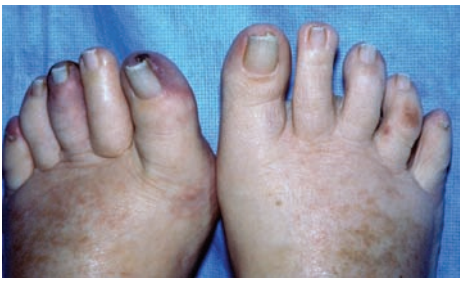
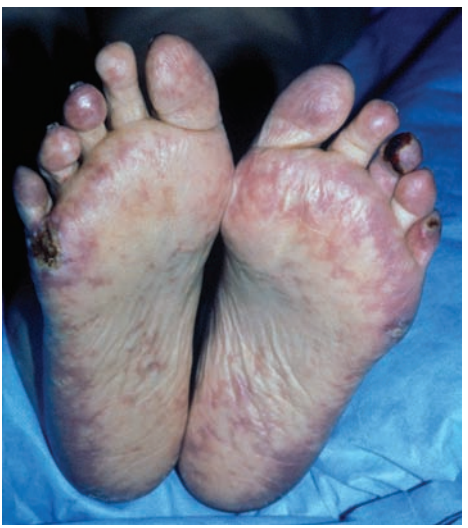




The skin lesions shown here developed in a 45-year-old man with hypertension after percutaneous renal angiography. Complete blood count shows eosinophilia. What is this syndrome?

a. Antiphospholipid antibody syndrome
b. Warfarin necrosis syndrome
c. Scleroderma renal crisis
d. Hepatorenal syndrome
e. Blue toe syndrome

Atheroemboli, Cholesterol Emboli, or Blue Toe Syndrome

Answer: e

- After angiography, atheroembolism is estimated to occur in up to 30% of patients, according to autopsy studies. However, the syndrome may occur without an obvious precipitating event. Many cases remain clinically silent
- Worsening renal function (67%-85%), increasing hypertension (43%-66%), skin changes (65%), abdominal pain (24%-37%), and leg pain (15%-28%) are the most common clinical sequelae
- Blue or purple toes and livedo reticularis are the most common skin changes, both of which are visible on the images shown on the previous page. Necrosis of digits may occur
- Fever, myalgias, headache, and weight loss are not uncommon. Gross hematuria, hematochezia, penile gangrene, and spinal cord infarction have been described in this syndrome
- Eosinophilia, eosinophiluria, hypocomplementemia, and increased erythrocyte sedimentation rate have been noted on laboratory investigations
- Definitive diagnosis is made by finding cholesterol clefts on biopsy of affected tissue
- There is no clearly effective treatment. Recurrences are common
- Prognosis is poor. Most deaths result from coronary artery disease within several years of embolization

References

Applebaum RM, Kronzon I. Evaluation and management of cholesterol embolization and the blue toe syndrome. *Curr Opin Cardiol*. 1996;11:533-542.
Robson MG, Scoble JE. Atheroembolic disease. *Br J Hosp Med*. 1996;55:648-652.

Shown is the urine specimen from a 72-year-old man who recently underwent coronary artery bypass grafting and mitral valve replacement. He reports no pain. Which of the following is the *most likely* cause of the change in urine color?

a. Urinary tract malignancy
b. Warfarin anticoagulation
c. Nephrolithiasis
d. Hemoglobinuria due to valve hemolysis
e. Myoglobinuria

Hemoglobinuria Due to Valve Hemolysis

Answer: d

- Postsurgical causes of traumatic hemolysis resulting in hemoglobinuria include the following:

  Patch repairs of atrial and ventricular septal defects
  Mitral and aortic valve replacements

- Older-model artificial valves are more likely to cause hemolysis
- Mechanisms of valve hemolysis are thought to be the following:

  Regurgitant jet impact on the prosthetic ring
  Regurgitant jet fragmentation by a dehisced prosthetic ring
  Rapid jet acceleration through prosthetic channels
  Direct trauma from older valves (blender effect)

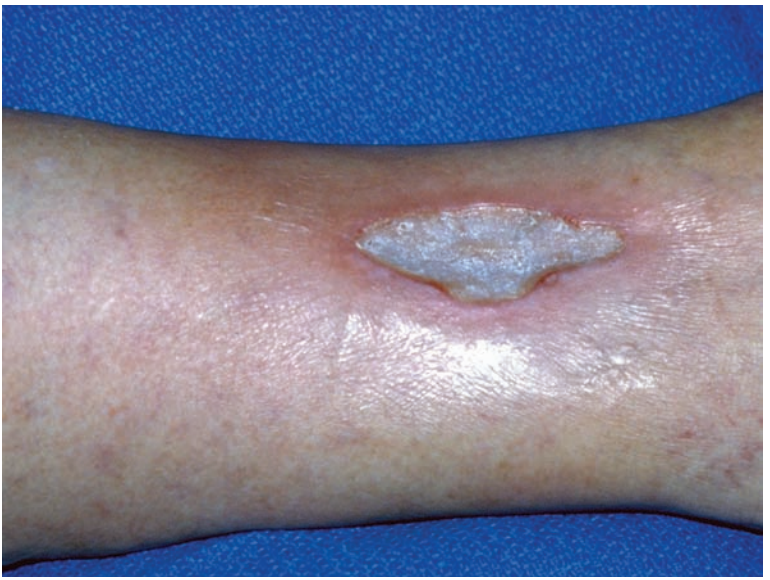
- If valve hemolysis is severe, reoperation for valve replacement may be indicated

References

Cerfolio RJ, Orzulak TA, Pluth JR, et al. Reoperation after valve repair for mitral regurgitation: early and intermediate results. *J Thorac Cardiovasc Surg*. 1996;111:1177-1183.

Shulman LN, Braunwald E, Rosenthal DS. Hematological-oncological disorders and heart disease. In *Heart Disease: A Textbook of Cardiovascular Medicine*. 5th ed. Edited by E Braunwald. Philadelphia: WB Saunders Company; 1997:1790.

Ward RP, Sugeng L, Weinert L, et al. Images in cardiovascular medicine: hemolysis after mitral valve repair. *Circulation*. 2000;101:695-696.



A 67-year-old man had a long history of hypertension, now controlled with angiotensin-converting enzyme inhibitor and diuretic therapy. The ulcer shown here developed on the lateral anterior shin and was extremely painful. Each of the following may be indicated in the care of this ulcer *except*:

a. Narcotic pain control
b. Smoking cessation
c. Excision
d. Addition of a β-adrenergic blocker to the antihypertensive regimen
e. Skin grafting

Arteriolar (Hypertensive) Ulcer or Martorell's Ulcer

Answer: d

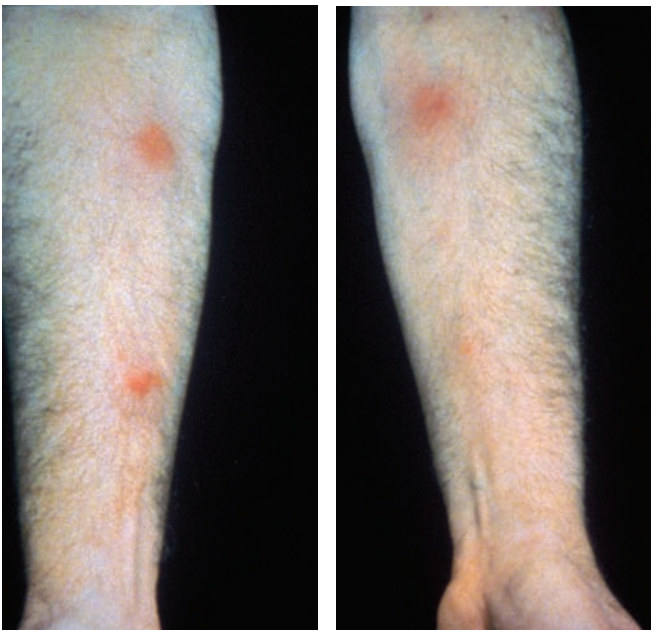
- Described by Martorell in 1945
- Hypertensive ulcers are characteristically located in the lateral supramalleolar region of the lower extremities
- Characteristically, the ulcers have a punched-out appearance and serpiginous borders
- Ulcers result from small artery and arteriolar narrowing and occlusion
- The ulcers are often extremely painful
- A history of hypertension is the sine qua non of diagnosis, but the patient may be receiving treatment and be normotensive at presentation
- Peripheral vascular disease, edema, and stasis pigmentation are usually absent
- Antihypertensive therapy, smoking cessation, and pain control are important in the management of these ulcers
- Excision and skin grafting are often needed. Lumbar sympathectomy is sometimes needed to allow healing
- Addition of β-adrenergic blocker therapy is relatively contraindicated in patients with arteriolar ulcers. If β-blocker therapy is being used, it should be stopped, if possible, to allow for maximal peripheral vasodilation

References

Goodfield M. Optimal management of chronic leg ulcers in the elderly. *Drugs Aging*. 1997;10:341-348.

Habermann TM. *Mayo Clinic Internal Medicine Board Review 2004-2005*. Philadelphia: Lippincott Williams & Wilkins; 2004:1030.

Henderson CA, Highet AS, Lane SA, et al. Arterial hypertension causing leg ulcers. *Clin Exp Dermatol*. 1995;20:107-114.



A 25-year-old man presents with a 3-month history of crampy abdominal pain and bloody diarrhea and a 3-week history of multiple red and tender lesions of the forearms. His past medical history is notable for low back pain, worse in the morning. What is the most likely underlying cause for his skin condition?

a. Sarcoidosis
b. Psoriasis
c. *Yersinia enterocolitica* infection
d. Medication
e. Inflammatory bowel disease

Erythema Nodosum

Answer: e

- Erythema nodosum (EN) presents as tender, red, quarter-sized subcutaneous nodules
- It is usually localized to the pretibial areas
- The nodules consist of plaques of infiltrating mononuclear cells
- The lesions may be acute and self-limited, or chronic
- The most common cause of EN is streptococcal pharyngitis
- Other infectious agents associated with EN include the following:

  Viruses
  Yersinia enterocolitica
  Coccidioides
  Histoplasma

- The drugs most commonly associated with EN are the following:

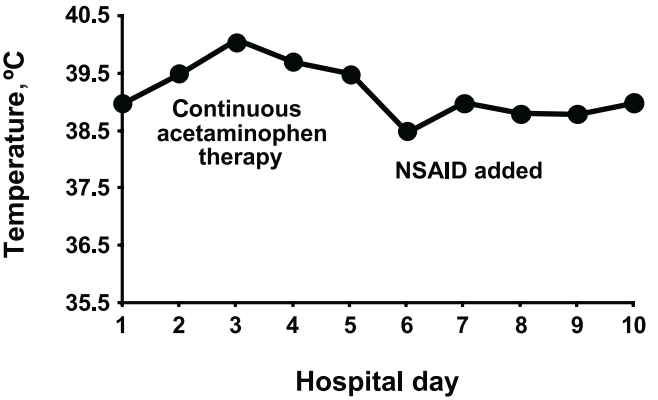
  Sulfonamides
  Oral contraceptive pills

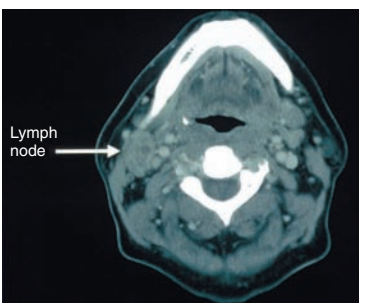
- EN also is associated with the following:

  Sarcoidosis
  Inflammatory bowel disease (as in this case)
  Behçet's syndrome

Reference

Habermann TM. *Mayo Clinic Internal Medicine Board Review 2004-2005*. Philadelphia: Lippincott Williams & Wilkins; 2004:30, 175, 178.

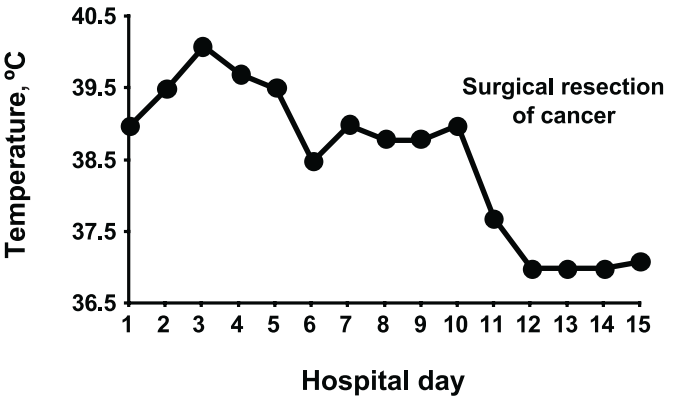




A 30-year-old man presents with a 4-week history of fevers (as high as 40°C), night sweats, and swelling of the right side of the neck. He did not respond to an empiric course of antibiotics. His fever curve and response to acetaminophen and a nonsteroidal anti-inflammatory drug (NSAID) are shown. What is the most likely cause of the fevers?

a. Abscess
b. Endocarditis
c. Neoplasm
d. Medication allergy
e. Chronic viral infection

Neoplasm-Associated Fever

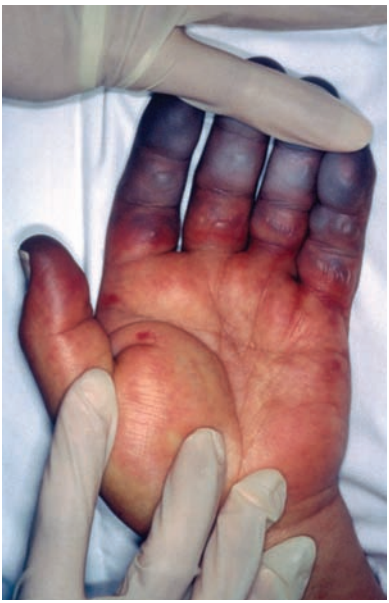


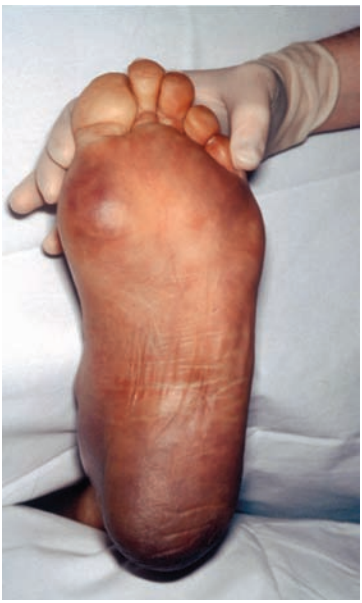
Answer: c

- Neoplasm-associated fever often is associated with hematologic malignancies (lymphoma in this case)
- Proposed diagnostic criteria for neoplastic fever include the following:

  Daily fevers of more than 37.8°C for more than 2 weeks
  Lack of evidence for infection
  Lack of evidence for allergic reaction
  Lack of response to antibiotics
  Lysis of fevers with NSAIDs (i.e., "naproxen test")
- Neoplastic fever is thought to be due to neoplastic production of pyrogenic cytokines (e.g., tumor necrosis factor, interleukin-1, and interleukin-6)
- Neoplasm should be included in the differential diagnosis of fever of unknown origin

Reference
Chang JC. Neoplastic fever: a proposal for diagnosis. *Arch Intern Med*. 1989;149:1728-1730.





A 58-year-old man was treated with tissue plasminogen activator after an anterior myocardial infarction and then admitted to the hospital. He was given intravenous unfractionated heparin. Four days later, discoloration of his feet and hands is noted. Which one of the following is true of this condition?

a. It is associated with concurrent use of acetylsalicylic acid
b. Cessation of smoking prevents progression of this illness
c. Urine eosinophils are common
d. Antigen-antibody interactions may be causative
e. Optimal treatment includes continuation of heparin therapy

Heparin-Induced Thrombocytopenia

Answer: d

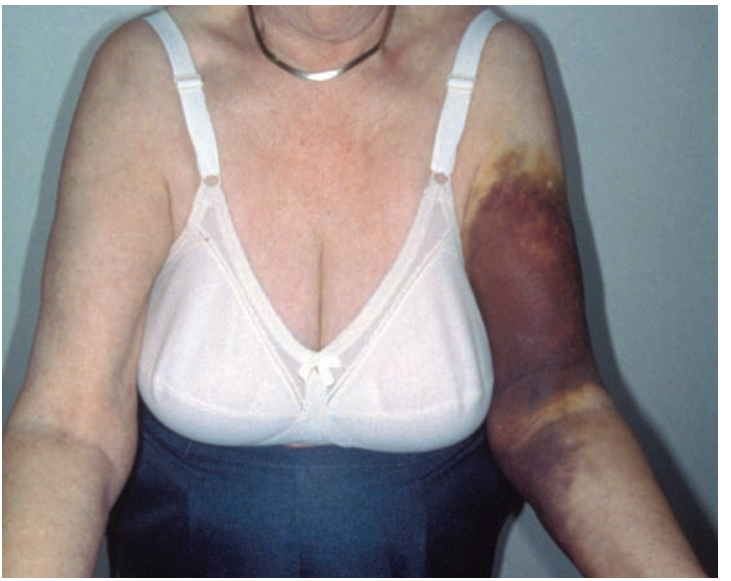
- The prevalence of heparin-induced thrombocytopenia (HIT) during heparin therapy has been reported to be 5%-10%
- The severity of disease varies, and the frequency of thrombosis with HIT is less than 1% to 2%
- The diagnosis may be supported by the presence in serum of heparin-associated antibodies found by enzyme-linked immunosorbent assay or heparin-dependent platelet-stimulating activity with a 3H-serotonin-release assay
- Onset is usually 4 to 10 days after initiation of therapy
- The cause of the phenomenon has not been completely elucidated, although it may be related to an antibody-antigen complex–mediated interaction
- Complications can include either thrombosis or hemorrhage
- Thrombosis can be severe, including bilateral deep venous thrombosis, pulmonary embolism, sagittal sinus thrombosis, and venous gangrene
- Treatment includes discontinued use of all heparin products, including intravenous flushes. If anticoagulation is critical to prevent further progression of thrombus, alternative therapies such as danaparoid or lepirudan may be initiated
- Use of warfarin is contraindicated in HIT because of the risk of worsening thrombosis and venous limb gangrene

References

Habermann TM. *Mayo Clinic Internal Medicine Board Review 2004-2005*. Philadelphia: Lippincott Williams & Wilkins; 2004:1025.

Kadidal VV, Mayo DJ, Horne MK. Heparin-induced thrombocytopenia (HIT) due to heparin flushes: a report of three cases. *J Intern Med*. 1999;246:325-329.

Warkentin TE, Elavathil LJ, Hayward CP, et al. The pathogenesis of venous limb gangrene associated with heparin-induced thrombocytopenia. *Ann Intern Med*. 1997;127:804-812.



Which one of the following physical maneuvers against resistance likely will be difficult for this patient?

a. Wrist flexion
b. Wrist extension
c. Forearm supination
d. Forearm pronation
e. Shoulder flexion

Rupture of the Distal Long Biceps Tendon

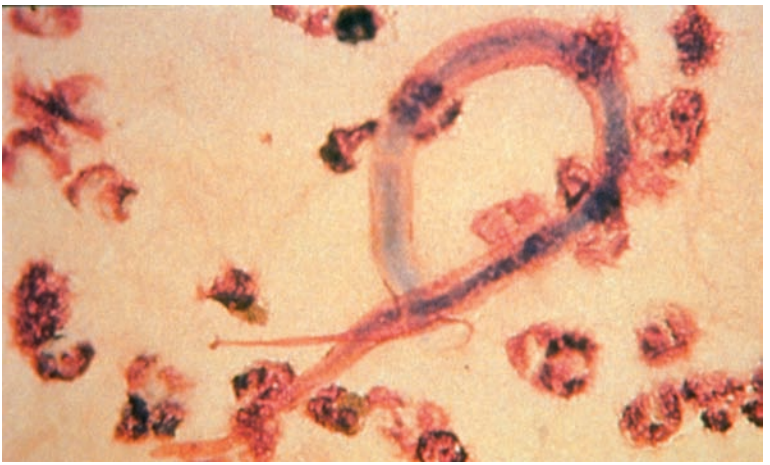
Answer: c

- Complete rupture of the distal biceps tendon is an unusual event; 96% of cases involve the long head
- This may be preceded by a traumatic or inflammatory event
- The dominant extremity is most often involved (86% of patients)
- Pain at the biceps tendon insertion site (proximal volar forearm) and progressive soft tissue swelling are key findings
- On physical examination, exquisite pain may be noted with forearm supination and flexion against resistance
- Computed tomography and magnetic resonance imaging are appropriate diagnostic methods to visualize the extent of tendon rupture. Contrast enhancement consistent with synovitis, bursal lesions, and tenosynovitis also may be readily visualized
- Management for partial tears or for patients without range-of-motion limitations may be conservative (i.e., analgesic therapy, casting, post-isometric relaxation)
- For complete tears and in patients with considerable limitations of arm function, surgical revision is appropriate
- Tobacco smoking increases the risk of distal biceps tendon rupture

References

Durr HR, Stabler A, Pfahler M, et al. Partial rupture of the distal biceps tendon. *Clin Orthop*. 2000;374:195-200.

Safran MR, Graham SM. Distal biceps tendon ruptures: incidence, demographics, and the effect of smoking. *Clin Orthop*. 2002;404:275-283.



A 35-year-old woman from eastern Africa presents with unilateral lower extremity edema. Her peripheral blood smear is shown here. At what time of day was the blood sample most likely obtained?

a. 6 AM to 12 noon
b. 12 noon to 6 PM
c. 6 PM to 12 midnight
d. 12 midnight to 6 AM
e. 9 AM to 3 PM

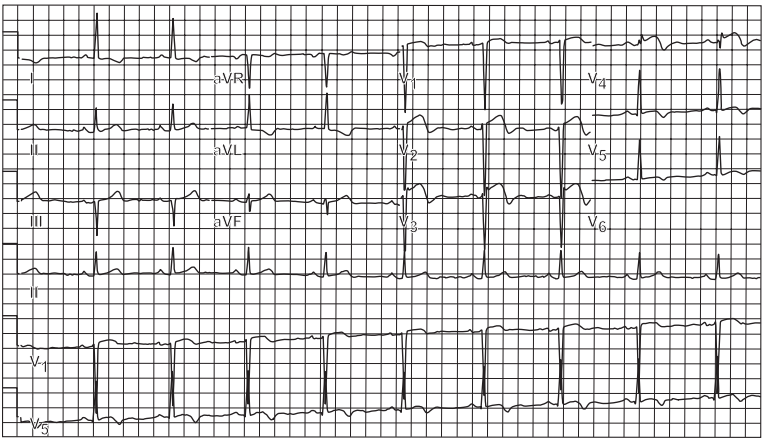
Answer: d

- *Wuchereria bancrofti* is the most common human filarial parasite
- Persons living in tropical regions across the world are at risk. Nearly 100 million people are infected worldwide
- Humans are hosts, and mosquitoes are vectors
- Living worms cause minimal tissue reaction, which may be asymptomatic
- Death of adult worms leads to granulomatous inflammation and permanent fibrosis, which may obstruct lymphatic channels
- The most serious consequence is elephantiasis (pachyderma)
- Bancroftian fever may occur at regular 24-hour intervals, reflecting the nocturnal periodicity of bancroftian filariasis
- Therefore, peripheral smear is most likely to be positive after midnight and before daybreak
- Also noted are hypereosinophilia, increased serum IgE, and positive antifilarial antibody
- Plain radiographs may show calcified, dead filarial worms. Ultrasonography may show live worms. Lymphoscintigraphy may have a role in clinically diagnosed cases to identify patients at risk for elephantiasis
- Other clinical consequences include filarial monoarthritis and tropical pulmonary eosinophilia, which may present as nocturnal coughing and wheezing
- Treatment is with diethylcarbamazine, ivermectin, or albendazole
- All treatments clear microfilariae, not adult worms

References

Dunn IJ. Filarial diseases. *Semin Roentgenol*. 1998;33:47-56.

Lymphatic filariasis: the disease and its control. Fifth report of the WHO Expert Committee on Filariasis. *World Health Organ Tech Rep Ser*. 1992;821:1-71.



A 49-year-old woman had a myocardial infarction 7 weeks ago but is otherwise recovering and feeling well. The electrocardiogram obtained in the office is shown here. An echocardiogram most likely will reveal which one of the following?

a. Large pericardial effusion
b. Biatrial enlargement
c. Calcified pericardium
d. Ventricular aneurysm
e. New anterolateral wall motion abnormalities

Ventricular Aneurysm

Answer: d

- Ventricular aneurysm is a localized protrusion of the ventricular cavity during systole and diastole. Wall motion is typically dyskinetic to akinetic
- Ventricular aneurysm often develops after acute transmural myocardial infarction, especially in situations involving occluded proximal or mid-left anterior descending coronary vessels
- Other causes for ventricular aneurysm include Chagas' disease and cardiac infiltration by sarcoidosis
- Electrocardiographic findings suggestive of ventricular aneurysm include the following:

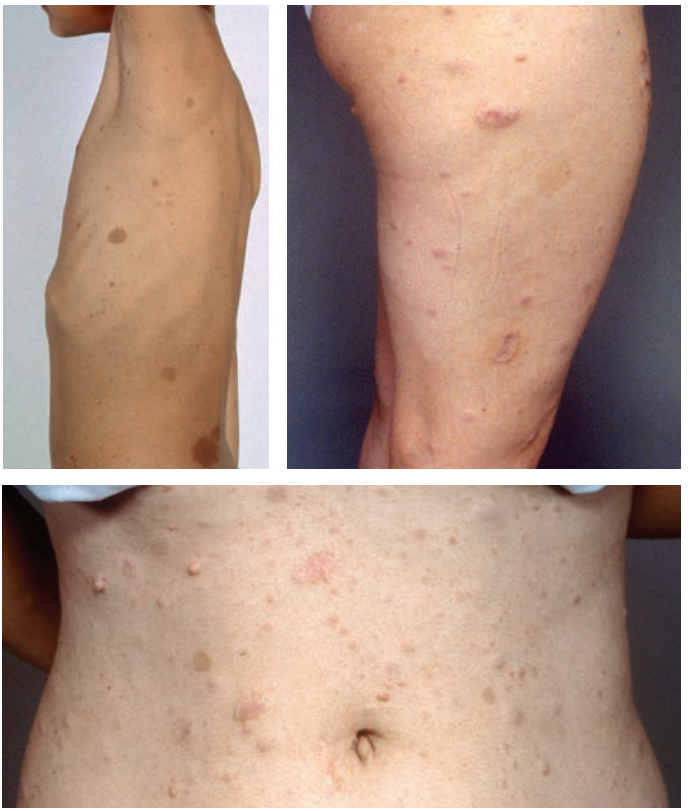
  Precordial leads showing large Q waves
  Persistent ST elevation (longer than 2 weeks) after acute myocardial infarction

- Diagnosis with echocardiography is considered more sensitive than ventriculography for detecting mural thrombus in the left ventricular aneurysmal cavity
- Complications of large aneurysms include the following:

  Congestive heart failure (due to poor left ventricular function)
  Angina pectoris (presence of ischemic but viable myocardium)
  Ventricular arrhythmias
  Rupture (especially if reinfarction occurs at aneurysm border)
  Thromboembolism (mural thrombi develop in 50% of patients, and 5% may embolize)

Reference

Ba'albaki HA, Clements SD Jr. Left ventricular aneurysm: a review. *Clin Cardiol*. 1989;12:5-13.



A pair of siblings (brother and sister) have multiple brown macules and nodular lesions on their skin. Their father has similar lesions. All of the following are part of the diagnostic criteria for this disease *except*:

a. Axillary or inguinal freckling
b. Two or more Lisch nodules of the iris
c. Positive family history
d. Six or more café au lait macules
e. Adenoma sebaceum

Neurofibromatosis Type 1

Answer: e

- Neurofibromatosis type 1 is autosomal dominant
- The incidence is 1 per 3,000 to 4,000 persons
- In 50% of cases, it is a new mutation
- Diagnosis is based on 2 or more of the following:

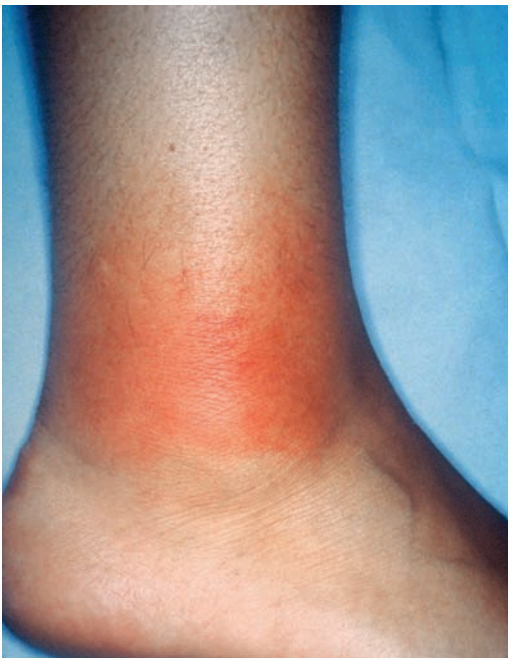
  Six or more café au lait macules
  Axillary or inguinal freckling
  Two or more Lisch nodules of the iris
  Two or more neurofibromas
  One plexiform neurofibroma
  A positive family history

- Uncommon characteristics that help make the diagnosis are the following:

  Orbital or sphenoid wing dysplasia
  Central nervous system glioma
  Renal artery dysplasia
  Tibial pseudofracture
  Abdominal aortic coarctation
  Pheochromocytoma
  Scoliosis

- Malignancy develops in less than 10% of patients, most often neurofibrosarcoma
- Multiple mutations in a gene of a GTPase involved in the *ras* signaling process are causative

Reference
Habermann TM. *Mayo Clinic Internal Medicine Board Review 2004-2005.* Philadelphia: Lippincott Williams & Wilkins; 2004:367.



A 13-year-old Sephardic Jewish boy had recurrent fever, abdominal pain, arthritis, pleuritic chest pain, and the skin lesions shown here. What is the treatment of choice?

a. Nafcillin
b. Cephalexin
c. Colchicine
d. Prednisone
e. Acetaminophen

Familial Mediterranean Fever (FMF, or Familial Paroxysmal Polyserositis)

Answer: c

- FMF is an autosomal recessive disease that affects mostly patients of non-Ashkenazi (e.g., Sephardic) Jewish, Armenian, Turkish, or Arabic descent
- FMF is marked by recurrent paroxysmal episodes lasting 12 to 72 hours involving inflammation of serosal tissues (e.g., pleura, peritoneum, and synovium). The commonly reported symptoms include fever (96%-100%), abdominal pain (89%-96%), chest pain (33%-57%), arthritis or arthralgias (21%-76%), erysipelas-like rash (12%-41%), and amyloidosis (2%)
- Laboratory abnormalities during attacks often include increased sedimentation rate, increased leukocyte count (neutrophilic predominance), increased fibrinogen and other acute-phase reactants, and microscopic hematuria and proteinuria. These abnormalities usually resolve after the attack
- Onset usually is before age 20 years, but late occurrences have been reported
- The predominant gene mutation is thought to have originated from a single common ancestor who lived about 2,500 years ago, before the Babylonian captivity of the ancient Jews
- Colchicine prophylaxis (1-2 mg per day) greatly reduces the number of attacks and may eliminate them altogether. This therapy has been instrumental in decreasing the frequency of amyloidosis in FMF

References

Ben-Chetrit E, Levy M. Familial Mediterranean fever. *Lancet*. 1998;351:659-664.

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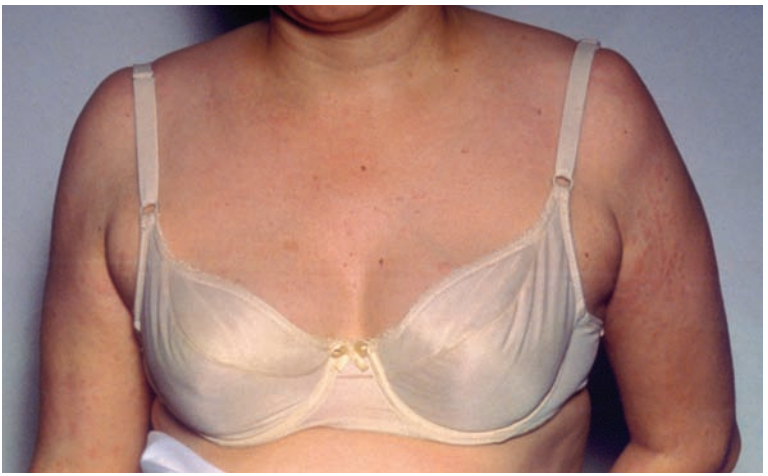

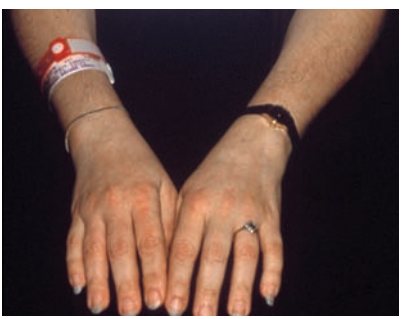

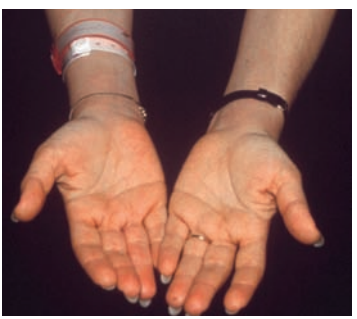

A 34-year-old woman complains of daily fevers, evanescent rash, and arthritis. Rheumatoid factor, antinuclear antibody, and Lyme serologic results are all negative. The ferritin value is 1,200 µg/L. What is the most likely diagnosis?

a. Infectious endocarditis
b. Adult-onset Still's disease
c. Bancroftian fever
d. Relapsing fever
e. Quotidian fever

Adult-Onset Still's Disease

Answer: b

- The disease was first described by Bywaters in 1971. The pathogenesis is unknown
- Multiple diagnostic criteria exist, but Yamaguchi's criteria have more than 92% sensitivity across multiple population groups:

  Major criteria: fever, arthralgia, typical rash, and leukocytosis
  Minor criteria: sore throat, lymphadenopathy, splenomegaly, liver dysfunction, and absence of rheumatoid factor and antinuclear antibody
  Diagnosis is established if 5 or more criteria are present, including 2 or more major criteria
- The rash is often described as evanescent and maculopapular and of salmon-pink coloration
- The arthritis is most common in the wrists, shoulders, hips, and knees
- Patients also may have serositis
- Although not part of the diagnostic criteria, the ferritin value is characteristically increased and is a marker of active disease
- Infectious, malignant, and other rheumatologic diseases should be excluded
- Treatment is with nonsteroidal anti-inflammatory drugs

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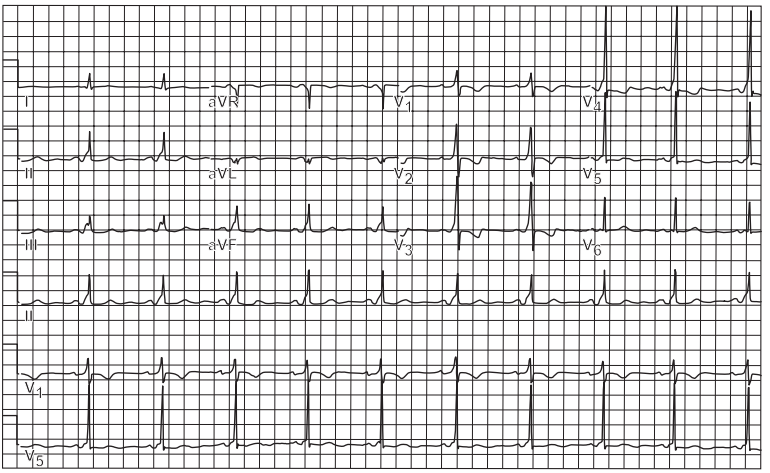

An 18-year-old man presents to your office for medical clearance before participating in fall sports. The baseline electrocardiogram is shown here. This pattern is most commonly associated with which one of the following congenital abnormalities?

a. Atrial septal defect
b Ventricular septal defect
c. Tetralogy of Fallot
d. Dextrocardia
e. Ebstein's anomaly

Wolff-Parkinson-White Pattern

Answer: e

- Electrocardiographic (ECG) features of Wolff-Parkinson-White (WPW) *pattern* which are indicative of preexcitation include the following:

  Short PR interval (<0.12 second)
  Wide QRS complex (>0.12 second)
  Gradual slurred upstroke of the QRS complex (delta wave)

- WPW *syndrome* may be diagnosed if patient has a history of recurrent tachyarrhythmias and evidence of preexcitation on ECG
- Most cases of WPW syndrome occur sporadically, although several familial patterns of inheritance have been described
- Approximately 7% to 20% of patients with WPW syndrome have associated congenital abnormalities. Ebstein's anomaly is the most common
- Accessory paths in WPW syndrome may be the following:

  Orthodromic: Conduction occurs antegrade through atrioventricular node, then retrograde through the accessory path. These are considered "concealed" paths because the QRS complex appears normal
  Antidromic: Conduction occurs antegrade through the accessory path, then retrograde through the atrioventricular node. This results in prominence of the delta wave and a widened QRS complex. This is less common than orthodromic conduction

- Most common cause of sudden death in patients with WPW syndrome is atrial fibrillation with rapid ventricular response that may degenerate into lethal ventricular fibrillation

References

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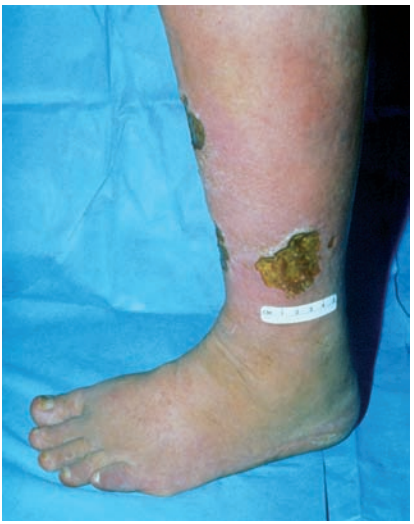

The ulcers shown here, hypertension, arthralgias, fatigue, abdominal pain, and mononeuritis multiplex developed in a 30-year-old woman. Biopsy showed acute necrotizing vasculitis.

1. Which of the following tests is most likely to have abnormal results?
   1a. Antineutrophil cytoplasmic antibody (c-ANCA) test
   1b. Blood eosinophil test
   1c. Chest radiography
   1d. Urinalysis
   1e. Electrocardiography

2. If this condition were due to an infectious disease, which of the following conditions would be most likely?
   2a. Bacterial endocarditis
   2b. *Mycoplasma* pneumonia
   2c. Leptospirosis
   2d. Syphilis
   2e. Viral hepatitis

Polyarteritis Nodosa

Answer 1: d

Answer 2: e

- Polyarteritis nodosa (PAN) is an acute necrotizing vasculitis that affects medium-sized and small arteries
- PAN is a systemic disease that commonly involves the kidneys, peripheral nerves, skin, and gastrointestinal tract
- PAN uncommonly involves the heart, central nervous system, lungs, and eyes
- Perinuclear-staining antineutrophil cytoplasmic antibody (p-ANCA) test may be positive but it is not specific for the disease
- Hepatitis B and C virus infections have been associated with some cases of ANCA-negative PAN. Treatment for these cases does not include immunosuppressive agents but rather antiviral medications and interferon alfa
- Angiography typically shows focal stenoses and microaneurysms of visceral arteries, especially the renal and mesenteric arteries
- For PAN that is not associated with hepatitis B or C virus, treatment is with corticosteroids and cytotoxic agents such as cyclophosphamide and azathioprine
- The 5-year survival rate is 5% without treatment and 80% with treatment

References

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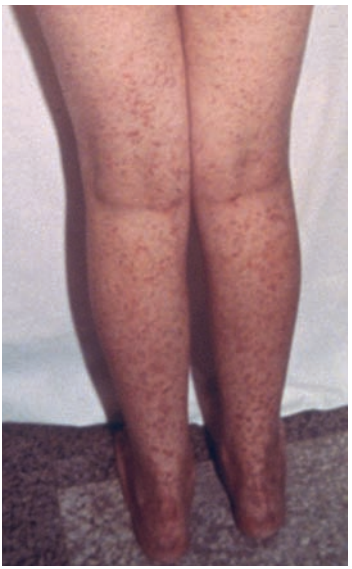
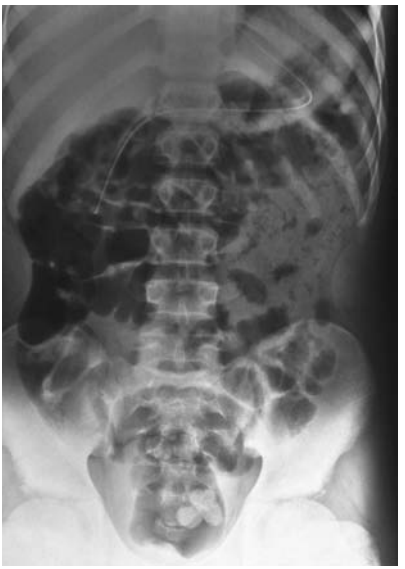




A previously healthy 26-year-old man presents with a 3-week history of abdominal pain and arthralgias. On physical examination, hypertension and palpable purpura of the lower extremities are found. Urinalysis shows hematuria. What is the most likely diagnosis?

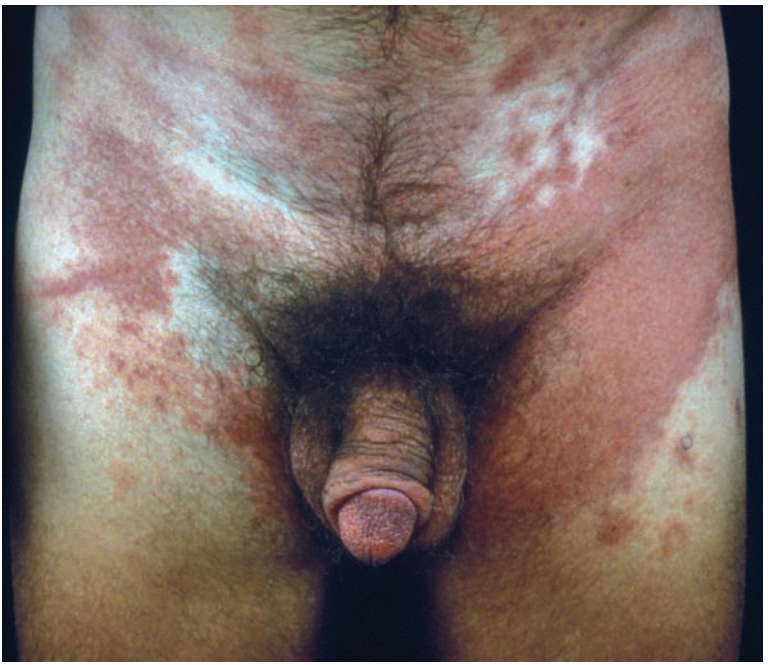
a. Henoch-Schönlein purpura
b. Cryoglobulinemia
c. Hypocomplementemic vasculitis
d. Rheumatoid vasculitis
e. Systemic lupus erythematosus

Henoch-Schönlein Purpura

Answer: a

- Henoch-Schönlein purpura (HSP) is a systemic hypersensitivity vasculitis
- Virtually all patients with HSP have palpable purpura
- Other classic signs and symptoms on presentation are abdominal pain, arthritis, and hematuria
- Biopsy shows vasculitis with IgA deposits; complement levels are normal
- Complications of HSP include hypertension, glomerulonephritis, intussusception, and gastrointestinal hemorrhage
- HSP usually resolves spontaneously after 1 week, although it may recur on several occasions over weeks to months before complete remission or after reexposure to the offending antigen
- Treatment is supportive only
- Prognosis is usually good, especially in children, but worsens with increasing age

Reference
Habermann TM. *Mayo Clinic Internal Medicine Board Review 2004-2005*. Philadelphia: Lippincott Williams & Wilkins; 2004:667, 969-970.



A 32-year-old man was not wearing a bathing suit when he walked through some weeds on the way to swim in a river. Within 24 hours, the severely pruritic lesions shown here developed. What is the likely cause?

a. Type I hypersensitivity reaction
b. Type II hypersensitivity reaction
c. Type III hypersensitivity reaction
d. Type IV hypersensitivity reaction
e. Type V hypersensitivity reaction

Allergic Contact Dermatitis (Rhus *Dermatitis)*

Answer: d

- The genus *Rhus* (*Toxicodendron*) includes poison ivy, poison oak, and poison sumac
- The plant oil urushiol is highly allergenic
- The delayed (type IV) hypersensitivity reaction is mediated by memory T cells
- In sensitive individuals (50%-70% of the U.S. population), an itchy, red, papulovesicular rash develops within 8 hours to 2 weeks after exposure. In severe cases, bullae may develop
- Even smoke from burning *Rhus* plants can result in skin lesions
- Common sites of involvement are the skin of the face and exposed extremities
- Prevention is the best course. If urushiol is washed off in 10 to 30 minutes, the cutaneous reactions can be avoided. Adequate clothing in high-risk areas is important for avoidance
- Clothing should be removed and washed as soon as possible after exposure
- Topical glucocorticoids may provide great relief if distribution is limited
- With facial, genital, or widespread involvement, systemic corticosteroids may be used. Prednisone equivalent of 1 mg/kg has been recommended, with tapers lasting at least 2 to 3 weeks. Shorter tapers risk rebound exacerbations
- The reaction usually resolves within 3 weeks with or without treatment. Secondary infection is the most frequent complication, but most cases resolve without complication

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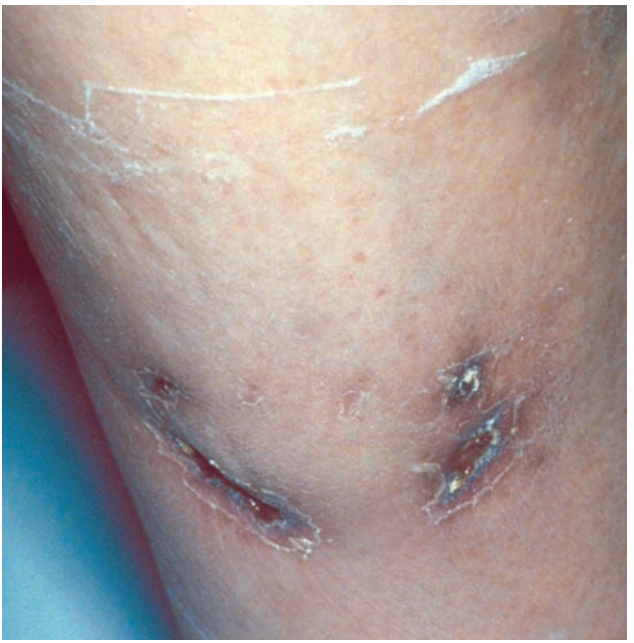

A patient was bitten by a cat. All of the following may be appropriate treatment in the setting of resulting infections, *except*:

a. Human rabies diploid vaccine
b. Tetanus toxoid booster
c. Human rabies immune globulin
d. Cephalexin
e. Amoxacillin/clavulanate

Answer: d

- Local infection develops in more than 80% of cat bites. Long, slender teeth that penetrate deeply but produce a largely closed wound predispose to inoculation and infection at the site of the bite wound
- *Pasteurella multocida*, the most common cause of infection after cat bites, is a gram-negative coccobacillus that is part of the oral flora of cats. Other causes of local infection include *Staphylococcus aureus*, *Streptococcus viridans*, and anaerobes
- Beware: *P. multocida* is often resistant to dicloxacillin, cephalexin, and clindamycin. Therefore, avoid these drugs for cat bites. Amoxacillin/clavulanate is acceptable treatment
- Rabies may manifest with hydrophobia, copious salivation, encephalitis, or myelitis. Definitive diagnosis requires the presence of Negri bodies on biopsy of the hippocampus. There is no known curative treatment for this invariably fatal disease. Therefore, human rabies immune globulin and human diploid vaccine should be given for prevention *before* the onset of clinical disease
- Tetanus is a risk for any contaminated wound. A tetanus-diphtheria booster should be given if the last booster was received more than 5 years ago

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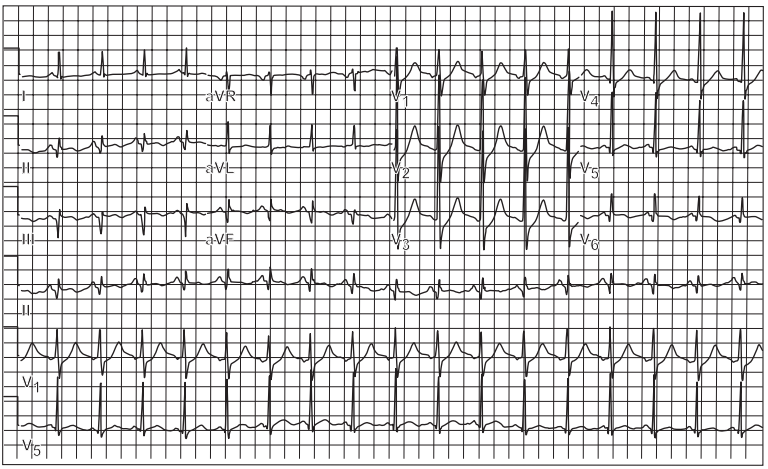

A 77-year-old man arrives in the emergency department complaining of nausea and vomiting. The electrocardiogram obtained is shown here. Which one of the following do you conclude?

a. Acute pericarditis
b. Lateral myocardial infarction
c. Anterior myocardial infarction
d. Posterior myocardial infarction
e. Noncardiac cause for the patient's symptoms

Posterior Myocardial Infarction

Answer: d

- Approximately 20% of acute myocardial infarctions (MIs) are posterior MIs due to left circumflex or right coronary artery lesions. They are the most commonly missed among practitioners for 2 reasons:

  1. Posterior MI may not be reflected by ST elevation (≥1 mm elevation in 2 or more contiguous leads) on standard electrocardiographic (ECG) leads. In 1 study, up to 11% of patients with posterior MI did not have ST elevation on standard ECG leads but did with the application of posterior leads
  2. ECG criteria suggestive of acute posterior MI are not commonly known

- Posterior MI commonly occurs in association with acute lateral or inferior MI, which worsens the prognosis of these latter conditions. Isolated posterior MI is considered unusual

- ECG criteria suggestive of this diagnosis include 1 or more of the following findings in lead V_1, V_2, or V_3:

  1. Tall, wide R wave
  2. Tall, upright T wave
  3. Horizontal ST-segment depression
  4. R/S wave ratio greater than 1.0 in lead V_2

- Posterior leads are placed in the following manner:

  V_7: fifth intercostal space at left posterior axillary line
  V_8: fifth intercostal space at left midscapular line
  V_9: fifth intercostal space at left paravertebral line

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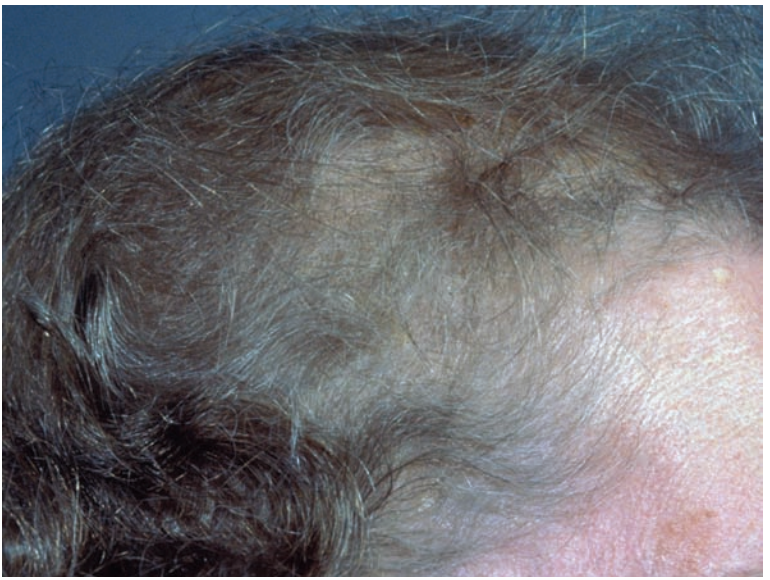

A 36-year-old man has recovered from a 6-week stay in the intensive care unit for severe pancreatitis. Now he presents with this hair loss. What is the most likely cause?

a. Androgenic alopecia
b. Telogen effluvium
c. Alopecia areata
d. Traction alopecia
e. Tinea capitis

Telogen Effluvium

Answer: b

- Diffuse shedding of normal hair after severe stress, such as the following:

  Shock
  Childbirth
  Surgery
  High fever
  Any severe illness
  Extreme grief
  Use of heparinoids
  Dieting with marked weight loss

- Hair follicles are usually randomly arranged in anagen (growing phase) or telogen (dying phase)
- Severe stress causes large numbers of anagen follicles to enter telogen, thus synchronizing hair growth and shedding; 100 to 1,000 hairs can be lost in a day
- Axillary and pubic hair also are affected
- Some patients (30% in some series) report trichodynia (a "pain in the hair" or paresthesia of the follicles)
- No treatment is necessary. The condition reverses over time when stress is resolved (usually 2-3 months)

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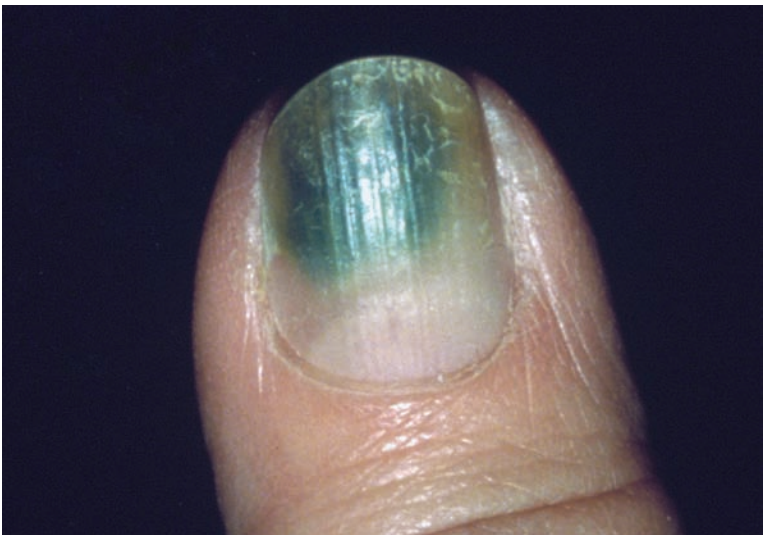

A 66-year-old woman who washes her dishes with ungloved hands presents with this nail finding. What is the likely cause?

a. *Proteus mirabilis* infection
b. *Pseudomonas aeruginosa* infection
c. Subungual hypersensitivity reaction to the dishwashing detergent
d. Cutaneous *Neisseria* infection
e. Cutaneous *Listeria* infection

Green Nails: Pseudomonas *Nail Infection*

Answer: b

- *Pseudomonas aeruginosa* is a motile gram-negative aerobe that produces blue (pyocyanin) and green (pyoverdin) pigments
- *P. aeruginosa* was previously known as *Bacillus pyocyaneus*, emphasizing its pigment production
- Chronic wet nails (as in "dishpan hands") promote the infection
- Usually, nail trauma is required for the infection to begin
- Concurrent fungal nail infection is often present
- Treatment may include the following:

  Keeping nails dry (protective gloves if immersion of hands cannot be avoided)
  Gentamicin topical drops up to four times daily
  Polymyxin B-acetic acid soaks
  Bleach (1:4 dilution) up to three times daily topically
  Oral ciprofloxacin
  Physical drainage

- Recurrence is possible
- *Pseudomonas* nail infection in health care workers can be a source of nosocomial infections

References
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Hall JH, Callaway JL, Tindall JP, et al. *Pseudomonas aeruginosa* in dermatology. *Arch Dermatol*. 1968;97:312-324.
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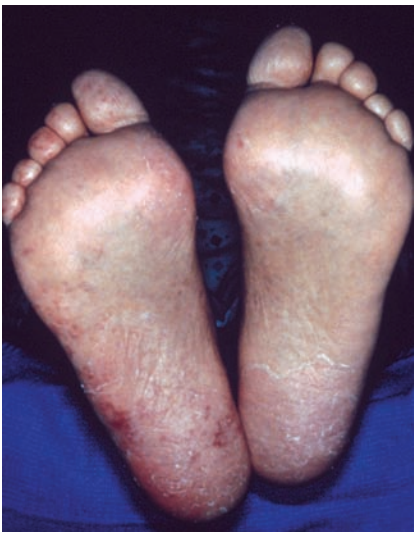
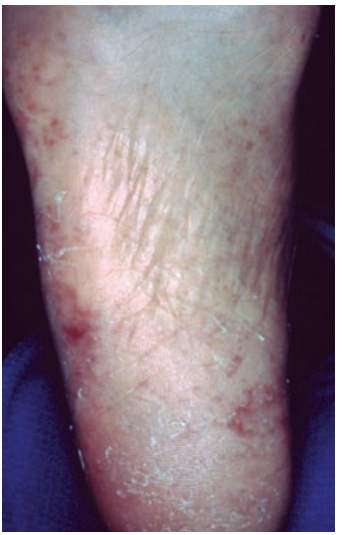




A 57-year-old man presents with new-onset fever, shortness of breath, lower extremity swelling, and weakness of his entire left side. If retinal examination is abnormal, which one of the following is a likely abnormality?

a. Papilledema
b. Sausage-shaped appearance of arterioles
c. Fundal hemorrhage
d. Background diabetic retinopathy
e. Absence of red reflex

Endocarditis

Answer: c

- Signs of endocarditis include:

Osler's nodes:	Painful lesions typically on pads of fingers or toes
Janeway lesions:	Painless distal cutaneous lesions
Roth's spots:	Fundal hemorrhages
Splinter hemorrhages:	Proximal nailbed hemorrhages

- Manifestations may include the following:

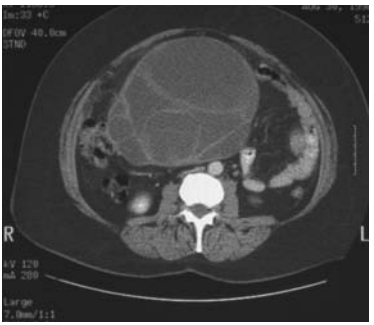
  Septic emboli
  Hematuria and renal failure
  Pulmonary infiltrates
  Intracerebral or epidural abscesses

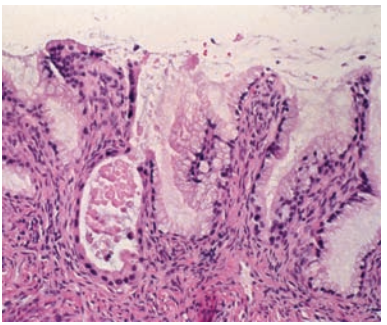
- Most common causative organism for native valve endocarditis is viridans group streptococci
- Injection drug users are more likely to have right-sided endocarditis caused by *Staphylococcus aureus* (60%)
- The most common cause of "culture-negative" endocarditis is previous antibiotic use
- HACEK organisms are other possible causes of culture-negative endocarditis:

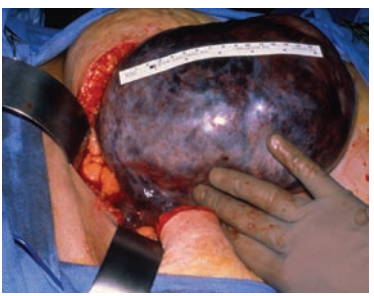
  Haemophilus
  Actinobacillus
  Cardiobacterium
  Eikenella
  Kingella

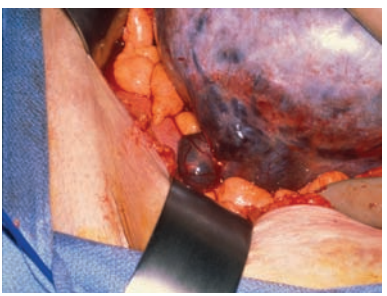
Reference

Habermann TM. *Mayo Clinic Internal Medicine Board Review 2004-2005*. Philadelphia: Lippincott Williams & Wilkins; 2004:581-583.









A 42-year-old woman presented with a 2-week history of dull pain in the left lower abdomen. Treatment with nonsteroidal anti-inflammatory drugs had been unsuccessful. On the day of presentation, she had the sudden onset of severe abdominal pain, nausea, and vomiting. A computed tomogram, intraoperative photographs, and pathologic results are shown. How long can this patient be expected to survive?

a. <6 months
b. 6 months-1 year
c. 1-5 years
d. 5-10 years
e. Survival is not altered by this disease

Mucinous Cystadenoma

Answer: e

- The most common form of mucinous ovarian tumor accounts for 20% of all ovarian neoplasms, but the overall incidence is low (6/100,000 women older than 40 years)
- Histologically, this tumor is identified by a single, tall, columnar epithelium with clear mucinous cytoplasm and uniform basally arranged nuclei resembling colonic epithelium
- 95% of the tumors are unilateral
- Usually asymptomatic, the tumor frequently becomes large before diagnosis. Record-setting masses weigh 150 to 300 pounds
- The large size may result in the following:

  Abdominal discomfort
  Dyspnea with recumbency
  Compression of the inferior vena cava with lower extremity edema
  Malnutrition as the woman diets to reduce the size of her abdomen

- This is a benign mass with a good prognosis
- Torsion of the 3-kg mass on the ovarian stalk caused the acute pain in this patient

References

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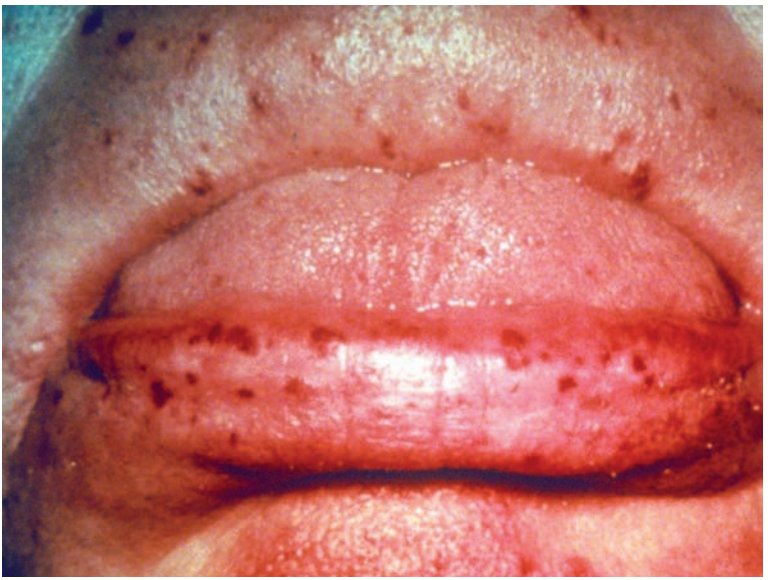


A 41-year-old man presents with hematochezia that has been present for 2 weeks. His past medical history is notable for recurrent epistaxis. His father died of stroke at a young age. Which one of the following is a common pulmonary manifestation of his disease?

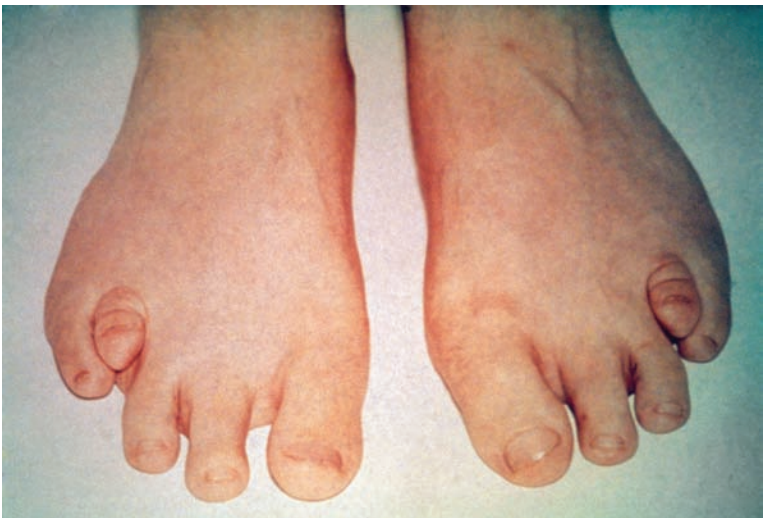
a. Pulmonary fibrosis
b. Arteriovenous malformation
c. Obstructive lung disease
d. Pleural effusion
e. Mesothelioma

Hereditary Hemorrhagic Telangiectasia

Answer: b

- Hereditary hemorrhagic telangiectasia (HHT) is also known as Osler-Weber-Rendu disease
- HHT is an inherited autosomal dominant disorder characterized by telangiectasia of the skin and mucous membranes
- Patients with HHT are predisposed to epistaxis and gastrointestinal bleeding
- In 20% of patients with HHT, pulmonary arteriovenous malformations (AVMs) develop
- AVMs cause shunting of blood, which in turn causes dyspnea, cyanosis, and clubbing
- AVMs also can lead to paradoxic embolism and stroke

Reference
Habermann TM. *Mayo Clinic Internal Medicine Board Review 2004-2005*. Philadelphia: Lippincott Williams & Wilkins; 2004:178, 365.



Abnormal laboratory values in this patient might include which one of the following?

a. Increased calcium
b. Decreased calcium
c. Decreased phosphate
d. Decreased parathyroid hormone
e. Decreased 1,25-dihydroxyvitamin D

Pseudohypoparathyroidism

Answer: b

- Pseudohypoparathyroidism is characterized by end-organ resistance to parathyroid hormone (PTH) at the receptor or postreceptor level
- On a molecular level, PTH activity can be affected by defects in the PTH receptor, mutations in adenylate cyclase, or alteration of G proteins. Distinguishing features of pseudohypoparathyroidism include the following:

  Type I: Decreased urinary cyclic adenosine monophosphate after administration of PTH
  Type Ia: Commonly known as Albright's hereditary osteodystrophy. Stimulatory G protein activity is decreased
  Type Ib: No phenotypic abnormalities, although renal resistance to PTH exists
  Type Ic: Similar to type Ia with hormone resistance, but no defects in G protein are noted

- Physical findings include short stature, obesity, round face, short metacarpals and metatarsals, mild mental retardation, and subcutaneous calcification
- High levels of PTH predispose to accelerated bone resorption and thus risk of fracture
- Laboratory findings include the following:

  Increased PTH
  Decreased calcium
  Increased phosphate

- Patients with pseudopseudohypoparathyroidism have similar physical findings without the biochemical abnormalities (i.e., normal levels of PTH, calcium, and phosphate)

References

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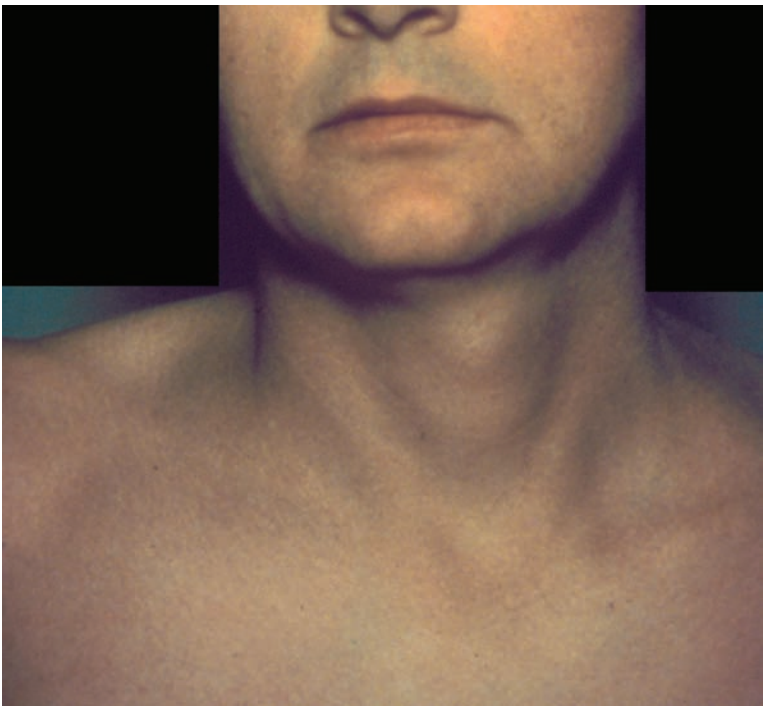
Habermann TM. *Mayo Clinic Internal Medicine Board Review 2004-2005*. Philadelphia: Lippincott Williams & Wilkins; 2004:215-216.



A 43-year-old man presents with polyuria and arthralgias. Hepatomegaly is found on physical examination. Laboratory findings include fasting glucose 296 mg/dL, alkaline phosphatase 390 mg/dL (normal, 98-251 mg/dL), and transferrin saturation 99%. Which one of the following is the treatment for this condition?

a. Phlebotomy
b. Cholestyramine
c. Deferoxamine
d. a + b
e. a + c

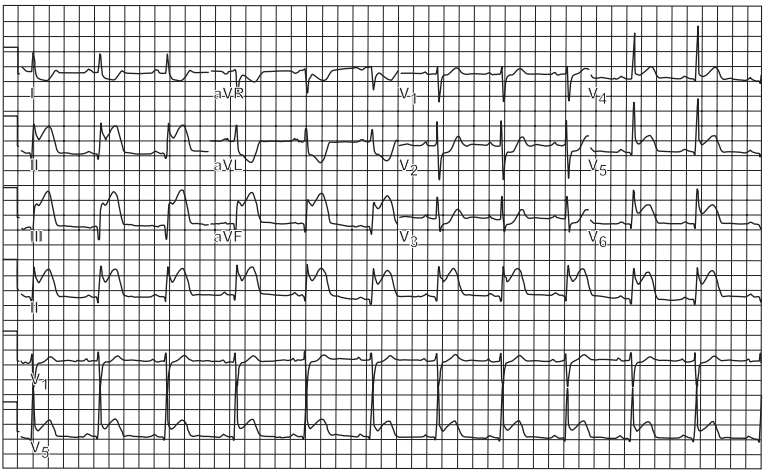
Hereditary Hemochromatosis

Answer: e

- Hereditary hemochromatosis is one of the most prevalent genetic disorders among persons of European ancestry (gene prevalence, 1:300)
- It is an autosomal recessive disease; the clinical disease develops in homozygotes
- The causative mutation results in unregulated iron uptake in the duodenum
- Men present with clinical symptoms earlier (4th-5th decade) than women because women's total body iron stores decrease with menstruation
- 50% of patients present with diabetes mellitus
- Other manifestations include hypogonadism, congestive heart failure, atrial fibrillation, abdominal pain, cirrhosis, hepatomegaly, arthropathy, and bronzed slate-gray skin
- The most advocated screening test for hereditary hemochromatosis is the transferrin saturation test; transferrin saturation more than 55% on 2 occasions suggests the disease
- Liver biopsy establishes the diagnosis; *HFE* gene testing is available
- Treatment with phlebotomy has best results before organ damage has occurred. Chelation with deferoxamine also has been used
- Hepatocellular carcinoma develops in 30% of patients with cirrhosis

Reference

Habermann TM. *Mayo Clinic Internal Medicine Board Review 2004-2005.* Philadelphia: Lippincott Williams & Wilkins; 2004:57, 303-304, 464-465.



A 54-year-old woman with diabetes arrives in the emergency department complaining of light-headedness and confusion. Her electrocardiogram is shown here. What do you conclude?

a. Acute pericarditis
b. Chronic pulmonary hypertension
c. Acute pulmonary embolism
d. Acute inferolateral myocardial infarction
e. Acute posterolateral myocardial infarction

Inferolateral Myocardial Infarction

Answer: d

- ST elevation (>0.1 mV) in inferior leads (II, III, avF) and lateral leads (I, avL, V_5, V_6) is suggestive of acute inferolateral myocardial infarction (MI)
- The presence of lateral electrocardiographic (ECG) abnormalities in addition to inferior, posterior, or inferoposterior ECG changes is highly sensitive for multivessel disease
- As a corollary to the above, the absence of lateral ECG changes in the presence of other ischemic abnormalities is strongly associated with single-vessel disease. This may assist in determining the need to pursue invasive investigations
- ST-segment elevation MI implies occlusion of epicardial coronary arteries
- 90% of myocardium supplied by an occluded coronary artery may infarct within 3 hours of the occlusion
- 25% to 30% of MIs are "silent"
- Silent MIs occur more often in patients with diabetes and in the elderly

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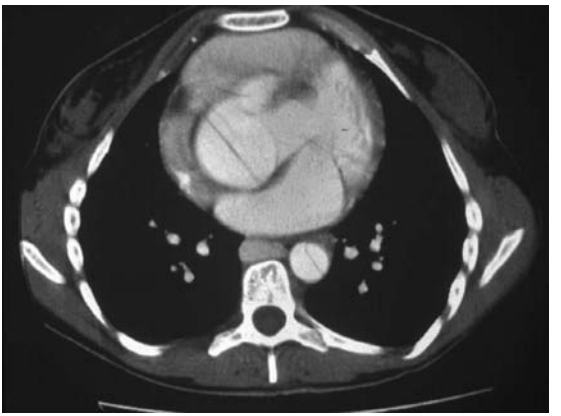
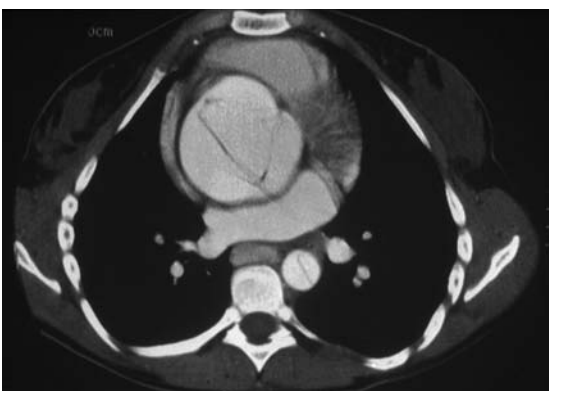
Habermann TM. *Mayo Clinic Internal Medicine Board Review 2004-2005*. Philadelphia: Lippincott Williams & Wilkins; 2004:101-109.

Mongiardo R, Schiavoni G, Mazzari M, et al. Significance of electrocardiographic abnormalities in the "lateral" leads in patients with acute inferior myocardial infarction. *Cardiologia*. 1988;33:681-690.





A 24-year-old woman whose height is 6 feet 5 inches complains of back pain. On examination, you notice her long, slender fingers, which easily hyperextend. Other clinical manifestations associated with this syndrome include all the following *except*:

a. Ectopia lentis
b. Highly arched palate
c. Mitral valve prolapse
d. Violaceous truncal striae
e. Mitral regurgitation

Acute Aortic Dissection in Marfan Syndrome

Answer: d

- Marfan syndrome is an autosomal dominant disorder; up to 20% of cases represent new mutations
- The syndrome has an incidence of at least 1:10,000
- Fundamental defect is in the gene for fibrillin-1 or fibrillin-2, found on chromosome 15
- The major cause for premature death is early dilatation of the ascending aortic root, leading to aortic incompetence and aortic dissection
- Multiorgan involvement includes the following:

  Cardiovascular system
  - Mitral valve prolapse with or without regurgitation
  - Dilatation of ascending aorta
  - Aortic or mitral regurgitation
  - Acute aortic dissection

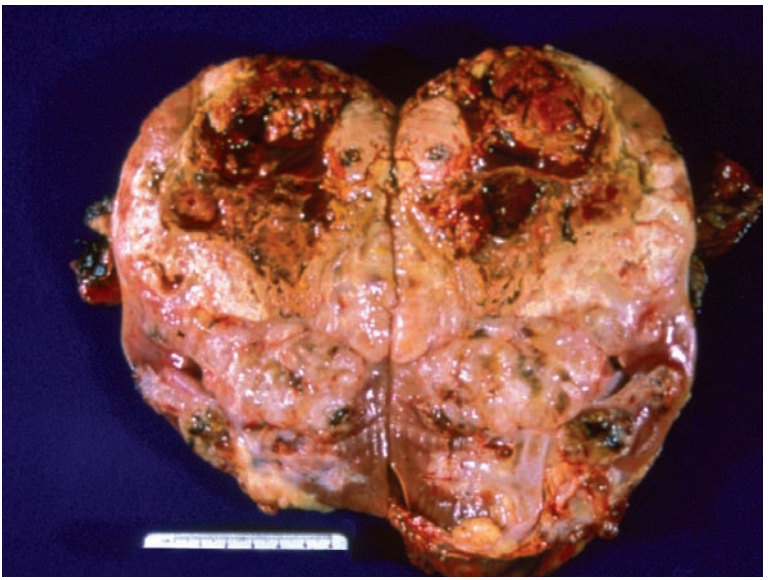
  Musculoskeletal
  - Tall stature with a low upper:lower segment ratio
  - Hyperextensibility of joints
  - Arachnodactyly
  - Scoliosis (60% of patients)
  - Pectus excavatum or carinatum
  - High arched palate
  - Long, slender facies

  Ocular
  - Ectopia lentis: subluxation of lens in 80% of patients
  - Retinal detachment
  - Myopia

References

Habermann TM. *Mayo Clinic Internal Medicine Board Review 2004-2005*. Philadelphia: Lippincott Williams & Wilkins; 2004:366-367.

Robinson PN, Godfrey M. The molecular genetics of Marfan syndrome and related microfibrillopathies. *J Med Genet*. 2000;37:9-25.



All of the following have been associated with an increased risk of renal cell carcinoma *except*:

a. Cigarette smoking
b. Severe obesity
c. Urinary tract infections
d. Use of oral contraceptives
e. Use of thiazide diuretics

Answer: d

- Up to 85% of all primary malignant kidney tumors are renal cell carcinomas. Up to 2% of renal cancers are hereditary
- Incidence is 6 to 12 per 100,000 per year in some studies
- Male:female ratio is 2:1
- Almost half of cases are discovered by chance during ultrasonography
- 5-year survival rate is more than 88% for localized tumors but less than 20% for metastatic disease
- Renal cell carcinoma may be characterized by hematuria, flank pain, and a palpable flank mass. However, this triad is not common in early disease
- Stauffer syndrome is characterized by liver function abnormalities in the setting of renal cell carcinoma without liver metastases
- Risk factors include the following:

  Tobacco use
  Severe obesity
  Other kidney disease
  Occupational exposure
  Thiazide medications
  Urinary tract infections

- Protective effects have been noted in women with moderate alcohol intake and oral contraceptive use

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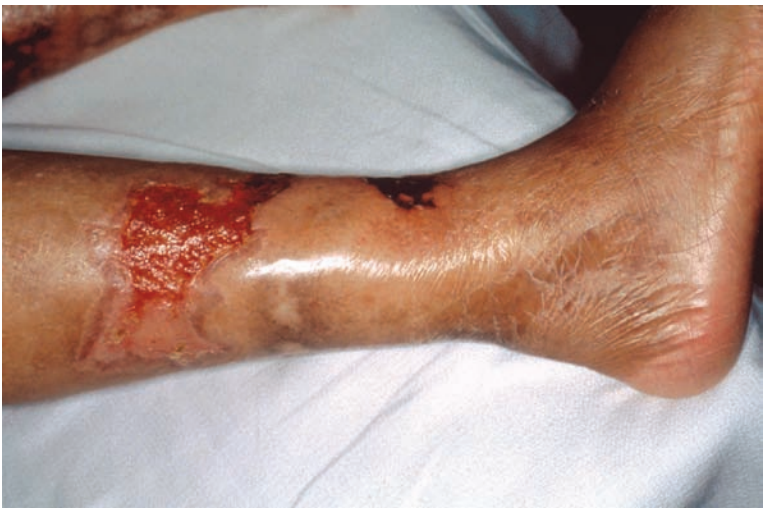
Dhôte R, Pellicer-coeuret M, Thiounn N, et al. Risk factors for adult renal cell carcinoma: a systematic review and implications for prevention. *BJU Int*. 2000;86:20-27.

Turner KJ. Inherited renal cancer. *BJU Int*. 2000;86:155-164.



Painful ulcers developed on the thighs, abdomen, and legs (shown here) of a 47-year-old obese woman with end-stage renal disease who was receiving dialysis. She has secondary hyperparathyroidism. What is the diagnosis?

a. Hypertensive ulcers
b. Venous stasis ulcers
c. Atheroemboli syndrome
d. Mixed cryoglobulinemia
e. Calciphylaxis

Calciphylaxis

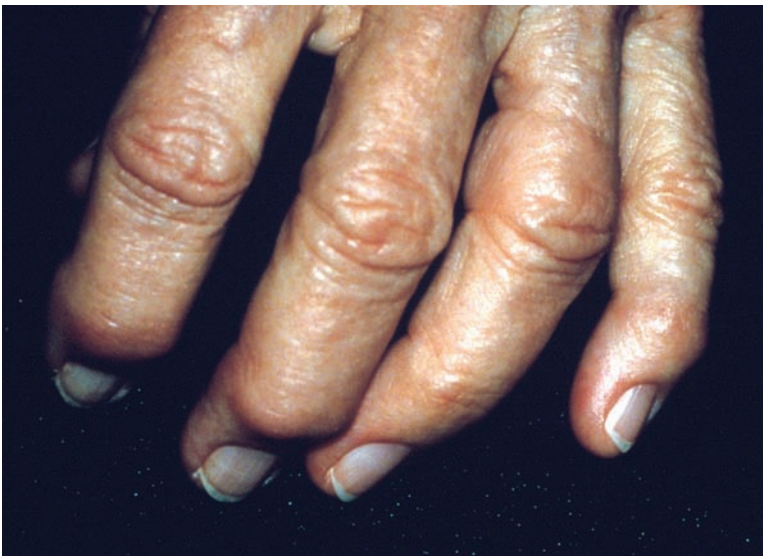
Answer: e

- Calcification of the media of small arteries and arterioles leads to ischemic necrosis and ulceration
- Initially the lesions appear violaceous. These often become hard, indurated, and ulcerated with overlying eschar as the subcutaneous fat becomes necrotic
- Lesions are very often extremely painful
- Increased calcium phosphate value is common at some point in the disease, but lesions may occur without either increased serum calcium or phosphate
- After lesions begin, serum calcium and phosphate values may actually decrease into the normal range
- Obese women with chronic renal failure and hypertension are the prototypic patients
- This condition develops in up to 4% of patients with chronic renal failure
- Parathyroidectomy and warfarin anticoagulation have not been uniformly successful for treatment
- Low-phosphate diet, phosphate binders, limitation of calcium and vitamin D intake, and weight loss may help
- Secondary sepsis is the unfortunate cause of the mortality (37%-77%) associated with this condition, despite therapy
- Exquisite skin care is warranted

References

Janigan DT, Hirsch DJ, Klassen GA, et al. Calcified subcutaneous arterioles with infarcts of the subcutis and skin ("calciphylaxis") in chronic renal failure. *Am J Kidney Dis*. 2000;35:588-597.

Kriskovich MD, Holman JM, Haller JR. Calciphylaxis: is there a role for parathyroidectomy? *Laryngoscope*. 2000;110:603-607.



In a 76-year-old woman, both hands have the changes shown here. The joints of her hands are painful. Which of the following is true concerning this condition?

a. 50% of people older than 60 years have this condition
b. Bouchard's nodes are exostoses of the distal interphalangeal joints
c. Heberden's nodes are exostoses of the proximal interphalangeal joints
d. Nonsteroidal anti-inflammatory drugs are the preferred treatment
e. The hand changes are not a result of normal wear and tear

Degenerative Joint Disease (Osteoarthritis)

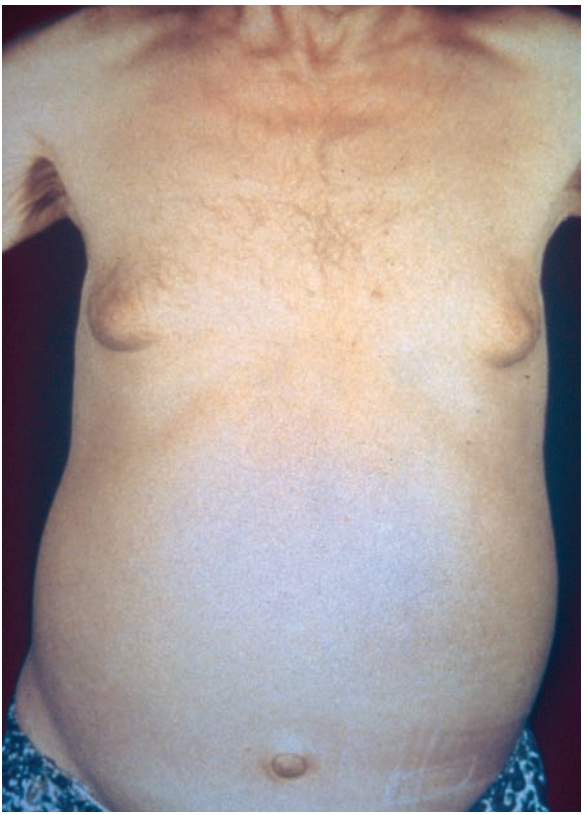
Answer: e

- Heberden's nodes: distal interphalangeal swelling, exostoses
- Bouchard's nodes: proximal interphalangeal swelling, exostoses
- 10% of people older than 60 years have degenerative joint disease
- Degenerative joint disease is not a result of normal wear and tear
- Mutations in collagen genes likely cause familial predisposition to osteoarthritis
- Pathologically, articular cartilage degenerates and new bone forms at joint margins (osteophytes)
- Commonly affected joints include the following:

  Distal interphalangeal joints
  Proximal interphalangeal joints
  First carpometacarpal joints
  Hips
  Knees
  Spine

- Deep aching improves with rest
- Acetaminophen is preferred over nonsteroidal anti-inflammatory agents for symptomatic treatment because it causes fewer gastrointestinal and renal side effects

References
Habermann TM. *Mayo Clinic Internal Medicine Board Review 2004-2005*. Philadelphia: Lippincott Williams & Wilkins; 2004:952-958.
Holderbaum D, Haqqi TM, Moskowitz RW. Genetics and osteoarthritis: exposing the iceberg. *Arthritis Rheum*. 1999;42:397-405.



A 70-year-old man who is an alcoholic is admitted to the hospital with diffuse abdominal pain and confusion. He has a history of hematemesis. If he continues to drink alcohol, what is his chance of 5-year survival?

a. 82%
b. 56%
c. 43%
d. 34%
e. 17%

Answer: d

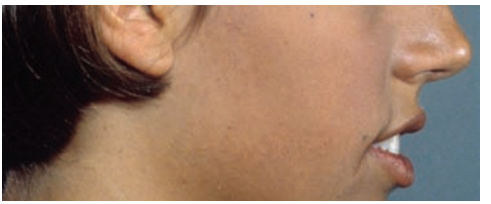
- History and physical examination evidence for portal hypertension due to cirrhosis of the liver include the following:

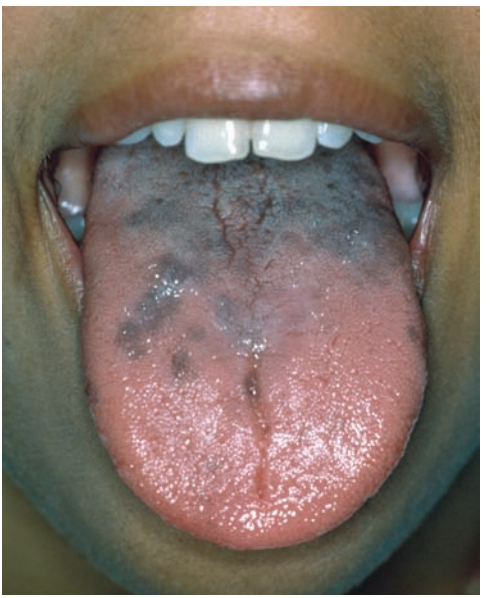
  Jaundice
  Hematemesis
  Ascites
  Gynecomastia
  Encephalopathy

- Diffuse abdominal pain in the presence of ascites suggests spontaneous bacterial peritonitis
- Women are more prone to alcoholic cirrhosis
- Liver enzymes may be normal
- Hepatitis C virus infection is common
- Liver transplantation is allowed if the patient maintains abstinence from alcohol
- The 5-year survival rate of patients with alcoholic cirrhosis *without* ascites, jaundice, or hematemesis *and* who abstain from alcohol is 89%
- The 5-year survival rate of patients who have alcoholic cirrhosis *with* ascites, jaundice, or hematemesis *and* who continue to drink alcohol is 34%

References

Habermann TM. *Mayo Clinic Internal Medicine Board Review 2004-2005*. Philadelphia: Lippincott Williams & Wilkins; 2004:302.
Prakash UBS. *Mayo Internal Medicine Board Review 2000-2001*. Philadelphia: Lippincott Williams & Wilkins; 2000:334.





An 18-year-old woman reports a 6-week history of fatigue, nausea, involuntary weight loss, and darkening of the skin. Which one of the following tests is the most helpful for establishing the diagnosis?

a. Adrenocorticotropic hormone (ACTH) stimulation test
b. Serum prolactin test
c. Serum potassium test
d. 24-Hour urine free cortisol test
e. Dexamethasone suppression test

Addison's Disease

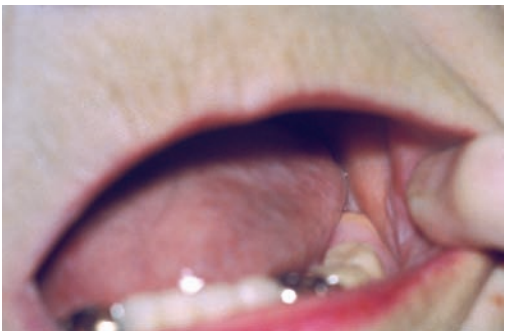
Answer: a

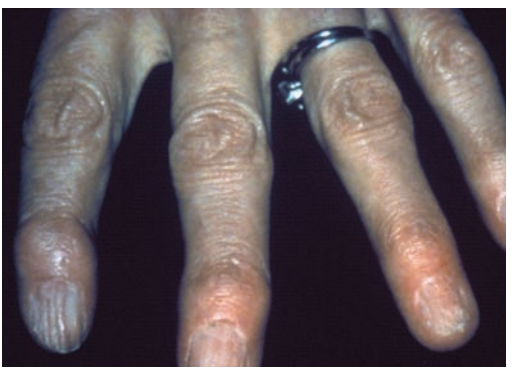
- Worldwide, tuberculosis is the most common cause of adrenocortical failure (Addison's disease)
- In the United States, autoimmune adrenalitis and bilateral adrenal hemorrhage are the most common causes
- Clinical presentation may include depression, fatigue, muscle weakness, anorexia, weight loss, nausea, vomiting, diarrhea, orthostatism
- Common laboratory manifestations include the following:

  Hyponatremia
  Hyperkalemia
  Fasting hypoglycemia
  Anemia
  Eosinophilia

- ACTH prohormone production is increased (propiomelanocortin), which results in increased melanocyte-stimulating hormone level and hyperpigmentation
- Hyperpigmentation may be most notable on the elbows, knees, and buccal mucosa and at surgical scars
- Addison's disease is diagnosed most reliably with the ACTH stimulation test
- Primary adrenocortical failure is treated with both glucocorticoids and mineralocorticoids. Glucocorticoid treatment should be augmented in acute illnesses

Reference
Habermann TM. *Mayo Clinic Internal Medicine Board Review 2004-2005*. Philadelphia: Lippincott Williams & Wilkins; 2004:219-220.





A woman with 3 children presents for a routine physical examination. She has no substantial past medical history. She is currently nursing her 13-month-old daughter and reports regular menstrual periods with normal flow. Review of systems is remarkable for curving and splitting of the nails and for craving ice chips. Which one of the following would you recommend?

a. Calcium carbonate
b. Folic acid
c. Iron sulfate
d. Vitamin B_{12}
e. Oral contraceptive pills

Iron Deficiency Anemia

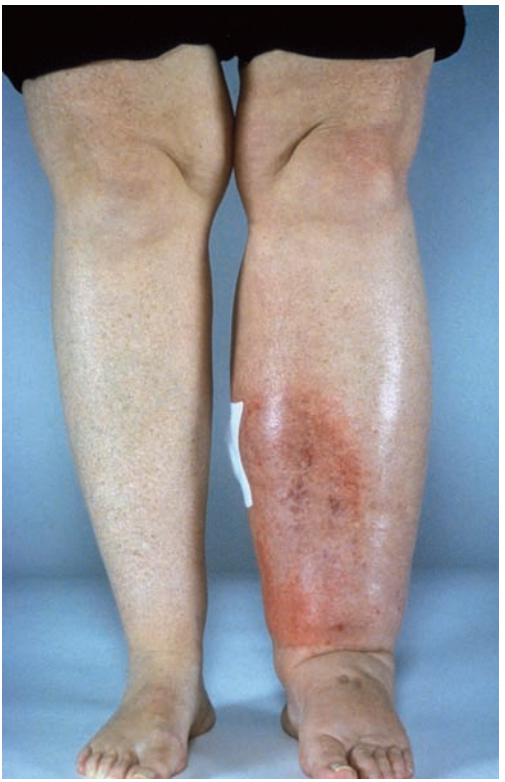
Answer: c

- Iron deficiency is the most common cause of anemia worldwide
- Fatigue and pallor are the most common symptoms and signs of iron deficiency anemia
- Pica is the compulsive desire to consume nonnutritive substances
- Pagophagia is the compulsive desire to eat ice
- Koilonychia, curving of the nails, occurs in 4% of patients with iron deficiency
- Nursing mothers have high iron requirements and are predisposed to iron deficiency
- The serum ferritin test is the most useful initial test for determining iron deficiency
- Iron sulfate 325 mg orally 3 times a day usually corrects anemia in 6 weeks and replenishes bone marrow reserves in 6 months
- Unless an obvious cause is present (e.g., menstruation), further evaluation for occult blood loss, especially gastrointestinal losses, may be warranted

Reference

Habermann TM. *Mayo Clinic Internal Medicine Board Review 2004-2005*. Philadelphia: Lippincott Williams & Wilkins; 2004:184, 411-413.

Next Hem case, p. 217



Chronic swelling developed in a 36-year-old man after recurrent infectious bouts of cellulitis. He currently does not have an infectious disease and ultrasonography is negative for deep venous thrombosis. Which of the following is most likely to be beneficial?

a. Antibiotics
b. Anticoagulants
c. Diuretics
d. Surgery
e. Compression stockings

Chronic Lymphedema

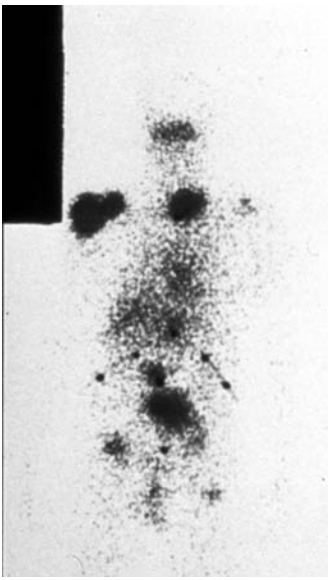
Answer: e

- Chronic lymphedema is caused by hypoplasia, dysfunction, or obstruction of lymphatics
- Primary forms are often familial (e.g., autosomal dominant Milroy's disease and Meige's disease) and often present after puberty with swelling in the feet or ankles
- Secondary forms are the most common
- In Western countries, cancer therapy (e.g., radiation or surgery) is the most common cause of lymphedema
- Worldwide, filarial infections are the most common cause
- Elephantiasis refers to characteristic skin changes:

  Thickening
  Enhanced skin creases
  Hyperkeratosis
  Papillomatosis in a swollen leg

- Lymphoscintigraphy is the best diagnostic test
- Treatment is with compression stockings, sequential compression pumps, or massage
- Prevention and prompt treatment of infection in limbs are very important
- Diuretics generally are not beneficial

References
Habermann TM. *Mayo Clinic Internal Medicine Board Review 2004-2005*. Philadelphia: Lippincott Williams & Wilkins; 2004:1026-1027.
Mortimer PS. Swollen lower limb-2: lymphoedema. *BMJ*. 2000;320:1527-1529.



A patient with follicular thyroid cancer has bone pain at various sites. Factors associated with a poor prognosis with this malignancy include all of the following *except*:

a. Advanced age
b. Female sex
c. Size of the primary tumor
d. Presence of distant metastases
e. Higher histologic grade

Follicular Thyroid Cancer

Answer: b

- About 20% of all thyroid cancers are follicular type
- Follicular thyroid cancer spreads preferentially by the hematogenous route and may rarely present with thyrotoxicosis
- Other types of thyroid cancers include the following:

  Papillary cancer: 50%-60% of all thyroid cancers, usually spreads to lymph nodes and has the best prognosis
  Anaplastic carcinoma: has the worst prognosis, typically presents in elderly individuals as a rapidly growing thyroid mass
  Medullary carcinoma: develops in childhood and usually begins as C-cell hyperplasia. It is also the most common manifestation of multiple endocrine neoplasia (type IIA)

- Poor prognostic factors include the following:

  Advanced age
  Male sex
  Higher histologic grade
  Size and invasiveness of tumor
  Presence of distant metastases

- Unlike follicular type, anaplastic and medullary thyroid carcinoma tissue do not readily pick up iodine 131
- Thyroid hormone therapy may be useful for tumor suppression

Reference
Habermann TM. *Mayo Clinic Internal Medicine Board Review 2004-2005*. Philadelphia: Lippincott Williams & Wilkins; 2004:212-213.

The arachnid shown here was removed from a 16-year-old girl with long black hair. Ascending flaccid paralysis had developed during the past 2 days.

1. When do you expect her symptoms to begin to resolve?

1a. 4-24 Hours
1b. 24-48 Hours
1c. 2-3 Days
1d. 3-7 Days
1e. Never. This condition is uniformly fatal

2. What is the most likely diagnosis?

2a. Guillain-Barré syndrome
2b. Tick fever
2c. Tick paralysis
2d. Myasthenia gravis
2e. Botulism

Answer 1: a

Answer 2: c

- Tick fever and tick paralysis are species-nonspecific reactions to tick bites
- Tick fever: not necessarily associated with tick paralysis. Manifests as fever, headache, nausea, and malaise. Resolves within 36 hours of tick removal
- Tick paralysis: rare condition caused by a toxin in the bite of a pregnant female tick

  First recognized in humans in the 19th century
  A common veterinary problem in Australia and South Africa
  Associated primarily with *Dermacentor andersoni* in the Pacific Northwest and Rocky Mountain regions of the United States, but 43 other species around the world are known to cause the disorder
  More common in children with dark hair (which hides the tick)
  After the tick has been attached from 4 to 7 days, progressive weakness develops over 2 days, which may progress to full ascending paralysis and areflexia
  Pupil reactions, sensorium, and sensation remain intact unless compounded by other illness
  Fever is rare
  The tick should be removed with blunt forceps at the head of the tick after application of petroleum jelly
  Symptoms usually begin to resolve within the first 24 hours of removal of the tick
  If undiagnosed, mortality rates are 10% to 12%
  Differential diagnosis includes Guillain-Barré syndrome, myasthenia gravis, botulism, porphyria, and transverse myelitis

References

Doan-Wiggins L. Tick-borne diseases. *Emerg Med Clin North Am*. 1991;9:303-325.
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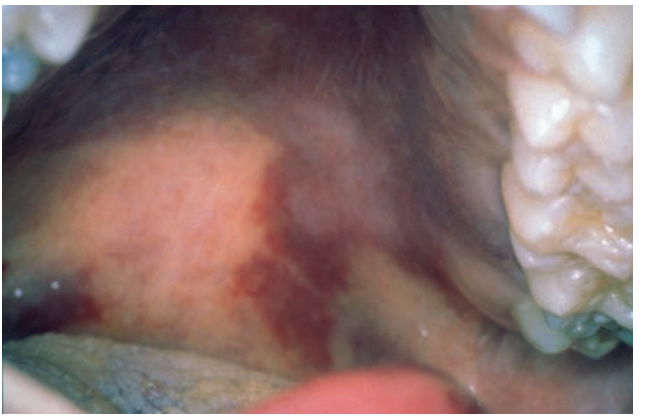

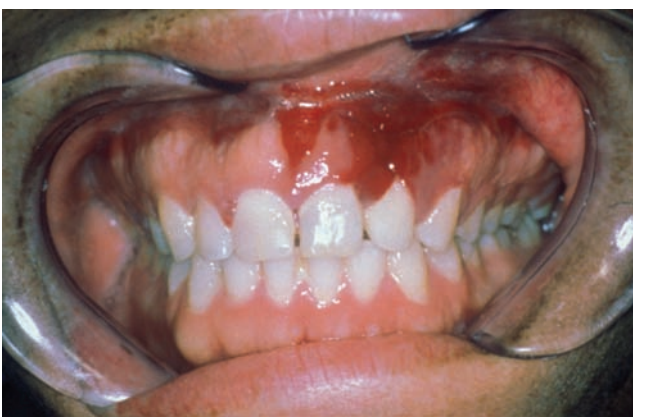

A 58-year-old man presents with dyspnea, cough, and the new onset of the oral lesions shown here. Which one of the following viruses is most likely present in this tissue?

a. Epstein-Barr virus
b. Human papillomavirus, types 16, 18
c. Varicella zoster virus
d. Human herpesvirus 8
e. Adenovirus

Answer: d

- Endemic (African) and classic Kaposi's sarcoma (KS) are the two major subtypes. Both are angioproliferative
- Endemic KS typically presents with plaques or papules of the trunk, extremities, face, and oral mucosa. More likely than classic KS to spread to lymphatics and viscera
- Classic KS typically involves the anterior tibial surface in an older patient population
- High prevalence in patients with human immunodeficiency virus (HIV) infection, especially in men who contracted HIV through anal sex with other men
- Human herpesvirus 8 has been consistently detected in all forms of KS lesions
- In HIV-infected patients, the presence of antibodies to this virus is predictive of future development of KS
- The most common symptoms of pulmonary KS are dyspnea and cough. In a series of 30 patients with acquired immunodeficiency syndrome (AIDS) who had pulmonary KS, 47% had chest pain
- Pulmonary KS, when it occurs, almost always follows mucocutaneous involvement
- Advanced pulmonary KS may be managed with cytotoxic agents such as vinca alkaloids, anthracyclines, bleomycin, and etoposide. Highly active antiretroviral therapy (HAART) also has been shown to be effective

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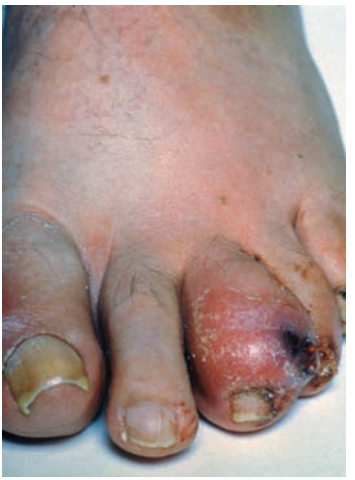

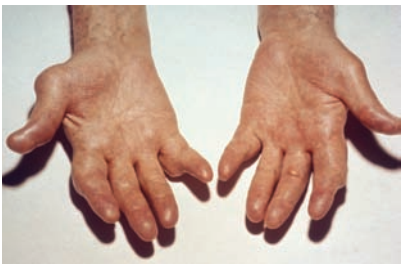

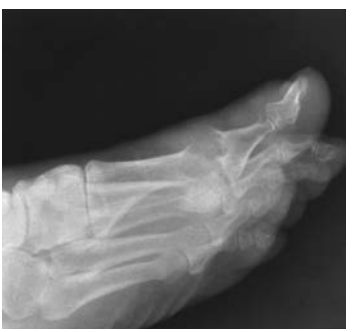

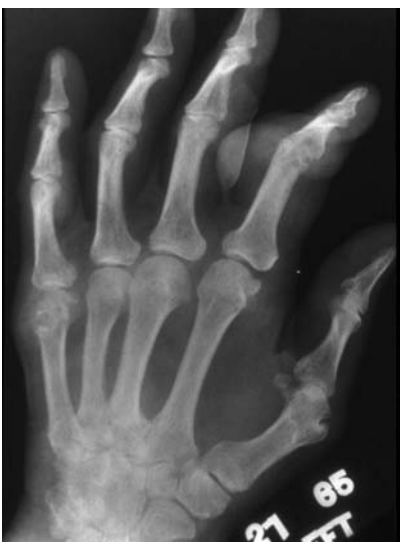

A 60-year-old man has an acutely painful third toe and the chronic hand findings shown here. All of the following treatments would be appropriate in this setting *except*:

a. Allopurinol
b. Colchicine
c. Indomethacin
d. Intra-articular corticosteroid injection
e. Ibuprofen

Answer: a

- Gout characteristically causes extreme pain with inflammation of a single joint in the lower extremity, but other joints can be involved
- Diagnosis: joint fluid aspiration shows negatively birefringent needle-shaped uric acid crystals under polarized light
- Nonsteroidal anti-inflammatory drugs (NSAIDs) are used for initial treatment. Colchicine is another option
- Intra-articular corticosteroids, intramuscular adrenocorticotropic hormone, and systemic corticosteroids can be used for patients unable to take NSAIDs
- Shifts in uric acid concentrations are more important than absolute levels for flare development
- Colchicine has been used for many years to prevent gout attacks and is effective in about 85% of patients, but it is not as popular now because of the side effect profile and the availability of other preventive options
- Allopurinol inhibits xanthine oxidase and is generally the prophylactic medication of choice
- Although allopurinol is effective for tophaceous gout in the intercritical period, it should not be used during an acute flare because the disease may worsen
- Uricosuric agents (probenecid, sulfinpyrazone, benzbromarone) occasionally are used for prophylaxis but less so than allopurinol because of multiple daily dosings, inhibition by salicylates, decreasing effectiveness with worsening renal function, and inappropriateness for use in patients who have had nephrolithiasis
- Allopurinol has none of the disadvantages listed above

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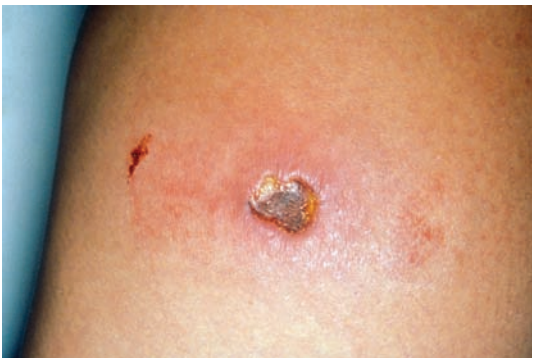

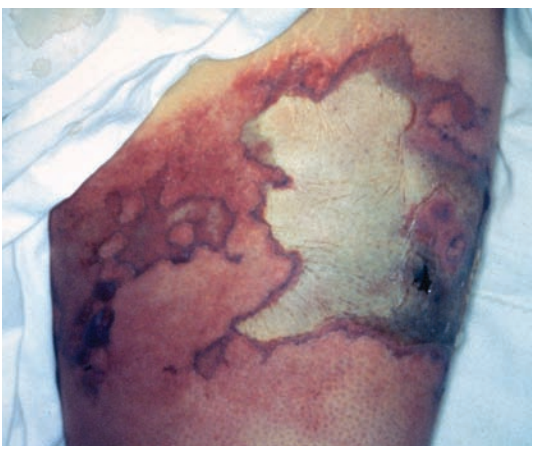

A patient felt a stinging sensation on his arm while dressing during an October morning. The area became painful and pruritic during the next few hours. During the next several days, hemorrhagic necrosis evolved. What is the likely cause?

a. Disseminated intravascular coagulation
b. Scorpion sting
c. Bee sting
d. Ecthyma gangrenosum
e. Spider bite

Brown Recluse (Loxosceles reclusa, *or Fiddle Spider) Bite*

Answer: e

- Brown recluse spiders live in dark areas and often enter homes during the fall months, hiding in, for example, clothing, closets, and storage rooms. Bites often occur during dressing
- Sphingomyelinase D2, hyaluronidase, and lipase within the venom produce dermonecrosis and chemotaxis of neutrophils
- Most cases resolve within 72 hours without treatment
- Severe cases may result in large areas of necrosis, requiring 3 months to 3 years to recover
- Acute intravascular hemolysis may result, but usually resolves within 1 week
- Treatment includes cleansing, cold compresses, elevation, immobilization of affected limbs, analgesics, antihistamines, and tetanus prophylaxis
- Dapsone administered within the first 72 hours may halt the progression of necrosis
- Immediate debridement is harmful but may be necessary after the acute inflammation is complete

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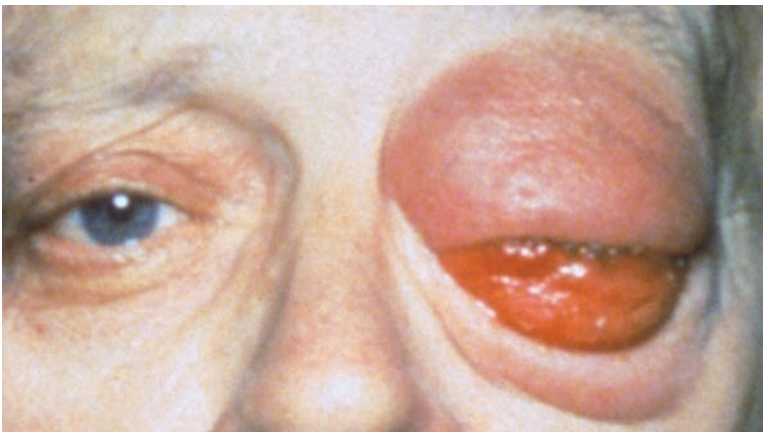

A 50-year-old man presents with the physical findings shown here and a subacute history of hematuria. This patient most likely has which one of the following?

a. Systemic lupus erythematosus
b. Scleroderma
c. Giant cell arteritis
d. Wegener's granulomatosis
e. Multiple sclerosis

Retro-orbital Pseudotumor in Wegener's Granulomatosis

Answer: d

- Wegener's granulomatosis most commonly is associated with granulomatous inflammation of the upper and lower respiratory tracts and renal biopsies consistent with focal segmental glomerulonephritis
- Cytoplasmic-staining antineutrophil cytoplasmic antibody (c-ANCA) is positive in more than 90% of active cases of Wegener's granulomatosis. Perinuclear-staining ANCA (p-ANCA) is found in less than 10% of patients with Wegener's granulomatosis
- Incidence is approximately 1 per 100,000 in the United States; male:female ratio is equal
- Ophthalmologic involvement is not uncommon in Wegener's granulomatosis
- Eye findings may include the following:

  Conjunctivitis
  Scleritis
  Retinal vasculitis
  Uveitis
  Retro-orbital pseudotumor

- Granulomatous involvement of the orbits may encase the optic nerve and is considered a late manifestation of the illness
- Orbital involvement by granulomatous masses may be managed initially with glucocorticoids. If unresponsive, other treatments include radiotherapy, retrobulbar alcohol injections, or surgical removal of the involved eye

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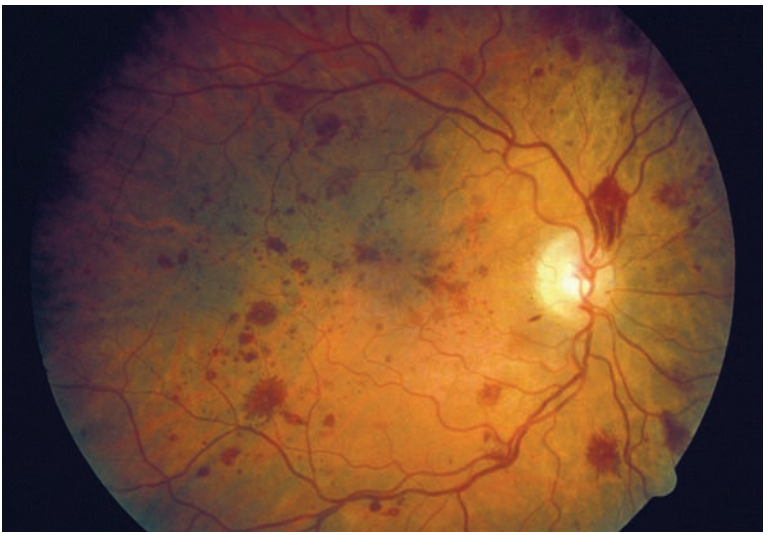

A 62-year-old man presents to your office with blurry vision, dyspnea, and occasional oral mucosal bleeding. Funduscopic examination shows tortuosity of the retinal veins. Which one of the following should most likely be in your differential diagnosis?

a. Diabetes mellitus
b. Non-Hodgkin's lymphoma
c. Acute pancreatitis
d. Waldenström's macroglobulinemia
e. Wilson's disease

Hyperviscosity Syndrome

Answer: d

- Hyperviscosity syndrome is characterized by fatigue, blurry vision, confusion, and mucosal membrane bleeding
- Symptoms usually do not arise until the blood viscosity is at least 4 times normal
- Funduscopic findings include characteristic "sausage-shaped" retinal veins (dilatation and segmentation of retinal veins) and papilledema
- In some cases, central retinal vein occlusion may occur, resulting in a "blood-and-thunder" appearance to the retina
- Increase in paraproteins (especially IgM and IgA) or in cellular constituents (leukocytes) can cause pathologic increases in serum viscosity
- Hyperviscosity syndrome may be present in the following:

  Waldenström's macroglobulinemia (IgM)
  Multiple myeloma (IgA)
  Hyperleukocytosis (as in acute lymphoblastic leukemia)
  Polycythemia

- Treatment of the underlying illness is indicated
- Plasmapheresis is effective therapy in Waldenström's macroglobulinemia and multiple myeloma
- Leukapheresis also may be used in cases of hematologic malignancy when chemotherapy is ineffective

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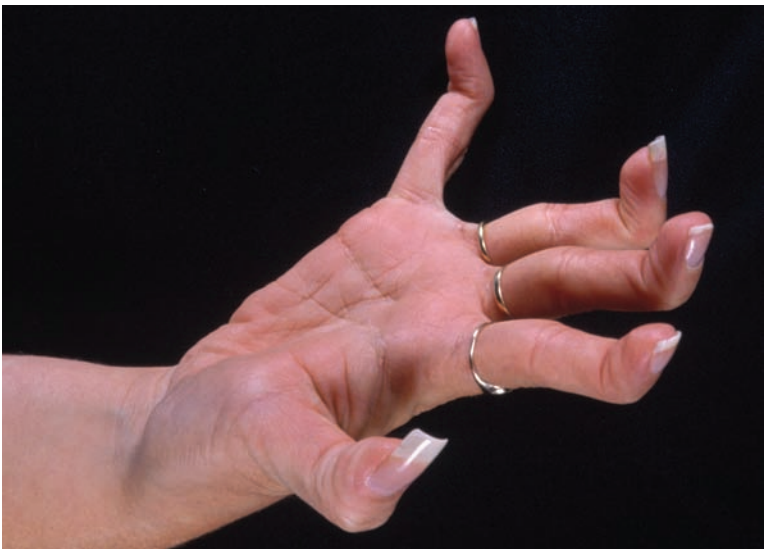

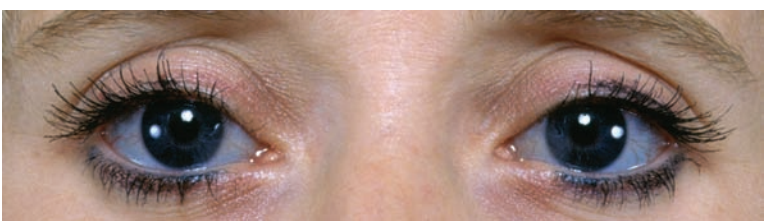

A 32-year-old woman presents to your office with complaints of hand pain. She has a history of multiple long-bone fractures, and previous evaluation with bone densitometry was consistent with osteoporosis. Which one of the following is the gene defect?

a. Type II collagen
b. Type I collagen
c. Type IV collagen
d. Fibrillin
e. Fibroblast growth factor

Osteogenesis Imperfecta

Answer: b

- Osteogenesis imperfecta is a congenital disorder also known as "brittle bone disease," characterized by defects in synthesis of type I collagen
- Of all the heritable disorders of connective tissue, osteogenesis imperfecta is the most common (1:20,000)
- Four subtypes have been described (I-IV). Type I is the most common and is also known as osteogenesis imperfecta tarda
- There is variable expressivity both within and between families. May have either autosomal dominant or autosomal recessive inheritance
- Clinical manifestations include the following:

  Multiple bone fractures
  Opalescent teeth
  Blue sclerae (shown in this case)
  Scoliosis
  Hearing loss
  Growth retardation
  Joint laxity

- Cardiac findings may include mitral valve prolapse and slightly increased aortic root diameter. The latter may progress to aortic regurgitation in 1% to 2% of patients. Chordae tendineae stretch also can occur and lead to considerable atrioventricular regurgitation

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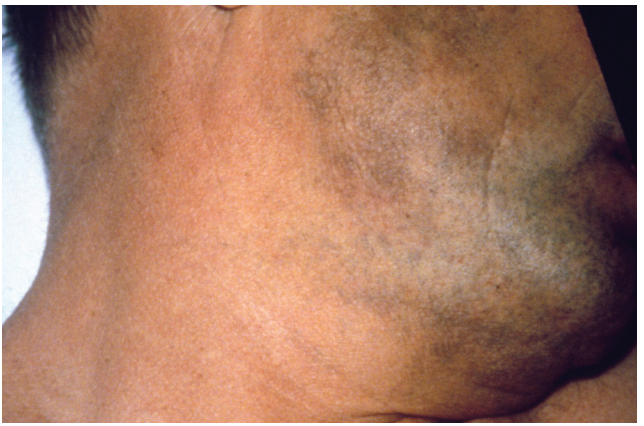

These lesions developed on sun-exposed skin of a 52-year-old man with atrial fibrillation. Biopsy shows yellow-brown granules in macrophages. Which of the following drugs is likely being used to treat his atrial fibrillation and is causing these findings?

a. Amiodarone
b. Bisoprolol
c. Coumadin
d. Diltiazem
e. Esmolol

Amiodarone Skin Reaction

Answer: a

- Adverse effects of amiodarone may affect many patients and various organs, such as the following:

  Skin (up to 57%)
  Liver (up to 55%)
  Lungs (up to 13%, including pulmonary fibrosis and interstitial pneumonitis)
  Thyroid (up to 11%)
  Corneal microdeposits
  Central nervous system abnormalities

- Skin effects include brown, blue-gray discoloration and photosensitivity
- Amiodarone and metabolites are concentrated 500 times more in fatty tissues than in serum
- Amiodarone association with intralysosomal lipids is thought to account for the yellow-brown granules seen in macrophages on biopsy
- The phototoxic reaction manifests as exaggerated burns in sun-exposed areas
- The skin reactions may be related to both dose and duration of therapy. Resolution of discoloration has been reported with lowering the dose of amiodarone
- Younger patients (<60 years) are at higher risk for amiodarone-related skin reaction

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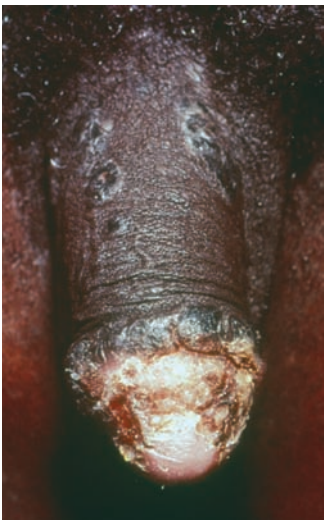

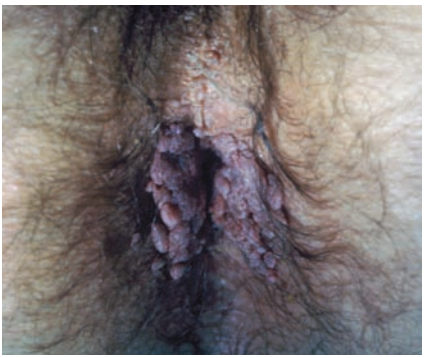

The lesions shown are recurrent. All of the following are correct regarding this condition *except*:

a. The causative agent is human papillomavirus
b. Without treatment, the lesions will not resolve
c. The presence of these lesions increases the risk of cervical carcinoma in women
d. Recurrences after treatment are common
e. Transmission is predominantly sexual

Condyloma Acuminata (Genital Warts)

Answer: b

- Condyloma acuminata are caused by human papillomavirus (HPV)
- Transmission is predominantly sexual
- Pointed, cauliflower-like lesions are typical on moist surfaces
- Thickened, keratotic lesions appear on drier surfaces, such as the penile shaft
- HPV types 6 and 11 are more commonly associated with visible warts and laryngeal papillomas
- HPV types 16 and 18 (among others) are more frequently associated with cervical carcinoma
- Untreated lesions typically resolve spontaneously but often recur
- Latent, subclinical infection may persist
- The disease is extremely common: 1% of sexually active U.S. adults have visible lesions; 15%-20% have latent infection
- Treatments include cryosurgery, topical fluorouracil, electrodessication, and topical podophyllin preparations. None are curative
- Recurrences after treatment are common

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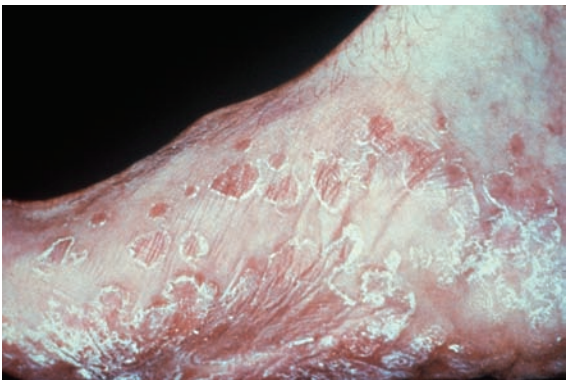

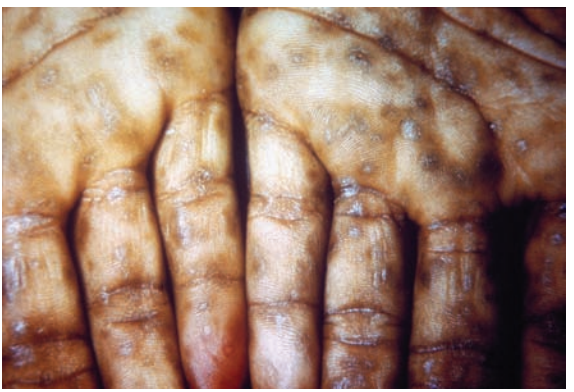

A 27-year-old man presents with the skin lesions shown here, fever, and lymphadenopathy. Six weeks previously he had a painless penile ulcer that resolved spontaneously without treatment. After appropriate therapy, what should be followed as a marker of treatment success?

a. Chest radiograph
b. Serologic results
c. Resolution of skin lesions
d. Resolution of fever
e. Resolution of lymphadenopathy

Secondary Syphilis

Answer: b

- Symptoms occur with hematogenous spread of *Treponema pallidum*
- Symptoms include the following:

  Constitutional findings
  Rash
  Mucocutaneous lesions (classically on the palms and soles)
  Alopecia
  Condylomata lata
  Lymphadenopathy

- Onset is usually 2 to 8 weeks after the chancre appears
- Skin lesions are highly infectious
- The clinical manifestations resolve spontaneously without treatment, but the patient remains infected with latent syphilis that may remain asymptomatic for years
- Fluorescent treponemal antibody absorption (FTA-ABS) test is positive in 100%, and VDRL test is positive in 99% of cases of secondary syphilis
- Treatment of choice is benzathine penicillin 2.4 million units intramuscularly
- Follow serologic results for a decrease in titers as evidence of treatment success

Reference

Habermann TM. *Mayo Clinic Internal Medicine Board Review 2004-2005*. Philadelphia: Lippincott Williams & Wilkins; 2004:592-593.

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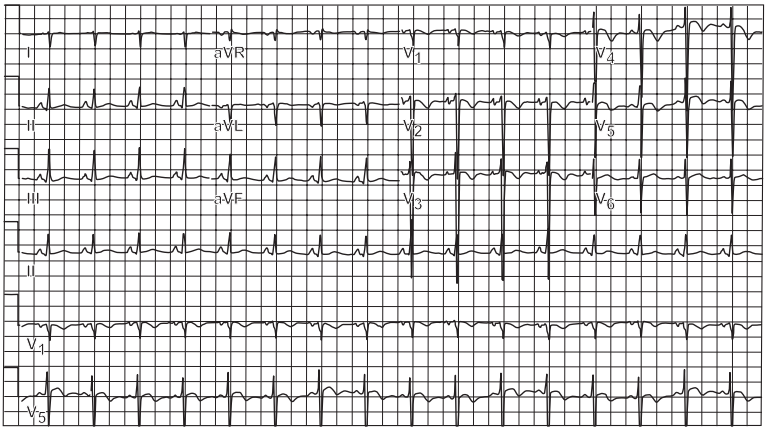

An asymptomatic 33-year-old woman comes to your office with the electrocardiogram shown here. Which of the following may explain the findings?

a. Subclinical pericarditis
b. Ventricular aneurysm
c. Vertically displaced heart
d. Left ventricular hypertrophy
e. Bicuspid aortic valve

Right Axis Deviation

Answer: c

- Electrocardiographic features include a negative QRS deflection in lead I and positive QRS deflection in lead aVF with a resultant QRS axis of more than 100°
- Conditions associated with right axis deviation include the following:

  Lateral wall acute myocardial infarction
  Pulmonary embolism
  Pulmonary hypertension
  Pulmonic stenosis
  Right ventricular hypertrophy
  Right bundle branch block
  Left posterior fascicular block
  Dextrocardia
  Vertically displaced heart (a normal variant)

- Isolated hereditary patterns of right axis deviation within families also have been described

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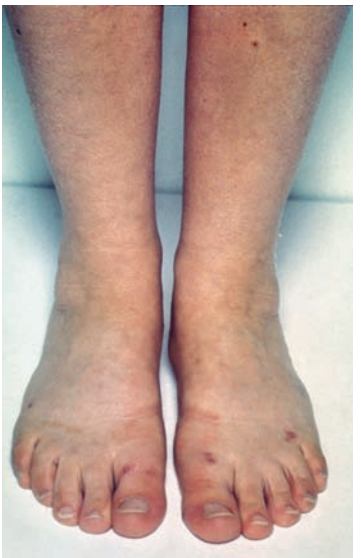

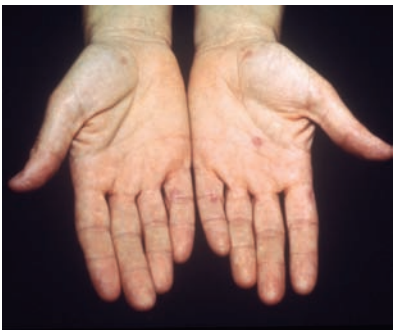

A 25-year-old menstruating woman with a painful right knee has the skin lesions shown here. Which type of culture is most likely to be positive?

a. Blood
b. Synovial fluid
c. Pharyngeal
d. Skin lesion
e. Cervical

Disseminated Gonococcal Infection

Answer: e

- Disseminated gonococcal infection occurs in 1% to 3% of gonococcus-infected patients. It is 3 to 5 times more common in women than men. Menstruation may predispose to disseminated gonococcal infection
- It is the most common form of infectious arthritis in community and teaching hospitals
- Bacteremic phase often manifests as tenosynovitis around the wrists and ankles, skin lesions (usually <30), and polyarthralgias
- Nonbacteremic phase occurs about 1 week later with monoarticular arthritis (knee, wrist, ankle)
- Most patients have no fever or chills (unlike other types of infectious arthritis)
- Blood and synovial fluid cultures are positive in 15% to 30% of patients and are mutually exclusive (i.e., one or the other may be positive, but they rarely are both positive)
- Pharyngeal and rectal cultures are positive in 10% to 20% of patients. However, cervical cultures are positive in 80% to 90% of women with disseminated gonococcus and urethral cultures are positive in 50% to 60% of men with disseminated gonococcus
- Treatment is with ceftriaxone or penicillin G if susceptible. Treatment with doxycycline or azithromycin is also included for *Chlamydia trachomatis*, which often coexists with *Neisseria gonorrhoeae*

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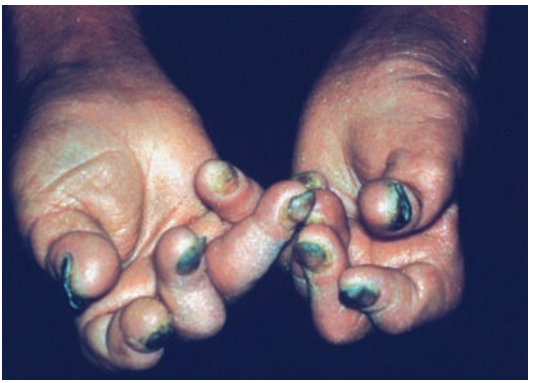

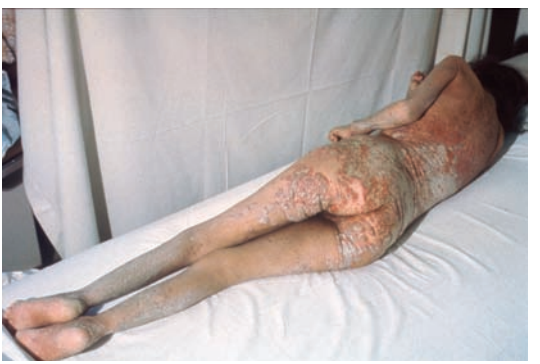

A 33-year-old woman has the skin lesions shown here. All of the following statements are true with regard to this condition *except*:

a. "Sausage" appearance to fingers is characteristic
b. The incidence of uveitis is high
c. The complication shown in the top figure develops in less than 7% of patients with this condition
d. Involvement of distal interphalangeal joints is common
e. Treatment includes nonsteroidal anti-inflammatory drugs, methotrexate, and azathioprine

Psoriatic Arthritis (Arthritis Mutilans)

Answer: b

- Between 1% and 2% of the U.S. population has psoriasis
- Onset of psoriasis is usually in the third decade
- A third of patients have a family history of psoriasis
- Arthritis develops in less than 7% of patients with psoriasis
- Severe skin involvement is associated with a higher risk of arthritis
- Nail pitting is associated with increased incidence of joint disease
- "Sausage" digits and involvement of the distal interphalangeal joints are characteristic
- Five clinical groups:

  Asymmetric oligoarthritis (70% of patients with psoriatic arthritis)
  Symmetric polyarthritis (rheumatoid factor–negative) (15%)
  Arthritis mutilans (5%)
  Distal interphalangeal joint–predominant (5%)
  Psoriatic spondylitis (often HLA-B27–positive) (5%)

- Treatment of psoriatic arthritis is with nonsteroidal anti-inflammatory drugs, methotrexate, or azathioprine

References

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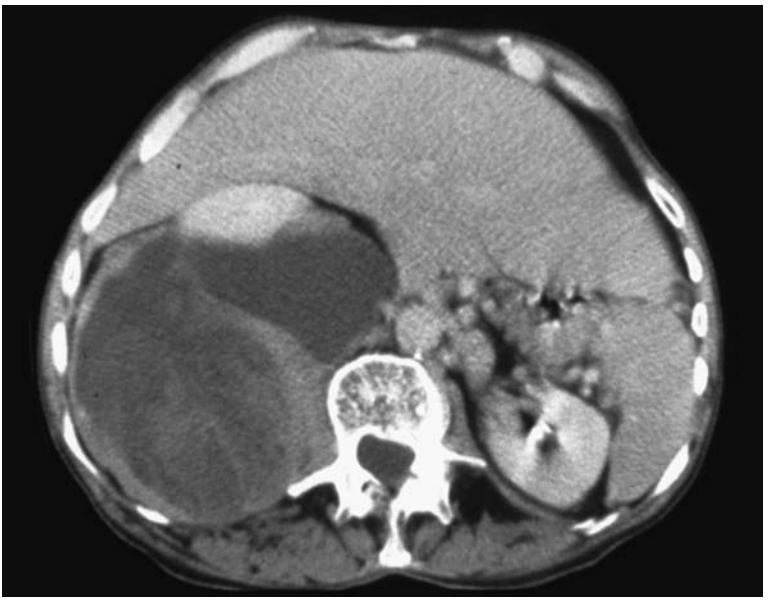
McGonagle D, Conaghan PG, Emery P. Psoriatic arthritis: a unified concept twenty years on. *Arthritis Rheum*. 1999;42:1080-1086.



A 51-year-old man presents with abdominal pain in the right lower quadrant and confusion. His confusion resolves with intravenous glucose. A large right-sided retroperitoneal mass is seen on computed tomography. After 17 hours of fasting, confusion again develops. His plasma glucose concentration was 23 mg/dL, and his confusion resolved with glucose infusion. What causes his hypoglycemia?

a. Excess tumor-produced insulin
b. Metabolic demand of the tumor
c. Insulin-like growth factor I
d. Insulin-like growth factor II
e. Glucagon deficiency

Answer: d

- Whipple's triad:

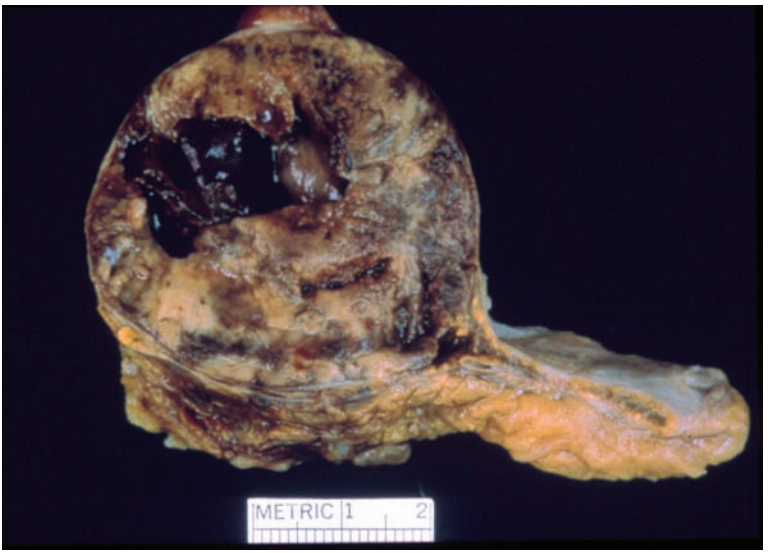
  Symptoms of hypoglycemia
  Low plasma glucose documented simultaneously with the symptoms of hypoglycemia
  Prompt resolution of symptoms when plasma glucose level is normalized

- Hypoglycemia may be insulin-mediated (e.g., insulinoma, exogenous insulin, sulfonylureas, autoimmune hypoglycemia) or non–insulin-mediated (e.g., alcohol, cortisol deficiency, renal failure, liver failure, sepsis, insulin-like growth factor II–secreting tumors)
- Extrapancreatic tumors that cause hypoglycemia are rare; they include those of mesodermal, epithelial, and hematopoietic origin
- These tumors are usually large and slow-growing and can be benign or malignant
- Hypoglycemia in affected patients is thought to be caused by insulin-like growth factor II, a polypeptide with 50% homology with pro-insulin and insulin-like activity
- During episodes of hypoglycemia, patients have suppressed plasma levels of insulin

References

Habermann TM. *Mayo Clinic Internal Medicine Board Review 2004-2005*. Philadelphia: Lippincott Williams & Wilkins; 2004:239-240.

Strauss G, Christensen L, Zapf J. Tumour-induced hypoglycaemia due to 'big' IGF-II. *J Intern Med*. 1994;236:97-99.



A 70-year-old woman presents after resection of a tumor, shown here, from her adrenal gland. Clinical presentations of her illness include all of the following *except*:

a. Anhidrosis
b. Hypertension
c. Weight loss
d. Headaches
e. Palpitations

Pheochromocytoma

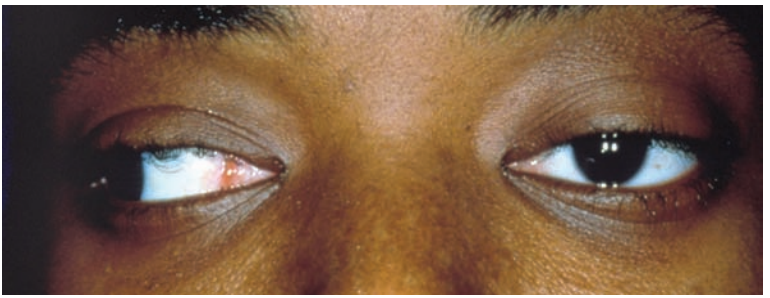
Answer: a

- The clinical presentation of pheochromocytoma may include headaches, palpitations, hypertension, sweating, and weight loss
- Pheochromocytoma causes less than 0.3% of all cases of hypertension but must be aggressively evaluated if it is suspected because progression to cardiovascular collapse and cerebrovascular accidents is not uncommon
- Most common location is adrenal medulla; other locations include along the sympathetic chain in the abdomen, chest, and neck
- Increased values of plasma and urinary catecholamines and their metabolites are suggestive of pheochromocytomas
- The tumors may be radiologically localized with computed tomography or magnetic resonance imaging. ^{123}I-metaiodobenzylguanidine (MIBG) scintigraphy is an additional method
- α-Adrenergic blockade is the mainstay of therapy. Phentolamine is appropriate in hypertensive emergencies. Surgical removal is curative
- β-Adrenergic blockade without α-adrenergic blockade risks hypertensive crisis because of unopposed α activity
- Pheochromocytoma may be a component of multiple endocrine neoplasia type IIA (pheochromocytoma, medullary thyroid carcinoma, primary hyperparathyroidism) or type IIB (pheochromocytoma, medullary thyroid carcinoma, mucosal neuromas, marfanoid habitus)

References

Habermann TM. *Mayo Clinic Internal Medicine Board Review 2004-2005*. Philadelphia: Lippincott Williams & Wilkins; 2004:223-224.

Witteles RM, Kaplan EL, Roizen MF. Sensitivity of diagnostic and localization tests for pheochromocytoma in clinical practice. *Arch Intern Med*. 2000;160:2521-2524.



The 28-year-old man shown here is attempting to gaze to the right. This nerve palsy is associated with all of the following *except*:

a. Diabetic ischemic neuropathy
b. Postoperative complication from vertebrobasilar aneurysm surgery
c. Compressive effects of intracranial aneurysms
d. Mass effect from intracranial lymphoma
e. Paget's disease

Third Nerve Palsy

Answer: e

- Most common reason for third nerve (oculomotor) palsy is ischemic diabetic neuropathy
- Other associated findings include the following:

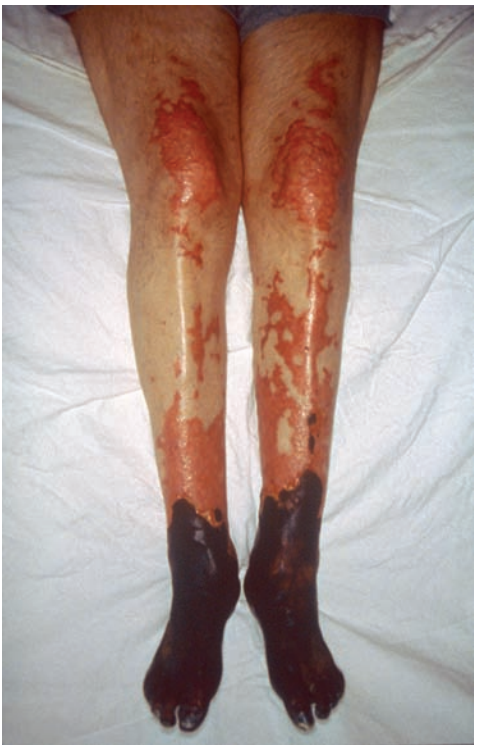
  Ptosis of the involved eye
  Lack of upward ocular movement
  Lack of inward ocular movement
  Lack of downward ocular movement

- On physical examination, the size of the pupil on the involved eye can assist in determining the cause for third nerve compromise
- Extrinsic compression of the third nerve (e.g., by an intracranial aneurysm) usually causes dilatation of the involved pupil and loss of light reactivity
- Ischemic damage to the third nerve (e.g., diabetes) generally does not result in such alterations in the pupil because the iris sphincter is unaffected by ischemic injury

References

Jacobson DM. Pupil involvement in patients with diabetes-associated oculomotor nerve palsy. *Arch Ophthalmol*. 1998;116:723-727.

Jacobson DM, Broste SK. Early progression of ophthalmoplegia in patients with ischemic oculomotor nerve palsies. *Arch Ophthalmol*. 1995;113:1535-1537.



A 63-year-old man with a history of hypertension and diabetes required a stay in the intensive care unit with vasopressors for severe pneumonia and disseminated intravascular coagulopathy. Many years ago, he had frostbite injury to his lower extremities. Which of the following is *not* a risk factor for the condition shown here?

a. Hypertension
b. Diabetes
c. Vasopressor therapy
d. Disseminated intravascular coagulopathy
e. History of frostbite injury

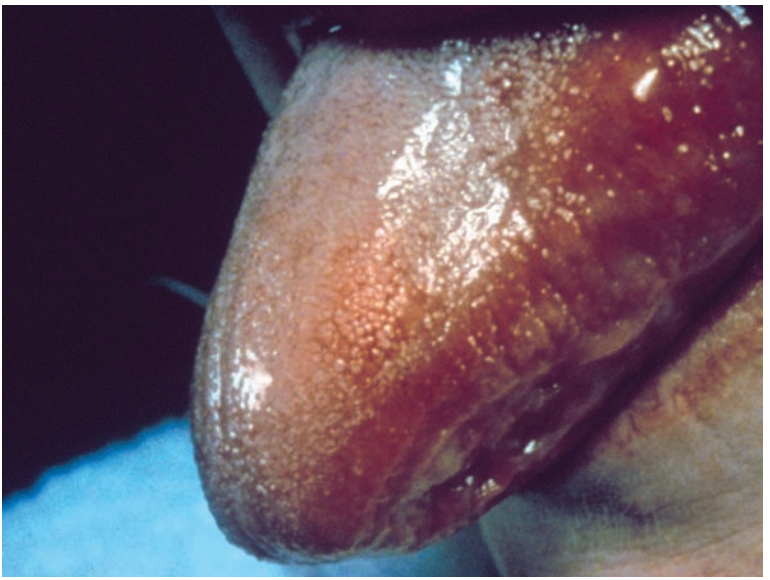
Symmetric Peripheral Gangrene

Answer: a

- The condition was first described in 1891 as symmetric distal ischemic damage without large-vessel obstruction
- "Gangrene" is from the Greek *gangraina*, meaning "an eating sore"
- Symmetric peripheral gangrene (SPG) is most often associated with infections that result in cardiogenic or hypovolemic shock
- SPG is now noted to have occurred in patients who have required vasopressor therapy
- SPG often involves acral distributions, including the distal extremities, tip of the nose, ears, and genitalia
- Risk factors include sepsis, disseminated intravascular coagulopathy, history of frostbite, scleroderma, polymyalgia rheumatica, Raynaud's phenomenon, renal insufficiency, splenectomy, diabetes, immunosuppression, alcoholism, systemic malignancy
- SPG should be suspected if symmetric acral areas become cold, pale, cyanosed, or painful
- Use of vasopressors should be discontinued, if possible, and any underlying disorders treated aggressively. If the patient has disseminated intravascular coagulopathy, heparinization is recommended
- Progression may lead to dry gangrene, which is often shriveled (as shown in this case)
- Amputation is necessary for treatment of extensive involvement

Reference

Knight TT Jr, Gordon SV, Canady J, et al. Symmetrical peripheral gangrene: a new presentation of an old disease. *Am Surg*. 2000;66:196-199.



A 51-year-old man presents with a 3-month history of loose, watery stools and dyspnea on exertion. Laboratory studies show renal insufficiency, and cardiomegaly is found on chest radiography. His tongue is huge. What is the diagnosis?

a. Whipple's disease
b. Primary systemic amyloidosis
c. Gaucher's disease
d. Hemochromatosis
e. Type 1 diabetes mellitus

Primary Systemic Amyloidosis With Gastrointestinal Involvement

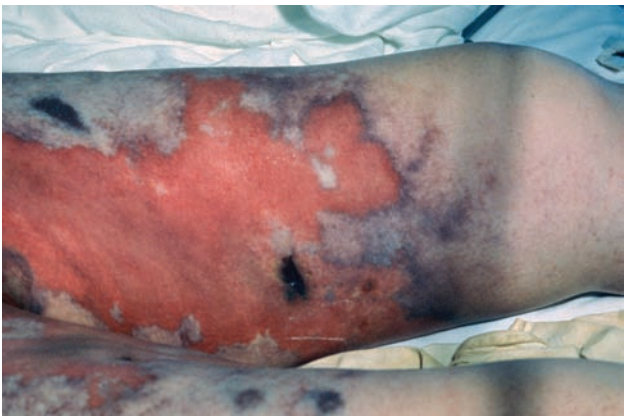
Answer: b

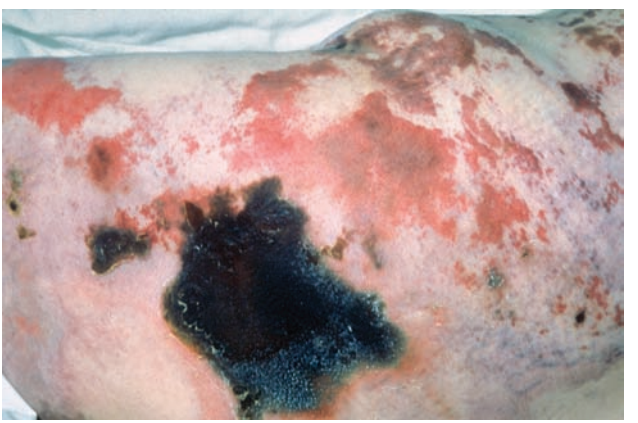
- Macroglossia is a well-described manifestation of primary systemic amyloidosis
- Primary systemic amyloidosis is a multisystem disease that also may present with the following:

  Fatigue
  Weight loss
  Hepatomegaly
  Renal insufficiency (most common presentation)
  Proteinuria
  Nephrotic syndrome
  Congestive heart failure (second most common, 25% of patients)
  Orthostatic hypotension
  Carpal tunnel syndrome (20% of patients)
  Peripheral neuropathy
  Posttraumatic purpura (e.g., postproctoscopic periorbital purpura)

- Amyloid involvement of the gastrointestinal tract may cause malabsorption and intestinal dysmotility, the latter of which may cause diarrhea, constipation, megacolon, and fecal incontinence
- Congo red staining of an aspirated sample of subcutaneous fat shows amyloid fibrils in 90% to 95% of patients, whereas rectal biopsy shows amyloid in 70% to 85% of patients

References
Gertz MA, Kyle RA. Primary systemic amyloidosis: a diagnostic primer. *Mayo Clin Proc*. 1989;64:1505-1519.
Habermann TM. *Mayo Clinic Internal Medicine Board Review 2004-2005*. Philadelphia: Lippincott Williams & Wilkins; 2004:273-274, 435-436.





In a 63-year-old woman with gram-negative sepsis, blood began oozing around her vascular access sites and the lesions shown here developed. What is the treatment priority?

a. Continuous heparin infusion
b. Administration of cryoprecipitate
c. Administration of fresh frozen plasma
d. Platelet transfusion
e. Treatment of the gram-negative sepsis

Disseminated Intravascular Coagulopathy

Answer: e

- Disseminated intravascular coagulopathy is a dynamic process of microvascular clotting leading to consumption of coagulation factors and subsequent bleeding
- Diagnosis depends on clinical setting, but screening test results include the following:

  Decreased platelets (90%)
  Increased international normalized ratio (90%)
  Decreased fibrinogen (70%)

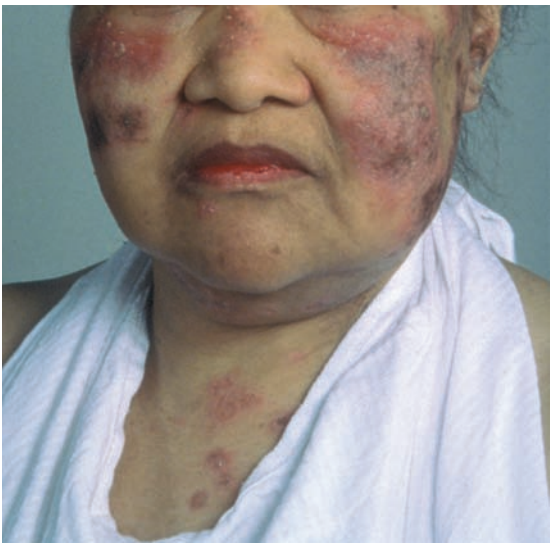
- Confirmatory tests include D-dimer (a measure of fibrin degradation) and soluble fibrin monomers (a measure of coagulation activation)
- Most common causes are, in decreasing order, the following:

  Malignancies
  Infections
  Trauma

- Others include liver disease, pregnancy, retained fetus, transfusion reactions, burns, and acute renal failure
- The underlying disease should be treated
- If life-threatening bleeding persists, deficiencies are treated with cryoprecipitate, fresh frozen plasma, and platelets
- Heparin is used for severe refractory cases, but it is contraindicated with central nervous system lesions

Reference

Habermann TM. *Mayo Clinic Internal Medicine Board Review 2004-2005*. Philadelphia: Lippincott Williams & Wilkins; 2004:446-447.



A 42-year-old woman has the rash shown here, photosensitivity, pleuritis, and lymphopenia. Drug-induced forms of this disease have a 95% association with which of the following?

a. Increased frequency of central nervous system and renal involvement
b. Antihistone antibody
c. Anticentromere antibody
d. Hepatotoxicity
e. Positive extractable nuclear ribonucleoprotein antigen

Answer: b

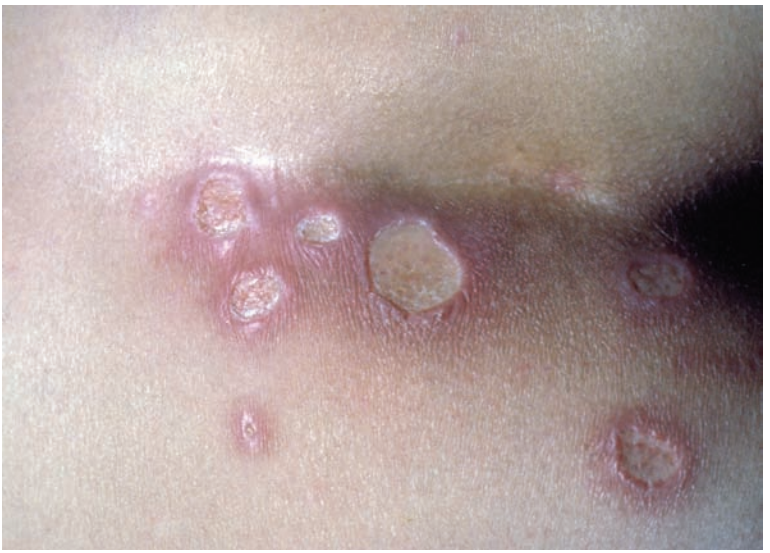
- Systemic lupus erythematosus is a relapsing and remitting disease with a wide variety of clinical manifestations
- At least 4 of the following are needed for diagnosis:

  Immunologic (e.g., anti-dsDNA, anti-Sm [Smith], false-positive VDRL, positive antiphospholipid antibodies)
  Malar rash
  Antinuclear antibody (ANA)-positive (95%)
  Neurologic change (e.g., psychosis, seizures, depression, anxiety)
  Oral ulcers (usually painless)
  Photosensitivity
  Discoid lupus
  Renal disease (e.g., proteinuria, casts, glomerulonephritis)
  Arthritis (usually nonerosive, reducible deformities)
  Serositis (e.g., pleuritis, pericarditis)
  Hematologic abnormalities (e.g., anemia, thrombocytopenia, leukopenia)
- Antihistone antibody is associated with drug-induced lupus (procainamide and hydralazine are common associations, many others probably are associated)
- There is an increased frequency among African Americans, Native Americans, and Asian Americans, but it is unusual among African blacks
- Treatment is based on clinical manifestations and severity

Reference

Habermann TM. *Mayo Clinic Internal Medicine Board Review 2004-2005*. Philadelphia: Lippincott Williams & Wilkins; 2004:988-993.

Next Rheum case, p. 255



An 80-year-old nursing home resident has been immobilized because of a hip fracture. The lesions shown here were noted in the sacral region on a recent hospital admission. What is the most important risk factor for development of these ulcers?

a. Age-related skin change
b. Nutritional deficiency
c. Urinary incontinence
d. Immobility
e. Shearing (i.e., dragging instead of lifting the patient)

Answer: d

- Risk factors for pressure ulceration:

  Immobility (most important)
  Age-related skin changes
  Nutritional deficiencies
  Urinary incontinence

- Pressure ulcers are promoted by the following:

  Pressure
  Shearing (i.e., dragging instead of lifting)
  Friction
  Moisture

- Persistent pressure that is greater than capillary pressure which lasts for 2 hours can cause ischemia and resultant ulceration. Therefore, immobilized patients should be turned at least every 2 hours

- The most common sites of pressure ulcers are the sacrum, thighs, heels, and lateral malleoli

- Treatment involves the following:

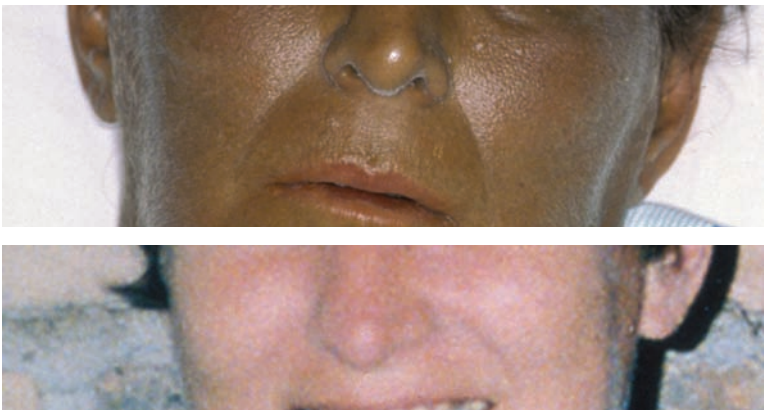
  Removing the pressure source (e.g., with a specialized bed or extremity protective devices)
  Debridement of devitalized tissue
  Cleansing initially and at each dressing change
  Prevention and treatment of any secondary infections
  Appropriate wound dressing that ideally keeps the ulcer tissue moist but the surrounding intact skin dry

References

Cervo FA, Cruz AC, Posillico JA. Pressure ulcers: analysis of guidelines for treatment and management. *Geriatrics*. 2000;55:55-60.

Goodfield M. Optimal management of chronic leg ulcers in the elderly. *Drugs Aging*. 1997;10:341-348.

Habermann TM. *Mayo Clinic Internal Medicine Board Review 2004-2005*. Philadelphia: Lippincott Williams & Wilkins; 2004:398-399.



A 45-year-old woman was admitted with cardiogenic shock and was supported with an intra-aortic balloon pump and left ventricular assist device. She experienced considerable intravascular hemolysis, requiring more than 180 units of packed red blood cells. You are called for a dermatology consultation. The lower photograph was taken after your advised treatment. What was the most likely treatment?

a. Systemic corticosteroids
b. Plasma exchange
c. Hemodialysis
d. Chelation therapy
e. Interferon alfa

Secondary Hemochromatosis

Answer: d

- Hemochromatosis can be either primary or secondary
- Primary hereditary hemochromatosis results from overabsorption of iron from gastrointestinal tract and has been linked to a genetic defect on the short arm of chromosome 6
- Frequency of the gene mutation in Western populations has been estimated to be 1 in 300 persons (includes carriers)
- The classic clinical triad is diabetes, bronze skin, and hepatic cirrhosis. Early clinical signs and symptoms include arthralgias, arthritis, fatigue, and abnormal results of liver function tests
- Determination of the transferrin saturation percentage is a reasonable screening test for this disorder. A transferrin saturation percentage more than 50% identifies approximately 98% of affected persons
- Definitive testing includes liver biopsy for stainable iron, which is markedly concentrated in the periportal hepatocytes
- Secondary: various underlying causes

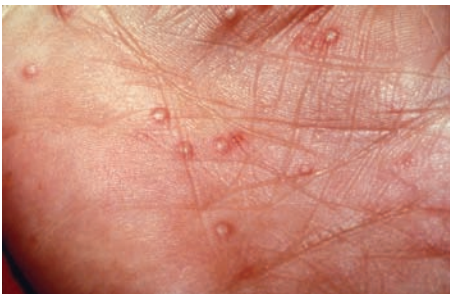
  Iron overload in the setting of chronic anemia
  Iron overload in the setting of cirrhosis
  Multiple transfusions (as in this case)
  Sideroblastic anemia

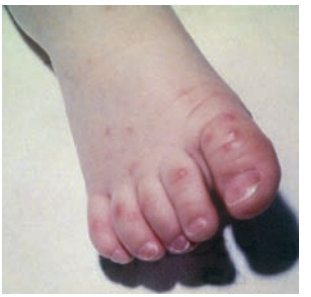
- Treatment strategies include chelation therapy, commonly deferoxamine, and maintenance phlebotomies. Chelation therapy does not reverse hepatic cirrhosis, hypogonadism, and arthropathy

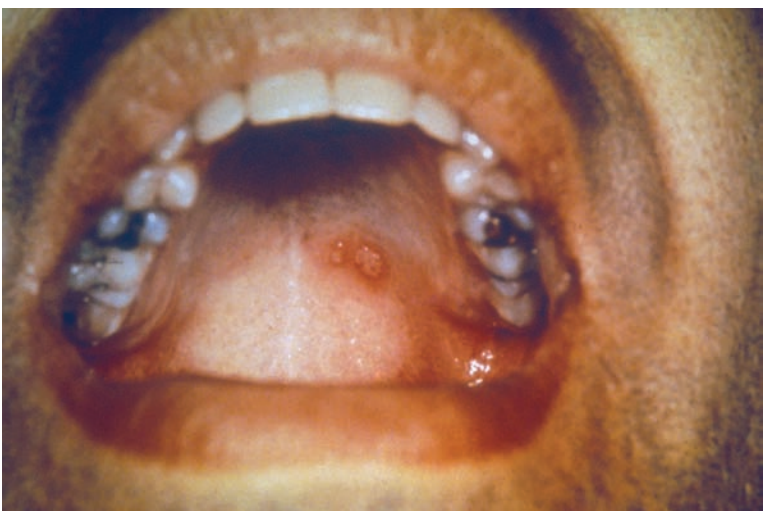
References

Habermann TM. *Mayo Clinic Internal Medicine Board Review 2004-2005*. Philadelphia: Lippincott Williams & Wilkins; 2004:464-465.

Powell LW, George DK, McDonnell SM, et al. Diagnosis of hemochromatosis. *Ann Intern Med*. 1998;129:925-931.







The lesions shown here were tender and occurred on the palms and feet and in the mouth of a 4-year-old child and his father. What is the causative agent?

a. Herpesvirus
b. Epstein-Barr virus
c. Coxsackievirus
d. Varicella-zoster virus
e. Smallpox virus

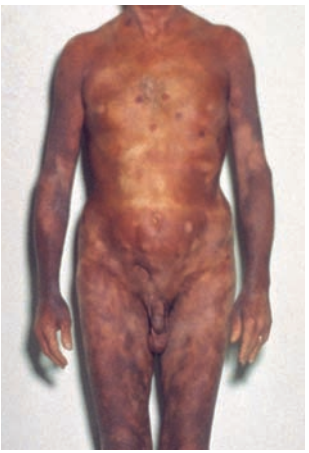
Answer: c

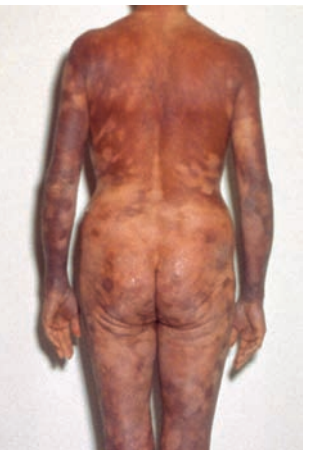
- Coxsackievirus A16, an enterovirus, is the most common cause of hand, foot, and mouth disease
- In 1998, enterovirus 71 was involved in an epidemic affecting nearly 1.5 million people in Taiwan, a small percentage of whom died of the disease
- Transmission is by fecal-oral and respiratory routes, most commonly in summer or early autumn
- Incubation period is usually less than 10 days
- Although it is more common in children, it can occur in adults
- Characteristic findings (present in about 85% of patients) are vesicles with a red halo on the mouth, palms, and soles
- Oral lesions are commonly found on the palate, tongue, and buccal mucosa. When the vesicles rupture, the resultant erosions and ulcerations are painful
- Lesions occur less commonly on other areas of the extremities and buttocks
- Associated signs and symptoms include fever, malaise, anorexia, cervical lymphadenopathy, and rhinitis
- Diagnosis is usually made by recognition of the clinical syndrome. Viral culture is possible but not usually practical because of the delay and expense
- The disease usually resolves in 1 to 2 weeks. Immunity is permanent after resolution

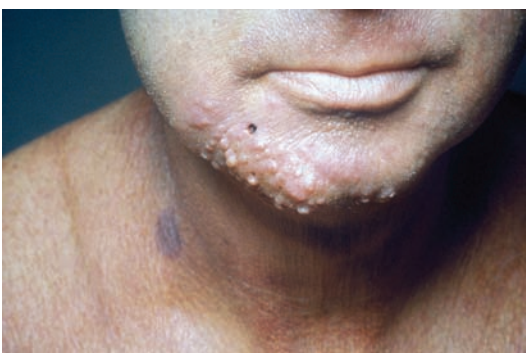
References

Haring JI. Case study: hand, foot, and mouth disease. *RDH*. 1999;19:22-24.

Ho M, Chen ER, Hsu KH, et al., for the Taiwan Enterovirus Epidemic Working Group. An epidemic of enterovirus 71 infection in Taiwan. *N Engl J Med*. 1999;341:929-935.







The skin lesions shown here and lymphadenopathy developed in a 41-year-old man. Biopsy of the lesions showed lymphocytes with hyperchromic and convoluted nuclei. Which of the following treatment options is appropriate?

a. Oral doxycycline
b. Intravenous third-generation cephalosporin
c. Plasma exchange
d. Psoralen with ultraviolet A light
e. Highly active antiretroviral therapy

Mycosis Fungoides or Cutaneous T-Cell Lymphoma

Answer: d

- The disease occurs in phases:

  The premycotic-patch phase lasts a few years to decades. Itching is common. Patches may be inconspicuous, but classically they are described as large, flat, brown-pink patches with stippling on the trunk, thighs, and upper arms

  The infiltrative-plaque phase is characterized by thickening of the patches with violaceous or reddened discoloration, often taking bizarre patterns. Itching continues. Alopecia may develop

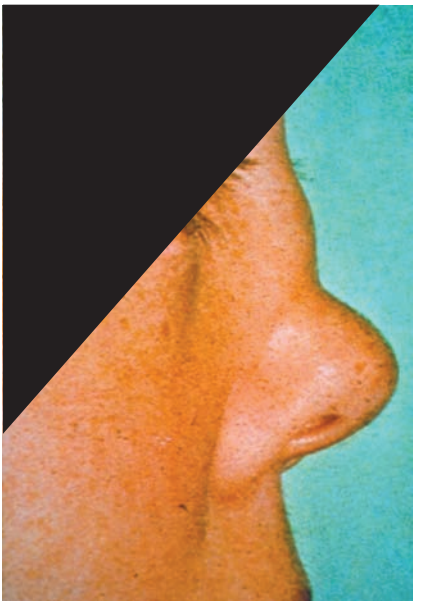
  The tumor phase is the most characteristic, with necrotic, ulcerating lesions, originally described as resembling mushrooms, hence the name "mycosis fungoides"

- Sézary syndrome accounts for about 5% of presentations of cutaneous T-cell lymphoma (CTCL). Sézary cells are atypical T cells with characteristic hyperconvoluted nuclei and periodic acid-Schiff–positive vacuoles. These cells involve the epidermis, dermis, and, later, peripheral blood
- Average age at presentation is 50 years. Blacks are twice as likely as whites, and men are twice as likely as women, to have CTCL
- Median duration of survival with the cutaneous form is 12 years. Once extracutaneous involvement occurs, median survival decreases to 2.5 years
- Treatment includes careful skin care, psoralen with ultraviolet A light, topical nitrogen mustard, radiation therapy, and chemotherapy

References

Habermann TM. *Mayo Clinic Internal Medicine Board Review 2004-2005*. Philadelphia: Lippincott Williams & Wilkins; 2004:170.

Lorincz AL. Cutaneous T-cell lymphoma (mycosis fungoides). *Lancet*. 1996;347:871-876.



The deformity shown here may occur in patients with Wegener's granulomatosis and is indicative of which one of the following?

a. Cartilaginous hypoplasia
b. Nasal septal perforation
c. Ethmoid sinus deformity
d. Osteoblastic hyperactivity
e. Osteoclastic hyperactivity

Saddle-Nose Deformity

Answer: b

- Saddle-nose deformity results from destruction of the nasal cartilage
- This deformity may occur in patients with Wegener's granulomatosis
- Additional findings may include nasal septal perforation due to ulceration and easy mucosal friability
- Nasal and chronic sinus congestion and epistaxis may be present
- More than 90% of patients with Wegener's granulomatosis present to a physician with complaints of upper or lower respiratory tract symptoms (e.g., rhinorrhea, nasal discharge, sinus pain, otitis media)
- Other symptoms and involved organs include the following:

  Constitutional symptoms (fever, malaise, weight loss)
  Skin changes (40%-50% of patients; e.g., urticaria, petechiae, papules, vesicles, ulcers, pyoderma, livedo reticularis)
  Eyes (43%; e.g., conjunctivitis, uveitis, proptosis)
  Nervous system (25%; e.g., sensory neuropathy, mononeuritis multiplex, cranial nerve palsies)
  Arthralgias (58%)
  Arthritis (28%)
  Pulmonary parenchyma (60%; e.g., cough, hemoptysis, or dyspnea)
  Renal disease (85%; e.g., focal segmental necrotizing vasculitis)
  Ears (e.g., hearing loss, otitis)

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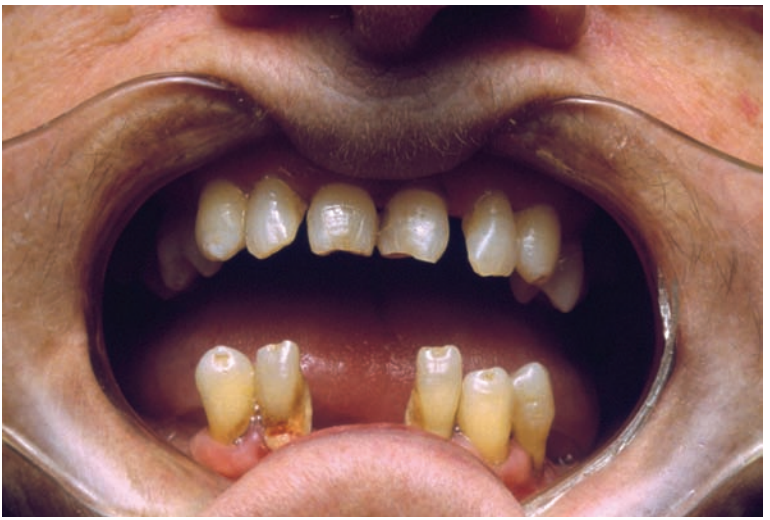

A child was born 1 year after his mother was infected with a sexually transmitted disease. Shown here are the abnormal teeth of the child. Which one of the following actions might have prevented this abnormality?

a. Maternal avoidance of tetracycline treatment for the sexually transmitted disease
b. Maternal avoidance of alcohol ingestion
c. Maternal avoidance of cocaine use
d. Maternal avoidance of tobacco use
e. Maternal treatment with penicillin

Hutchinson's Teeth of Congenital Syphilis

Answer: e

- Hutchinson's teeth are characterized by notching of the incisors due to defects in early tooth development
- Fetal infection with *Treponema pallidum* most likely occurs after the fifth gestational month in women who have been infected for less than 2 years
- Both mother and fetus may be cured with penicillin treatment of the mother before birth of the child, although some abnormalities may persist depending on the stage of development of the fetus when treatment was begun
- Congenital syphilis, once rare, has increased in incidence in association with sexual promiscuity and illegal drug use, often in urban areas
- If not treated, multiple stigmata, in addition to Hutchinson's teeth, may result, including the following:

  Interstitial keratitis
  Frontal bossing
  Saber shins (anterior tibial bowing)
  Mulberry molars (with small multiple cusps on the first molar)
  Saddle-nose deformity
  Nasal septal perforation
  Scaphoid scapulae
  High palatine arch
  Epiphyseal enlargement
  Eighth nerve deafness
  Rhagades (linear scars around the mouth, as if perpetually pursing the lips)
  Clutton's joints (symmetric spontaneous hydroarthroses, usually of the knees)
  Fusiform dactylitis
  Mental deficiencies

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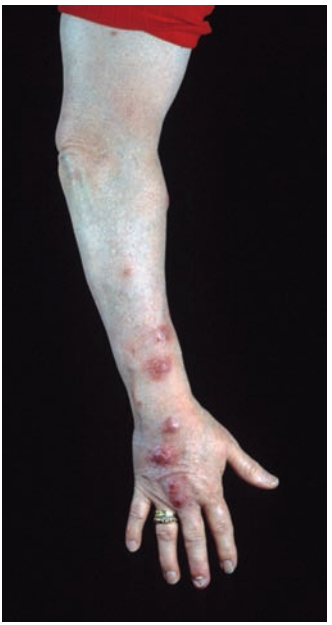

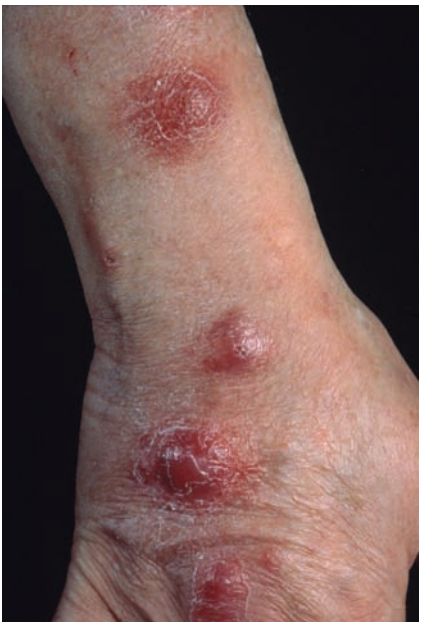

A 52-year-old woman presents with an erythematous, scaly, and nodular eruption of the upper extremity of 3 weeks in duration. She has pet fish. Which one of the following is the most likely diagnosis?

a. Sporotrichosis
b. Nocardiosis
c. Leishmaniasis
d. *Francisella tularensis* infection
e. *Mycobacterium marinum* infection

Nodular Lymphangitis Due to Mycobacterium marinum

Answer: e

- *Mycobacterium marinum* is an atypical mycobacterium that causes cutaneous infections after aquatic-related inoculation
- *M. marinum* lesions are sometimes referred to as swimming pool granuloma
- *M. marinum* infection also often occurs after owners of pet fish clean aquariums
- This condition often presents with a chronic indurated nodule of the extremities (e.g., fingers and hands)
- Lesions caused by *M. marinum* may ulcerate and leave scars. This infection also may progress to nodular lymphangitis
- Treatment may include observation (simple lesions) or antimicrobial agents:

  Doxycycline
  Trimethoprim-sulfamethoxazole
  Rifampin and ethambutol

- Surgical excision also may be effective
- Other causes of nodular lymphangitis:

  Sporotrichosis
  Nocardiosis
  Leishmaniasis
  Tularemia

References

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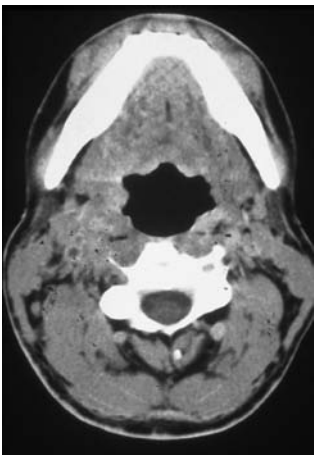
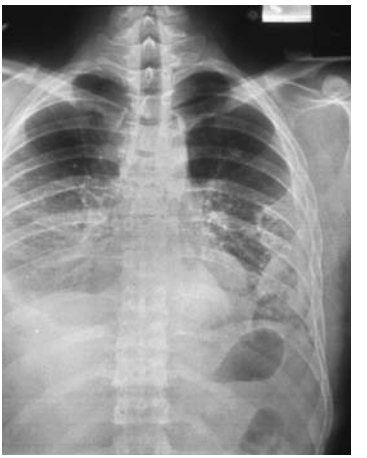
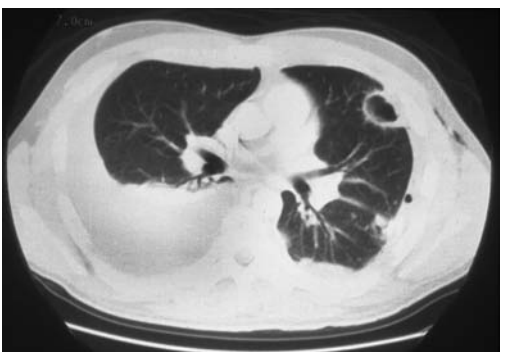






A homeless person had a toothache for several months before development of a foul-smelling cough and dyspnea. This disorder is most commonly associated with septic thrombosis of which one of the following?

a. Internal jugular vein
b. External jugular vein
c. Internal carotid artery
d. Cavernous sinus
e. Subclavian vein

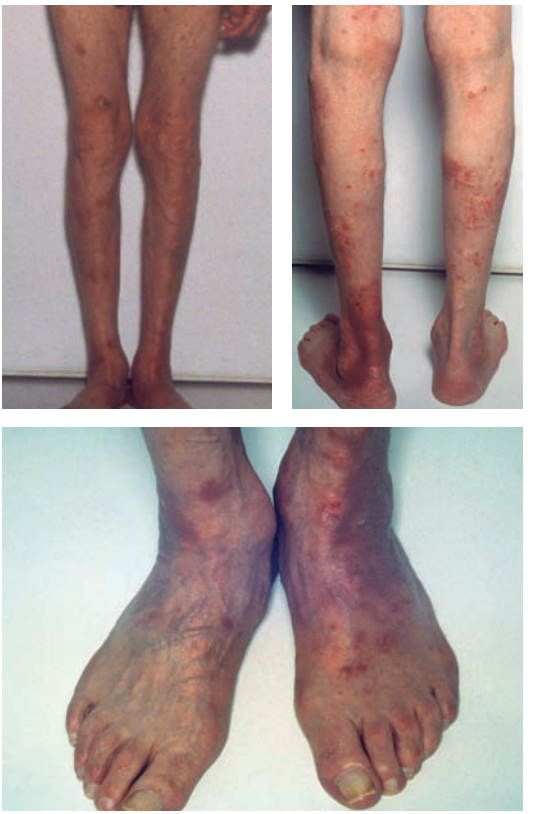
Lemierre's Syndrome

Answer: a

- Lemierre's syndrome, or jugular vein suppurative thrombophlebitis, is an unusual illness that may occur as a complication after an episode of pharyngotonsillitis
- It is characterized by the sequence of septic thrombosis of the internal jugular vein, bacteremia, septic pulmonary emboli, and metastatic abscess formation
- *Fusobacterium necrophorum* is an anaerobic, gram-negative rod that has been associated with Lemierre syndrome
- Treatment includes use of clindamycin, penicillin, or chloramphenicol
- Metronidazole is advocated for its bacteriostatic properties and should be administered for at least 1 month in combination with the chosen antibacterial regimen
- Embolectomy may be necessary to remove the septic thrombus and to prevent further seeding of infection
- Ligation of the internal jugular vein may be necessary to prevent further embolization of thrombotic debris
- Mortality ranges from 4% to 12% even with appropriate antibiotics. In the pre-antibiotic era, mortality was estimated to be as high as 90%
- There is no consensus regarding the use of anticoagulants in this disorder

Reference

Stokroos RJ, Manni JJ, de Kruijk JR, et al. Lemierre syndrome and acute mastoiditis. *Arch Otolaryngol Head Neck Surg*. 1999;125:589-591.



A 69-year-old man with a history of chronic loose stools presents with intense burning and itching due to symmetrically distributed groups of vesicles and papules. How should this skin condition be treated?

a. Gluten-free diet
b. Acyclovir
c. Topical corticosteroids
d. Psoralen with ultraviolet-A (PUVA) light
e. Topical bacitracin

Dermatitis Herpetiformis

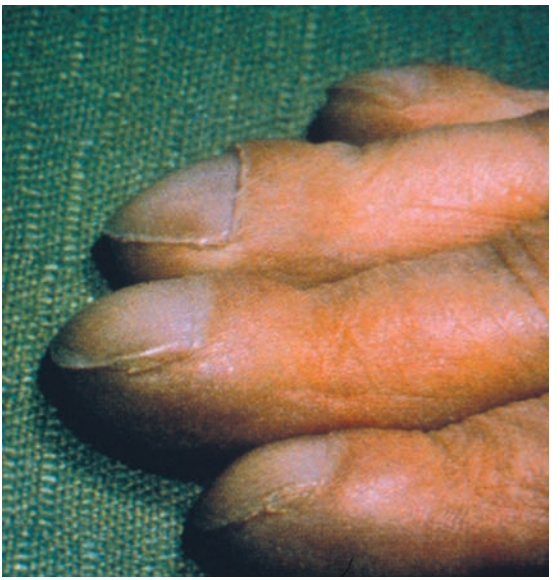
Answer: a

- Dermatitis herpetiformis (DH) is an immune-mediated bullous disease
- The onset of DH is usually during the third or fourth decade of life
- Most patients have some degree of gluten-sensitive enteropathy (often low-grade or subclinical)
- Skin lesions are often very pruritic, grouped vesicles
- Common sites of involvement include the following:

  Elbows
  Knees
  Buttocks
  Back of neck
  Scalp
  Low back

- IgA anti-endomysial antibodies are present in 70% of patients with DH
- Direct immunofluorescence finding of IgA deposits in skin is the hallmark of diagnosis
- DH may respond to a gluten-free diet; if not, dapsone and a gluten-free diet usually help
- Corticosteroids are not helpful in treating DH

Reference
Habermann TM. *Mayo Clinic Internal Medicine Board Review 2004-2005.* Philadelphia: Lippincott Williams & Wilkins; 2004:173-174, 179.



The physical finding shown here may be associated with all of the following *except*:

a. Lung abscess
b. Emphysema
c. Pulmonary fibrosis
d. Small cell lung carcinoma
e. Ulcerative colitis

Clubbing

Answer: b

- Clubbing is a physical finding characterized by increased soft tissue mass in the distal segment of a digit. This results in loss of the normal nail angle
- Five grades of clubbing:

  Grade 1: Softening of the nail bed
  Grade 2: Loss of the normal 15° angle between the nail and the cuticle
  Grade 3: Exaggerated convexity of the nail
  Grade 4: Distal segment of digit assumes a clubbed appearance
  Grade 5: Nail and adjacent skin have a glossy appearance with nail striations

- Hereditary, idiopathic, humoral, or neurogenic mechanisms have been implicated, but no cause has been definitively determined
- Associated with various conditions, including the following:

  Lung abscess
  Lung cancer (non–small cell more often than small cell lung carcinoma)
  Pulmonary fibrosis
  Bronchiectasis

- Clubbing is *not* independently associated with chronic obstructive pulmonary disease (emphysema or bronchitis)
- Clubbing is part of the triad that defines hypertrophic pulmonary osteoarthropathy. Painful periosteal hypertrophy of long bones and symmetric arthralgias of large joints are 2 additional features of this arthropathy

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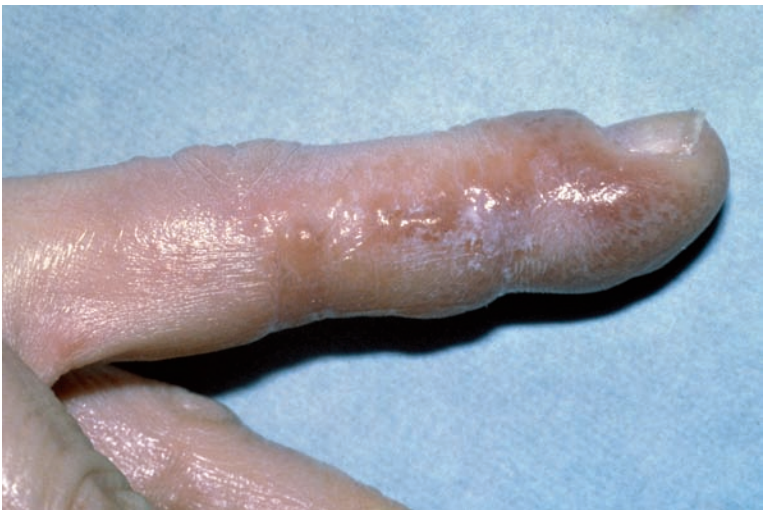

The painful lesions shown here developed in a 31-year-old surgeon in an area where she had been stuck by a contaminated needle. What is the diagnosis?

a. Digital cellulitis
b. Acute cutaneous human immunodeficiency virus
c. Contact dermatitis
d. Herpetic whitlow
e. Bacterial felon

Herpetic Whitlow

Answer: d

- Herpes simplex virus (HSV) is the cause of herpetic whitlow
- Most often, HSV 1 is involved, but HSV 2 results in identical lesions
- Precautions should be taken by health care workers when treating patients with HSV, especially those with respiratory infection undergoing invasive procedures
- Incubation takes 2 to 14 days after exposure. This period is followed by throbbing pain and vesicle development, which typically last about 2 weeks
- Diagnosis is suspected from the history and physical examination and can be confirmed by culture or polymerase chain reaction of the vesicle fluid or a positive Tzanck smear
- Like all HSV infections, whitlow can be chronic and recurring
- If recurrences are frequent, treatment with acyclovir or another antiviral medication may be warranted
- Use of suppressive antiviral medications may decrease recurrences by more than 80%
- Because herpetic whitlow is not a whitlow or felon in the classic sense, incision and drainage is not indicated

References
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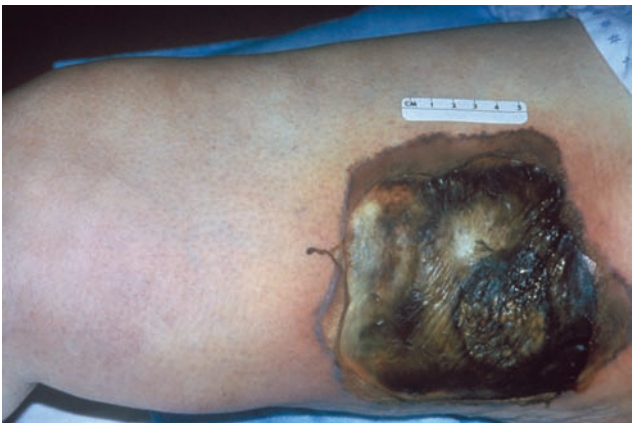
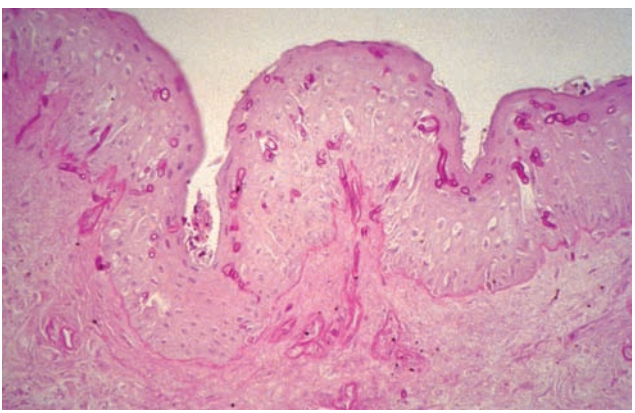
Habermann TM. *Mayo Clinic Internal Medicine Board Review 2004-2005*. Philadelphia: Lippincott Williams & Wilkins; 2004:571-572.





The lesion shown here developed in a neutropenic patient after hematogenous spread from a nasal source. Biopsy result of the lesion is also shown. What is the most likely diagnosis?

a. Aspergillosis
b. Blastomycosis
c. Histoplasmosis
d. Mucormycosis
e. Coccidioidomycosis

Mucormycosis

Answer: d

- The causative agent is *Rhizopus* species, Zygomycetes, resulting in invasive fungal disease
- Risk factors include the following:

  Diabetic ketoacidosis
  Neutropenia
  Renal failure
  Deferoxamine therapy

- Mucormycosis often involves nasal passages or sinuses
- Patients with pulmonary or gastrointestinal mucormycosis rarely survive beyond 2 weeks
- Diagnosis is based on typical black necrotic lesions and is confirmed by biopsy
- Management requires treating the underlying condition, if possible, surgical debridement, and amphotericin B
- Differential diagnosis should include ecthyma gangrenosum caused by *Pseudomonas* infection in patients with granulocytopenia

References

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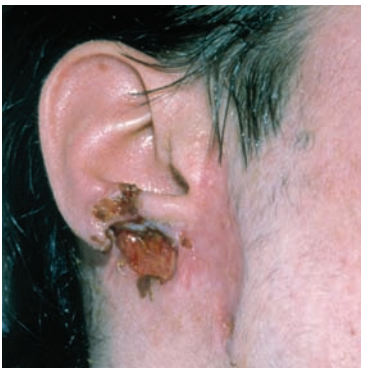
Strasser MD, Kennedy RJ, Adam RD. Rhinocerebral mucormycosis: therapy with amphotericin B lipid complex. *Arch Intern Med*. 1996;156:337-339.



A 26-year-old man with a history of seasonal allergies, frequent bouts of sinusitis, and cigarette smoking presents with cough, epistaxis, and a nonhealing ear wound. Which of the following tests would be the most helpful for making the diagnosis?

a. Perinuclear-staining antineutrophil cytoplasmic antibody
b. Cytoplasmic-staining antineutrophil cytoplasmic antibody
c. Antinuclear antibody
d. Antibody to double-stranded DNA
e. Erythrocyte sedimentation rate

Wegener's Granulomatosis

Answer: b

- Wegener's granulomatosis is a systemic granulomatous vasculitis involving arteries and veins
- Wegener's granulomatosis primarily involves the following:

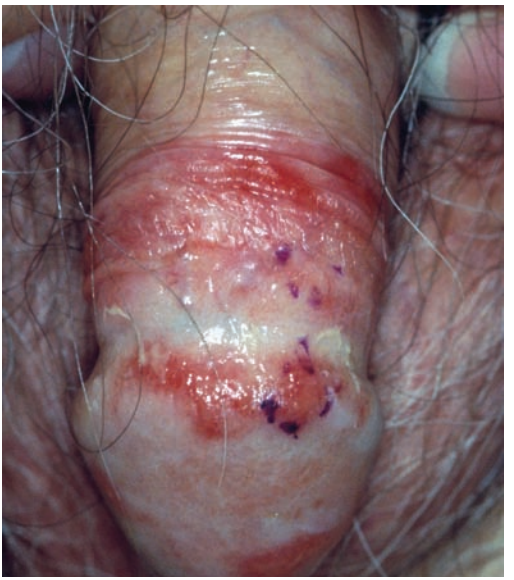
Ear, nose, throat	90%
Kidney	80%
Lungs	60%
Skin	40%-50%
Eyes	43%
Joints	28%-58%
Central nervous system	25%

- The differential diagnosis of saddle-nose deformity:

  Leprosy
  Relapsing polychondritis
  Wegener's granulomatosis
  Congenital syphilis

- The cytoplasmic-staining antineutrophil cytoplasmic antibody test generally is considered specific for Wegener's granulomatosis, but a positive result without clinical evidence of disease does not establish the diagnosis
- The combination of cyclophosphamide and prednisone results in complete remission in more than 90% of patients
- Trimethoprim-sulfamethoxazole is used to prevent relapses

Reference
Habermann TM. *Mayo Clinic Internal Medicine Board Review 2004-2005*. Philadelphia: Lippincott Williams & Wilkins; 2004:904-905.



The circumscribed, nonpainful lesions shown here developed without any intercurrent illness in a 60-year-old man. Biopsy reveals a plasma cell infiltrate. What is the lesion?

a. Circinate balanitis of Reiter's syndrome
b. Erythroplasia of Queyrat
c. Syphilis
d. Lichen planus
e. Zoon's balanitis

Zoon's Balanitis

Answer: e

- Zoon's balanitis also is known as balanitis circumscripta plasmacellularis and plasma cell balanitis
- It was first described by J.J. Zoon in 1952. In 1954, Zoon also described the vulvar equivalent in women
- Clinically, it usually presents with a yellow or blood-tinged sterile discharge and painless, glistening, maculopapular lesions. Multiple pinpoint, bright-red "cayenne pepper" spots often are noted
- Histologic results include a bandlike (lichenoid) inflammatory infiltrate with plasma cell predominance and hemosiderin deposition in the subcutaneous tissues. Keratinocytes are thinned and "lozenge-shaped" with "watery spongiosis" or edematous widening of the intercellular spaces
- The condition is benign. No treatment is necessary, but topical corticosteroids have been used
- Classically described in elderly uncircumcised men, circumcision has been reported to result in resolution of the lesions
- Differential diagnosis for papulosquamous penile lesions includes psoriasis, lichen planus, lichen sclerosis, cicatricial pemphigoid, secondary syphilis, drug reactions, erythroplasia of Queyrat, bowenoid papulosis, and circinate balanitis of Reiter's syndrome

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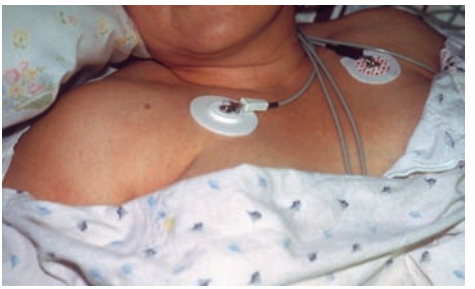
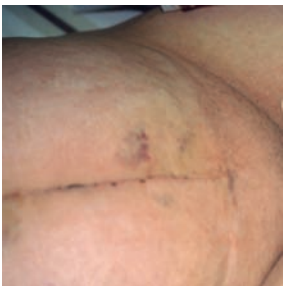
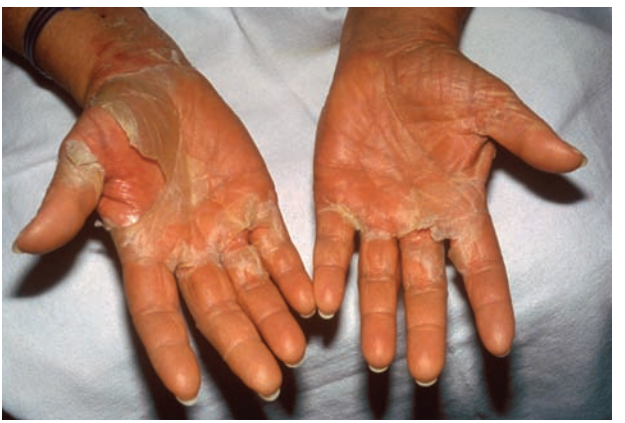
Kavanagh GM, Burton PA, Kennedy CT. Vulvitis chronica plasmacellularis (Zoon's vulvitis). *Br J Dermatol*. 1993;129:92-93.

Yoganathan S, Bohl TG, Mason G. Plasma cell balanitis and vulvitis (of Zoon): a study of 10 cases. *J Reprod Med*. 1994;39:939-944.







Wound infection developed after cesarean section in a 34-year-old woman. She had the skin lesions shown here, hypotension, and fever. Blood cultures were negative. Five days later, her palms desquamated as shown here. All of the following are true regarding this syndrome *except*:

a. It is caused by a toxin-producing strain of *Staphylococcus aureus* in a nonimmune person
b. Treatment includes supportive care and antibiotics
c. Blood cultures are usually positive for *S. aureus*
d. It may occur in menstruating women with prolonged tampon use
e. The mortality rate is 5% to 10%

Staphylococcal Toxic Shock Syndrome

Answer: c

- The syndrome is caused by staphylococcal toxin (toxic shock syndrome toxin-1, TSST-1) and staphylococcal enterotoxins B and C
- It is associated with the following:

  Menstruating women with prolonged tampon use
  Staphylococcus aureus pneumonia developing after influenza
  Childbirth
  Tracheitis
  Wound infections
  Nasal packing
  Barrier contraceptives
  Localized abscesses

- Diagnosis is made according to the following criteria: fever, exfoliative erythroderma, reddened mucous membranes, hypotension, and multiorgan involvement
- Other findings may include injected conjunctivae and strawberry tongue
- Blood cultures are usually negative (unlike streptococcal toxic shock syndrome)
- Treatment is supportive, although β-lactam antibiotics are often given. It is important to identify and remove the source of the *S. aureus* if possible (e.g., drain abscesses, remove tampon, remove nasal packing)
- The syndrome recurs in up to 40% of cases because of an inability to generate appropriate immunity
- The mortality rate is 5% to 10%

References

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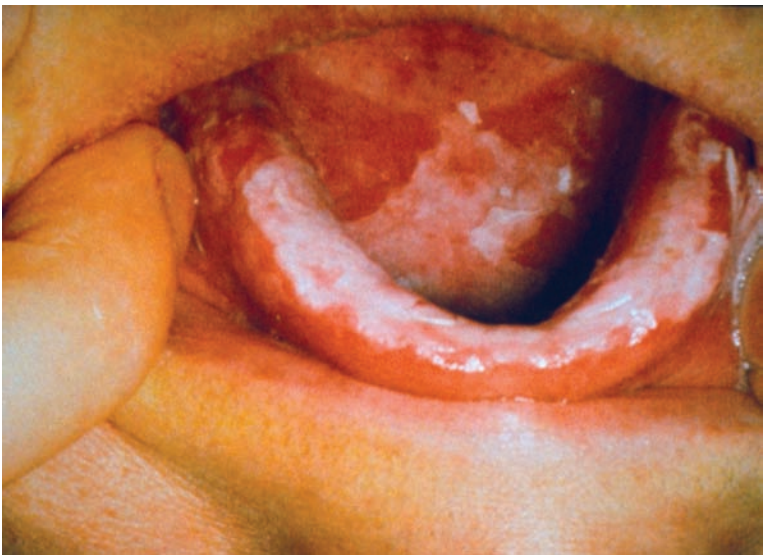

A 47-year-old heterosexual, edentulous man presents with fever, fatigue, swollen cervical nodes, and the oral lesions shown here. All of the following may predispose a patient to this condition *except*:

a. Diabetes mellitus
b. Acute pancreatitis
c. Human immunodeficiency virus infection
d. Immunosuppressive chemotherapy
e. Chronic corticosteroid use

Oral Thrush

Answer: b

- Oral thrush is caused by *Candida*, most commonly *Candida albicans*
- Risk factors for development of oral candidiasis:

  Smoking
  Recent use of antibiotics
  Recent use of corticosteroids
  Chemotherapy
  Immunocompromising illnesses (e.g., human immunodeficiency virus, hematologic malignancy)
  Diabetes
  Xerostomia

- Variants of oral candidiasis include the following:

  Pseudomembranous candidiasis (thrush)
  Chronic hyperplastic candidiasis (can progress to malignancy)
  Angular cheilitis
  Erythematous candidiasis (associated with use of dentures and antibiotics)

- The whitish exudates of thrush may be easily scraped off the tongue to reveal underlying erythema. In contrast, oral hairy leukoplakia (Epstein-Barr virus infection) commonly involves the lateral sides of the tongue and is adherent
- If odynophagia is coexistent, *Candida* esophagitis must be suspected

References
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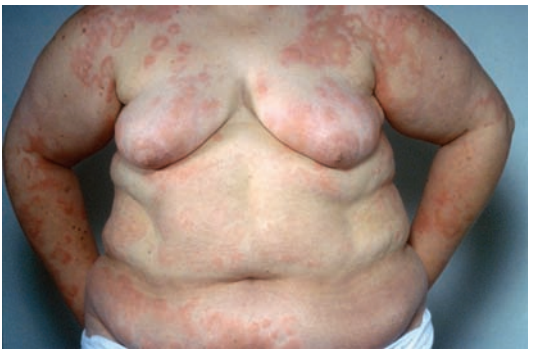

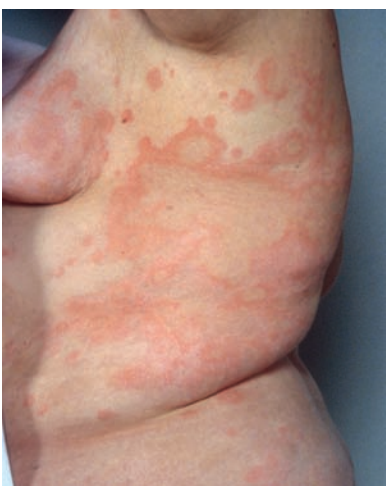

The skin lesions shown here developed 1 week into an upper respiratory tract infection in a 55-year-old woman. What are these lesions called?

a. Erythema nodosum
b. Erythema multiforme
c. Erythema infectiosum
d. Erythema migrans
e. Necrolytic migratory erythema

Erythema Multiforme

Answer: b

- This acute, self-limited phenomenon is characterized by "iris" or "target" lesions
- Erythema multiforme is part of the spectrum of conditions, including Stevens-Johnson syndrome (SJS) and, most severely, toxic epidermal necrolysis (TEN)
- It may be associated with infections, including *Mycoplasma pneumoniae*, *Yersinia enterocolitica*, and viral infections, especially herpes simplex
- Drug associations include sulfonamides, penicillins, phenylbutazone, barbiturates, phenytoin and other anticonvulsants
- Erythema multiforme is associated less commonly with underlying connective tissue diseases or malignancies
- The differential diagnosis is multiple and includes tinea corporis, urticaria, pityriasis rosea, dermatitis herpetiformis, pemphigus, pemphigoid, lichen planus, and systemic lupus erythematosus
- Treatment of mild cases involves removing offending agents, careful observation, and antimicrobials for superinfections
- Severe cases (SJS or TEN) necessitate hospital admission, aggressive fluid replacement, antimicrobials for superinfections, aggressive supportive care, and ophthalmologic and dermatologic consultations
- Corticosteroid use is controversial

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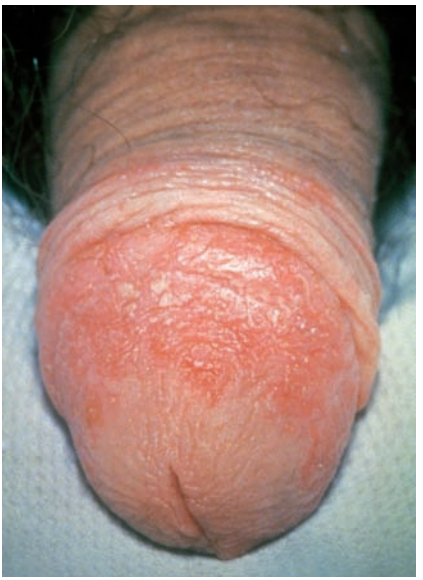

A 35-year-old man with ankylosing spondylitis who is recovering from a diarrheal illness complains of blurred vision, arthritis, and the lesions shown here. What is the diagnosis?

a. Circinate balanitis of Reiter's syndrome
b. Erythroplasia of Queyrat
c. Syphilis
d. Lichen planus
e. Zoon's balanitis

Circinate Balanitis (in setting of Reiter's Syndrome)

Answer: a

- Classic Reiter's syndrome consists of arthritis, conjunctivitis (30%), and urethritis (45%). Reactive arthritis is the hallmark, other associations being less common
- Other commonly reported associations are circinate balanitis (25%-70% of males), uveitis (12%-20%), oral lesions (14%), nail changes (13%), and keratoderma blennorrhagicum (i.e., a papulosquamous skin eruption most commonly on the palms and soles, 23%)
- The penile lesions of circinate balanitis in the setting of arthritis are virtually diagnostic of Reiter's syndrome
- Nongonococcal urethritis is the most common infection associated with Reiter's syndrome (46%). Enteritis also may be associated
- *Chlamydia trachomatis* can be isolated from up to 70% of men who show signs of urethral infection. However, the skin lesions of circinate balanitis do not appear infected
- The penile lesions are usually painless
- There is a strong association with HLA-B27 (75%)
- Most noninfectious symptoms resolve within 3 to 12 months and can be treated conservatively with nonsteroidal anti-inflammatory drugs. There is a recurrence in about 15% of patients, and chronic arthritis develops in about 15% of patients

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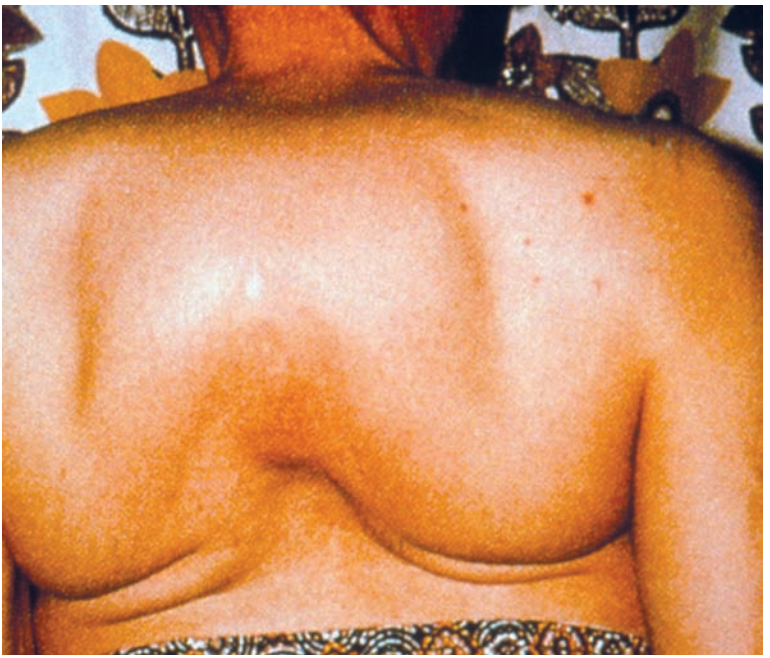

Associated characteristics of the condition shown here include which one of the following?

a. Pulmonary limitations when the scoliotic angle is more than 45°
b. Arterial hypercapnia
c. Increases in total lung capacity
d. Increases in vital capacity
e. Arterial hypoxemia

Kyphoscoliosis

Answer: e

- Kyphoscoliosis is the most common mechanical spinal deformity associated with pulmonary compromise
- Limitations on pulmonary function begin when the scoliotic angle exceeds 70°
- Characteristic findings:

  Decreased total lung capacity
  Decreased vital capacity
  Arterial hypoxemia
  Normal arterial CO_2

- Pulmonary hypertension and cor pulmonale are critical complications in severe and long-standing kyphoscoliosis
- Indications for surgical management in the younger patient include the following:

  Scoliotic angle more than 60° with chronic pain
  Physical deformity unacceptable to the patient

- In the older patient, surgical management may be pursued if:

  The scoliotic angle has progressed
  Radiculopathy or spinal stenosis is present
  Loss of pulmonary function is not believed to be due to a primary pulmonary process

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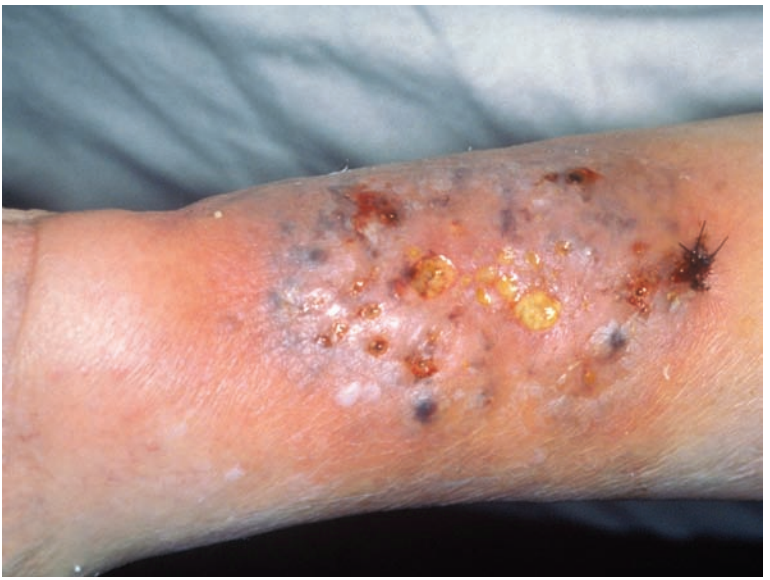

The lesion shown here developed in a 36-year-old man with ulcerative colitis after he bumped his shin on a coffee table. Biopsy shows neutrophilic predominance.

1. This skin finding is associated with all of the following *except*:
   1a. Inflammatory bowel disease
   1b. Rheumatoid arthritis
   1c. Acute myelogenous leukemia
   1d. Paraproteinemias
   1e. Non–insulin-dependent diabetes mellitus

2. All of the following may be indicated for treatment *except*:
   2a. Systemic corticosteroids
   2b. Azathioprine
   2c. Cyclophosphamide
   2d. Cyclosporine
   2e. Surgical debridement

Pyoderma Gangrenosum

Answer 1: e

Answer 2: e

- Pyoderma gangrenosum (PG) is an idiopathic, inflammatory, ulcerative disease that is neither infectious ("pyoderma") nor gangrenous. Histopathologic findings are nonspecific; PG is a diagnosis of exclusion. Early lesions often show neutrophilic predominance
- PG is classic for pathergy (lesions occurring at sites of trauma), which occurs in up to 50% of patients. Therefore, debridement is contraindicated
- PG presents with tender papules that develop into painful ulcers with "rolled-up" edges and surrounding edema
- Multiple lesions are common; the lower extremity is the most commonly involved site (75%-80%)
- At least 50% of patients have an associated condition. Common associations are inflammatory bowel disease (ulcerative colitis is more common than Crohn's disease), rheumatoid and nonrheumatoid arthritis, hematologic malignancies, and paraproteinemias
- PG was first described and the association with ulcerative colitis reported at Mayo Clinic in 1930. Even then it was noted that the skin lesions improved when the underlying disorder improved
- Immunosuppression is the mainstay of treatment
- Lesions heal with cribriform scarring: 47% within 6 months, 69% within 1 year, 95% within 3 years

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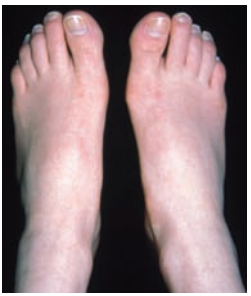

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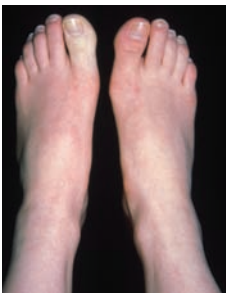
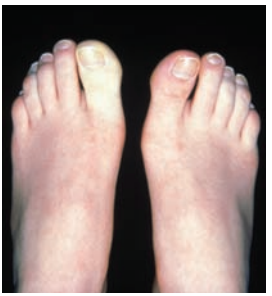


Elevated and cooled 1 minute



Elevated and cooled 5 minutes

A 26-year-old woman complains of pain and numbness in her left great toe with cold exposure. If the predisposing condition is primary rather than secondary, all of the following are true *except*:

a. Women are more often affected than men
b. Digital ulceration is a rare complication
c. Livedo reticularis frequently is present
d. Attacks are frequent
e. This condition is often associated with future connective tissue disease

Raynaud's Phenomenon (Superimposed on Fixed Obstruction)

Answer: e

- In this case the fixed obstruction was thought to be due to recent trauma to the left great toe
- Raynaud's phenomenon is more likely to be associated with future connective tissue disease if the patient is male or has positive results for antinuclear antibody
- *Primary* Raynaud's phenomenon usually:

  Affects females
  Involves all digits
  Is of mild-to-moderate severity
  Has onset at menarche
  Involves frequent attacks

  In addition, it:
  May be precipitated by emotional distress
  Is not associated with future connective tissue disease
  Rarely involves edema or periungual erythema
  Rarely involves digital ulceration
  Is frequently associated with livedo reticularis

- *Secondary* Raynaud's phenomenon:

  Is associated with connective tissue disease (e.g., systemic sclerosis, polymyositis, mixed connective tissue disease)
  Affects males or females
  Often begins in a single digit
  Is moderate to severe
  Includes digital ulceration in 30% to 50% of cases
  Is uncommonly associated with livedo reticularis
  Has onset in the mid 20s
  Involves infrequent attacks
  Is not precipitated by emotional distress
  Is frequently associated with edema or periungual erythema

Reference
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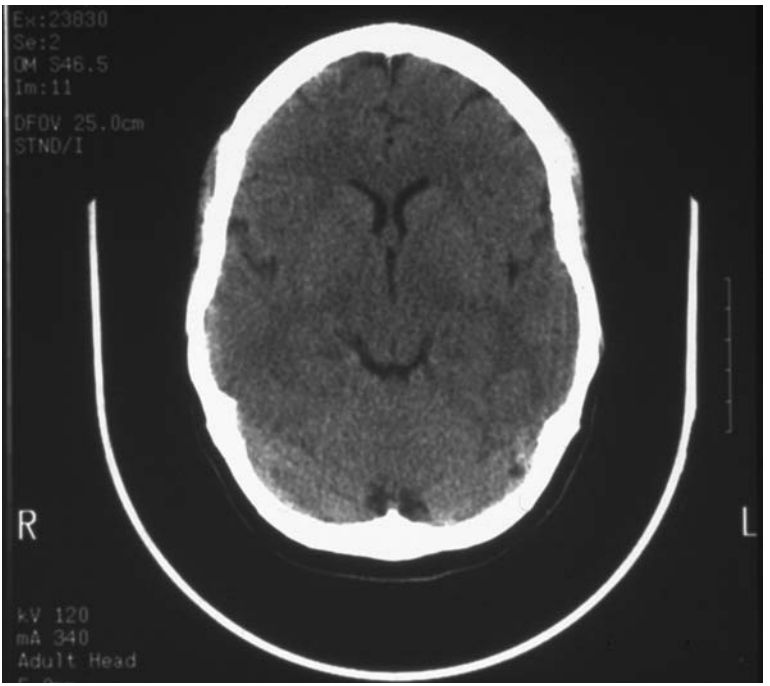
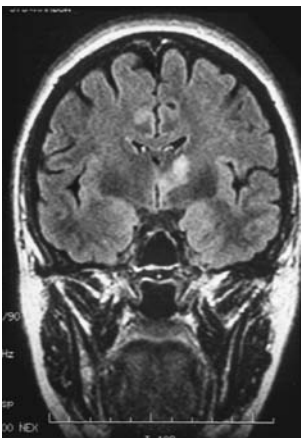


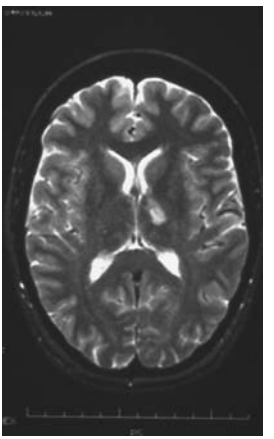
A 60-year-old man with a history of hypertension presented 4 hours after the onset of speech deficits. Which of the following statements is correct?

a. The negative computed tomogram shown here effectively rules out acute stroke
b. T2-weighted magnetic resonance imaging (MRI) would be expected to be positive at 4 hours if a stroke has occurred
c. Diffusion-weighted MRI may be positive 4 hours after a stroke has occurred
d. If a stroke has occurred, the lesion is likely in the right paramedian thalamic region
e. Diffusion-weighted MRI will show an acute thalamic stroke after T2-weighted MRI

Left Thalamic Stroke

Answer: c





- Computed tomography (CT) is often negative in acute stroke. Use of CT in the setting of acute stroke is to exclude intracranial hemorrhage and mass effect. A negative CT does not rule out acute stroke
- Diffusion-weighted MRI may be positive in acute thalamic stroke as soon as 4 hours after symptoms
- T2-weighted MRI may show hyperintensity at the site of stroke within 24 hours of the event (see images above, obtained 1 day after the CT on previous page)
- Physical manifestations of thalamic stroke depend on the location of the infarct. Language deficits would be expected from left paramedian lesions

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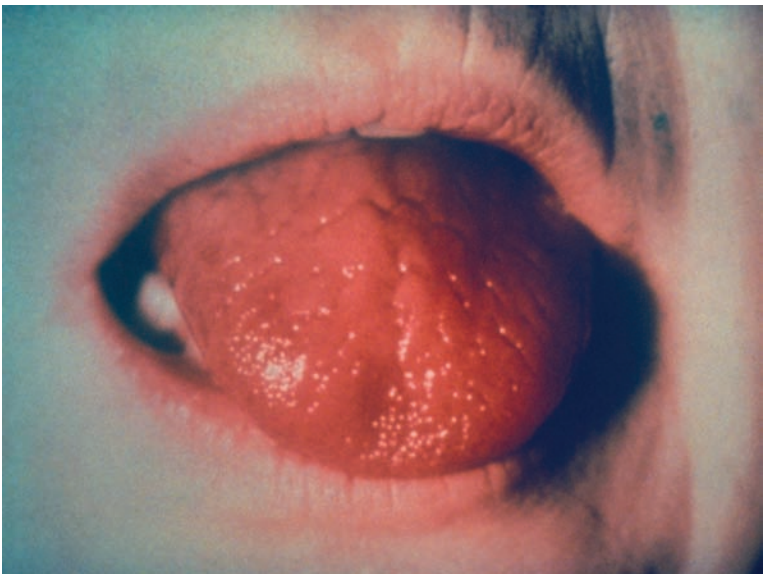

A 65-year-old woman presents with fatigue and a sore tongue. She has a history of diabetes mellitus and hypothyroidism. She is anemic. What is the most likely diagnosis?

a. Vitamin B_{12} deficiency
b. Folic acid deficiency
c. Iron deficiency
d. Beriberi
e. Primary systemic amyloidosis

Pernicious Anemia

Answer: a

- The causes of vitamin B_{12} deficiency are many and include achlorhydria, pernicious anemia (lack of intrinsic factor), gastrectomy, ileal resection, bacterial overgrowth, chronic pancreatitis, and long-term strict vegetarian diet
- Signs, symptoms, and laboratory findings that may be present with vitamin B_{12} deficiency:

  "Beefy," atrophic tongue
  Diarrhea
  Paresthesias
  Gait disturbance
  Mental status changes
  Position and vibratory sense impairment
  Decreased ankle reflexes
  Anemia with increased mean corpuscular volume
  Howell-Jolly bodies and hypersegmented neutrophils on peripheral smear
- Pernicious anemia is the most common cause of vitamin B_{12} deficiency
- Patients with pernicious anemia often have other immune-mediated conditions, including type 1 diabetes mellitus, hypothyroidism, and vitiligo
- Antibodies to intrinsic factor are highly specific to patients with pernicious anemia

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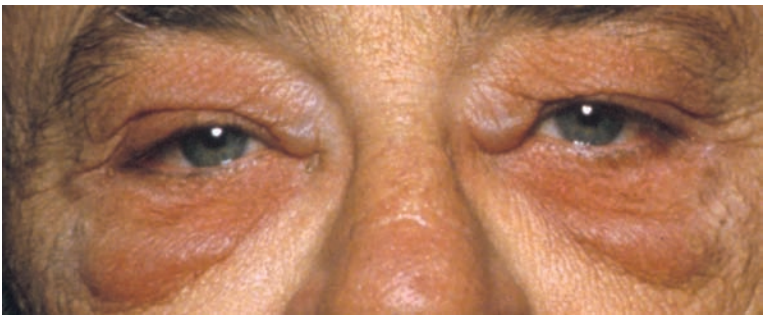

Five days previously, ramipril was prescribed for a 60-year-old man with type 2 diabetes mellitus. He now presents with the condition shown here. What is the diagnosis?

a. Nephrotic syndrome
b. Hypothyroidism
c. Ramipril allergy
d. Angioedema associated with angiotensin-converting enzyme inhibitor
e. Superior vena cava syndrome

Angioedema Associated With Angiotensin-Converting Enzyme Inhibitor

Answer: d

- Angioedema associated with angiotensin-converting enzyme (ACE) inhibitor is not an allergic reaction
- Bradykinin and substance P are degraded by ACE inhibitors
- Bradykinin and substance P may accumulate in patients taking ACE inhibitors, resulting in angioedema
- Angioedema usually occurs within the first week of therapy; however, late reactions have been reported
- Patients in whom this condition develops are likely sensitive to all ACE inhibitors
- Angioedema also has been reported in patients receiving angiotensin II receptor antagonists
- Treatment consists of stopping use of the medication; antihistamines, epinephrine, and corticosteroids have been used

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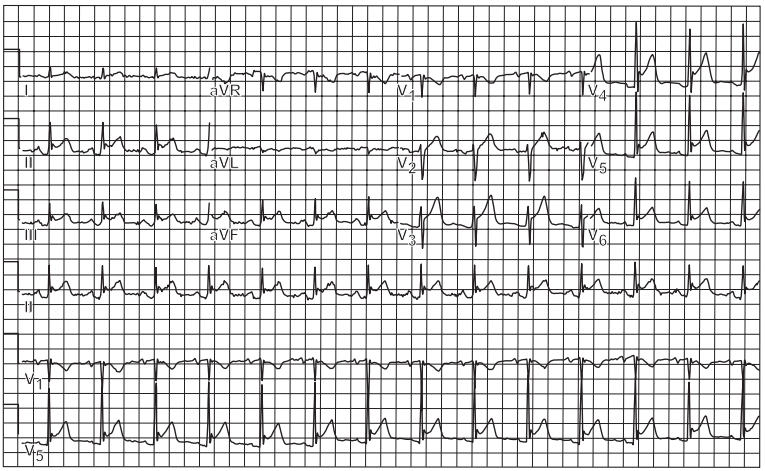

A 45-year-old man returns to your office complaining of chest pain while trying to sleep the night before. He says he feels better when he bends over to tie his shoes. His electrocardiogram is shown here. You conclude that he should try discontinuing the use of which one of the following medications?

a. Metoprolol
b. Acetaminophen
c. Indomethacin
d. Hydralazine
e. Diltiazem

Acute Pericarditis

Answer: d

- The numerous causes of pericarditis include the following:

  Infection
  Viral: coxsackievirus
  Bacterial: *Staphylococcus*, *Pneumococcus*, tuberculosis
  Fungal: histoplasmosis
  Rheumatologic: sarcoidosis, systemic lupus erythematosus
  Neoplastic: breast, lung
  Drugs: hydralazine, procainamide, methyldopa
  Trauma
  Renal: uremia
  Vascular: myocardial infarction, post–myocardial infarction syndrome (Dressler's syndrome)
  Idiopathic

- Patients commonly present with complaints of centrally located chest pain made worse with recumbency and alleviated with leaning forward
- A 3-component pericardial friction rub may be auscultated with the diaphragm of the stethoscope at the left lower sternal border. It is best heard with the patient leaning forward after exhalation
- Findings on electrocardiography include diffuse concave upward ST elevation across precordial and limb leads (except aVR and V_1). Convex upward ST elevation is more indicative of ischemic insult
- Occasionally, PR-segment depression may be noted (prominent in lead II and usually absent in aVR and V_1)

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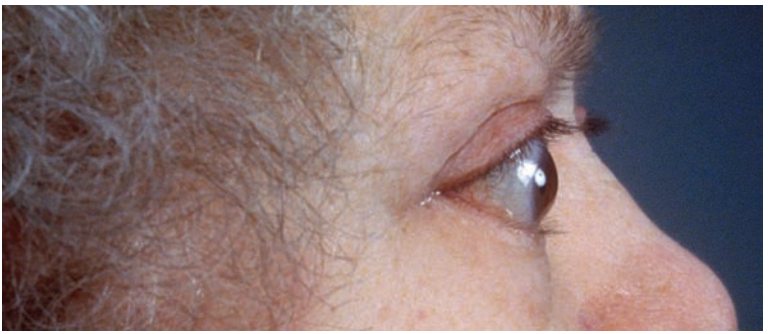
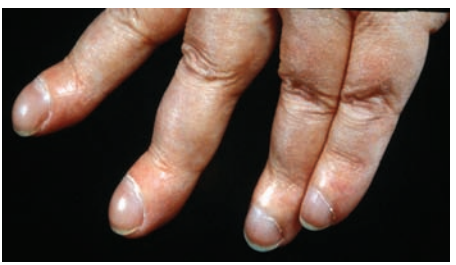
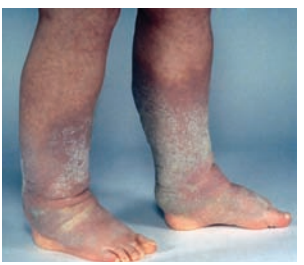






A 51-year-old woman presents with lower-extremity swelling, pruritus, blurred vision, and swollen fingertips. Which of the following treatments is *not* used for this condition?

a. Thioamides (e.g., methimazole and propylthiouracil)
b. Radioiodine therapy (^{131}I)
c. L-Thyroxine
d. Subtotal thyroidectomy
e. β-Adrenergic receptor antagonists

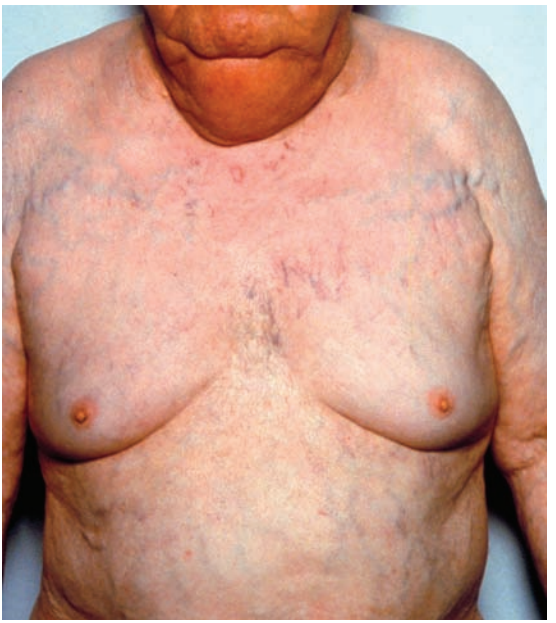
Graves' Disease

Answer: c

- Graves' disease is a multisystem autoimmune disease with a strong familial predisposition
- Characteristic triad:

  Hyperthyroidism
  Diffuse goiter
  Mesenchymal extrathyroidal effects (e.g., ophthalmopathy and pretibial myxedema)
- Ophthalmopathy and dermopathy are caused by accumulation of hyaluronic acid
- Clubbing is a well-described manifestation
- Autoantibodies that stimulate the thyroid-stimulating hormone–receptor cause the hyperthyroidism
- Thioamides, radioiodine therapy, and surgery are used to treat the disease
- β-Adrenergic receptor antagonists are used to control adrenergic manifestations of the disease
- L-Thyroxine is used only after a patient is rendered hypothyroid by definitive therapy (surgery or radioactive iodine)
- Eye and skin changes are not affected by treatment of the hypothyroidism

Reference
Habermann TM. *Mayo Clinic Internal Medicine Board Review 2004-2005*. Philadelphia: Lippincott Williams & Wilkins; 2004:208-210.



A 62-year-old ex-smoker presents with complaints of a 10-kg weight loss, night sweats, occasional hemoptysis, and light-headedness. Physical findings associated with this syndrome include all of the following *except*:

a. Facial flushing
b. Periorbital edema
c. Prominent venous channels on anterior chest wall
d. Increased jugular venous pressure
e. Hepatomegaly

Superior Vena Cava Syndrome

Answer: e

- Extrinsic compression of the pliable superior vena cava (SVC) may produce:

  Venous engorgement of the upper torso
  Facial flushing and edema
  Shortness of breath
  Blurred vision
  Hoarseness and stridor
  Orthopnea
  Light-headedness or syncope

- Approximately 80% of cases involving SVC syndrome are caused by underlying malignancy
- With bronchogenic carcinoma (most common), lymphoma, and other cancer that has metastasized to the thorax, 3% to 20% of cases will be complicated by SVC syndrome
- Benign SVC syndrome is characterized by an underlying nonmalignant cause, such as mediastinal lymphadenitis, iatrogenic causes (e.g., venous thrombosis of indwelling central line catheters), syphilitic aortic aneurysm, and tuberculous mediastinitis
- SVC syndrome can progress rapidly to tracheal obstruction or critically increased intracranial pressures
- In malignancy, early chemotherapy and radiation therapy are indicated. However, because of the possibly slow response, other reasonable options include surgical bypass of the obstruction or placement of endovascular stents

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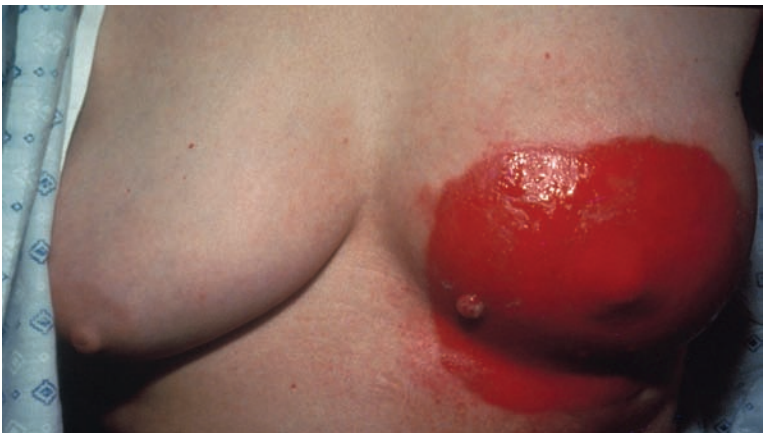

A 45-year-old woman presents with an erythematous, weeping, eczematous rash on the left breast (shown here). It has been present for 1 month and has not responded to topical corticosteroids. What should be done next?

a. Chest radiography
b. Biopsy
c. Topical antifungal therapy
d. Mammography
e. Test for *BRCA1* and *BRCA2* genes

Paget's Disease of the Breast

Answer: b

- Paget's disease of the breast usually begins as a reddened, scaly eczema of the areola. The case shown on the preceding page is particularly advanced
- Virtually all patients with Paget's disease of the breast have an underlying ductal carcinoma, whereas 50% of patients with extramammary Paget's disease have an underlying malignancy
- Paget and cancer cells have the same origin
- Any eczematous lesion of the breast that does not respond to topical corticosteroids should undergo biopsy
- Women with Paget's disease of the breast are the same average age as women with other forms of breast cancer
- Mastectomy is the usual treatment
- In recent years, breast-conserving surgery with radiotherapy has been used with some success

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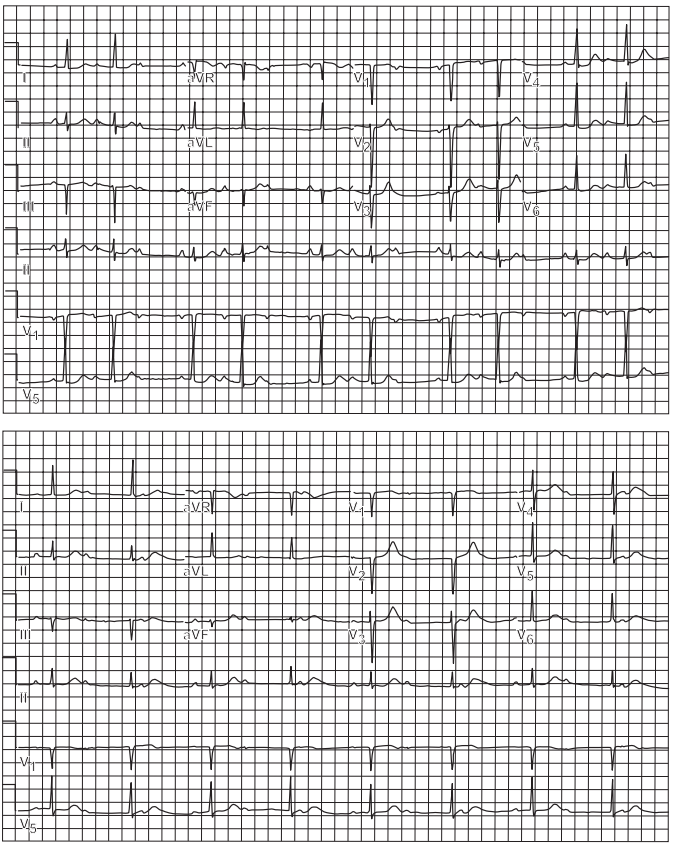

The electrocardiograms (ECGs) shown here were obtained from 2 asymptomatic patients who were having routine physical examinations. Which of these ECGs shows a potential class II indication for pacemaker placement according to the American Heart Association (AHA) and the American College of Cardiology (ACC)?

a. The top ECG
b. The bottom ECG

Mobitz Type I Atrioventricular Block and Third-Degree Atrioventricular Block

Answer: b

- Three types of atrioventricular (AV) conduction block:

  First-degree
  PR interval is more than 0.2 second and is constant
  Second-degree
  Mobitz type I: Also known as Wenckebach block. Characterized by a progressive increase in the PR interval until a P wave fails to conduct (top ECG)
  Mobitz type II: No progressive prolongation of PR interval. However, abrupt conduction failure occurs
  Third-degree
  Complete heart block with AV dissociation (bottom ECG). If the QRS complex is narrow, this is likely AV nodal block, whereas a widened QRS likely implies His-Purkinje system block

- According to the AHA and the ACC, implantation of permanent pacemakers in asymptomatic patients is according to the following guidelines:

  Class I (universally accepted):
  Third-degree AV block with documented asystole 3 seconds or more, or escape rate less than 40 beats per minute while awake
  Third-degree AV block or second-degree Mobitz type II block with chronic trifascicular or bifascicular block
  Class II (potential indications):
  Third-degree AV block with escape rate more than 40 beats per minute
  Second-degree AV block with no evidence of trifascicular or bifascicular block
  Class III (pacing not indicated):
  Asymptomatic first-degree and second-degree Mobitz type I AV block

References

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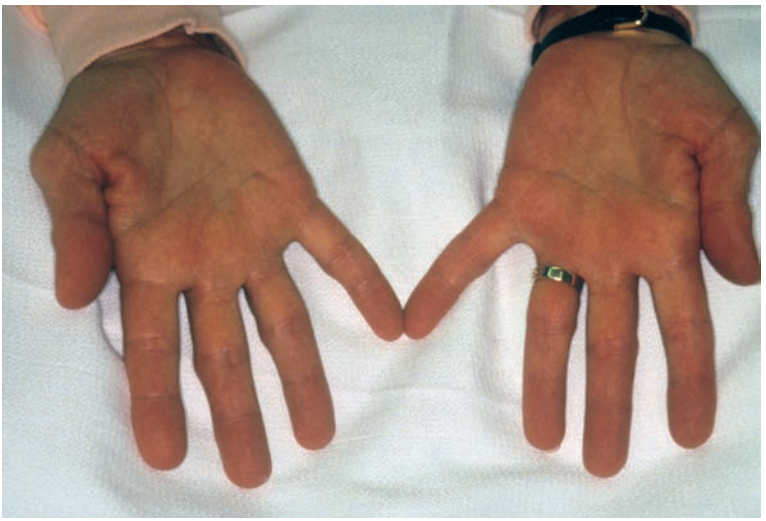

The skin coloration shown here developed in an asymptomatic 26-year-old woman. The soles of her feet have a similar coloration. Her sclerae are white. What is the likely cause?

a. Acute intravascular hemolysis
b. Cirrhosis of the liver
c. Splenic vein thrombosis
d. Primary biliary cirrhosis
e. Carotenemia

Carotenemia

Answer: e

- Xanthoderma (yellow-orange skin discoloration) is caused by eating large amounts of foods high in β-carotene, such as:

Oranges	Sweet potatoes
Mangoes	Milk fat
Apricots	Egg yolk
Carrots	All green vegetables

(This patient ate a large spinach salad every day for many weeks)

- Unlike jaundice, the sclerae remain white in carotenemia
- Serum also may be more orange-yellow than usual, and the serum β-carotene level is usually increased
- Other clues to the diagnosis include normal results of liver function tests, normal bilirubin and hemoglobin values, and no evidence of hemolysis
- Most carotene is converted to vitamin A aldehyde (retinol) in the intestine. Nonconverted carotene is absorbed without change and gives serum its normal yellow color
- There are no long-term sequelae. The skin discoloration resolves quickly (within 4-6 weeks) after decreased ingestion of foods rich in β-carotene
- Uncontrolled diabetes mellitus may result in carotenemia as a result of extreme hyperlipidemia. Differentiation from benign carotenemia usually is not difficult

References

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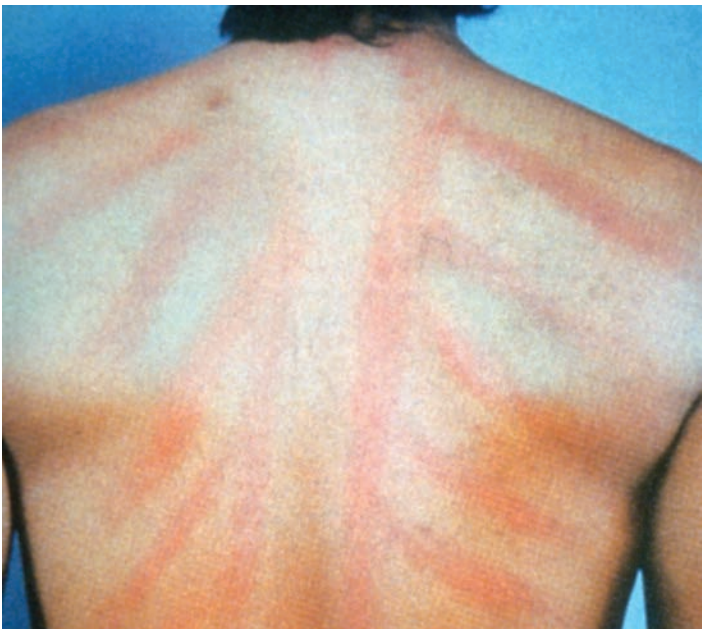

A 27-year-old Southeast Asian man presented with a history of headaches. The lesions shown here were found on examination. What is the likely cause?

a. "Christmas tree" pattern of pityriasis rosea
b. Linear erythema multiforme
c. Traditional medicine treatment
d. Subcutaneous parasitic infection
e. Abusive trauma

Figure from Crutchfield CE III, Bisig TJ. *N Engl J Med*. 1995;332:1552. By permission of the Massachusetts Medical Society.

Coining

Answer: c

- Coining is a common traditional healing practice among many Asian cultures
- It is also known as skin scraping, kua-sha, coa gio, and spooning
- It may involve spreading hot oil on the trunk. Scraping also may be applied to the neck, nose, forehead, elbows, and knees
- A coin is then used to mark the skin of the back to promote healing of illness
- The traditional theory is that coining creates an area for disease-causing wind to leave the body. This is thought to improve circulation and decrease inflammation
- It most often is used for minor ailments of fever, headache, cough, flu, and heat stroke
- This patient had migraine headaches
- The lesions usually heal, but scarring and hyperpigmentation can occur
- In children, it is sometimes misdiagnosed as child abuse
- Occasionally, serious complications (e.g., full-thickness burns) occur
- Overall frequency is thought to be underreported because of suspicion of the medical "establishment"

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Crutchfield CE III, Bisig TJ. Images in clinical medicine: coining. *N Engl J Med*. 1995;332:1552.

Look KM, Look RM. Skin scraping, cupping, and moxibustion that may mimic physical abuse. *J Forensic Sci*. 1997;42:103-105.

A 77-year-old woman is admitted for recurrent anemia, jaundice, and gross hematuria. She underwent aortic valve replacement for severe aortic stenosis 1 month ago. Which of the following is true?

a. The patient's serum plasma is likely pink-red
b. The patient's serum bilirubin value is likely normal
c. Urine dipstick test is negative for heme
d. The patient's creatine kinase level is more than 10,000 U/L
e. The patient's plasma haptoglobin level is increased

Hemoglobinuria

Answer: a

- Hemoglobinuria is associated with both inherited and acquired hemolytic disorders

  Inherited: glucose-6-phosphate dehydrogenase deficiency, sickle cell disease, thalassemias, hereditary spherocytosis
  Acquired: autoimmune hemolytic anemia, transfusion mismatch, traumatic (e.g., prosthetic valve as in this case, march hemoglobulinuria), thermal injury, infection (e.g., malaria)

- Serum hemoglobin binds to haptoglobin. When the haptoglobin levels decrease, the free hemoglobin (methemoglobin) is cleared by the kidneys
- Serum free hemoglobin concentration must exceed at least 25 mg/100 mL before hemoglobinuria ensues
- Urine dipstick test is positive for heme in both myoglobinuria and hemoglobinuria, but only in hemoglobinuria is the serum plasma pink-red. This occurs because haptoglobin binds hemoglobin as opposed to myoglobin
- In patients with a mechanical valve prosthesis, perivalvular leakage, even if hemodynamically insignificant, can induce hemolysis. The irregular valvular contour may generate enough shear stress to damage erythrocytes
- In cases of traumatic hemolysis due to valve replacements, reoperation is often necessary and may lead to immediate reversal of the hemolysis and consequent resolution of hemoglobinuria

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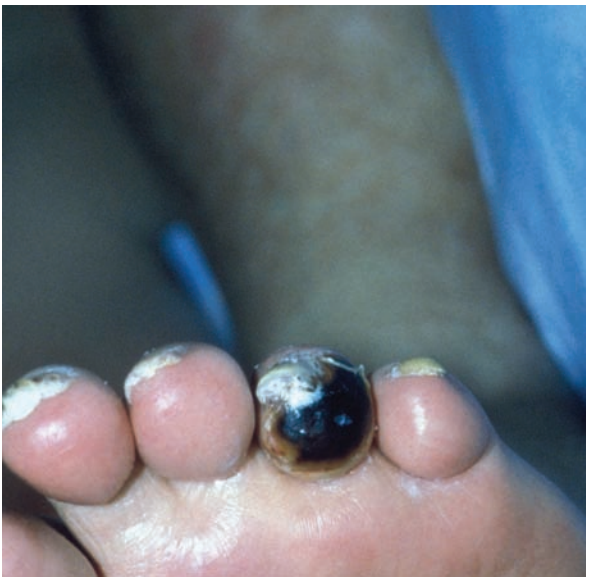

The painful ulcer shown here developed after minor trauma in a 60-year-old smoker with known coronary artery disease. His pedal pulses are very diminished. What is the likely cause?

a. Venous ulcer
b. Arteriolar (hypertensive) ulcer
c. Neurotrophic ulcer
d. Arterial (ischemic) ulcer
e. Cryoglobulinemic ulcer

Ischemic Ulcer (Arterial Sclerotic Occlusive Disease)

Answer: d

- Arterial sclerotic occlusive (ASO) disease reduces blood flow below the viability threshold
- Clinical signs suggestive of ASO disease and risk of ulceration include calf muscle atrophy, loss of hair over the toes and feet, thickened toenails, delayed capillary refill, and diminished or absent pedal pulses
- Ulcers are often distal in distribution. Toes, anterior shins, and posterior calves are common sites
- Most ischemic ulcers are painful unless associated with neuropathy. Surrounding skin may be cold or pale or have fixed erythema
- Ankle-brachial index (ABI) less than 0.8 is suggestive of significant arterial stenosis, and patients may be candidates for surgical intervention
- Arteriosclerosis must be considered a generalized disease. ABI less than 0.9 is associated with a 2-fold increased risk of coronary artery disease and a 4-fold increased risk of cerebrovascular accident or transient ischemic attack
- Risk factors for ASO include cigarette smoking (6-fold increase), diabetes (6-fold increase), hypercholesterolemia, and hypertension

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Goodfield M. Optimal management of chronic leg ulcers in the elderly. *Drugs Aging*. 1997;10:341-348.

Hafner J. Management of arterial leg ulcers and of combined (mixed) venous-arterial leg ulcers. *Curr Probl Dermatol*. 1999;27:211-219.

Kiehlmann I, Lechner W. Complications in the treatment of leg ulcers. *Curr Probl Dermatol*. 1999;27:170-173.

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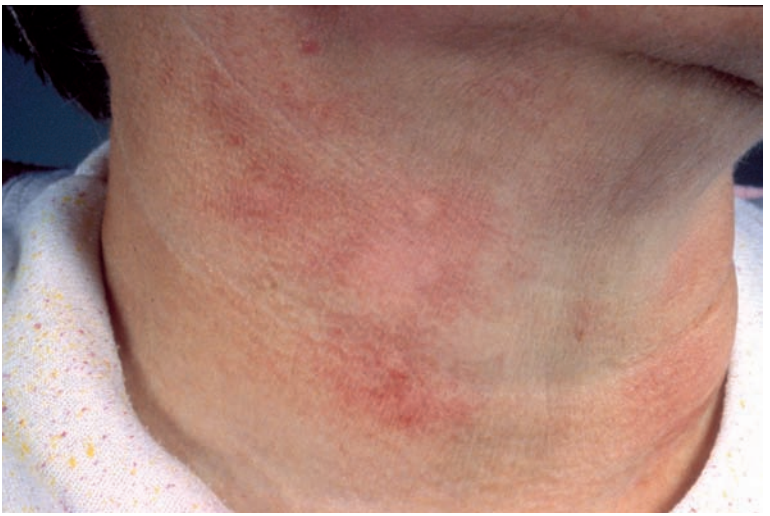

A 24-year-old woman has the lesions shown here in cold-exposed areas and wheezes in the cold. What should you counsel her to avoid?

a. Cyproheptadine
b. Swimming
c. Ice cream
d. Angiotensin-converting enzyme inhibitors
e. Pregnancy

Cold Urticaria

Answer: b

- Cold urticaria is classic wheal-and-flare urticaria in cold-exposed areas
- Typical syndromes may be shown diagnostically by applying an ice cube to the skin; a wheal or angioedema develops after the skin rewarms
- Cold exposure triggers IgE-mediated mast cell degranulation, which causes histamine release
- Cold urticaria may be associated with wheezing or syncope
- Most cases are idiopathic
- Secondary cases may be associated with the following:

  Cryoglobulins
  Cold hemolysins
  Chronic lymphocytic leukemia
  Mononucleosis
  Syphilis
  Various other disorders

- Immersion in cold water can lead to angioedema, generalized urticaria, and shock. Therefore, patients should be advised against swimming
- In addition to avoidance of precipitating cold exposures, cyproheptadine (histamine$_1$ blocker) has been useful for treatment

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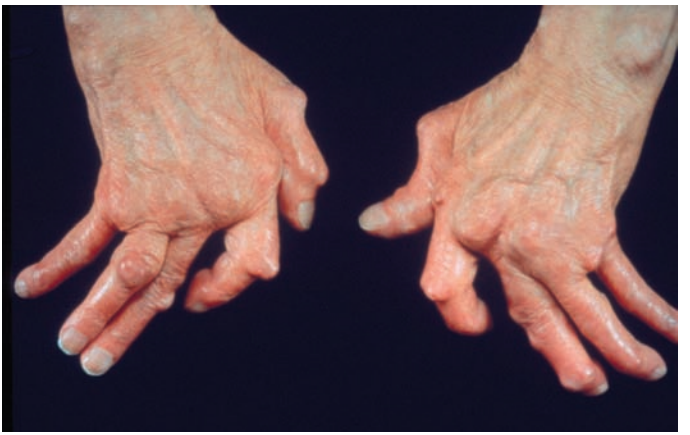

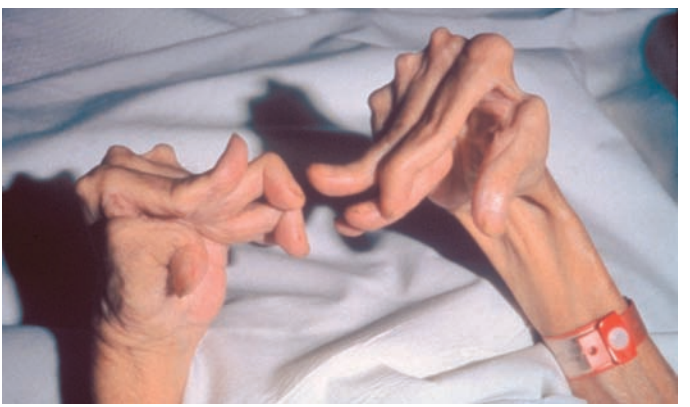

These elderly women present with complaints of chronic polyarticular arthritis. Musculoskeletal deformities of this disease include all of the following *except*:

a. Swan-neck deformity of digits
b. Boutonnière deformity of digits
c. Ulnar deviation of metacarpophalangeal joints
d. Heberden's nodes
e. Pes planus

Answer: d

- Rheumatoid arthritis is a systemic inflammatory disease involving the synovial lining of joints
- Most commonly affected joints include the metacarpophalangeal and proximal interphalangeal joints
- Boutonnière deformity is hyperextension of the distal interphalangeal joint and flexion of the proximal interphalangeal joint
- Swan-neck deformity is hyperextension of proximal interphalangeal joint and flexion of distal interphalangeal joint
- Involvement usually predominates in small joints and is bilateral
- Patients may complain of prolonged morning stiffness and of symptoms that worsen with repetition
- Complications include cervical spine instability, especially atlantoaxial subluxation. Preoperative radiographs may include cervical spine flexion and extension views
- Treatment options other than physical medicine, nonsteroidal anti-inflammatory drugs, and corticosteroids include use of disease-modifying agents of rheumatic disease:

  Methotrexate
  Hydroxychloroquine
  Intramuscular injection of gold salts
  Penicillamine
  Sulfasalazine
  Leflunomide
  Azathioprine
  Cyclophosphamide

Reference

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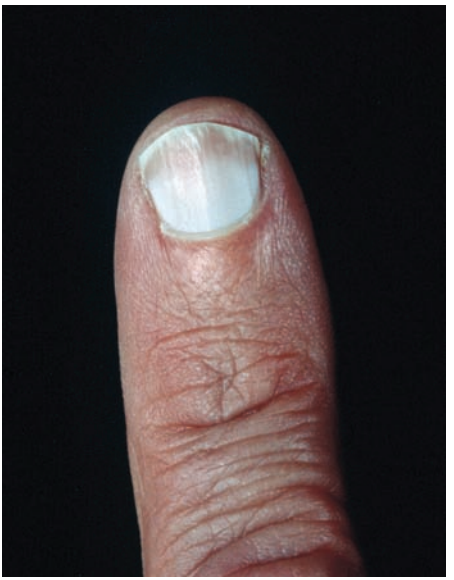

All of the following are associated with this nail change *except*:

a. Cirrhosis
b. Adult-onset diabetes mellitus
c. Congestive heart failure
d. Age
e. Ulcerative colitis

Terry's Nails

Answer: e

- Terry's nails is a condition characterized by whitening, ground-glass appearance of the proximal or entire nail
- It results from changes in the nail bed, not the nail itself
- Often, a 0.5- to 3.0-mm zone of normal appearance remains in the distal portion of the nail
- The distal band is now known to be due to telangectasias
- In 1954, Terry first described the nail changes in association with cirrhosis
- The condition also is associated with chronic congestive heart failure, adult-onset diabetes, and age
- The odds ratio for the presence of cirrhosis, congestive heart failure, or diabetes is 2.7 for all patients
- The odds ratio for cirrhosis, congestive heart failure, or diabetes is 5.3 for patients younger than 50 years
- Thus, the presence of Terry's nails is a more important diagnostic sign in younger patients

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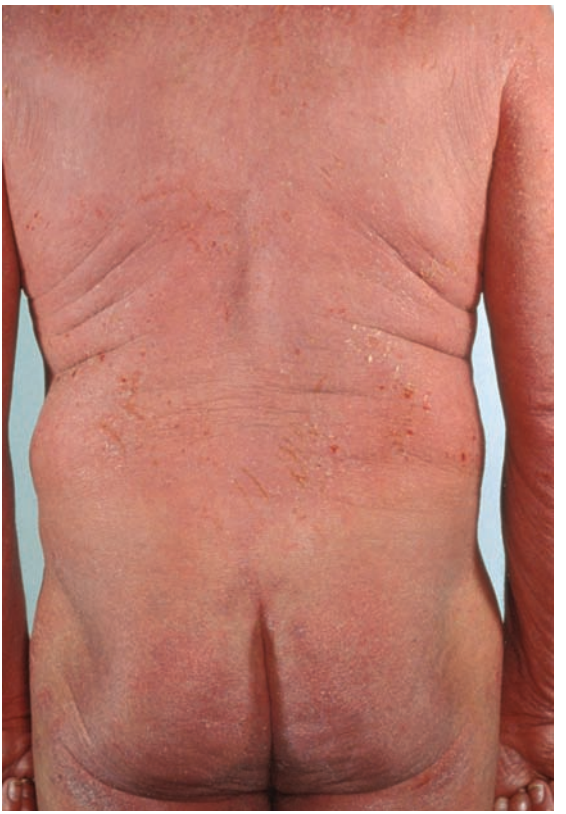


A 45-year-old man presents with redness, edema, and scaling on his body. This problem initially started as multiple red, pruritic patches, which progressed over the course of 1 to 2 weeks. Which of the following is known to cause this condition?

a. Drugs
b. Flare of cutaneous disease
c. Malignancy
d. Acquired immunodeficiency syndrome
e. All of the above

Erythroderma

Answer: e

- Erythroderma (also known as exfoliative dermatitis) is the result of inflammation and rapid turnover of the epidermis, resulting in erythema and scaling
- Patients experience:

  Malaise
  Pruritus
  Chills (due to heat loss)
  Hypotension
  Congestive heart failure

- The most common causes of erythroderma:

  Preexisting cutaneous diseases
  Drug reactions
  Malignancies

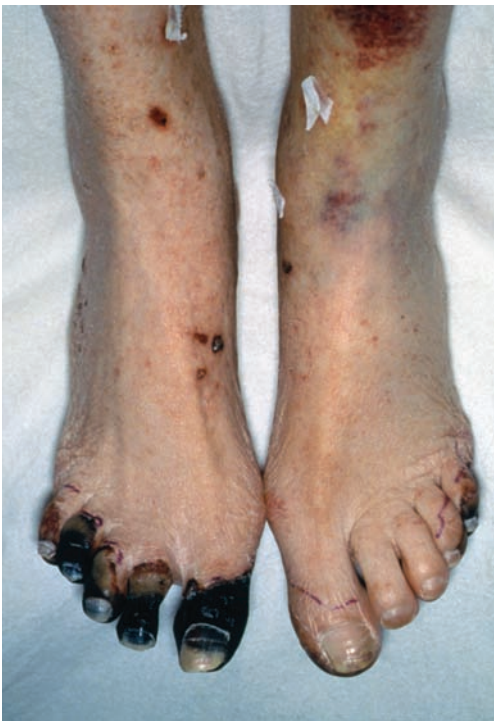
- Theoretically, any drug can cause erythroderma
- The most commonly associated malignancies:

  Cutaneous T-cell lymphoma (mycosis fungoides)
  Acute leukemia
  Chronic leukemia

- A specific underlying cause is not found in approximately 25% of patients with erythroderma

Reference

Karakayli G, Beckham G, Orengo I, et al. Exfoliative dermatitis. *Am Fam Physician*. 1999;59:625-630.



Type II cryoglobulinemia is most commonly associated with which of the following?

a. Primary sclerosing cholangitis
b. Hepatitis C
c. Hepatitis B
d. Scleroderma
e. Hemochromatosis

Cryoglobulinemia

Answer: b

- Cryoglobulins are proteins (immunoglobulins) that precipitate at lower temperatures
- "Essential" cryoglobulinemia has no clearly identifiable cause, and "secondary" cryoglobulinemia may be associated with various disorders
- Type II or III mixed cryoglobulinemia may occur in up to 50% of patients with hepatitis C
- More than 70% of patients with cryoglobulinemia will have cutaneous purpura, arthralgias, or weakness. Collectively, this is known as Meltzer's triad
- Other clinical features include headaches, visual disturbances, ischemic digital ulceration or infarction, Raynaud's phenomenon, stroke, hematochezia, myocarditis, and polyneuropathy
- Renal failure may occur with acute nephritic or nephrotic syndrome
- Diagnosis is based on sampling venous blood. The sample must first be completely coagulated before cold temperature (4°C) exposure for 96 hours. One then observes for precipitation of the cryoproteins
- Vasculitis is more prominent in type II cryoglobulinemia and results from immune complex deposition on the vascular endothelium. Hypocomplementemia is a supportive laboratory marker and occurs in up to 90% of patients with active cryoglobulinemic vasculitis

References

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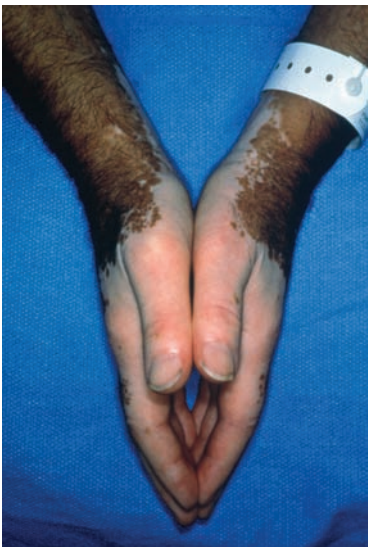
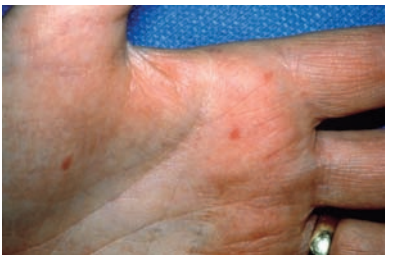




The hands shown here are those of a 59-year-old man who presents with heartburn and Raynaud's phenomenon. Which of the following is *not* part of this syndrome?

a. Skin thickening of the trunk
b. Sclerodactyly
c. Esophageal dysmotility
d. Telangiectasias
e. Calcinosis cutis

CREST Syndrome

Answer: a

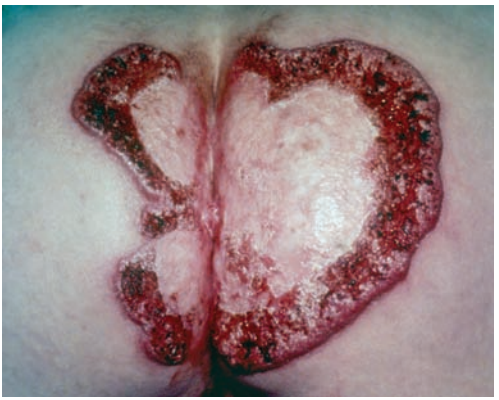
- CREST syndrome is also known as "limited scleroderma"
- CREST syndrome is a multisystem disorder characterized by the following:

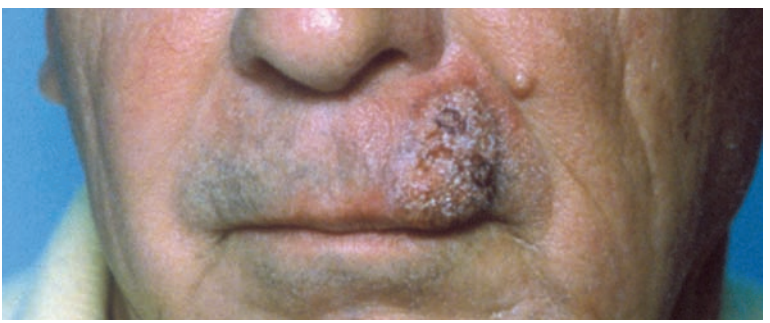
  *C*alcinosis cutis
  *R*aynaud's phenomenon
  *E*sophageal dysmotility
  *S*clerodactyly
  *T*elangiectasias

- Skin involvement in CREST syndrome is limited to the extremities (whereas it is much more diffuse in scleroderma)
- Involvement of joints and tendons is rare
- Lung involvement (e.g., reduced diffusing capacity, pulmonary hypertension) occurs in 70% of patients
- Anticentromere antibody is found in 70% to 90% of patients, and antiscleroderma-70 antibody in 10%
- Patients with CREST are at increased risk for primary biliary cirrhosis
- Raynaud's phenomenon usually occurs before the other changes

Reference

Habermann TM. *Mayo Clinic Internal Medicine Board Review 2004-2005*. Philadelphia: Lippincott Williams & Wilkins; 2004:996.





The painless skin lesions shown here developed in a 62-year-old gardener from the upper midwestern United States. Chest radiography showed a perihilar mass worrisome for carcinoma. Biopsy of the lesions showed "broad-based buds." What is the diagnosis?

a. Histoplasmosis
b. Blastomycosis
c. Coccidioidomycosis
d. Mucormycosis
e. Actinomycosis

Blastomycosis

Answer: b

- Blastomycosis is a dimorphic fungus with "broad-based buds" on biopsy specimens stained with periodic acid-Schiff or methenamine silver. Mycelial form grows in the laboratory
- It is endemic to the southeastern and upper midwestern United States (Ohio and Mississippi river valleys)
- Soil exposure is common
- The primary infection is usually pulmonary and may be relatively mild. However, perihilar adenopathy, pleural fibrosis, and cavitation may occur mimicking carcinoma or tuberculosis. Granulomas on biopsy are noncaseating
- Extrapulmonary sites are usually secondarily involved. Most common areas are skin, bone, urogenital tract, and central nervous system
- Skin lesions are most often painless and nonpruritic with a sharp border that spreads
- Diagnosis is by biopsy, stains, and culture. Serologic results are not reliable
- Initial treatment is with amphotericin B for severe cases or immunocompromised patients. Itraconazole given for 6 months is the treatment of choice for follow-up after amphotericin B therapy and for monotherapy of non–life-threatening cases

References

Habermann TM. *Mayo Clinic Internal Medicine Board Review 2004-2005*. Philadelphia: Lippincott Williams & Wilkins; 2004:568.

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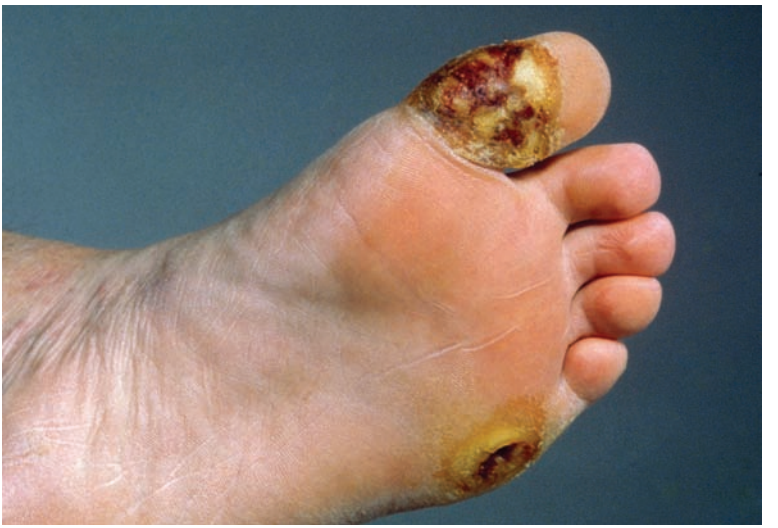

The painless ulcers shown here developed in a 55-year-old man with diabetes who did not recall any antecedent trauma. What type of ulcer is this?

a. Venous
b. Arterial
c. Arteriolar
d. Neurotrophic
e. Embolic

Neurotrophic Ulcer

Answer: d

- Neurotrophic ulcers tend to develop in sites of chronic trauma in patients with neuropathies, especially diabetes
- Pressure points are common sites of involvement:

  Soles of the feet under the metatarsal heads
  Under the heel
  Over the toes
  On the medial side of the first metatarsal
  Over the malleoli
- The ulcers are usually painless, unlike most vascular ulcerations
- The surrounding skin is often pale
- Treatment involves:

  Protecting the area
  Increasing the blood supply
  Debriding calluses and dead tissues
  Treating secondary infection
- After wound healing, specialized footwear is often indicated to protect bony prominences from further trauma
- Vigilant podiatric care is a necessity for patients with diabetes

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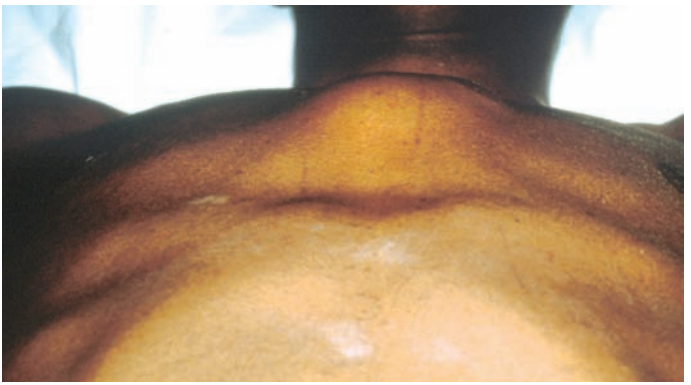

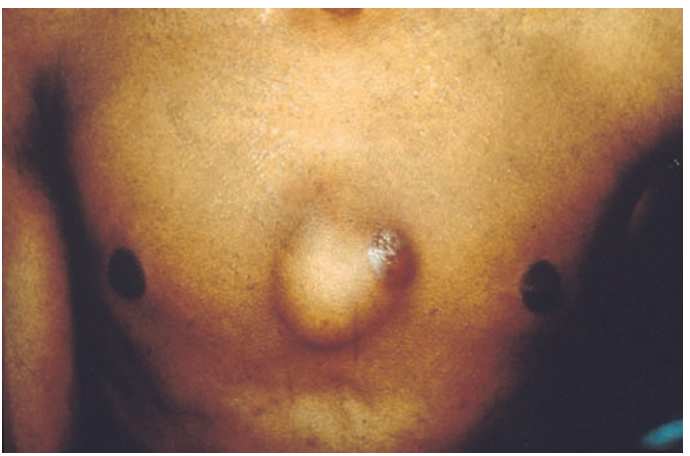

In patients who are positive for human immunodeficiency virus, primary tuberculosis develops after exposure to *Mycobacterium tuberculosis* in approximately what percentage?

a. 10%
b. 40%
c. 60%
d. 80%
e. 95%

Sternal Tuberculoma

Answer: b

- The leading cause of death from infection worldwide remains tuberculosis
- Three million estimated deaths per year are attributed to this disease, and the overwhelming majority of cases are in third-world developing countries
- The most common mode of transmission is by inhalation of droplet nuclei from expectorated sputum
- The efficacy of the bacille Calmette-Guérin vaccine ranges from 0% to 80% for primary tuberculosis, and the vaccine is not effective for preventing reactivation of illness
- Among immunocompetent individuals exposed to tuberculosis, 30% will become infected, but less than 5% of this group will progress to active disease
- In patients with human immunodeficiency virus exposed to *Mycobacterium tuberculosis*, primary tuberculosis will develop in approximately 40%
- Tuberculous lymphadenitis (scrofula) is the most common type of extrapulmonary tuberculosis, but tuberculosis can involve almost any organ, including the skin, as in this case
- Cervical lymph nodes are commonly affected. Abscess and sinus tracts may develop with subsequent cutaneous discharge of pus
- Worldwide, about 10% of cases are resistant to at least one antituberculous medication. Primary multidrug resistance remains low, at approximately 0.2%

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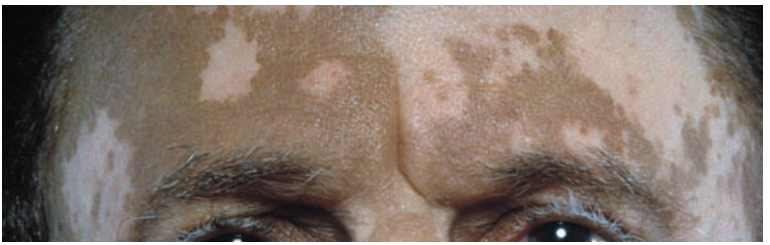

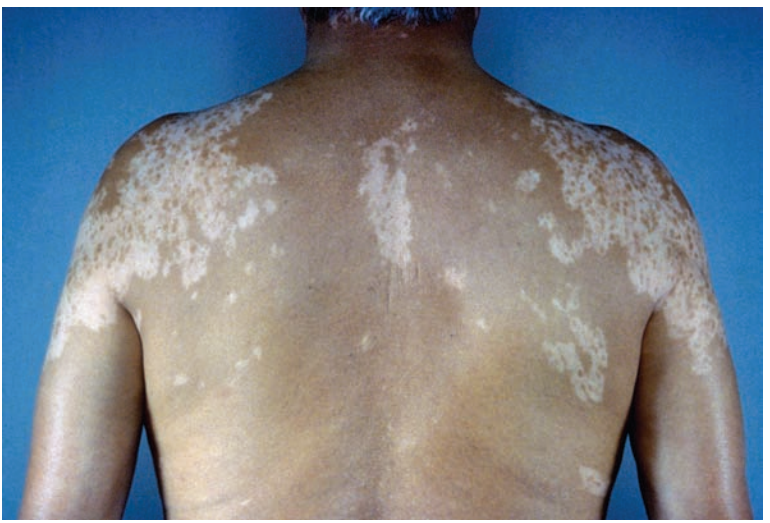

A 35-year-old man has recurrent aseptic meningitis, blurred vision, painful reaction to loud noises, and the skin changes shown here. What is this syndrome called?

a. Achard-Thiers syndrome
b. Bamberger-Marie syndrome
c. Dejerine-Klumpke syndrome
d. Rokitansky-Küster-Hauser syndrome
e. Vogt-Koyanagi-Harada syndrome

Answer: e

- Diagnosis requires:

  No history of eye trauma or surgery
  At least 3 of the following:
  Chronic bilateral iridocyclitis
  Posterior uveitis, often associated with retinal detachments
  Tinnitus, neck stiffness, cranial nerve palsies, cerebrospinal fluid pleocytosis, or other central nervous system disturbance
  Alopecia, poliosis, or vitiligo (face or scalp most commonly)

- In 90% of patients, poliosis or clumps of whitened hair develop in the midst of normal-colored hair. Piebald eyelashes, i.e., clumps of white eyelashes in the midst of normal eyelashes, are shown in this case
- Patients also may have hearing loss, vertigo, dysacusis, headache, drowsiness, nausea, vomiting, dysphagia, and confusion
- Most patients present between the 2nd and 5th decades of life
- Ophthalmologic consultation should be obtained
- Pathogenesis is unknown but is thought to be related to T-cell–mediated autoimmunity to uveal or retinal antigens
- With immunosuppressive treatment, two-thirds of patients obtain vision of 20/40 or better

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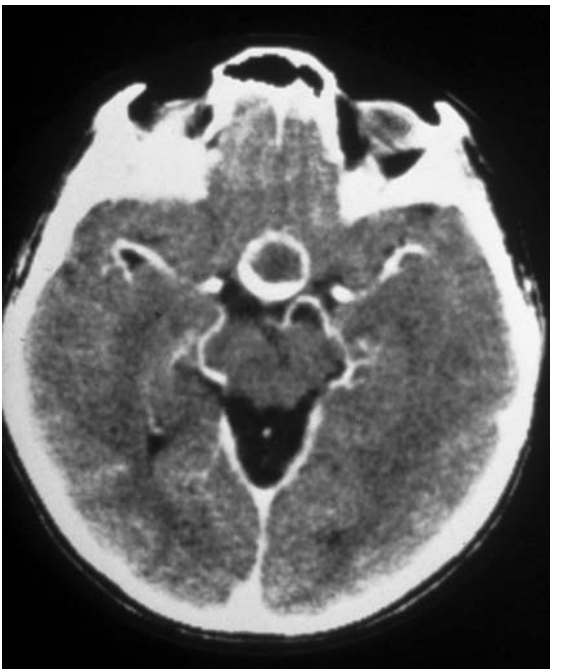

The condition shown here is associated with which of the following?

a. Weight loss
b. Truncal ataxia
c. Hyperprolactinemia
d. Syndrome of inappropriate secretion of antidiuretic hormone
e. Right arm weakness

Craniopharyngioma

Answer: c

- Craniopharyngioma is the most common tumor in the pituitary region in childhood, although it may occur at any age
- It derives from remnants of Rathke's pouch and evolves as a slow-growing, encapsulated, squamous cell tumor
- Clinical presentation may include the following:

  Obstructive hydrocephalus
  Diabetes insipidus
  Hyperprolactinemia
  Hypopituitarism

- Computed tomography of the head commonly shows a cystic mass or enlarged sella turcica with suprasellar calcification
- Small craniopharyngiomas may be amenable to surgical excision, whereas larger tumors necessitate decompression. Radiotherapy is an additional management
- Preoperative factors such as lethargy and visual deterioration are associated with poor postoperative outcomes
- Tumors that are mainly sellar are reported to have good postoperative outcomes
- Hypothalamic injury is one of the most dangerous complications that may occur after tumor resection

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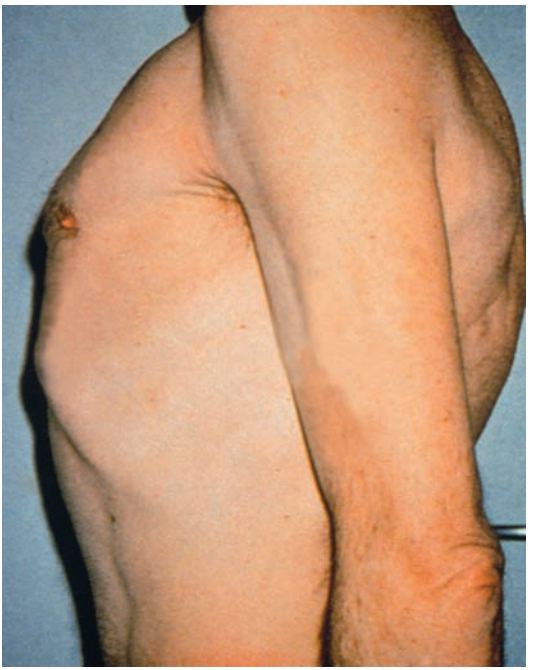

The barrel chest in patients with emphysema is caused by which of the following?

a. Excess right ventricular strain
b. Associated intrinsic rib cage deformity
c. Pulmonary hyperinflation and underlying bullous disease
d. Pleural inflammation
e. Associated kyphoscoliosis

Emphysema

Answer: c

- Emphysema is characterized by dilatation of terminal bronchial air spaces. Compromise of lung elastin in the pulmonary interstitium is contributive
- α_1-Antitrypsin deficiency (panlobular, lower-zone emphysema) occurs in 5% of patients with emphysema
- Most emphysema is the centrilobular form and caused by smoking
- Patients generally appear of thin frame and maintain near normal Pa_{O_2}
- Work of breathing is typically increased, and pursed-lip breathing may be evident ("pink puffer")
- Chest radiography may show hyperinflation
- Severe emphysema can lead to CO_2 retention
- Complications include bullous rupture and pneumothorax
- Treatment may include the following:

  Bronchodilators
  Pulmonary rehabilitation
  Tobacco cessation
  Oxygen therapy
  Lung-volume reduction surgery (remains controversial)

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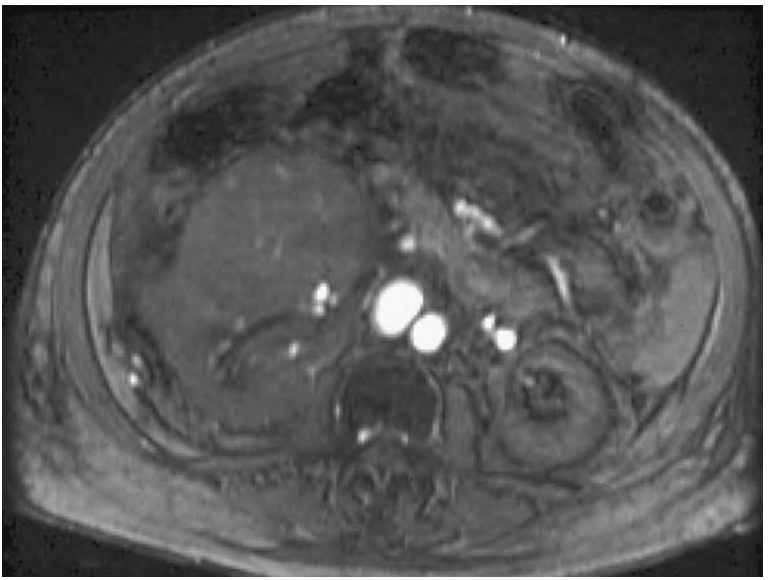

A 65-year-old woman presents with fatigue, right flank pain, and ankle edema. Laboratory studies show increased liver enzyme values and microscopic hematuria. What is the diagnosis?

a. Choledocholithiasis
b. Nephrolithiasis
c. Hepatocellular carcinoma
d. Meigs' syndrome
e. Renal cell carcinoma

Renal Cell Carcinoma

Answer: e

- Renal cell carcinoma originates in the renal cortex
- Risk factors for renal cell carcinoma include the following:

  Male sex
  Older age
  Smoking
  Obesity
  Hypertension
  Polycystic kidney disease
  Environmental toxins (e.g., asbestos, heavy metals, petroleum products)
  Unopposed estrogen therapy
  von Hippel-Lindau syndrome

- The classic triad (hematuria, abdominal pain, and a palpable mass) occurs in only 10% of patients
- Stauffer syndrome occurs in the setting of renal cell carcinoma; it is marked by increased values on liver function tests. It is not due to metastasis, but rather cholestasis
- Patients can present with only constitutional symptoms (e.g., fevers, sweats, weight loss, malaise); indeed, this tumor can present in so many ways it is called the "internist's tumor"
- Surgery is the mainstay of treatment; hormonal and chemotherapeutic agents have little or no benefit

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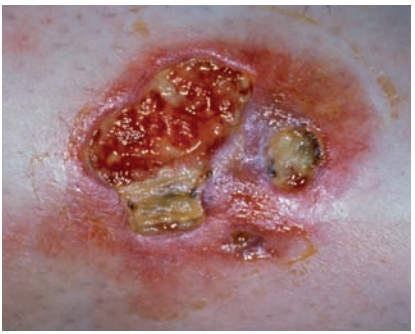

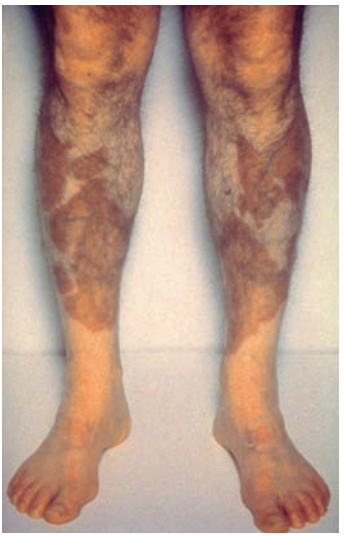

A 38-year-old man with a chronic illness presents with skin changes of the shins, as shown here. One of the plaques has ulcerated. What is the most likely chronic underlying disease?

a. T-cell lymphoma
b. Diabetes mellitus
c. Sarcoidosis
d. Psoriasis
e. Vasculitis

Necrobiosis Lipoidica Diabeticorum

Answer: b

- The plaques of necrobiosis lipoidica diabeticorum classically are described as:

  Occurring on the shins
  Yellow-brown
  Atrophic
  Telangiectatic
  Occasionally ulcerating

- Two-thirds of patients with necrobiosis lipoidica diabeticorum have diabetes mellitus
- However, only 0.3% of patients with diabetes mellitus have necrobiosis lipoidica diabeticorum
- Treatment consists of the following:

  Exquisite skin care
  Whirlpool therapy
  Antibiotics only if superinfection occurs
  Topical psoralen and ultraviolet light A (PUVA) therapy may be of some benefit

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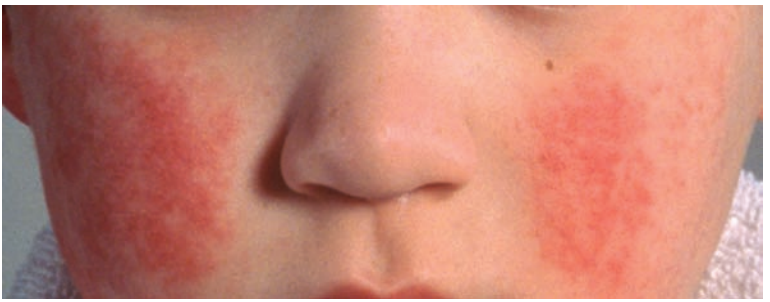

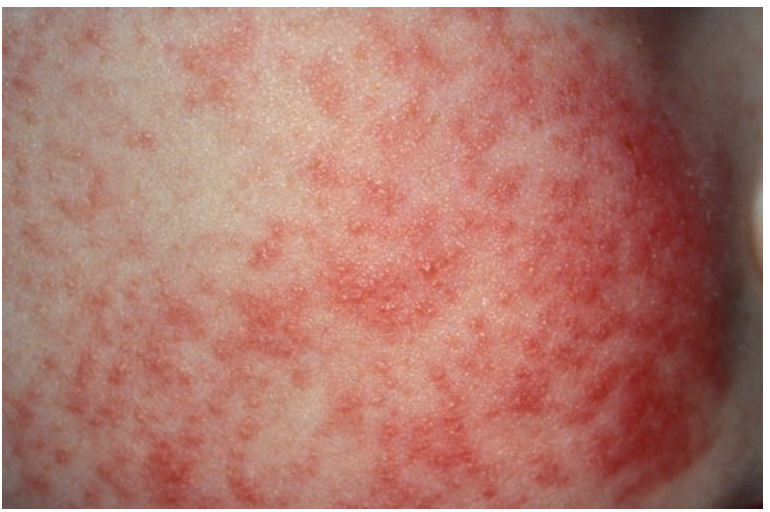

A 5-year-old boy has a 2-day history of fever, malaise, and the rash shown here. The virus that caused this exanthem also causes which one of the following?

a. Aplastic crisis
b. Myalgia, arthralgia, and arthritis
c. Hydrops fetalis
d. Red cell aplasia
e. All of the above

Erythema Infectiosum (Fifth Disease)

Answer: e

- Parvovirus B19 is a very common virus: 50% are seropositive by age 15 years, 80% to 100% are seropositive by age 70 years
- Parvovirus B19 is associated with 5 major syndromes:

  Exanthems (erythema infectiosum, or fifth disease) in children and adults, usually self-limiting
  Aplastic crisis in patients with hemolytic anemia
  Myalgia, arthralgia, and arthritis in adults (women more than men)
  Hydrops fetalis or fetal death if pregnant mother is infected (5% chance)
  Red cell aplasia or chronic anemia can develop in immunosuppressed patients (therapy is with intravenous immunoglobulin)
- Less common (and less definite) associations include meningitis, encephalopathy, brachial plexus neuropathy, polyarteritis nodosa, Wegener's granulomatosis, giant cell arteritis, acute hepatitis, Kawasaki's disease, myocarditis, and Raynaud's phenomenon
- Diagnosis of acute infection is by detection of specific IgM antibodies or by polymerase chain reaction
- Historically, the other 4 childhood exanthems were (in order): measles, scarlet fever, rubella, and exanthem subitum

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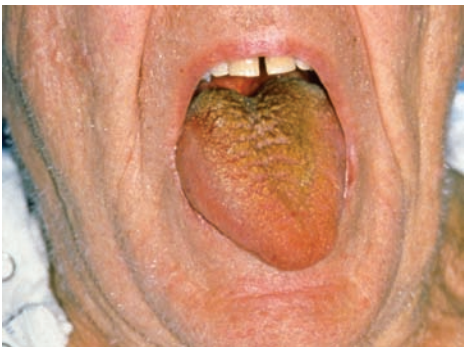

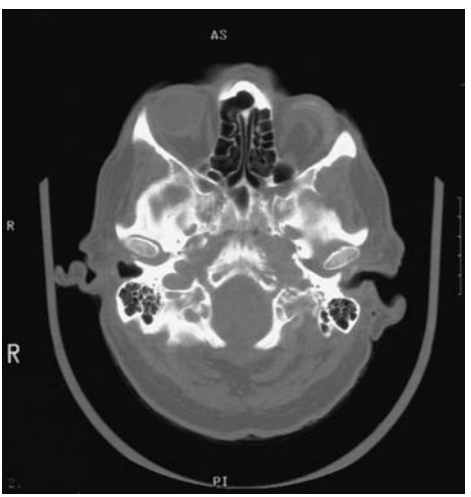

A 78-year-old male smoker presents with a 6-month history of left neck pain and a 2-week history of slurred speech and difficulty swallowing. Where is the lesion?

a. Right cranial nerve XI
b. Left cranial nerve XI
c. Right cranial nerve XII
d. Left cranial nerve XII
e. None of the above

Cranial Nerve XII Palsy

Answer: d

- With cranial nerve XII palsy, the tongue deviates to the "bad," or affected, side
- The hypoglossal canal is the site of the lesion
- Ipsilateral neck pain strongly suggests the lesion is due to metastatic carcinoma (in this case, lung cancer)
- Indeed, in some series of cranial nerve XII palsy, nearly half of cases are due to malignancy
- Because paralysis of cranial nerve XII can be an ominous sign, thorough evaluation is warranted
- Other than malignancy, causes of cranial nerve XII palsy include the following:

  Benign tumors
  Vertebrobasilar infarcts
  Internal carotid artery dissection
  Trauma
  Postsurgical complication in head and neck surgery
  Infection
  Inflammatory diseases

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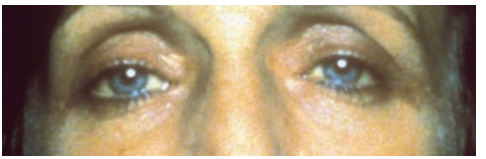

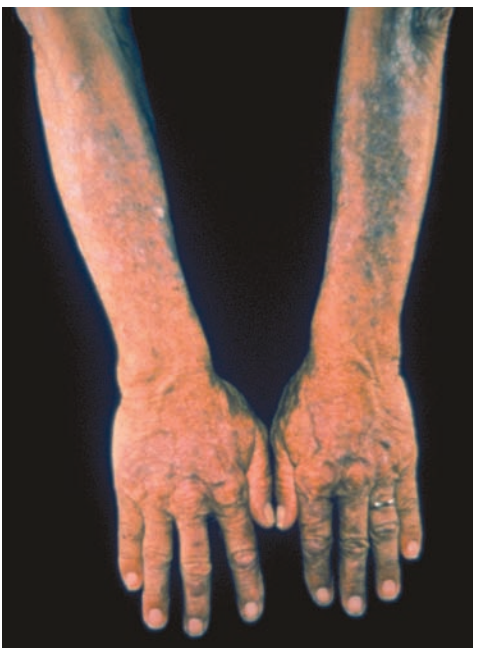

A 55-year-old woman reports an 8-month history of fatigue, malodorous loose stools, diffuse pruritus, and nontender prominence of the right abdomen. Which of the following blood tests is the most useful for establishing the diagnosis?

a. Antimitochondrial antibodies
b. Anti-smooth muscle antibodies
c. Antinuclear antibodies
d. Anti–double-stranded DNA antibodies
e. Ferritin

Primary Biliary Cirrhosis

Answer: a

- Primary biliary cirrhosis (PBC) is a chronic, cholestatic liver disease
- Middle-aged women are primarily affected
- Symptoms and signs of PBC include the following:

  Fatigue
  Pruritus
  Loose stools (due to cholestasis)
  Jaundice
  Xanthelasmas
  Hepatomegaly

- An asymptomatic increase in the level of alkaline phosphatase may be the first identifiable abnormality
- Some patients also have Hashimoto's thyroiditis or sicca syndrome
- Circulating antimitochondrial antibodies occur in 90% to 95% of patients
- Liver biopsy reveals granulomatous infiltration and destruction of small bile ducts
- Ursodeoxycholic acid delays the need for liver transplantation
- Cholestyramine may alleviate pruritus

Reference

Habermann TM. *Mayo Clinic Internal Medicine Board Review 2004-2005*. Philadelphia: Lippincott Williams & Wilkins; 2004:302-303.

A 45-year-old man presents with dyspnea, and pleural effusion is noted on chest radiography. Thoracentesis results in the fluid shown here. All of the following are possible causes for the effusion *except*:

a. Pulmonary infarction
b. Asbestos
c. Malignancy
d. Recent motor vehicle accident
e. Obstructive sleep apnea

Hemothorax

Answer: e

- Major causes for hemothorax are the following:

  Malignancy
  Pulmonary infarction
  Trauma (e.g., penetrating injury, postoperative)
  Asbestosis
  Pancreatitis

- Technically, a bloody pleural effusion is classified as a hemothorax if the hematocrit value of the pleural fluid is greater than half that of the patient's blood
- Most traumatic hemothoraces can be managed with tube thoracostomy
- In a patient with known lung cancer, a hemorrhagic effusion usually signals pleural metastasis
- Complications that may result from residual hemothoraces after conservative drainage include evolution of fibrothorax and empyema
- Empyema may occur because the chest tube may act as a nidus for tracking infection into the pleural cavity
- Residual hemothoraces may be managed definitively with surgical evacuation
- Upright and lateral decubitus chest radiographs are appropriate for initial diagnosis of pleural effusions
- Additional confirmatory method is computed tomography of the chest to best differentiate fluid and parenchymal disease

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APPENDIX
Cases by Specialty

Categories are not mutually exclusive. A case may be classified under more than one specialty.

Core Internal Medicine

Allergy/Immunology (Allergy/Immunol)

Cardiovascular Diseases (CV)

Dermatology (Derm)

Endocrinology (Endo)

Gastroenterology (GI)

Hematology (Hem)

Infectious Diseases (ID)

Musculoskeletal Diseases (Musc)

Nephrology (Neph)

Neurology (Neuro)

Oncology (Oncol)

Pulmonary Diseases (Pulm)

Rheumatology (Rheum)

Vascular Diseases (Vasc)

Cross-Content and Non-Core Internal Medicine

Complementary and Alternative Medicine (CAM)

Critical Care (Crit Care)

Ear, Nose, and Throat (ENT)

Genetics (Genet)

Obstetrics and Gynecology (Ob/Gyn)

Ophthalmology (Ophth)

Toxicology (Toxicol)

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