She has twice recently presented to another emergency room complaining of chest pain. Following each evaluation, she was told that there was no obvious explanation for her pain.

Her initial presentation of SLE consisted of arthritis, rash, mucosal ulcers, and hair loss. She has been treated continuously with hydroxychloroquine and required low dose prednisone as well. She has been pregnant twice; she delivered a healthy child at 36 weeks and had a spontaneous fetal loss at eight weeks.

Physical examination reveals a patient who is pale and in severe pain. Her blood pressure is 100/60 mmHg, pulse is 88, respiratory rate is 18, and temperature is 98.8°F (37.1°C).

Chest is clear to auscultation. The heart sounds are normal without murmurs. There is no soft tissue or costochondral joint pain. There is no evidence of active synovitis, mucosal ulcers, or skin lesions.

Laboratory results

CBC, ESR, electrolytes, creatinine and urinalysis are all normal
CPK = 620 IU/L [10.3 µkat/L] (normal = 10 to 70 IU/L [0.17 to 1.17 µkat/L])
Chest x-ray: normal

Electrocardiogram reveals a sinus tachycardia with ST elevation in the anterior leads.

During the examination the patient develops a rapid ventricular tachycardia that proceeds to ventricular fibrillation and cardiac arrest. The patient dies and an autopsy is performed.

The most likely finding which led to the patient's demise is?

A. A diffuse vasculitis with involvement of the coronary vessels
B. Diffuse, non-inflammatory bland thromboses in the small myocardial vessels with patent coronary arteries
C. A large pericardial effusion with cardiac tamponade
D. Extensive coronary atherosclerosis
E. Libman-Sacks endocarditis with coronary artery embolism

Question 81

A 30-year-old woman presents with a one-year history of progressive fatigue. The fatigue was first noted following an upper respiratory tract infection seven months earlier. She describes diffuse muscle and joint pain, intermittent fevers, sore throat, and headaches. She quit her job as a receptionist.

Her past medical history is notable for an episode of depression during her college years that was treated with electroconvulsive therapy. She sleeps up to 12 hours each day without feeling refreshed.

The physical examination is normal.

Laboratory results:

Hemoglobin = 12.7 g/dL [7.9 mmol/L] (normal = 12 to 16 g/dL [7.4 to 9.9 mmol/L])
WBC = 3,500/mm3 with 22 percent lymphocytes (normal = 4,300 to 10,800/mm3)

Serum electrolytes, liver function studies and TSH are normal.

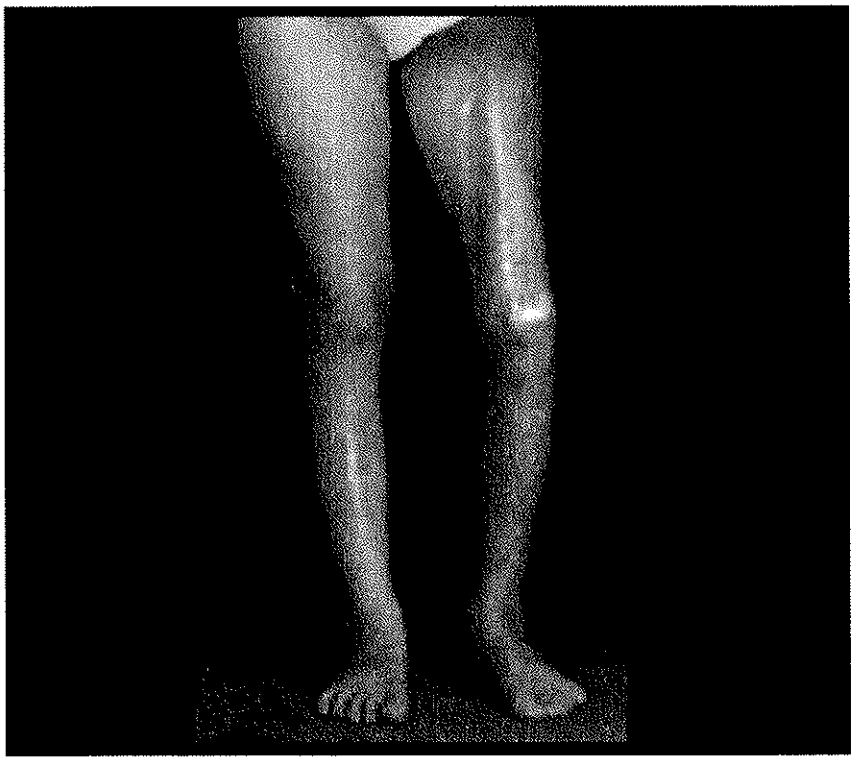
Which of the following tests or procedures should you order next?

A. Flow cytometry to evaluate lymphocyte subsets
B. ELISA test to measure antibodies to Borrelia burgdorferi
C. Brain MRI
D. Psychiatric consultation

E. Epstein-Barr virus (EBV) antibody titers

Question 82

A 29-year-old nurse is referred to you for further management of leg atrophy. She has noticed the gradual change in the appearance of the skin and soft tissues of the entire left leg for at least five years (see figure). She is concerned because she is starting to limp.



Courtesy of Carol M Black, MD.

Which of the following statements regarding this patient's condition is CORRECT?

A. This lesion may be quietly progressive for a long period of time.
B. This patient most likely has ingested supplements containing L-tryptophan.
C. This condition will likely progress into a systemic illness.
D. This condition spares the face and oral cavity.
E. Involvement of the contralateral extremity is likely.

Question 83

A 76-year-old retired executive is referred to you by his internist for further evaluation of an elevated erythrocyte sedimentation rate (ESR). This was noted as part of his routine annual lab tests.

He had the following lab results:

Hemoglobin = 14.2 g/dL [8.8 mmol/L] (normal = 13 to 18 g/dL [8.1 to 11.2 mmol/L])
Hematocrit = 42 percent (normal = 42 to 52 percent)
MCV = 88 fl (normal = 86 to 98 fl)
Platelets = 367,000/mm3 (normal = 130,000 to 400,000/mm3)
WBC = 7,700 with normal differential (normal = 4,300 to 10,800/mm3)

ESR = 68 mm/hr (normal = 0 to 20 mm/hr)

He denies all musculoskeletal complaints. He is healthy and takes ASA 81 mg/day and vitamin E 400 IU/day. His exam is remarkable for some osteoarthritic changes in both hands. A serum protein electrophoresis demonstrates a monoclonal protein in the gamma region. You are considering the diagnosis of a monoclonal gammopathy of undetermined significance.

Which of the following features is not associated with this condition?

A. Fewer than 10 percent plasma cells in the bone marrow
B. A serum monoclonal protein concentration of 1.5 g/dL
C. Normal magnetic resonance imaging (MRI) of the thoracolumbar spine
D. Absence of light chain proteinuria in the urine
E. Serum creatinine of 2.1 mg/dL

Question 84

A 32-year-old woman with systemic lupus erythematosus who is now 28-weeks pregnant presents with bilateral leg swelling. Five years earlier, she was successfully treated with six months of pulse cyclophosphamide and oral steroids for diffuse proliferative glomerulonephritis. Two years ago, a urinalysis revealed new proteinuria, microscopic hematuria, and RBC casts. Cyclophosphamide was reinstituted monthly for six months, then quarterly for two years. Eight months ago, a 24-hour urine collection demonstrated 1 g of protein; there were no cells on urinalysis and the serum creatinine concentration was 1.0 mg/dL. One month later, she informed you that she was pregnant. Serologies for anti-DNA and antiphospholipid antibodies were negative.

Current exam shows a blood pressure of 148/98 mmHg. She has no hair loss, oral lesions, pulmonary findings, arthritis, or rash. She has 2+ edema in both legs and arms.

A complete blood count is normal. A blood smear does not reveal any evidence of intravascular hemolysis. Analysis of an untimed urine sample suggests at least moderate proteinuria.

Other laboratory tests have been ordered including a serum creatinine, uric acid, and 24-hour urine collection to quantitate protein and calcium excretion.

Which one of the following combinations of test results is most compatible with the diagnosis of preeclampsia?

A. Serum creatinine 1.0 mg/dL [88.4 µmol/L], serum uric acid 3.0 mg/dL [178 mol/L], 24 hour urine calcium of 200 mg [5 mmol], urine sediment has large numbers of red cells and cellular casts.
B. Serum creatinine 4.5 mg/dL [398 µmol/L], serum uric acid 3.0 mg/dL [178 mol/L], 24 hour urine calcium of 200 mg [5 mmol], urine sediment has large numbers of red cells and cellular casts
C. Serum creatinine 1.0 mg/dL [88.4 µmol/L], serum uric acid 8.0 mg/dL [475.8 mol/L], 24 hour urine calcium of 60 mg [1.5 mmol], urine sediment has no red or white cells or cellular casts
D. Serum creatinine 4.5 mg/dL [398 µmol/L], serum uric acid 8.0 mg/dL [475.8 mol/L], 24 hour urine calcium of 200 mg [5 mmol], urine sediment has a few white blood cells but no red cells or cellular casts

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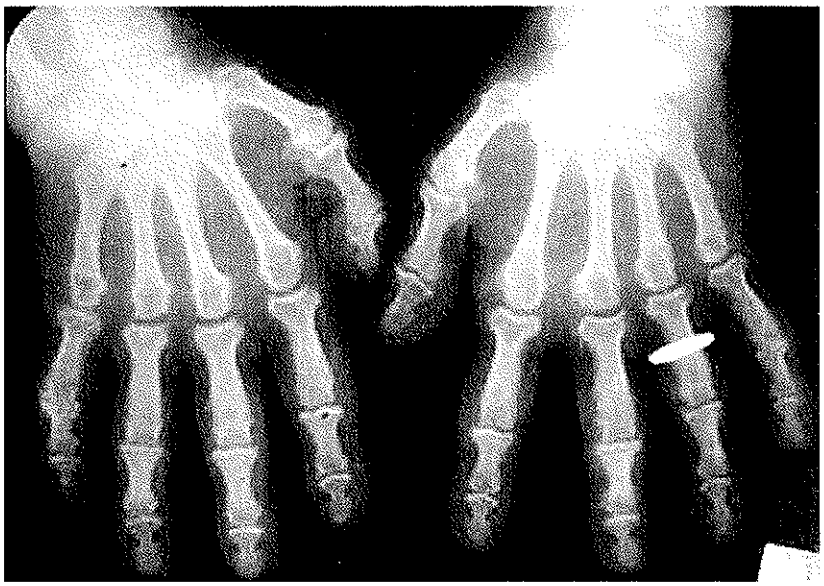
Question 85

A 54-year-old man is referred for evaluation of joint pain. Over the past five years he has noted the insidious progression of pain in the knees, shoulders, elbows, wrists, fingers, and lumbar spine. Pain is worse with activity, with associated swelling predominantly of the hands and fingers. Past medical history is remarkable for hypertension and borderline diabetes.

In a review of systems he notes excessive sweating. He has intermittent numbness of the thumb and 2nd and 3rd fingers bilaterally.

Physical examination reveals macroglossia, multiple skin tags, and hyperhidrosis of the palms. Tinel's sign is positive bilaterally. Musculoskeletal exam shows decreased shoulder range of motion with bilateral glenohumeral crepitus, elbow tenderness, bogginess, and limited range of motion of the wrists, diffuse soft tissue swelling of the fingers, bilateral knee effusions, and limited range of lumbar spine motion.

Hand radiographs are shown (see figure).



Courtesy of Verna Wright, MD, FRCP.

The patient's most likely diagnosis is?

A. Amyloidosis
B. Hypothyroid arthropathy
C. Hyperparathyroidism
D. Acromegaly
E. Idiopathic calcium pyrophosphate deposition disease (CPPD)

Question 86

A 33-year-old accountant with ankylosing spondylitis (AS) diagnosed 12 years ago is seen for a follow-up visit. His disease has been characterized by the presence of back pain and stiffness with limited lumbar mobility, sausage-like swelling of two toes, and left hip arthritis. He has had a variable response to indomethacin, ketoprofen, and naproxen. The ESR ranges between 40 and 65 mm/hr (normal = 0 to 15 mm/hr).

Which of the following features of this patient correlates most strongly with a worse outcome?

A. Elevated ESR
B. Hip arthritis
C. Sausage-like toe swelling
D. Poor response to nonsteroidal antiinflammatory drugs (NSAIDs)
E. Limitation in spinal mobility

Question 87

A 55-year-old woman with a 10-year history of rheumatoid arthritis complains of increasing pain and weakness. Pain is localized over her shoulders and thighs. She notes increasing difficulty with climbing stairs, getting up from low chairs, and combing her hair. Her rheumatoid arthritis has been previously well controlled on prednisone 7.5 mg/day, methotrexate 10 mg/week, and penicillamine 500 mg/day.

Despite her rheumatoid arthritis, she is very active and working full-time as a nurse but, with her recent difficulties, she is afraid she may have to quit her job.

Past medical history is significant for hypothyroidism, elevated cholesterol, and surgical menopause. Other medica-

tions include levothyroxine 0.1 mg/day, nicotinic acid, estrogen replacement, folic acid, calcium supplements, and a multivitamin.

Physical examination demonstrates chronic stable joint deformities of the hands, wrists, and elbows. Range of motion of the shoulders, hips, and knees is not painful. Other than synovial hypertrophy of the MCP joints, no active synovitis is noted.

With testing against resistance, there is weakness in the upper arms and thighs measured as 4/5 bilaterally. No muscle tenderness is noted. Deep tendon reflexes are intact.

Laboratory results:

Hematocrit = 35 percent (stable) (normal = 37 to 48 percent)
MCV = 104.7 FL (normal = 80 to 100 FL)
White blood count = 5,200/mm3 (normal = 4,300 to 10,800/mm3)
Platelets = 230,000/mm3 (normal 130,000 to 400,000/mm3)
Sedimentation rate = 33 mm/hr (normal = 0 to 30 mm/hr)
Rheumatoid factor = 76 IU/mL (normal <20 IU/mL)
ANA = 1:640 (speckled pattern)
Anti-DSDNA: negative
TSH = 5.9 IU/mL (normal = 0.4 to 5 IU/mL)
SGOT (AST) = 56 U/L (normal = 0 to 42 U/L)
SGPT (ALT) = 78 U/L (normal = 0 to 42 U/L)
Creatine kinase (CK) = 1225 U/L [20.4 µkat/L] (normal = 40 to 150 U/L [0.67 to 2.5 µkat/L])
Cholesterol = 214 mg/dL (desirable = <200 mg/dL)

Repeat CK one week later continues to be elevated at 1330 U/L.

In addition to ordering an electromyelogram study (EMG), which one of the following interventions would be appropriate at this time?

A. Stop nicotinic acid and observe
B. Increase levothyroxine to 0.125 mg/day and observe
C. Stop penicillamine and increase prednisone to 60 mg/day
D. Stop methotrexate and check folate levels
E. Rapidly taper prednisone and increase methotrexate

Question 88

A 22-year-old college student with a three-year history of rheumatoid arthritis is well controlled on a combination of cyclosporine, methotrexate, and low-dose prednisone. In the past year, she has also been started on ranitidine for acid reflux symptoms, an oral contraceptive, and supplemental calcium with vitamin D.

Despite being maintained on a constant dose of prednisone, she is concerned about recent weight gain. She inquires whether any of her medications might be altering the metabolism of the prednisone.

Which one of her medications may increase the concentration of corticosteroids?

A. Cyclosporine
B. Methotrexate
C. Ranitidine
D. Oral contraceptives
E. Calcium

Question 89

A 66-year-old patient with osteoarthritis of the knees is seen for follow-up. She has been treated with nabumetone 750 mg twice daily for three years. Her son, a gastroenterologist, has suggested to her that she discuss with you the use of the selective cyclooxygenase-2 (COX-2) enzyme inhibiting nonsteroidal anti-inflammatory drugs (NSAIDs).

Which of the following statements regarding these agents is true?

A. They have superior analgesic and anti-inflammatory effects compared to traditional NSAIDs.
B. COX-2 inhibition may promote colon tumor growth.
C. COX-2 inhibitors are contraindicated in patients who are allergic to sulfonamides.
D. Nabumetone and etodolac are also relatively selective for the COX-2 enzyme isoform at low doses.
E. The incidence of hypertension does not increase with higher doses of rofecoxib.

Question 90

A 27-year-old stagehand presents to you with low back and heel pain. He has had chronic low back pain for nine months, which he relates to his work. His symptoms are worse when he is trying to rest. He has had an episode of acute eye inflammation, which was treated by an ophthalmologist.

On examination, he is an African-American man with limited lumbar flexion and swollen Achilles tendons. His erythrocyte sedimentation rate is 44 mm/hr (normal = 0 to 15 mm/hr).

Which of the following procedures would be most likely to confirm his diagnosis?

A. Radionuclide bone scan
B. Oblique views of the sacroiliac joints
C. HLA-B27 test
D. CT scanning of the sacroiliac joints
E. Radiographs of the lumbar spine

Question 91

An 80-year-old man with a history of sick sinus syndrome with a permanent pacemaker and diabetes mellitus is seen for severe back pain. The pain has progressively worsened over the past three weeks and now keeps him awake at night. He has had a sore throat, dry cough, and fever for the past week. He denies a history of bowel or bladder incontinence or recent trauma.

On physical examination, his temperature is 100.9°F (38.3°C). There is marked tenderness to palpation which is localized to the mid thoracic area. Neurologic examination is normal.

An erythrocyte sedimentation rate (ESR) is 108 mm/hr (normal = 0 to 20 mm/hr). Chest radiograph is normal. Plain radiographs of the thoracic spine show multi-level disc space narrowing with osteophytes formation. Blood cultures are drawn.

What should be performed next?

A. CT scan
B. Indium-labelled leukocyte scan
C. Three-phase bone scintigraphy, and if positive, gallium imaging
D. Gadolinium-enhanced MRI

E. Contrast myelography

Question 92

A three-year-old boy is brought to the emergency room for evaluation of a fever and rash for the past four days. Three days after the onset of fever, his mother noted reddening of his conjunctivae, palms, and soles.

Physical examination reveals a temperature of 104°F (40.3°C).

ENT: There is bilateral conjunctival injection and diffuse erythema of the lips, oral cavity, tongue, and pharynx.

Skin: There is a morbilliform erythematous eruption over the palms and soles.

Cardiac: A II/VI systolic murmur is audible.

Laboratory evaluation:

WBC = 13,000 cells/mm3 (normal = 4,300 to 10,800/mm3)
ESR = 44 mm/hr (normal = 0 to 15 mm/hr)
AST = 34 IU/L (normal = 35 IU/L)
ALT = 24 IU/L (normal = 35 IU/L)

Anti-streptolysin antibody: negative
Rapid streptococcus screen: negative

Which of the following statements best describes this patient's current condition?

A. Diffuse lymphadenopathy is uncommon in this disease, although careful examination is likely to reveal splenomegaly.
B. Sudden death is a feared complication.
C. As in many other rheumatic diseases, corticosteroids are a mainstay of therapy.
D. Lipid abnormalities are commonly found, including elevations in the serum high-density lipoprotein levels.
E. Anticoagulation is often required during the acute illness.

Question 93

A 79-year-old woman presents to your office with a complaint of progressively worsening pain in her buttocks and thighs. She notes the pain when standing or walking. The pain is relieved by rest. She has had some degree of back pain for the past two years; however it has definitely worsened over the past month. Her past medical history reveals breast cancer treated with surgery 15 years ago without recurrence. She has smoked 10 cigarettes per day for the past 40 years.

On examination, you note limited lumbar spinal motion with flexion and especially extension. There is no restriction of straight leg raising. The knee reflexes are 2+ bilaterally but ankle reflexes are absent. There is some tenderness on palpation of the lower lumbar spine.

Plain radiographs show disc space narrowing and facet joint osteoarthritis at L4-L5. You suspect lumbar spinal stenosis.

Which of the following statements regarding this diagnosis in this patient is true?

A. An imaging study of the lumbar spine would likely show a herniated lumbar disc.
B. An imaging study of the lumbar spine would likely show metastatic breast cancer.
C. A positive Romberg sign and a wide-based gate are highly specific for spinal stenosis, but not very sensitive.

D. Without surgical decompression, there is a high risk for this patient developing the cauda equina syndrome.

Question 94

A 25-year-old man is referred for evaluation of progressive weakness, diffuse muscle spasms, and pain. He is currently unable to adequately perform his job as a landscaper; every time he pushes a lawn mower up a hill he develops muscle pain and a sensation of spasm that forces him to stop frequently and rest. Otherwise, he has felt well except for an episode of painless dark urine noted six months ago.

His examination today is completely normal, including a thorough evaluation of muscle strength. His muscle build and tone are normal.

Which of the following tests obtained at the visit is most likely to be abnormal?

A. Serum CK
B. Electromyelography (EMG)
C. Routine muscle biopsy
D. Forearm ischemic exercise test

Question 95

A 26-year-old nurse with a history of rheumatoid arthritis has developed idiopathic thrombocytopenic purpura (ITP). You have referred her to a hematologist for further treatment, however, the hematologist seeks your opinion regarding the management of this problem.

All of the following statements regarding the treatment of her ITP are correct EXCEPT?

A. If the patient's rheumatoid factor (RF) is markedly elevated, treatment with intravenous immune globulin (IVIG) may be relatively contraindicated.
B. If she is to be treated with prednisone, a response should be observed within the first few weeks.
C. If her platelet count is above 50,000/µL, she may not require therapy.
D. If required, a splenectomy carries a 95 percent success rate.
E. Anti-D therapy is effective only in patients who are Rh-positive.

Question 96

A 38-year-old African-American man presents to your clinic with a two-month history of dry cough and shortness of breath when climbing stairs. He has an 18-month history of severe Raynaud's phenomenon and a one-year history of rapidly progressive diffuse scleroderma associated with gastroesophageal reflux disease. He denies fever or productive cough. He denies tobacco use.

His oral temperature is 99.1°F (37.3°C). The skin over his face, chest, hands, forearms, feet, and legs is shiny, free of wrinkles, and cannot be pinched between the fingers. He has flexion contractures of the 2nd through 5th PIPs bilaterally and a shallow ulceration over the right second fingertip. There are crackles on inspiration at the bases of both lungs.

Laboratory studies reveal:

CBC: normal
Urea nitrogen, serum = 8.0 mg/dL [2.9 mmol/L] (normal = 2.5 to 8.0 mg/dL [0.9 to 2.9 mmol/L])
Creatinine = 0.8 mg/dL [70 mmol/L] (normal = 0.8 to 1.3 mg/dL [70 to 114 mmol/L])
ANA = 1:160 with a speckled pattern

Antibodies to SSA, SSB, U1-RNP. Smith antigens are negative. Antibodies to Scl-70 are positive. PA and lateral chest radiographs are unremarkable

Pulmonary function tests:

FEV1 = 50 percent predicted
FVC = 50 percent predicted
Diffusion capacity of carbon monoxide (DLCO) = 55 percent predicted

High resolution computed tomography (CT) reveals a ground glass opacification at both lung bases.

For a patient with these findings, what is the most likely outcome regarding his pulmonary disease?

A. He is likely to respond to therapy
B. He is unlikely to respond to therapy
C. He is likely to develop recurrent opportunistic pulmonary infections
D. He is likely to develop pulmonary hypertension
E. His pulmonary symptoms and disease are unlikely to progress regardless of treatment

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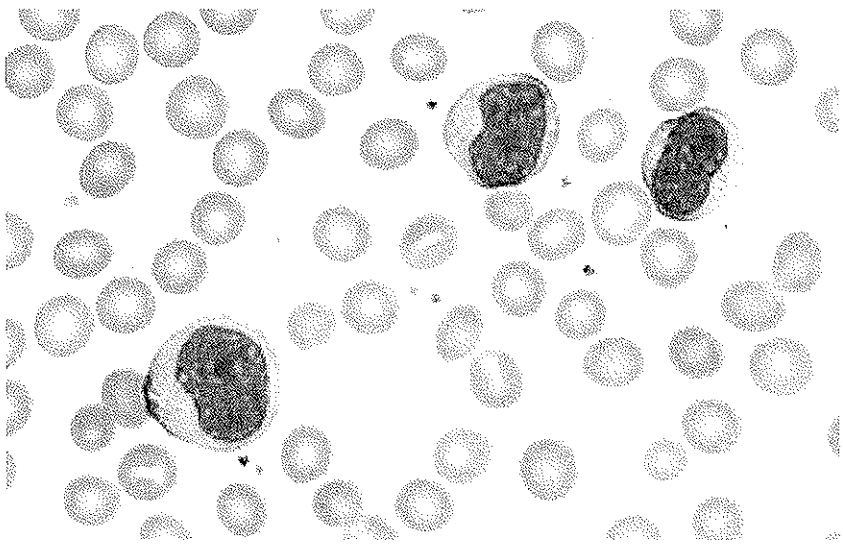
Question 97

A 20-year-old college student is seen in the emergency room because of sudden onset of increasing left upper quadrant pain, which began this morning after breakfast. The pain is localized and intense. He denies any recent trauma. He has not felt well lately and for the past few weeks has been tired. One week ago, he developed a sore throat and was diagnosed as having tonsillitis. He was prescribed ampicillin. Two days ago he developed a rash over his arms, face and chest. He complains of diffuse muscle achiness. The rheumatology service is being consulted now because of his lab results.

His laboratory studies reveal:

Hemoglobin = 12.1 g/dL [7.5 mmol/L] (normal = 13 to 18 g/dL [8.1 to 11.2 mmol/L])
WBC = 18,800/mm3 (differential pending) (normal = 4,300 to 10,800/mm3)
Platelets = 100,000/mm3 (normal = 130,000 to 400,000/mm3)
ESR = 77 mm/hr (normal = 0 to 15 mm/hr)
ANA: negative

A blood smear is shown (see figure).



Courtesy of Carola von Kapff, SH (ASCP).

On exam he is a pale-looking male in severe discomfort because of abdominal pain. His blood pressure is 140/90 mmHg, pulse is 100, respirations are 18, and temperature is 101.1°F (38.4°C).

There is diffuse cervical adenopathy. The tonsils are enlarged and covered with exudate. There are palatal petechiae. There is a macular eruption over the arms and chest.

The joints and proximal muscles are diffusely tender. There is no joint swelling. There is marked abdominal discomfort with rebound tenderness and guarding. The remainder of the exam is unremarkable.

What is the most likely diagnosis?

A. Adult Still's disease
B. Kawasaki's disease
C. Systemic lupus erythematosus
D. Infectious mononucleosis

Question 98

A 28-year-old woman presents to her physician with slowly progressive left knee pain. The pain is worse with activity and eases with rest. She notes mild swelling but no warmth or redness. She denies any trauma but is a gym teacher at the local high school. She competed in gymnastics in college and recalls several episodes of joint dislocations involving her shoulders and elbows.

Past medical history is notable for a curvature of the spine and an asymptomatic "heart murmur." Her 25-year-old sister has similar problems and a history of a retinal detachment. There is no other family history of joint problems, heart disease, or sudden death.

Upon hearing this history, you are concerned that she may have Marfan syndrome. Which of the following features would NOT be consistent with this diagnosis?

A. Aortic stenosis
B. Kyphosis
C. Mitral valve prolapse
D. Retinal detachment
E. Spinal dural ectasia

Question 99

A 42-year-old accountant is evaluated for progressive symptoms in her dominant hand consistent with carpal tunnel syndrome. One year ago, she had only nighttime pain and no weakness. She tried wrist splints at that time but found them uncomfortable to wear. She now experiences constant pain with worsening numbness. On examination, she demonstrates decreased pinch strength.

Regarding the surgical options of an open incision versus an endoscopic procedure for treatment of carpal tunnel syndrome, which of the following is TRUE?

A. An open incision demonstrates better long-term resolution of symptoms but has a higher complication rate.
B. An open incision demonstrates better long-term resolution of symptoms with a lower complication rate.
C. An endoscopic approach demonstrates better long-term resolution of symptoms but a higher complication rate.
D. An endoscopic approach demonstrates better long-term resolution of symptoms with a lower complication rate and a more rapid return to work.
E. Long-term outcomes are equal with either approach.

Question 100

A 54-year-old woman presents to your office with the complaint of hot flashes and vaginal dryness. Her last menstrual period was one year ago. A bone density study reveals a femoral neck T score of -2.3. Her cholesterol level is 250 mg/dL [6.5 mmol/L] (desirable <200 mg/dL [<5.17 mmol/L]). She does not want to resume menses.

She has no history of venous thrombosis.

Family history is positive for coronary artery disease and her mother had a hip fracture. There is no family history of breast cancer.

Which of the following therapies offers the best management plan for this patient?

A. Raloxifene
B. Alendronate
C. Daily oral estrogen and progesterone
D. Daily oral estrogen
E. Daily oral estrogen and cyclic progesterone

Question 101

A 35-year-old smoker is seen in the emergency room for evaluation of a painful left eye. For the past two weeks she has noticed a low grade fever, myalgias, loss of appetite, and difficulty hearing. She has lost function in both hands due to pain and swelling of the wrists and MCPs bilaterally. She is referred to you for further evaluation.

The patient has been in good health otherwise. Family history is notable for a sister and a first cousin with SLE.

On physical examination her temperature is 99.4°F (37.7°C), blood pressure is 145/95 mmHg, pulse is 80/min, and respiratory rate is 12/min.

The left eye is slightly red and there is reduced visual acuity measuring 20/40 OS versus 20/20 OD. Nasal exam reveals bilateral painless ulcers.

Chest exam: Some scattered rales at the right lung base. Heart sounds normal; there are no murmurs. There is synovitis of the wrists and MCPs bilaterally. Neurologic exam is unremarkable. There is no skin rash.

Laboratory results:

Hemoglobin = 9.3 g/dL [5.8 mmol/L] (normal = 12 to 16 g/dL [7.4 to 9.9 mmol/L])
WBC = 12,000/mm3 with a normal differential (normal = 4,300 to 10,800/mm3)
Platelets = 420,000/mm3 (normal = 130,000 to 400,000/mm3)
Creatinine = 1.2 mg/dL [106.1 µmol/L] (normal = 0.6 to 1.0 mg/dL [53 to 88 µmol/L])
AST = 33 IU/L (normal = 0 to 35 IU/L)
ALT = 41 IU/L (normal = 0 to 35 IU/L)
CK = 145 IU/L [2.4 µkat/L] (normal = 40 to 150 U/L [0.67 to 2.5 µkat/L])

Urinalysis reveals 5 RBC per high-powered field, no casts.

Chest x-ray: diffuse right lower lobe alveolar infiltrate.

Which test would most likely lead to the correct diagnosis?

A. Skin test for pathergy

B. Anti-double stranded DNA antibody
C. Anti-glomerular basement membrane antibody
D. ANCA
E. Renal arteriogram

Question 102

A 40-year-old man is referred to you for evaluation of osteoporosis. At a routine employment physical, his chest radiograph demonstrated two unsuspected vertebral compression fractures and diffuse osteopenia. He does not complain of back pain. He is on no medication and there is no past history of corticosteroid or anticonvulsant therapy. He does not consume alcohol. He eats a regular diet and has not lost weight. He runs several times weekly.

He denies any abdominal pain, bloating, or diarrhea. He has noted no change in libido. Family history is of note in that his mother has osteoporosis and chronic diarrhea.

His physical examination is entirely unremarkable except that his recorded height of 70 inches represents a 1inch loss from his peak height.

Dual x-ray absorptiometry (DXA) demonstrates an average T-score of -3.2 for the lumbar spine and -1.8 at the hip. The chest x-ray exhibits compression deformity of the T-11 and T-12 vertebral bodies. Lumbar spine films demonstrate osteopenia but no additional compression fractures.

Laboratory studies obtained (complete blood count, metabolic panel, thyroid stimulating hormone, testosterone level, 24-hour urinary calcium excretion, protein electrophoresis, and phosphorous level) are all normal except for a 25-hydroxyvitamin D level of 5.6 ng/mL [14.0 nmol/L] (normal = 8 to 42 ng/mL [20 to 105 nmol/L]).

After reviewing the data, which one of the following choices would be your next step?

A. Begin calcitriol 25 micrograms daily
B. Begin ergocalciferol 50,000 units weekly
C. Begin calcidiol 50 micrograms daily
D. Obtain a serum IgA endomysial antibody and small bowel biopsy
E. Obtain a serum IgG antigliadin antibody and small bowel biopsy

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Question 103

A 37-year-old woman with rheumatoid arthritis has failed therapy with hydroxychloroquine. You have recommended initiation of therapy with sulfasalazine. She is planning to have children and wants your advice regarding the use of this drug in this context.

What would you advise?

A. Sulfasalazine is unsafe for use during pregnancy and therefore should not be recommended.
B. Sulfasalazine is safe for use during pregnancy but should be avoided if she is planning to breastfeed her child.
C. Sulfasalazine is safe for use during pregnancy and can be used during breastfeeding.
D. There is an increased risk of prematurity and birth defects in children born to mothers taking sulfasalazine.
E. Sulfasalazine can be safely used during pregnancy, if the dose is reduced by 50 percent.

Question 104

A 29-year-old man presents with a two-year history of low back pain. The pain has progressed in the past few months to include his thighs bilaterally. He describes the pain as episodic "spasms" of the involved areas.

On examination, he stands in a fixed, lordotic posture. The lumbar paraspinal and thigh muscles are rigid.

Radiographs of the sacroiliac joints and lumbar spine are normal. Electromyogram (EMG) of the lower extremities reveals continuous motor unit activity without insertional irritability or spontaneous fibrillation.

The most likely diagnosis is?

A. Ankylosing spondylitis
B. Myotonic dystrophy
C. Stiff-person syndrome
D. Diffuse idiopathic skeletal hyperostosis
E. Fibromyalgia

Question 105

A 55-year-old office manager returns two weeks following carpal tunnel release on her dominant hand. While her open incision has healed well, she is concerned over some persistent hand pain and numbness. Her grip and pinch strength continue to be significantly decreased.

Which of the following is an appropriate expectation for this patient's surgical outcome?

A. Improvement in numbness and pain is generally seen at two weeks, and there is maximum recovery of strength by six weeks
B. Improvement in numbness and pain is generally seen at two weeks, and there is maximum recovery of strength by three months
C. Improvement in numbness and pain is generally seen at six weeks, and there is maximum recovery of strength by six months
D. Improvement in numbness and pain is generally seen at six weeks, and there is maximum recovery of strength by two years
E. Improvement in numbness and pain is generally seen at three months, and there is maximum recovery of strength by one year

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Question 106

A 64-year-old man is referred for evaluation of left foot and ankle swelling. He is two months post-cadaveric renal transplant.

Physical exam is remarkable for diffuse soft tissue swelling of the entire left foot extending proximally to the mid-calf. The extremity is warm and exquisitely tender, with evidence of hyperhidrosis.

Medications include cyclosporine 200 mg po bid, prednisone 20 mg qd, and azathioprine 150 mg qd. Cyclosporine trough level is 274 ng/mL.

Laboratory studies reveal:

Serum uric acid = 8.6 mg/dL [511.5 µmol/L] (normal = 2.8 to 8.0 mg/dL [150 to 480 µmol/L])
Creatinine = 1.3 mg/dL [115 mmol/L] (0.8 to 1.3 mg/dL [70 to 114 mmol/L])

Radiographs of the foot and ankle reveal areas of patchy demineralization. Technetium-99m pertechnetate bone scintigraphy demonstrates marked increased uptake of radiotracer in the left ankle and entire foot.

Aspiration of the left ankle was unsuccessful.

In addition to referring the patient to physical therapy, which of the following options would you recommend?

A. Increase prednisone dose to 20 mg bid
B. Begin colchicine 0.6 mg po q hour until pain and swelling resolve, maximum 4.8 mg/24 hours
C. Decrease cyclosporine to 100 mg po bid
D. Decrease azathioprine to 75 mg po qd
E. Intraarticular ankle injection of methylprednisolone

Question 107

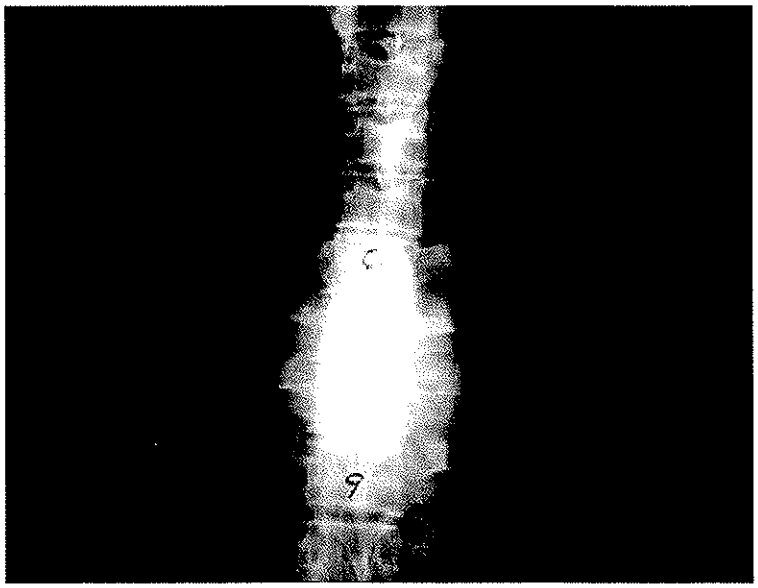
A 40-year-old man is referred for evaluation of mid-thoracic back pain. The pain began insidiously three months ago and has increased in severity of late. He initially noted relief with ibuprofen but it has lost its efficacy. He denies fever, chills, or weight loss.

He immigrated to the United States from Africa four years ago. At the time, he was noted to have a positive PPD and a normal chest radiograph. He was treated with isoniazid for six months. He worked as a house painter but has stopped recently because he feels that his legs are getting weaker.

On examination, he complains of spine pain in the region of T7 through T10. There is spinal tenderness and muscle spasm. There is weakness of both legs 3/5 bilaterally. The deep tendon reflexes are reduced in both legs. A radiograph of the thoracic spine is taken (see figure). You are concerned that he may have tuberculosis.

Which of the following statements regarding his condition is true?

A. His lack of constitutional symptoms would be unusual for spinal tuberculosis (TB).
B. A possible complication is cord compression resulting in paraplegia.
C. Plain radiographs of the spine characteristically demonstrate vertebral destruction with sparing of intervertebral disks.
D. This patient is likely to be HIV positive.
E. His chest radiograph is likely to show changes consistent with active tuberculosis.



Courtesy of Charles E Putnam, MD.

Question 108

A 27-year-old woman is referred to you by her endocrinologist for evaluation of abnormal laboratory findings. She was diagnosed with Graves' disease five years ago and has been treated with propylthiouracil (PTU) since then. Recently, she developed skin lesions over the ankles and feet; a skin biopsy revealed a leukocytoclastic vasculitis. Aside from the uncomfortable skin lesions, she feels well.

Laboratory results:

CBC: normal
ESR = 42 mm/hr (normal = 0 to 20 mm/hr)
Creatinine = 1.1 mg/dL [97.2 mmol/L] (normal = 0.6 to 1.0 mg/dL [53 to 88 mmol/L])

Liver function tests: normal
ANA: 1:80 homogeneous pattern
ANCA: "positive," full results pending

Which one of the following statements regarding her condition is true?

A. Her leukocytoclastic vasculitis is not associated with the development of a positive ANCA.
B. She has developed a drug-induced SLE.
C. If she has a PTU-induced vasculitis, it would be characterized by positive ANCA specificities to several different target antigens.
D. If she has a PTU-induced vasculitis, it would be characterized by a positive ANCA with specificity to myeloperoxidase (MPO).
E. Her skin lesions are due to her underlying Graves' disease.

Question 109

A 56-year-old post-menopausal woman with known primary biliary cirrhosis presents with acute midthoracic spine pain. Radiographs of the spine show a wedge-shaped deformity of the T-7 vertebral body. The bone densitometry T-scores are -2.6 and -1.4 for the lumbar spine and hip respectively.

Which one of the following statements is correct?

A. The patient most likely has a low-turnover osteoporosis.
B. High dose regimens of vitamin D provide effective therapy.
C. The risk of spontaneous fracture becomes reduced as soon as four months post liver transplant.
D. In most of these patients, the appendicular skeleton (cortical bone) is affected more than the axial skeleton (trabecular bone).

Question 110

A 42-year-old physician complains of right shoulder pain for the past several weeks. He has tried several non-steroidal anti-inflammatory agents with minimal response. An avid tennis player, the pain is now interfering with his game. He is interested in an injection for symptom relief since he is training for a local competition to be held in one month.

Examination of his shoulder is consistent with an impingement syndrome, and you offer to inject his shoulder using a corticosteroid preparation and a local anesthetic. He wants your advice regarding the risk for infection, tendon rupture, and post-injection flare up of pain.

Which of the following statements regarding the injection procedure is CORRECT?

A. The infection rate is considered to be 0.3 percent.
B. An iatrogenic joint infection should be suspected if there is a flare up of pain lasting longer than or beginning later than 48 hours after the injection.
C. Post-injection flares are most commonly associated with injections of corticosteroid preparations containing triamcinolone.
D. The addition of a local anesthetic may increase the risk of tendon rupture from the larger mass effect of the injected bolus.
E. The addition of a local anesthetic may increase the risk of a post-injection flare.

Question 111

A 67-year-old woman presents with a six-month history of progressive, painful numbness and tingling of the hands and forearms. This has begun to interfere with sleep. She also notes neck pain. In addition, she has developed bilateral shoulder pain aggravated by activities that require reaching motions.

Her past medical history is notable for end-stage renal failure requiring continuous ambulatory peritoneal hemodialysis (CAPD) for the past eight years.

On physical examination, there is soft tissue swelling around both shoulders with decreased range of motion. There is also decreased sensation to pinprick over the distal thumbs and index fingers. The Phalen's test is positive bilaterally. A joint aspiration revealed clear synovial fluid with a noninflammatory cell count.

The diagnosis of dialysis-related amyloidosis is considered. What next step would you take to help make a diagnosis?

A. Consider another diagnosis because dialysis-related amyloidosis does not develop in patients treated with CAPD
B. Obtain plasma concentration of ß2-microglobulin
C. Recommend that the patient switch to hemodialysis
D. Verify presence of amyloid by obtaining a skin biopsy
E. Stain joint aspirate for amyloid

Question 112

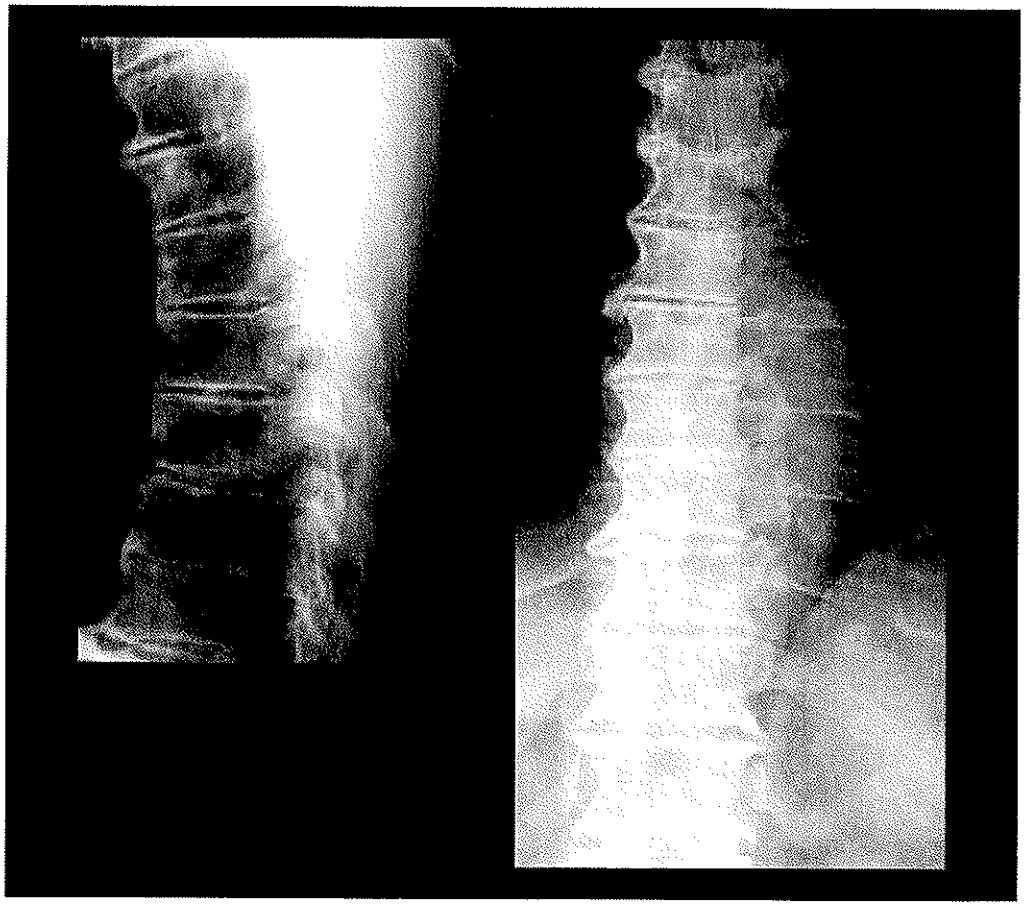
A 56-year-old man presents with a two-day history of a painful swollen right knee. He has had several similar episodes over the past few months. On examination, he has a swollen, warm knee and a nodule on the extensor surface of his left arm that appears to be a tophus.

Which of the following statements regarding treatment options for this patient is incorrect?

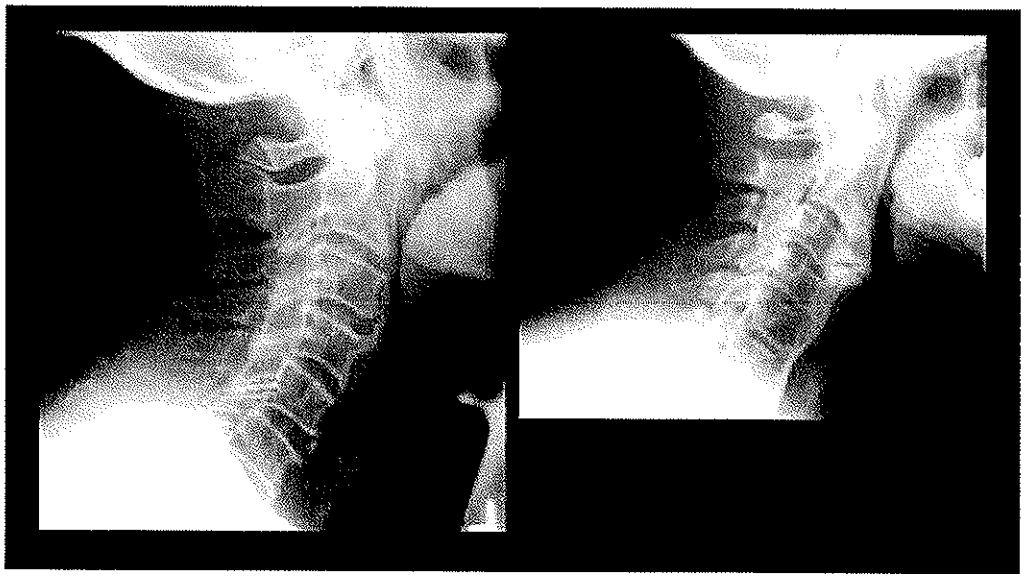
A. Leukopenia, renal insufficiency, or recent use of oral colchicine should be considered contraindications to the use of intravenous colchicine.
B. Since probenecid increases urinary calcium excretion in gouty patients, it should not be used in patients with prior nephrolithiasis.
C. Patients with mild renal insufficiency, who are treated with standard doses of allopurinol and a diuretic, are at increased risk for the allopurinol hypersensitivity syndrome.
D. Allopurinol is absolutely contraindicated for use in patients treated with mycophenolate, methotrexate, or azathioprine.

Question 113

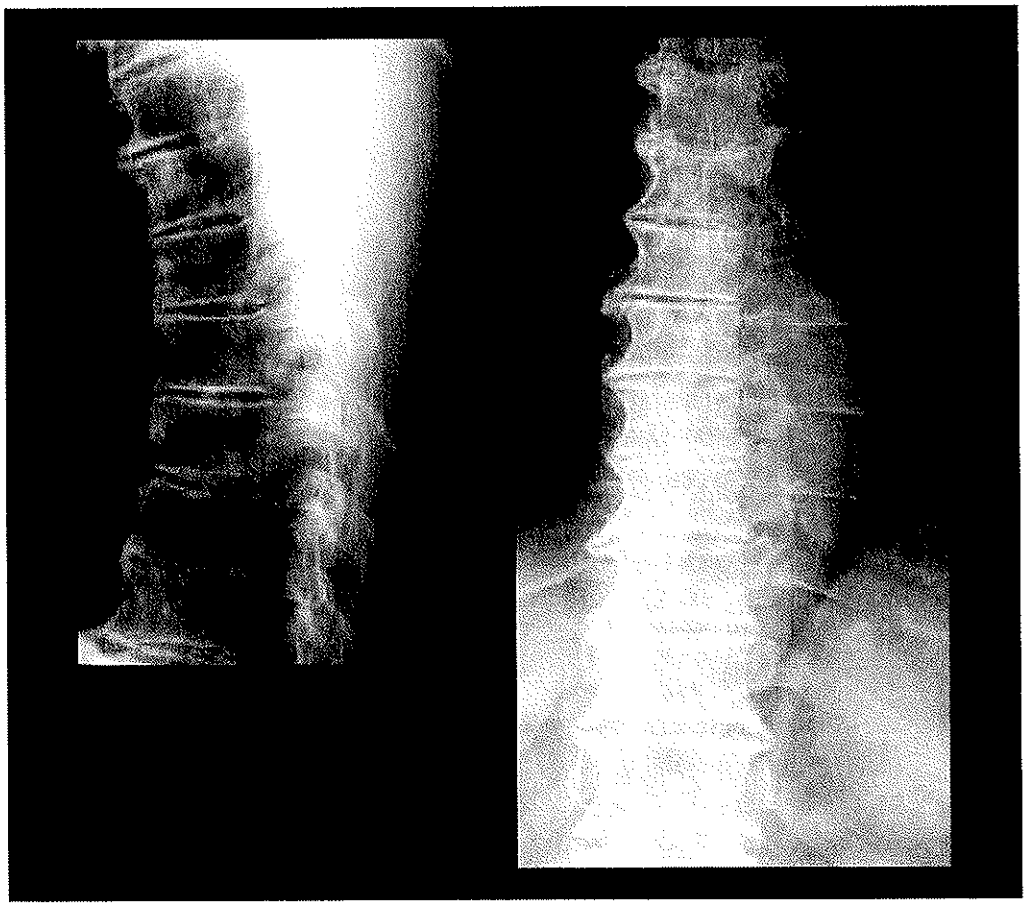
A 65-year-old man with back pain is seen by you for a second opinion regarding his condition. He brought along a series of spine radiographs obtained by his internist (see figures a, b, and c).



Courtesy of John Esdaile, MD.



Courtesy of John Esdaile, MD.



Courtesy of John Esdaile, MD.

Which one of the following features is a manifestation of his condition?

A. Vertebral fractures
B. Development of lumbar radiculopathy
C. Association with Stiff-man syndrome
D. Association with iritis
E. Extraspinal enthesopathy

Question 114

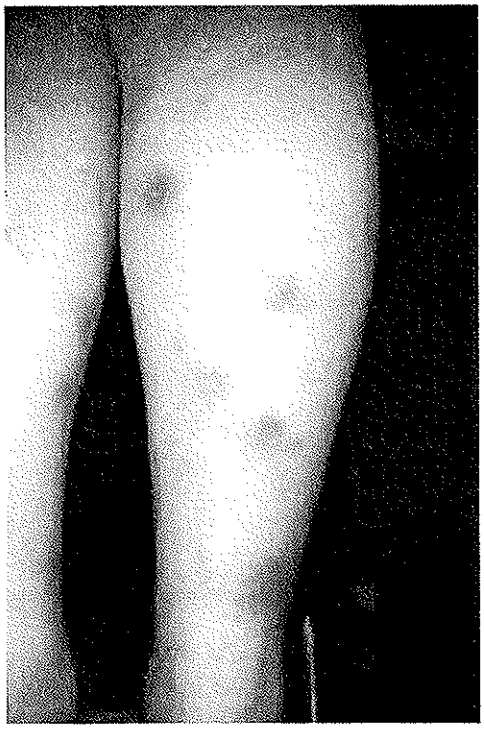
A 42-year-old woman reports the onset of several painful, lesions over both shins two weeks ago (see figure). In addition, she describes pain and swelling around her ankles and distal legs for the past week, such that it has become difficult to fit into her shoes and painful to walk. She is otherwise healthy and takes no medications.

Examination of the extremities reveals 2+ distal leg edema and swelling around each ankle, with tenderness to squeeze. There is pain with plantar and dorsiflexion of the ankles. A chest x-ray reveals bilateral hilar adenopathy.

She is alarmed about her precipitous decline in functional status and seeks your opinion regarding her condition and its prognosis.

What would you tell her?

A. It would be highly unusual for the arthritis to persist beyond several months.
B. This condition is more commonly seen in men.
C. The arthritis will probably resolve before the disappearance of her skin lesions.
D. The arthritic symptoms often recur.
E. Sarcoidosis is the likely diagnosis.



Reproduced with permission from The Skin and Infection: A Color Atlas and Text, Sanders, CV, Nesbitt, LT Jr (Eds), Williams & Wilkins, Baltimore. 1995.

Question 115

A 24-year-old college student from Thailand is sent to you by his internist for consultation regarding the abrupt onset of muscle weakness. Over the past week his legs have become weak and his CK is 468 IU/L [4.6 µkat/L] (normal = 60 to 400 IU/L [1.0 to 6.7 µkat/L]). The patient is wheeled into your office because he is unable to walk. He recalls having a diarrheal illness four weeks ago which resolved. He denies pain but complains of paresthesias in his hands and feet. He received various unknown immunizations prior to leaving for college, but he remembers being administered the BCG vaccine for tuberculosis.

He denies use of injected drugs or alcohol and has no tattoos. He has had several sexual partners including sex workers, but says he has used condoms regularly during sexual intercourse.

Examination of his lower extremities reveals normal muscle bulk but marked bilateral weakness of the extensor hallucis longus, anterior tibialis and quadriceps muscles. The deep tendon reflexes are unobtainable in the legs.

Laboratory tests, including an serologic test for syphilis, an ANA, HIV antibody, and an HTLV-1 antibody, are normal. Magnetic resonance imaging of the entire spine is also normal.

A lumbar puncture is performed, with cerebrospinal fluid analysis revealing no cells and a total protein of 80 mg/dL.

Which of the following statements regarding this patient's illness is correct?

A. Marked elevation of serum CK is common and associated with the development of acute renal failure.
B. The weakness may rapidly progress to involve the upper extremities.
C. He probably has a history of Raynaud's phenomenon, photosensitive rash, and dysphagia.
D. He likely has a family history of progressive muscle weakness.

Question 116

A 53-year-old woman is referred to you by her gastroenterologist for evaluation of achiness. She has noted some generalized discomfort in her lower back, pelvis, and legs. It does not seem to change with either rest or activity.

The patient has a history of celiac disease treated by a gluten-free diet.

Which of the following conditions has not been associated with celiac disease?

A. Polymyalgia rheumatica
B. Osteomalacia
C. Axial arthritis
D. Peripheral arthritis
E. Selective IgA deficiency

Question 117

A 43-year-old man is referred to you by his dermatologist for evaluation of chest wall pain. He notes a steady pain over the chest wall made worse by palpation. The pain is felt maximally over the sternum and the clavicles.

Past medical history is notable for acne and palmoplantar pustulosis. His only medication is doxycycline. There is no history of bowel disease, urethritis, or conjunctivitis. An HLA-B27 study ordered by his dermatologist is negative.

On exam the sternoclavicular joints are swollen and tender bilaterally. The sternum is diffusely tender to touch. The remainder of the joint exam is unremarkable. There is full mobility of the entire spine. There are scattered pustular lesions over the palms and soles and facial acne.

Plain radiographs of the sternoclavicular joints demonstrate enlargement and sclerosis of the medial clavicles; an osteolytic lesion may be evident on the right side. In view of these findings, he was also referred to an orthopedic surgeon who has arranged for a biopsy of the region.

Which one of the following statements regarding his condition is true?

A. Doxycycline is generally an effective therapy.
B. The biopsy findings would resemble a sterile osteomyelitis.
C. The biopsy findings would resemble a proliferative synovitis.
D. There is a strong association with HLA-B27.
E. A long-term complication is the development of osteosarcoma.

Question 118

A 28-year-old attorney is referred to you on an urgent basis because of her concerns regarding Lyme disease. She was vacationing in coastal Connecticut two weeks ago and was bitten by a tick. Yesterday she noted a new "bull's eye" rash over her thigh. She is ten weeks pregnant and her obstetrician suggested that she discuss the management of this problem with you.

Which of the following options do you recommend?

A. The patient should be informed that Lyme disease may predispose to congenital anomalies
B. Intravenous ceftriaxone 2 g daily for seven days
C. Doxycycline 200 mg qd for four weeks

D. Amoxicillin 2.0 grams po qd for four weeks
E. Await the results of Lyme antibody testing before recommending antibiotic therapy

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Question 119

A 55-year-old woman has recurrent muscle weakness. She presented seven months earlier with mild proximal muscle weakness in both her arms and legs. She had no skin rash, and her physical examination was otherwise remarkable for normal breast, pelvic, and rectal examinations. Stool was negative for occult blood. A screening colonoscopy done at age 50 was normal.

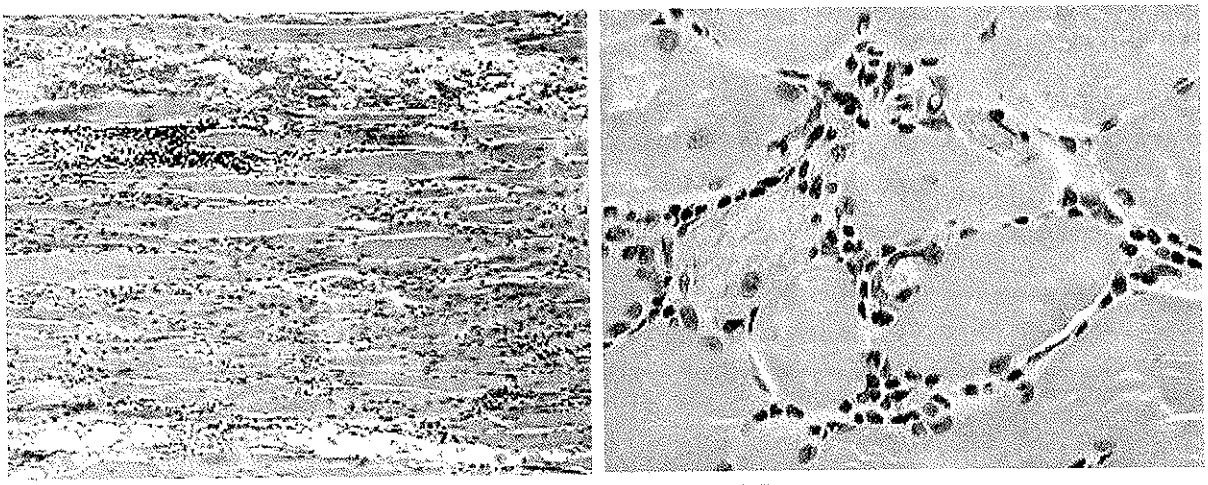
Relevant lab studies done previously included:

Creatine kinase (CK) = 825 U/L [13.8 µkat/L] (normal = 40 to 150 U/L [0.67 to 2.5 µkat/L])
ANA = 1:640 in a speckled pattern
TSH = 4.0 IU/mL (normal = 0.4 to 5 IU/mL)
Anti-Jo-1 antibodies: negative
Anti-Mi-2 antibodies: negative

Chest radiography and mammography were normal.

EMG was reported as having increased insertional activity, spontaneous fibrillations; abnormal myopathic low amplitude, short-duration polyphasic motor potentials; and high-frequency discharges.

A muscle biopsy of the right quadriceps is shown (see figure).



Courtesy of Cynthia Magro, MD and William W Pendlebury, MD.

She was treated with prednisone 60 mg per day and her CK and strength normalized. Two separate attempts to taper the prednisone below 20 mg/day resulted in weakness and increases in her CK to 750 IU/L.

What would be the most appropriate step at this time?

A. Begin oral methotrexate
B. Increase prednisone to 60 mg/day for three months and then repeat taper
C. Begin monthly intravenous immune globulin therapy
D. Recheck thyroid function
E. Initiate a more extensive workup for malignancy

Question 120

A 65-year-old man with a history of Wegener's granulomatosis (WG) presents to your office for follow-up. He was diagnosed with WG at age 59. He had sinus, pulmonary, and renal involvement and was treated with prednisone and cyclophosphamide. He received a total of 14 months of cyclophosphamide, approximately 2 mg/kg per day along with a tapering dose of prednisone and had a complete response to therapy. He has been off these medications for over three years and has been treated with prophylactic therapy consisting of daily trimethoprim-sulfamethoxazole since. The patient feels "great" and is completely asymptomatic with an unremarkable physical exam.

Laboratory studies reveal:

Hemoglobin = 10.4 g/dL [6.5 mmol/L] (normal = 13 to 18 g/dL [8.1 to 11.2 mmol/L])
ESR = 41 mm/hr (normal = 0 to 20 mm/hr)
Creatinine = 1.1 mg/dL [97.2 mmol/L] (normal = 0.8 to 1.3 mg/dL [70 to 114 mmol/L])
ANCA = 55 IU
Urinalysis = 25 RBC/HPF, no casts and trace protein
Liver function: normal
Chest x-ray: normal

The most appropriate management at this time would be?

A. Begin cyclophosphamide 2 mg/kg/day
B. Begin cyclophosphamide 2 mg/kg/day with prednisone 60 mg/day
C. Renal biopsy
D. Urological evaluation including cystoscopy
E. Begin methotrexate 20 mg/week

Question 121

A 70-year-old man is seen for evaluation of a progressively discolored and painful big toe. Over the past three weeks, the toe has become darker in appearance and more painful with weight-bearing activities. His primary care physician has prescribed hydrocodone for pain relief. Three months ago, he was started on warfarin therapy for atrial fibrillation. His other medications include simvastatin and atenolol.

Aside from having a bluish right big toe, the exam was notable for livedo reticularis over the thighs. The arterial pulses were present in both feet.

Laboratory studies are pending. You suspect that this patient has either a vasculitis or an atheroembolic lesion in his foot.

Which of the following laboratory features would you NOT expect to find if he has atheroembolic disease?

A. Eosinophiluria
B. Cryoglobulins
C. Hypocomplementemia
D. An inflammatory, perivascular infiltrate on biopsy
E. Elevated serum creatinine

Question 122

You are asked to consult on a 33-year-old woman admitted to the inpatient ophthalmology service for visual loss. Over the past three months, the patient has suffered multiple episodes of the rapid onset of decreased vision. She is

now completely blind in the left eye. She has a large blind spot in the right eye. There is no history of eye pain.

Review of systems reveals a history of episodic painful mouth sores.

Physical exam is unremarkable. Ophthalmologic examination is most consistent with a retinal vasculitis with evidence of vascular sheathing and vitreous hemorrhage.

CBC and liver and renal function studies are unremarkable. The ESR is 42 mm/hr (normal = 0 to 20 mm/hr). Other serologies are pending. A chest radiograph is unremarkable.

The differential diagnosis in this patient may include the following conditions EXCEPT?

A. Systemic lupus erythematosus (SLE)
B. Cogan's syndrome
C. Multiple sclerosis
D. Behcet's disease
E. Giant cell arteritis

Question 123

A 75-year-old man comes to your office for a first visit. He recently retired and moved to Greenwich, Connecticut. Since he will be living there full-time, he is concerned about his increased risk of Lyme disease and is inquiring about prevention. He denies a history of Lyme disease and feels completely well. A thorough review of symptoms is entirely negative.

Despite his lack of symptoms, he arranged through a physician friend to have a Lyme antibody test. The results include:

IgM Lyme ELISA: negative
IgG ELISA: Borderline positive
IgG Western blot: Displaying bands at 21, 31, 34, 39, 41, 66 and 93 Kd

What is the most appropriate interpretation of the data and therapeutic recommendation?

A. He was probably exposed to Borrelia burgdorferi in the past; however, he is asymptomatic and does not require treatment but Lyme vaccination would be useful.
B. He was definitely exposed to Borrelia burgdorferi and requires treatment with ceftriaxone 2 grams IV QD for 14 days.
C. He was exposed to Borrelia burgdorferi and requires treatment with amoxicillin and probenecid 500 mg po tid for three weeks.
D. He has not had Lyme disease and is a good candidate for the Lyme vaccine.
E. He has no evidence of active Lyme disease. Antibiotic treatment is of uncertain value in this setting. Vaccination would not be useful.

Question 124

A 75-year-old man with a history of rheumatoid arthritis presents with increasing pain in his right upper thigh over the past three months. He had a right total hip replacement four years previously without complications. He has had no problems with this hip until recently. His arthritis is otherwise doing well. He currently takes methotrexate 15 mg weekly and prednisone 5 mg daily.

He denies fever or chills, but recalls having undergone a dental procedure four months previously.

Which of the following statements regarding this patient is true?

A. The lack of fever or chills makes the diagnosis of a septic prosthetic joint extremely unlikely.
B. An infected prosthetic joint in this setting is most likely to be caused by an organism that contaminated the surgical field at the time of joint replacement.
C. Diagnosis of a septic prosthetic joint in this setting often requires biopsy of synovial tissue or periarticular bone.
D. Prophylactic antibiotics have been shown to reduce the risk of prosthetic joint infections in patients undergoing dental procedures.
E. Radiographs of a prosthetic joint of this age will usually allow for differentiation of aseptic loosening from infection.

Question 125

A 26-year-old, HIV-positive man presents with a two-month history of proximal muscle weakness. His CK is 2500 IU/L [41.7 μkat/L] (normal = 60 to 400 IU/L [1.0 to 6.7 μkat/L]). Among other medications, he is currently taking zidovudine (AZT). His internist asks you how to distinguish muscle disease due to HIV infection from that resulting from AZT.

Which one of the following would you recommend:

A. Electromyography
B. CD4+ T cell level
C. Repeat CK level
D. Muscle biopsy
E. Discontinue AZT

Question 126

A 33-year-old woman is referred to you by her obstetrician for your advice regarding the management of her pregnancy. She is currently 10 weeks pregnant and feels well. Last year, she had a spontaneous abortion at nine weeks. As part of her evaluation at the time, it was noted that she had an IgG antiphospholipid level of 38 U (GPL, normal <20 U). A recently repeated level was 43 U.

She denies any symptoms related to a possible connective tissue disease. She does not smoke and does not take any medications except multivitamins. She developed a deep venous thrombosis of the calf at age 27 and was maintained on anticoagulation for six months. Family history is strongly positive for osteoporosis. She is concerned about the safety of the treatment options and how they could impact upon the type of anesthesia she would receive at childbirth.

You suggest the initiation of anticoagulation therapy using heparin given subcutaneously.

Which of the following statements regarding the treatment of her condition is correct?

A. There is no need to start low dose aspirin as well as heparin.
B. Compared to unfractionated heparin, low molecular weight heparin is less frequently associated with the development of osteoporosis.
C. Low molecular weight heparin and unfractionated heparin carry similar risks for the development of epidural hematomas in women receiving regional anesthesia for childbirth.
D. This patient does not require anticoagulation therapy postpartum.

Question 127

A 34-year-old African-American man presents for rheumatologic evaluation; he was referred by his internist. He had

been diagnosed with scleroderma two years earlier when he presented with Raynaud's phenomenon, rapidly progressive skin changes, and arthralgias with tendon friction rubs. He has been treated with prednisone at low doses for his musculoskeletal complaints. The dose was recently increased to 10 mg qd to treat progressive hand stiffness. His past medical history is otherwise notable for peptic ulcer disease treated four years ago.

On examination, he is a thin man with sclerodermatous facies in no acute distress. His weight is 158 pounds (71 kg) and his blood pressure is 140/100 mmHg.

The cardiorespiratory exam was normal; his abdomen was nontender. Extremities revealed sclerodactyly with pits in the left 3rd and 4th digital tufts and acrocyanosis of the 3rd through 5th digits in both hands. Sclerodermatous skin changes extended proximally to the upper arms, the thighs, and over the chest wall. There were tendon friction rubs over the extensor tendons of the hands and knees.

Labs include a normal CBC and comprehensive metabolic profile. Creatinine was 1.5 mg/dL [132.6 mmol/L] (normal = 0.8 to 1.3 mg/dL [70 to 114 mmol/L]). Urinalysis revealed trace protein and 2-4 RBC/HPF without casts.

Which of the following options is most appropriate?

A. Begin nifedipine (long acting) 30 mg qd and aspirin qd to improve digital perfusion
B. Increase the prednisone dose to 15 mg qd
C. Begin captopril 25 mg tid and schedule a follow-up visit the next day
D. Begin celecoxib 200 mg qd and reduce the prednisone to 7.5 mg qd with plans for further taper
E. Do not start any new medications; arrange for follow-up visit in six weeks

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Question 128

You receive a telephone call from the daughter of one of your patients with rheumatoid arthritis (RA). She is concerned because her mother, a 64-year-old woman with longstanding RA, has new onset photophobia and severe, constant right eye pain of 16 hours' duration. Your patient thinks her vision may be somewhat reduced. She does not notice any ocular discharge, but says the white part of her eye is very red.

Ten years earlier, she was diagnosed with secondary Sjögren's syndrome. She has continued to use artificial tears on a regular basis. Current medications include hydroxychloroquine 400 mg qd, prednisone 5 mg qd, and atenolol 50 mg qd.

Given the information you have at the present time, what is the most likely diagnosis?

A. Primary open-angle glaucoma
B. Scleritis
C. Hydroxychloroquine related toxicity
D. Episcleritis
E. Vitreous hemorrhage

Question 129

A 50-year-old man received an allogeneic bone marrow transplant for chronic myelogenous leukemia seven years ago. Post-transplant, he developed features of chronic graft-versus- host disease (GVHD) which have been controlled with the use of prednisone and cyclosporine A. He was referred to you because of increasing skin tightness noted over his abdomen, thighs, and calves. He also complained of pain and stiffness affecting his shoulders, fingers, and knees.

Other medical problems include a history of dysphagia treated with a proton pump inhibitor and a history of bronchiolitis obliterans.

On examination, he has joint contractures affecting these joints.

Laboratory studies revealed:

Hemoglobin = 11.9 g/dL [7.4 mmol/L] (normal = 13 to 18 g/dL [8.1 to 11.2 mmol/L])
WBC = 5,400/mm3 with normal differential (normal = 4,300 to 10,800/mm3)
Platelets = 53,000/mm3 (normal = 130,000 to 400,000/mm3)
Creatinine = 4.7 mg/dL [415.6 mmol/L] (normal = 0.8 to 1.3 mg/dL [70 to 114 mmol/L])
AST = 44 IU/L (normal = 35 IU/L)
ALT = 54 IU/L (normal = 35 IU/L)
Alk phosphatase = 201 U/L [3.4 nkat/L] (normal = 30 to 120 U/L [0.5 to 2.0 nkat/L])
Anti-mitochondrial antibody: positive

Chronic graft-versus-host disease may explain all of the following features of this patient's illness EXCEPT?

A. Thrombocytopenia
B. Esophageal disease
C. Severe renal failure
D. Bronchiolitis obliterans
E. Antimitochondrial antibodies

Question 130

A 53-year-old Caucasian woman with fever is admitted to the hospital with shortness of breath for the past two weeks. She also complains of numbness and tingling in both feet, now extending up to the knees. For the past six weeks she has not felt well; she has noted progressive weakness, anorexia, and weight loss of 20 pounds.

On physical exam her temperature is 102.5°F (39.2°C), pulse is 100/min, respiratory rate is 20/min, and blood pressure is 126/80 mmHg. There are diffuse wheezes on expiration. The heart sounds are normal. The joint and skin exams are normal. The neurological exam demonstrates a reduction in sensation to vibration and pinprick along the soles of the feet extending halfway up the shins. The deep tendon reflexes are symmetric and 1+ at the knees and 0 at the ankles.

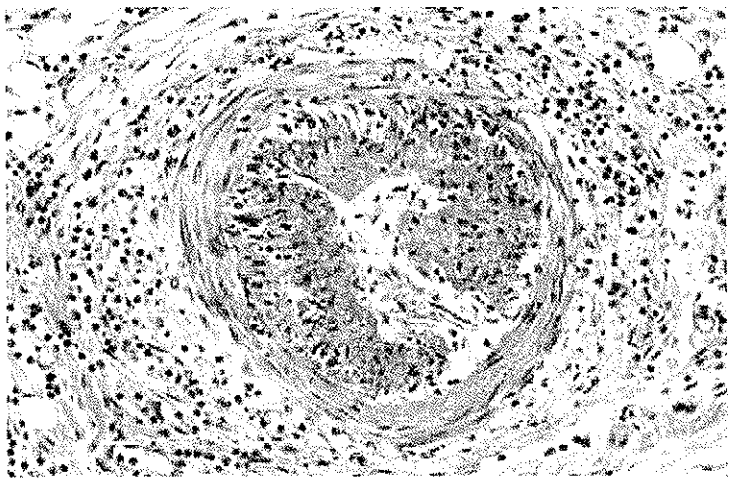
A chest radiograph shows bilateral pulmonary infiltrates.

Lab results:

Hemoglobin = 9.6 g/dL [6.0 mmol/L] (normal = 12 to 16 g/dL [7.4 to 9.9 mmol/L])
WBC = 20,000/mm3 with 40 percent neutrophils, 40 percent lymphocytes and 15 percent eosinophils (normal = 4,300 to 10,800/mm3)
ESR = 75 mm/hr (normal = 0 to 30 mm/hr)
BUN = 52 mg/dL [18.6 mmol/L] (normal = 10 to 20 mg/dL [3.6 to 7.1 mmol/L])
Creatinine = 2.6 mg/dL [230.0 mmol/L] (normal = 0.6 to 1.0 mg/dL [53 to 88 mmol/L]),
Urinalysis = 2+ protein, 2+ blood, no casts

Hepatitis B and C screen: negative
HIV: negative

The following day the patient was noted to have a right foot drop. A sural nerve biopsy was performed (see figure).



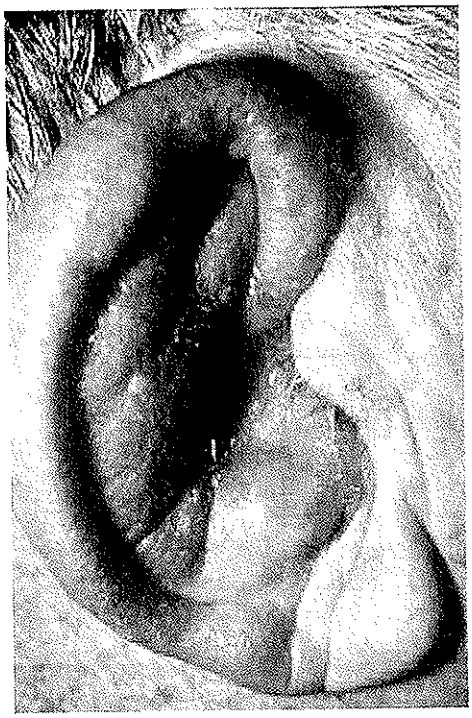
The patient's diagnosis is?

A. Hypereosinophilic syndrome
B. Churg-Strauss syndrome
C. Polyarteritis nodosa
D. L-tryptophan induced disease
E. Wegener's granulomatosis

Question 131

A 55-year-old man presents with a one-week history of redness, pain, and mild swelling of the ears and pain and stiffness of the fingers bilaterally. He reports three previous episodes of similar ear symptoms over the past eight months, each lasting one to two weeks.

Physical examination reveals an erythematous ear (see figure).



Courtesy of Jerome H Herman, MD.

The midline anterior neck is tender to palpation. Chest is clear to auscultation. Heart exam is normal. Joint examination shows mild tenderness of the proximal interphalangeal joints with normal range of motion.

Laboratory studies include a hematocrit of34.2 percent (normal = 42 to 52 percent) with normal MCV and MCH, urinalysis and chemistry panel, and a Westergren sedimentation rate of 65 mm/hr (normal = 0 to 20 mm/hr). Rheumatoid factor, anti-nuclear antibody, and anti-neutrophil cytoplasmic antibody are negative.

Which one of the following organs/tissues is most commonly involved in patients with this disorder?

A. Laryngotracheobronchial airway
B. Peripheral joints
C. Eye
D. Internal ear
E. Medium-sized blood vessels

Question 132

A 60-year-old man with a cardiac arrhythmia has been treated with procainamide for five years. He recently presented to his internist because of arthralgias, swollen joints, fatigue, fever of 101°F (38.6°C), and dyspnea. Upon examination, he was found to have a pleural effusion and a positive ANA at 1:320 (diffuse).

Which one of the following tests is least helpful in distinguishing SLE from drug-induced lupus?

A. Urinalysis
B. Serum complement levels
C. Anti-dsDNA antibodies
D. Anti-histone antibodies

Question 133

A 64-year-old man with ankylosing spondylitis (AS) whom you have known for many years comes to clinic with the complaint of weakness and numbness of the hands and difficulty climbing stairs for the past three days. He has painful paresthesias in his hands. He has noticed some urinary frequency and constipation. He denies any trauma to his neck or back.

On physical examination, his temperature is 98.8°F (37.4°C), pulse is 98 beats/min, respirations are 16/min, and blood pressure is 160/95 mmHg. General physical examination is unremarkable. His spine is unchanged compared to his baseline, namely rigid and nontender. Neurologic examination reveals bilateral grip weakness, decreased sensation of light touch in the hands and forearms, and decreased strength in the lower extremities. He is mildly hyperreflexic in the upper and lower extremities.

What is the most likely cause of the patient's symptoms?

A. Herniated cervical disc at C4-5
B. Cauda equina syndrome
C. Atlantoaxial subluxation
D. Syndesmophyte fracture at the C5-6 interspace
E. Compression fracture with secondary spinal stenosis

Question 134

A 48-year-old pharmacist with rheumatoid arthritis (RA) has been treated with hydroxychloroquine (HCQ) 400 mg per day for the past year. Her RA has been well controlled, however she is concerned about potential side effects related to the drug.

For example, she says her serum cholesterol had risen from 189 mg/dL [4.9 mmol/L] one year ago to 199 mg/dL [5.1 mmol/L] today (desirable <200 mg/dL [<5.17 mmol/L]). She read a review article suggesting that HCQ may cause a proximal muscle myopathy and chronic use may increase the risk for stroke and cardiomyopathy. Occasionally she notes a mild abdominal pain and bloating after taking the medication. Finally, she attributes her inability to focus on nearby objects as being a manifestation of HCQ toxicity.

Which of the following statements regarding hydroxychloroquine use is valid?

A. It may elevate the serum cholesterol.
B. It increases the risk for stroke.
C. It may cause gastric ulcer formation.
D. It may cause a proximal muscle myopathy but not a cardiomyopathy.
E. An inability to focus may be noted within the first few weeks of therapy but generally resolves on its own.

Question 135

A 34-year-old woman with seropositive rheumatoid arthritis of 11 months' duration presents to you for a second opinion regarding the management of her illness. She has been treated with several different nonsteroidal antiinflammatory agents with minimal relief of her symptoms.

On examination, you note active synovitis in the small joints of her hands and feet. Radiographs reveal small marginal erosions in her metacarpal heads and ulnar styloid. Methotrexate therapy had been recommended by her previous physician, but she is unwilling to use it because of fear of toxicity as well as potential teratogenicity.

She has a history of a skin rash with the use of sulfamethoxazole. You consider sulfasalazine therapy for her RA.

Which of the following statements regarding this decision is correct?

A. Therapy with sulfasalazine is not possible because of her history of allergy to sulfonamide antibiotics.
B. Although sulfasalazine could be used, the evidence that it is beneficial in this patient with erosive rheumatoid arthritis is equivocal.
C. In clinical trials, skin rash is the most common adverse reaction related to sulfasalazine.
D. The patient should be cautioned about possible infertility related to sulfasalazine use.
E. Sulfasalazine is an appropriate choice for therapy for her rheumatoid arthritis.

Question 136

A 48-year-old woman with a 12-year year history of rheumatoid arthritis (RA) presents with a four-week flare of painful swelling of her metacarpophalangeal (MCP) joints, wrists, metatarsophalangeal joints, and knees. This occurred after slowly tapering her prednisone from 10 mg/day to 5 mg/day over a two-month period. The dose had been increased after infliximab was discontinued because of the development of a severe itch and urticaria during her fifth infusion of the medication.

Aside from prednisone, other medications include: methotrexate 25 mg per week, folic acid 1 mg/day, and ibuprofen 600 mg TID.

The joint exam shows synovitis involving the wrists, small joints of the hands, both knees, and feet.

Her ESR is 65 mm/hr (normal = 0 to 30 mm/hr). Radiographs of the hands demonstrate joint space narrowing involving several of the MCP joints bilaterally and erosions are seen at the distal ulnae. These are new compared to the most recent radiographs taken two years earlier.

You have access to and are considering the use of an interleukin-1 receptor antagonist (IL-1ra) therapy. Which of the following statements regarding this therapy is true?

A. Similar to infliximab, this is a chimeric protein and thus should not be used in this patient with a previous allergic reaction to infliximab.
B. Based on data from clinical trials, you should advise her that slowing of radiographic progression would require at least two years of treatment.
C. The patient should be cautioned that the most common adverse effect is the development of transient injection site reactions.
D. Because IL-1ra inhibits prostaglandin production, her ibuprofen should be discontinued.

Question 137

A 23-year-old woman with systemic lupus erythematosus is scheduled to initiate therapy with intravenous pulse cyclophosphamide. She is currently being treated with prednisone 40 mg daily. She is concerned about potential toxicities of cyclophosphamide.

Laboratory studies reveal:

Serum creatinine = 2.2 mg/dL [194.5 mmol/L] (normal = 0.6 to 1.0 mg/dL [53 to 88 mmol/L])
Complete blood count: normal
Electrolytes: normal
Liver function: normal

Which of the following statements regarding therapy with cyclophosphamide in this patient is TRUE?

A. She requires a reduction in the initial intravenous (IV) pulse cyclophosphamide dose below the standard dose of 0.75 g/m2.
B. Ovarian failure is more likely to occur because she is young.
C. The risk for the development of bladder cancer would be approximately equal whether she was treated with intravenous pulse or oral cyclophosphamide.
D. The development of a marked leukopenia during cyclophosphamide therapy would require cessation of therapy in this patient.

Question 138

A 72-year-old woman presents with severe right-sided headaches radiating from the temple to occiput, severe myalgias of the hip and shoulder girdles, morning stiffness lasting three hours, and jaw pain. There is no history of vision loss or jaw claudication.

Physical examination reveals that the right temporal artery is tender to palpation. There is also some tenderness in the occipital area on the right side. There are normal pulsations of the left temporal artery without beading or bruits.

The patient has an extensive history of cigarette smoking and is status-post myocardial infarction. The erythrocyte sedimentation rate (ESR) is 93 mm/hr (normal = 0 to 30 mm/hr).

The patient is prescribed prednisone 60 mg per day. A right temporal artery biopsy performed five days later does not confirm the diagnosis of giant cell arteritis (GCA) despite an extensive evaluation of multiple sections.

Which of the following is the best strategy?

A. Continue prednisone at 60 mg/day for presumed giant cell arteritis since positive histopathologic findings were obscured by prednisone.
B. Discontinue prednisone
C. Obtain a biopsy of the left temporal artery
D. Lower prednisone dose to treat a clinical diagnosis of polymyalgia rheumatica
E. Continue prednisone and obtain a biopsy of the right occipital artery

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Question 139

A 23-year-old graduate student was referred for consultation because of acute hearing loss and an elevated ESR. There was sudden, bilateral loss of hearing without any other complaints being noted. She denied seizures, paresthesias, confusion, and new changes in vision.

She had a history of migraine headaches with visual aura for which she used an aspirin/caffeine medication.

Serial audiograms revealed fluctuating bilateral sensorineural hearing loss. Brain MRI showed high signal intensity (on T2 weighted images) in the periventricular white matter. Neuropsychiatric testing revealed a decrease in the patient's IQ from 130 to 101 with documented difficulty with executive function, retrieval and retention of data, and no depression.

The ESR was 69 mm/hr (normal = 0 to 20 mm/hr). The CBC, electrolytes, thyroid function, ANA and hepatitis serologies, and multiple blood cultures were all normal.

A lumbar puncture revealed the following cerebrospinal fluid findings: An elevated opening pressure; total protein 64 mg/dL; 4 wbc/hpf with 100 percent lymphocytes and glucose of 88 mg/dL.

You suspect an isolated vasculitis of the central nervous system (ICNSV). A cerebral angiogram is performed and demonstrates multiple occlusions with sharp cutoffs consistent with a vasculitis.

Which of the following options would you recommend next?

A. Positron emission tomography (PET) scan and serum ANCA antibody
B. Combined cortical and leptomeningeal biopsy
C. Temporal artery biopsy
D. Begin high dose prednisone
E. Begin high dose prednisone and cyclophosphamide

Question 140

A 30-year-old Caucasian woman with rheumatoid arthritis (RA) is seen for a routine follow-up visit. She has had RA for ten years, which has been recently treated with leflunomide 20 mg qd. She has decided to try and become pregnant. She usually has regular menses, but is three weeks late. She seeks your advice regarding therapeutic options for her RA.

Current medications include leflunomide 20 mg qd and naproxen 500 mg bid.

The joint exam is notable for some soft tissue swelling over both wrists, MCPs, and knees bilaterally.

Which one of the following options would you recommend?

A. Maintain current drug regimen until patient has a positive pregnancy test
B. Discontinue leflunomide
C. Discontinue leflunomide and begin cholestyramine
D. Discontinue leflunomide and begin etanercept
E. Advise patient to avoid becoming pregnant for at least one year

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Question 141

A 29-year-old teacher is seen in your office on an urgent basis with severe shortness of breath. She was seen by one of your associates one month earlier and described a two-month history of puffy hands, polyarthralgias, and Raynaud's phenomenon. The patient's internist had prescribed three different non-steroidal anti-inflammatory drugs which had been of no benefit and your partner, suspecting an early connective tissue disorder, advised initiation of prednisone at a dosage of 10 mg per day. This had been helpful in relieving her joint discomfort.

At the time of her visit to you, she described a one-week history of breathlessness on exertion and nighttime shortness of breath awakening her from sleep. She denied any fever, cough, or chest pain. Her only medication was prednisone.

On physical examination her temperature is 98.5°F (36.9°C), pulse is 110, respirations are 24, and blood pressure is 170/118 mmHg. Jugular venous pressure was not elevated. Bibasilar rales were present at the lung bases. Cardiac auscultation revealed an S3 without murmur or pericardial rub. Abdomen was nontender without masses or organomegaly. There was no lower extremity edema. There was mild sclerodactyly but no proximal skin thickening. Tendon friction rubs were present over the anterior tibialis tendons bilaterally.

You directed her to the Emergency Room and laboratory studies were obtained.

Hemoglobin = 10 g/dL [6.2 mmol/L] (normal = 12 to 16 g/dL [7.4 to 9.9 mmol/L])
Platelet count = 90,000/mm3 (normal = 130,000 to 400,000/mm3)
White cell count = 9,000/mm3 (normal = 4,300 to 10,800/mm3)
Creatinine = 2.0 mg/dL [176.8 mmol/L] (normal = 0.6 to 1.0 mg/dL [53 to 88 mmol/L])
Urinalysis = trace protein, no casts
Antinuclear antibody = 1:160 in a nucleolar pattern

Chest x-ray: Bibasilar infiltrates and cardiomegaly

Echocardiogram: Mild pericardial effusion
Electrocardiogram: Sinus tachycardia

The most likely diagnosis is?

A. Thrombotic thrombocytopenic purpura (TTP)
B. Scleroderma renal crisis
C. Systemic lupus erythematosus (SLE)
D. Interstitial lung disease
E. Hemolytic-uremic syndrome (HUS)

Question 142

A 43-year-old woman with hepatitis C liver disease and cryoglobulinemia has noted the onset of fatigue. She notes that this development has progressed slowly over the past month. She denies any joint pain, dysesthesia, fever, or chills.

Past history is notable for intravenous drug abuse while a college student. She denies any illicit drug use for the past two decades.

Treatment of her condition has been with interferon alpha-2a, 2 million units three times per week for the past six months. According to her gastroenterologist, the patient's last set of blood tests, which included a complete blood count and liver enzymes, was normal.

The most likely diagnosis is?

A. Interferon-induced hypothyroidism
B. Worsening cryoglobulinemia
C. Worsening liver disease
D. Illicit drug use
E. Interferon-induced myopathy

Question 143

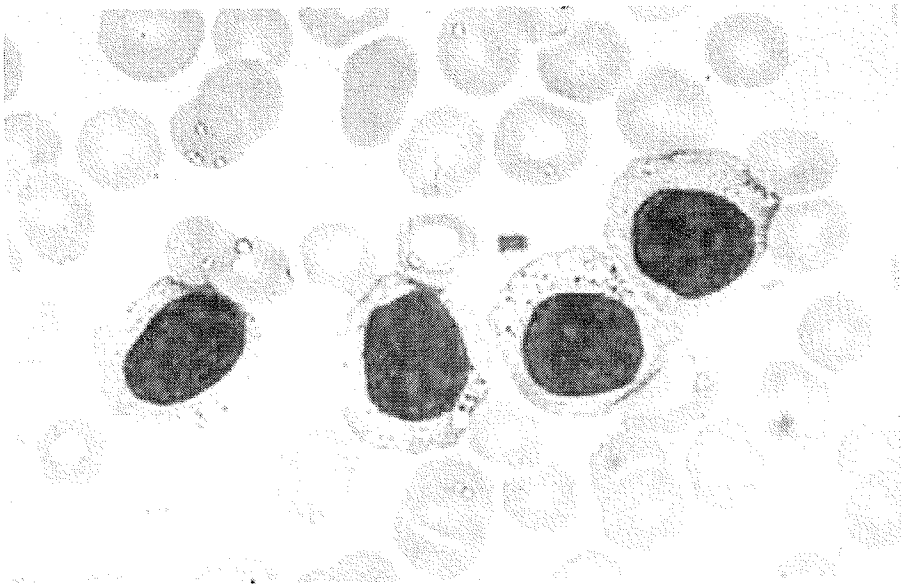
A 50-year-old veterinarian is referred to you for management of his polyarthritis. He had the onset of symmetric joint swelling involving the wrists, hands, and feet two years ago. He has been treated with hydroxychloroquine, sulfasalazine, and methotrexate. Each drug was discontinued at some point by his previous rheumatologist because of a persistently low white blood cell count. He has had several hospitalizations for treatment of sinus infections, pneumonia, and skin abscesses.

The physical exam reveals a chronically ill man. There is synovitis of the wrists and the metacarpophalangeal joints of both hands. There is no lymphadenopathy or organomegaly.

Laboratory studies reveal:

Hematocrit = 34 percent (normal = 42 to 52 percent)
Platelet count = 119,000/mm3 (normal = 130,000 to 400,000/mm3)
White blood cell count = 1,750 cells/mm3 with 70 percent lymphocytes and 25 percent neutrophils (normal = 4,300 to 10,800/mm3)
ANA: positive 1:640
RF: 220 U

A peripheral blood smear is shown (see figure).



Courtesy of Thomas Loughran, MD.

All of the following features would be observed in patients with this condition EXCEPT?

A. Infection is the leading cause of mortality
B. There is a mild to moderate lymphocytosis
C. The course of the polyarthritis will not affect the course of the hematologic disorder
D. Many patients carry the HLA-DR4 genotype
E. Rheumatoid factor (RF) positivity is universal

Question 144

A 53-year-old nurse with longstanding Sjögren's syndrome is seen for a routine follow-up visit. She has noted the recent onset of a generalized itch, which makes her very uncomfortable throughout the day. Additionally, she thinks that her skin color has darkened over the past year. She denies any weight change or fever. Her sicca symptoms have not changed.

Current medications include artificial tears and atenolol 50 mg qd for hypertension.

On examination, the skin is slightly hyperpigmented. There are several excoriations from scratching. There is hepatosplenomegaly without tenderness. No other abdominal masses are felt and there is no lymphadenopathy. The small joints of the hands show changes consistent with osteoarthritis.

Laboratory results:

Hemoglobin = 13.1 g/dL [8.1 mmol/L] (normal = 12 to 16 g/dL [8.1 to 11.2 mmol/L])
WBC = 7,600 cells/mm3 (normal = 4,300 to 10,800 /mm3)
Platelets = 125,000/mm3 (normal = 130,000 to 400,000/mm3)
Creatinine = 1.1 mg/dL [97.2 mmol/L] (normal = 0.6 to 1.0 mg/dL [53 to 88 mmol/L])
ALT = 33 IU/L (normal = 35 IU/L)
AST = 38 IU/L (normal = 35 IU/L)
Alkaline phosphatase = 613 IU/L [10.2 nkat/L] (normal = 30 to 120 IU/L [0.5 to 2.0 nkat/L])
Albumin = 3.7 g/dL (normal = 3.5 to 5.5 g/dL)
Hepatitis C antibody: negative

Hepatitis B surface antigen: negative

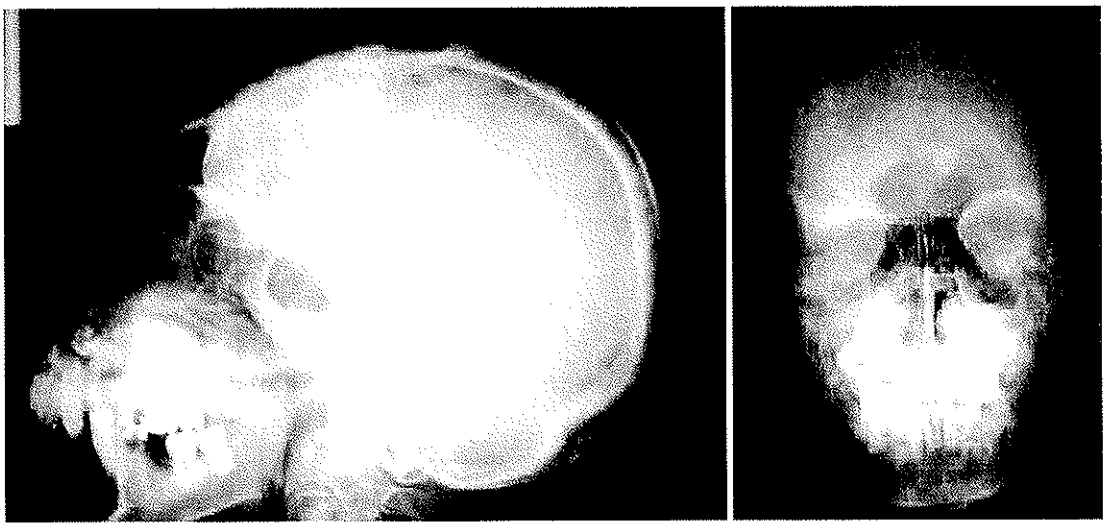
ANA: 1:640 speckled
Anti-Ro antibody: negative
Anti-La antibody: 95 IU

All of the following statements regarding this patient's condition are true EXCEPT?

A. This patient is at risk for the development of hepatocellular carcinoma.
B. In the early phase of this condition, there may be striking elevations of the serum high-density lipoproteins (HDL).
C. If positive, the titer of antimitochondrial antibodies (AMA) predicts disease progression.
D. Survival is decreased and cirrhosis is more likely to be present in patients with Sjögren's syndrome.
E. This condition has never been reported in childhood.

Question 145

A 52-year-old man presents with skull pain. The pain has been present for about one year and has been slowly progressive. A radiograph of the skull is taken (see figure).



Courtesy of Nicky Kelepouris, MD.

Which of the following statements regarding this condition is true?

A. This condition is common in Asia and Africa.
B. Hearing loss is a rare complication.
C. This patient may be at risk for the development of a facial palsy.
D. Treatment of these lesions with radiation therapy carries an excellent prognosis.
E. The serum parathyroid hormone level (PTH) is infrequently elevated.

Question 146

You are considering the initiation of infliximab therapy in a 57-year-old woman with longstanding rheumatoid arthritis (RA). She is interested but is concerned that infliximab has a murine component. A friend of hers developed urticaria after a number of infusions of infliximab. She is also concerned because of her history of allergies to pollen, grasses, and cats.

Which of the following statements regarding infliximab and its murine component is correct?

A. The development of a human anti-chimeric molecule antibody (HACA) response is detected in the majority of patients treated with infliximab.
B. A lower dose of infliximab will likely reduce the possibility of developing an anti-HACA antibody response.
C. Anti-HACA antibodies clearly impair the clinical response to infliximab.
D. Anti-HACA antibodies are associated with a higher overall incidence of side effects with infliximab.
E. Concomitant methotrexate use reduces the HACA response.

Question 147

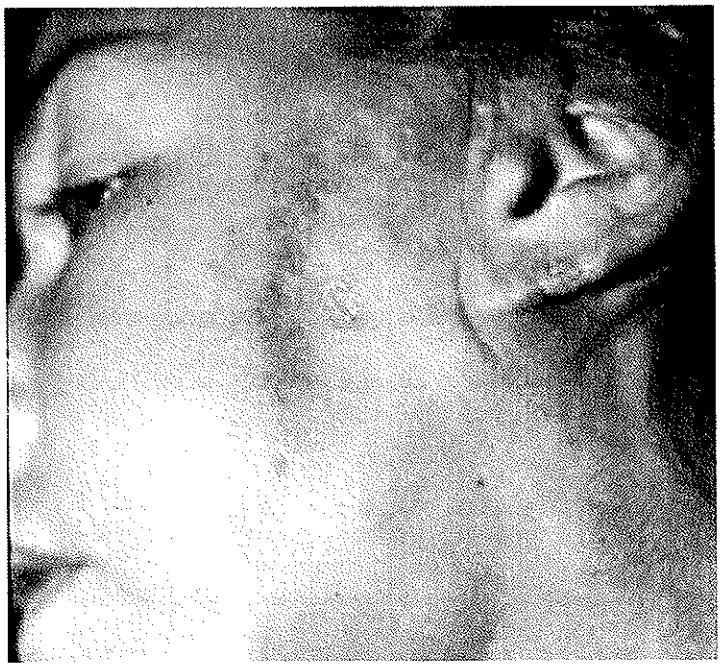
A 28-year-old African-American woman is referred from dermatology clinic because of the results of a recent skin biopsy from a new facial lesion. She developed an area of painful redness and induration over her cheek (see figure).

A review of systems is unremarkable. She has no other medical problems.

Laboratory evaluation reveals:

CBC: normal
ESR = 21 mm/hr (normal = 0 to 20 mm/hr)
ANA = 1:320

Other laboratory tests and serologies are pending.



Courtesy of Samuel Moschella, MD.

What do you recommend?

A. Reassure the patient that she is unlikely to have a systemic illness
B. Begin hydroxychloroquine 400 mg daily, informing her that she is at risk of developing systemic lupus erythematosus (SLE)
C. Begin doxycycline 200 mg qd for 28 days
D. Begin D-penicillamine 500 mg qd
E. Arrange for HIV testing

Question 148

A 74-year-old retired engineer is referred for evaluation of polyarthritis. He seeks a second opinion for his condition, which was labelled as rheumatoid arthritis by the first rheumatologist he saw. Three years ago he developed the onset of swelling and dysesthesia of both hands. He was told that he had carpal tunnel syndrome, but he refused surgery. For the past year he has noted bilateral shoulder swelling. There has been a gradual loss of motion but only slight pain. He is concerned about the loss of strength in both hands and his lessened ability to make a fist.

The musculoskeletal examination reveals bilateral swelling of both shoulders. There is slight loss of rotation but minimal discomfort with passive range of motion. There is fullness over the wrists bilaterally and thickening of the palmar fascia bilaterally with a moderate degree of flexion contractures present. There is soft tissue swelling around the metacarpophalangeal and proximal interphalangeal joints bilaterally. The hips show full range of motion, and the knees show moderate synovial thickening bilaterally.

Laboratory results:

Hemoglobin = 12.0 g/dL [7.4 mmol/L] (normal = 13 to 18 g/dL [8.1 to 11.2 mmol/L])
WBC = 7,700/mm3 (normal = 4,300 to 10,800/mm3)

Platelets = 232,000/mm3 (normal = 130,000 to 400,000/mm3)
ESR = 66 mm/hr (normal = 0 to 20 mm/hr)
RF: negative
ANA: negative

All of the following features may characterize this condition EXCEPT?

A. Subcutaneous nodules
B. Jaw claudication
C. Non-inflammatory synovial fluid
D. Joint erosions
E. Symmetric polyarthritis

Question 149

A 42-year-old African-American military officer based in Arizona was seen with a four-month history of progressive swelling in the left knee and right 5th DIP joint. Two months previously, he had experienced a two-week illness characterized by cough, fever, chills, and night sweats. He had three separate aspirations and steroid injections of the left knee in the past four months with minimal benefit, and cultures of the synovial fluid had been negative each time.

Physical examination revealed marked induration and swelling of the left knee and into the soft tissues above and below the knee. There was swelling of the right 5th DIP joint with serosanguineous drainage from the tip.

Synovial fluid left knee: WBC 27,000/mm3 with 84 percent polys and 16 percent mononuclear cells.

Radiographs of the knee and finger reveal soft tissue swelling with a suggestion of a lytic bony lesion involving the proximal interphalangeal joint.

Which of the following diagnoses is most likely?

A. Coccidiomycosis
B. Lyme disease
C. Mycobacteria marinarum
D. Histoplasmosis
E. Seronegative spondyloarthropathy

Question 150

A 35-year-old man is admitted to your service because of the new onset of a systemic illness characterized by fever, fatigue, and hypertension. He has been in good health except for a prior history of hepatitis B five years ago.

On examination, his blood pressure is 150/100 mmHg, pulse is 90, respirations are 17, and temperature is 100.1°F (38.4°C).

There is livedo reticularis over the arms and legs. The remainder of the exam is unremarkable.

Laboratory studies reveal:

Hemoglobin = 11.8 g/dL [7.3 mmol/L] (normal = 13 to 18 g/dL [8.1 to 11.2 mmol/L])
WBC = 7,500/mm3 with normal differential (normal = 4,300 to 10,800/mm3)
Platelets = 370,000/mm3 (normal = 130,000 to 400,000/mm3)
ESR = 77 mm/hr (normal = 0 to 15 mm/hr)
Creatinine = 2.8 mg/dL [247.5 mmol/L] (was 1.5 mg/dL [132.6 mmol/L] on admission) (normal = 0.8 to 1.3 mg/dL [70 to 114 mmol/L]

AST = 57 IU/L (normal = 35 IU/L)
ALT = 45 IU/L (normal = 35 IU/L)
ANCA: negative
Urinalysis: 1-3 RBC/HPF, 2+ protein, no casts

A renal biopsy is recommended by the renal service and they seek your opinion. What would you recommend and why?

A. Recommend EMG and nerve conduction study, and if indicated, proceed with a sural nerve biopsy since it is a less morbid procedure than renal biopsy
B. Repeat ANCA and await results of anti-PR3 antibody, as you suspect first result is an error
C. Do not recommend renal biopsy unless urinalysis demonstrates casts since renal biopsy findings may be nonspecific
D. Order ANA, C3, C4 and defer decision until results are available as you consider systemic lupus a significant possibility
E. Proceed with renal biopsy looking for evidence of arteriolitis or focal necrotizing glomerulonephritis

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RSAP2001: Rheumatology Self-Assessment Program

Part II: Answers

Answer to Question 1
Answer: E

This patient demonstrates features of the antiphospholipid antibody syndrome (APS) and SLE. In addition to the development of thrombotic events, patients may have other organ involvement including cardiac, pulmonary, renal, hematologic, and cutaneous. This patient has acutely developed a major thrombosis and a pulmonary embolus. Such patients should receive life-long warfarin therapy to achieve an INR of 3.0 or higher. Thus, choice E is correct.

The other choices are not appropriate:

- The administration of warfarin to achieve an INR of only 2.0 to 2.5 has been found to provide less protection against recurrent thrombosis than that achieved with higher INRs.

- Less than life-long warfarin therapy would place the patient at increased risk for recurrent thrombosis, particularly if her antiphospholipid antibody titers remain elevated.

Educational objective: To recognize the indications for chronic warfarin therapy in the patient with antiphospholipid antibody syndrome.

For more information see the following UpToDate topic review(s) in the RSAP2001 program:
1. Prognosis and therapy of the antiphospholipid antibody syndrome

Answer to Question 2
Answer: D

Endothelin is the most potent vasoconstrictor known. It is believed to play an important role in the pathogenesis of scleroderma and in patients with primary Raynaud's phenomena. The basal secretion of endothelin-1 from the endothelial cell may provide an important early link between endothelial cell damage and fibroblast activation. Elevated circulating levels of endothelin-1 have been found in patients with scleroderma or secondary Raynaud's phenomenon. Thus, choice D is correct.

Adrenergic mechanisms appear critical to the development of primary Raynaud's phenomena. The abnormal cold responsiveness is most likely to be mediated by the alpha-2 adrenergic response. Selective alpha-2 adrenergic blockade has been shown to be more effective in diminishing vasospastic attacks compared with selective alpha-1 adrenergic antagonists.

There appears to be reduced activity of nitric oxide in patients with systemic sclerosis. Endothelial derived nitric oxide inhibits vascular smooth muscle contraction, platelet aggregation, and leukocyte and platelet adhesion to endothelial cells.

Endothelial dysfunction is likely an early event with patients with Raynaud's phenomena and systemic sclerosis. Studies have demonstrated endothelial abnormalities in the uninvolved skin of patients with early systemic sclerosis.

Educational objective: To understand current concepts in the pathogenesis of Raynaud's phenomena.

For more information see the following UpToDate topic review(s) in the RSAP2001 program:
1. Pathogenesis of Raynaud's phenomena
2. Pathogenesis of Scleroderma

Answer to Question 3
Answer: C

The eosinophilic-myalgia syndrome was first identified when some patients taking supplements of the amino acid L-tryptophan developed severe muscle pain and eosinophilia eventually leading to its removal from the market. While

muscle pain was the hallmark, some patients developed neuropathic disease ranging from paresthesias to a sensory peripheral neuropathy to a devastating ascending polyneuropathy leading to respiratory failure. This was the most common cause of death reported to the Centers for Disease Control. Thus, answer C is correct.

The other answers are incorrect.

- Scleroderma skin changes including induration and hide-bound changes developed in some patients, even to the point of resembling eosinophilic fasciitis with the skin texture having the appearance of peau d'orange. Typically, nonpitting edema affecting the extremities was observed in 60 percent of patients.

- Myalgias and proximal myopathy were noted in many patients. Muscle histopathology revealed a perimyositis. EMG studies were usually normal; in some cases, nerve conduction studies did reveal evidence of a sensory motor neuropathy.

- Fasciitis sometimes involved the skin over large joints, and large joint polyarthralgias were common; however, synovitis was not observed.

- Although significant heart disease was uncommon, a few cases of myocarditis, coronary artery spasm, and sinus tachycardia were reported. Sudden death was associated with postmortem findings of fibrosis of the sinoatrial node and bundle of His.

Educational objective: To recognize the clinical manifestations of eosinophilic-myalgia syndrome.

For more information see the following UpToDate topic review(s) in the RSAP2001 program:
1. Pathogenesis and clinical manifestations of the eosinophilia-myalgia syndrome
2. Treatment and prognosis of the eosinophilia-myalgia syndrome

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Answer to Question 4
Answer: C

When evaluating patients with lower extremity pain, one should consider non-arthritic causes, particularly when the pain is not maximally present over a joint or there are sensory complaints. This latter symptom suggests a neurologic cause. Among those to consider are the syndromes caused by nerve entrapment, either at the level of the lumbosacral spine or more peripherally. The lateral femoral cutaneous nerve is a small sensory nerve that is a direct branch of the lumbar plexus. Entrapment of the nerve as it traverses below the inguinal ligament medial to the anterior superior iliac spine produces the very common syndrome of meralgia paresthetica. These patients typically present with complaints of paresthesias and pain that radiates down the lateral aspect of the thigh toward the knee. The pain can be quite significant. In more advanced cases, a fixed sensory loss on the lateral thigh occurs. There are no motor or deep tendon reflex abnormalities on exam.

The only known risk factor for this condition is the wearing of tight fitting belts by obese individuals. There is no increased incidence in patients with RA.

EMG/NCS generally have a limited role in the evaluation of this disorder. Studies may be relatively normal in mild cases. Needle electromyography is performed to help exclude radiculopathy or plexopathy.

Mononeuropathies of the obturator nerve are uncommon. When present, it is often secondary to pelvic trauma or surgery. Presenting features include pain, weakness in leg adduction, and sensory loss over a small area in the medial thigh.

Educational objective: To be familiar with non-arthritic causes of leg pain.

For more information see the following UpToDate topic review(s) in the RSAP2001 program:
1. Overview of lower extremity peripheral nerve syndromes

Answer to Question 5
Answer: B

This patient with scleroderma presents with features suggestive of scleroderma renal crisis. These include the abrupt onset of renal failure and hypertension, a microangiopathic hemolytic anemia, and a urinalysis showing only mild proteinuria without an active sediment.

In this situation, renal function may continue to deteriorate through the first several days of therapy with an angiotensin converting enzyme (ACE) inhibitor. Although the blood pressure should be lowered slowly, with a target reduction of 10 to 15 mm Hg per day, her control is not yet adequate. The dose of enalaprilat should be increased.

The use of other antihypertensive agents such as hydralazine or minoxidil are occasionally required to achieve blood pressure control, but these drugs should not be employed until the ACE inhibitor therapy has been increased to the maximal dosage.

There is no evidence that plasmapheresis is useful in the treatment of scleroderma renal crisis.

Educational objective: To review the management of scleroderma renal crisis.

For more information see the following UpToDate topic review(s) in the RSAP2001 program:
1. Scleroderma renal disease
2. Drug treatment of hypertensive emergencies

Answer to Question 6
Answer: D

This patient with longstanding RA has developed features of cervical myelopathy probably secondary to cervical subluxation, a complication of RA. These patients can develop symptoms insidiously, either with or without neck pain. This patient appears to be developing a spastic quadraparesis. Imaging studies of the neck are required to further evaluate this condition, in particular the atlantoaxial joint where subluxation can occur most frequently.

An MRI of the lumbar spine would not be indicated. The patient's brisk hyperreflexia would not be consistent with a lesion in the lumbar spine.

An EMG and nerve conduction study, although helpful, would not be as accurate as a cervical MRI in localizing the area of damage.

This patient's history and physical exam are not consistent with either a statin-induced myopathy or with a corticosteroid-induced proximal myopathy.

Educational objective: To be familiar with the presentation of cervical spine disease and its complications in patients with RA.

For more information see the following UpToDate topic review(s) in the RSAP2001 program:
1. Cervical subluxation in rheumatoid arthritis

Answer to Question 7
Answer: D

Bleomycin, an antitumor antibiotic, has been used to treat a variety of tumors including squamous cell carcinoma of the head and neck, cervix, esophagus, and the lymphomas. The major limitation of its use relates to the development of a dose dependent, life-threatening pneumonitis. In 10 percent of cases, this may progress to interstitial pulmonary fibrosis. In addition, several cases of scleroderma with Raynaud's phenomenon have been described in patients undergoing cancer chemotherapy with bleomycin. However, a positive ANA has not been observed. Thus, choice D is

correct.

Educational objective: To recognize the rheumatologic syndromes that may be associated with the prior use of chemotherapeutic agents, particularly bleomycin.

For more information see the following UpToDate topic review(s) in the RSAP2001 program:
1. Bleomycin-induced lung injury
2. Risk factors for and possible causes of scleroderma

Answer to Question 8
Answer: D

Colchicine is an effective therapy for the treatment of crystal-induced arthritis, such as gout or pseudogout. It appears to be equally effective in both conditions. Long-term use of colchicine has been associated with the development of a neuromyopathy. The histopathology demonstrates vacuole formation; however, the mitochondria are not involved.

Although the efficacy of prophylactic therapy is the subject of some controversy, oral colchicine is of value in reducing the frequency of recurrent acute gouty arthritis in at least some patients with a prior history of gout. Suppression of the influx of inflammatory cells into synovium may in part underlie the prophylactic action of colchicine.

The use of intravenous colchicine is associated with the same toxicities as seen with oral ingestion of the drug. These include gastrointestinal symptoms such as diarrhea, nausea, or anorexia; neurologic symptoms; and skin rash. The maximum intravenous dose should in no instance exceed 4 mg during any 24-hour period or during any attack.

Because of its sclerosing effects, one needs to avoid local infiltration of colchicine into the adjacent soft tissue. Thus, it is never given intramuscularly.

Educational objective: To be familiar with the use of colchicine.

For more information see the following UpToDate topic review(s) in the RSAP2001 program:
1. Treatment of gout
2. Drug-induced myopathies

Answer to Question 9
Answer: B

Henoch-Schönlein purpura is a systemic vasculitis characterized by the tissue deposition of IgA-containing immune complexes. The classic tetrad of clinical manifestations includes rash, arthralgias, abdominal pain, and renal disease. The disease primarily affects young children, usually under the age of five. It appears to be more severe in older individuals; adults have more severe and frequent renal involvement. However, the overall outcome is good in most patients; studies have noted complete recovery in 94 and 89 percent of children and adults, respectively.

All of the manifestations of active HSP usually resolve spontaneously. However, recurrences are common, occurring in approximately one-third of patients. Recurrent symptoms and signs, which tend to mimic the original episode but are less severe, are normally observed within four months of resolution of the initial symptoms. Recurrences are more likely to occur in patients with nephritis.

The renal prognosis is excellent in most patients, who tend to have focal glomerular involvement and transient hematuria and proteinuria that resolves within several months

Educational objective: To be familiar with the clinical aspects of Henoch-Schönlein purpura.

For more information see the following UpToDate topic review(s) in the RSAP2001 program:
1. Henoch-Schönlein purpura

Answer to Question 10
Answer: C

This patient presents with features suggesting diffuse alveolar hemorrhage (DAH). The presentation is often abrupt in onset with cough, dyspnea, hemoptysis, and fever the predominant findings. The chest radiograph demonstrates diffuse alveolar infiltrates and a normal heart size.

A variety of diseases may be associated with DAH including systemic necrotizing vasculitis (Wegener's granulomatosis, Churg Strauss syndrome, and microscopic polyangiitis), Goodpasture's syndrome, SLE, poststreptococcal infection, and drugs. The vasculitides are associated with a positive ANCA. A positive ANA is seen with SLE, and the anti-GBM antibody is positive in most patients with Goodpasture's syndrome. By comparison, classic polyarteritis nodosa usually spares the lung.

This patient has a positive ANA, but a negative ANCA. Although she lacks extrapulmonary findings, pulmonary hemorrhage may be the presenting finding in some patients with SLE. Thus, choice C is correct.

Educational objective: To recognize the possible presentations of systemic lupus erythematosus.

For more information see the following UpToDate topic review(s) in the RSAP2001 program:
1. The diffuse alveolar hemorrhage syndromes
2. Clinical manifestations and diagnosis of Wegener's granulomatosis and microscopic polyangiitis
3. Churg-Strauss syndrome (allergic granulomatosis and angiitis)
4. Pulmonary manifestations of systemic lupus erythematosus

Answer to Question 11
Answer: C

This patient with RA displays classic features of severe Felty's syndrome, including splenomegaly, leg ulcers, severe granulocytopenia, thrombocytopenia, and recurrent infections. An increase in the dose of methotrexate may be an effective approach to treat the patient's granulocytopenia and recurrent infections.

Recombinant granulopoietic growth factors are being used with increasing frequency to raise the granulocyte count in Felty's syndrome. Both granulocyte macrophage colony stimulating factor (GM-CSF) and granulocyte colony stimulating factor (G-CSF) effectively reverse granulocytopenia and reduce infectious complications in many patients. Therapy for prolonged periods with low doses of these agents may be associated with significant benefits. In general, however, the colony stimulating factors are primarily recommended for short-term use during periods of neutropenia with active infection.

Splenectomy is indicated as a last resort therapy in patients with severe granulocytopenia and recurrent infections in whom aggressive treatment has failed to elevate the white blood cell count and/or reduce the incidence of bacterial infections.

By comparison, the efficacy of tumor necrosis factor inhibitors, such as etanercept, in the treatment of Felty's syndrome is unknown. Moreover, this patient may be at increased risk for the development of sepsis with the use of this agent because of his cutaneous ulcers and buttock abscess. Thus, the use of etanercept is not recommended in this patient.

Educational objective: To recognize signs, symptoms, and treatment of Felty's syndrome.

For more information see the following UpToDate topic review(s) in the RSAP2001 program:
1. Clinical manifestations and diagnosis of Felty's syndrome
2. Drug therapy in Felty's syndrome
3. Indications for splenectomy in Felty's syndrome
4. Anticytokine therapies in rheumatoid arthritis