

## Systemic Lupus Erythematosus: Review Questions

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### QUESTIONS

Choose the single best answer for each question.

1. A 23-year-old woman was diagnosed with systemic lupus erythematosus (SLE) at age 13 years when she presented with polyarticular joint swelling. On a regimen of methotrexate and hydroxychloroquine, her joint pain and swelling have been relatively well controlled. The patient's fingers exhibit both boutonnière and swan-neck deformities, but she is able to make a normal closed fist. What will radiographs of the patient's hands likely reveal?

  - (A) Bone fragmentation
  - (B) Carpal fusion
  - (C) Erosions in the proximal joints
  - (D) Marked subchondral cysts
  - (E) Normal joints
2. A 32-year-old woman with longstanding SLE presents with severe headache. Her blood pressure is 200/110 mm Hg, the cardiac examination is remarkable for an  $S_4$ , and a new heart murmur is noted at the left sternal border. Her medical history includes 3 spontaneous abortions and multiple episodes of pleurisy, arthritis, and thrombocytopenia. Dipstick urinalysis is positive for protein and trace blood. Microscopic examination shows 3 to 5 erythrocytes per high power field and an occasional granular cast. Serum creatinine level is 4.0 mg/dL. What is a renal biopsy likely to show?

  - (A) Diffuse proliferative glomerulonephritis
  - (B) Focal segmental glomerulonephritis
  - (C) IgA deposits
  - (D) Mesangial deposits (seen by electron microscopy only)
  - (E) Thrombotic microangiopathy
3. A 20-year-old man with SLE presents to the emergency department with chest pain that started 10 days ago. He has become increasingly dyspneic over the last 48 hours and has been too exhausted to make meals. He will not lay flat on the examination table and insists on leaning slightly forward. Blood pressure is 94/50 mm Hg and heart rate is 100 bpm. The heart sounds are quiet, and tubular breath sounds are heard at the left scapular border. There is 1+ peripheral edema. Chest radiograph shows a possible cardiomegaly. What is the next step in this patient's management?

  - (A) Arterial blood gas
  - (B) Diuresis with intravenous furosemide
  - (C) Echocardiogram
  - (D) High-dose nonsteroidal medication
  - (E) Infusion of stress dose steroids
4. What is the most specific indicator of Raynaud's phenomenon commonly associated with SLE or a related connective tissue (CT) disease?

  - (A) Alopecia
  - (B) Diffuse myalgias and arthralgias
  - (C) Dilated capillary loops on capillary microscopy
  - (D) History of a first-degree relative with SLE
  - (E) Onset of severe fatigue in the last 2 months
5. A 31-year-old man with a history of lupus nephritis in remission is seen in the office for routine follow-up. Medications include hydroxychloroquine and naproxen. Laboratory tests are performed, and the hemoglobin level is noted to have dropped to 9.0 mg/dL since the last testing 3 months ago. Platelet counts, leukocyte counts, and renal function are normal. The erythrocyte sedimentation rate is 45 mm/h. Other than being more tired than usual, the patient has no complaints. What will the evaluation of anemia most likely show?

  - (A) Cold agglutinins
  - (B) Elevated haptoglobin
  - (C) Negative Coomb's testing
  - (D) Normal total bilirubin
  - (E) Spherocytes on peripheral smear

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## ANSWERS AND EXPLANATIONS

- (E) Normal joints.** The arthritis of SLE is classically described as nondeforming. Unlike rheumatoid arthritis (RA), boutonnière and swan-neck deformities are reducible (ie, the joints can be realigned by manually moving them into the correct position), but the tendons lack the integrity to hold the bones in alignment. Erosions are not seen in SLE arthritis but can be seen in RA, the spondyloarthropathies, and crystal disease. Subchondral cysts are a radiographic feature of degenerative arthritis. Bone fragmentation is seen in both Charcot and pseudo-Charcot joints. Carpal fusion is seen primarily in juvenile RA but also may occur in adult RA.
- (E) Thrombotic microangiopathy.** A renal biopsy is necessary in cases in which clinical features overlap. In this case, given the marked hypertension with clinical manifestations of antiphospholipid-antibody syndrome (APS)—multiple miscarriages, thrombocytopenia, and possibly Libman-Sacks endocarditis—and with the urinary sediment showing no erythrocyte casts, the most likely diagnosis is renal disease due to APS. In APS, pathologic examination can show thrombosis of the glomerular arterioles and capillaries. Unlike lupus nephritis, the lesions are noninflammatory. Mesangial deposition seen only on electron microscopy is associated with class I nephritis. At this level, clinical symptoms are generally absent. Diffuse and segmental glomerulonephritides are commonly associated with erythrocyte casts. IgA deposits are seen in IgA nephropathy and Henoch-Schönlein vasculitis. Patients with renal IgA deposits commonly have gross hematuria.
- (C) Echocardiogram.** Serositis in SLE can involve the pericardial, pleural, or gastrointestinal serosal surfaces. Pericarditis is the most frequent cardiac manifestation of SLE, and presentation can range from small to large effusions that can increase gradually or rapidly. Although tamponade is rare, it must be considered in any patient who presents with symptoms of cardiac compromise. Generally, the most accessible means of evaluation is echocardiogram. Treatment will include steroids, but in this degree of illness, the preferred dose is 1000 mg methylprednisolone daily for 3 days. Nonsteroidal anti-inflammatory drugs are an adjunctive therapy in serosal inflammation but are not sufficient for primary treatment. Diuresis would be dangerous, as cardiac output is dependent on the right ventricular filling pressure. Pulse oximetry would be sufficient to estimate oxygen saturation; an arterial blood gas is not necessary at this point.
- (C) Dilated capillary loops on capillary microscopy.** Raynaud's phenomenon is relatively common as a primary disorder, occurring in approximately 2% of the general population. As only a small percentage of people with Raynaud's have an associated CT disease, it is useful to know the prognostic factors. Dilated capillary loops observed by capillary microscopy and a positive antinuclear antibody test are the most important factors to indicate a possible connection. Younger age at onset also is associated with a higher likelihood of a CT disease. Alopecia, fatigue, and musculoskeletal symptoms are all features of CT disease but are too nonspecific to be helpful diagnostically unless they occur in the context of more specific signs and symptoms. A first-degree relative does increase the possibility of developing a CT disease but not to a degree that is helpful in making a diagnosis.
- (E) Spherocytes on peripheral smear.** The anemia of SLE is warm-antibody mediated, Coombs' positive, and is characterized by extravascular hemolysis. In extravascular hemolysis, as erythrocytes pass through the spleen, antibodies that are attached to the cell membrane are removed. Loss of a portion of the cell membrane results in loss of the normal biconcave shape and evolution to a spherical shape. Associated laboratory findings include hyperbilirubinemia and decreased haptoglobin. Cold agglutinins are associated with mycoplasma infections, lymphoid neoplasms, and paroxysmal cold hemoglobinuria.

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