Hepatic and Splenic Infarction in Systemic Lupus Erythematosus

September 14, 2005 | Pain [1], Cardiovascular Diseases [2], Rheumatoid Arthritis [3]
By Natascha Stone, MD [4]

Systemic lupus erythematosus (SLE) was diagnosed in an 18-year-old man who presented with polyarthritis, fever, hypoxia, fatigue, anemia, neutropenia, and abnormal urinary sediment. A renal biopsy showed diffuse mesangial proliferative glomerulonephritis (World Health Organization class II). Serologic tests were positive for fluorescent antinuclear antibody (FANA), SS-A, SS-B, anti-Sm and anti-dsDNA antibodies, and rheumatoid factor; a direct Coombs' test result was positive as well.

The patient had no history of smoking, alcohol or illicit drug use, or sexually transmitted disease. His mother and maternal aunt both had SLE. Prednisone therapy was initiated. The patient's hypoxia resolved, and renal function test results normalized.

Drs Sherif Nasef, Natascha Stone, and Stanley Kaplan of the University of Tennessee, Memphis, report that 1 month later, the patient returned with complaints of nonproductive cough, malaise, generalized weakness, dyspnea at rest, arthralgia, myalgia, and weight loss. He denied chest pain, palpitations, anorexia, nausea or vomiting, diaphoresis, headache, visual disturbances, rash, oral ulcers, and alopecia.

The patient's temperature was 38.1°C (100.6°F); pulse rate, 166 beats per minute; blood pressure, 80/50 mm Hg; and respiratory rate, 28 breaths per minute. Bitemporal wasting and dry mucosa were noted; his extremities were cyanotic. Breath sounds were decreased over the left lung base. Cardiovascular assessment found tachycardia with hyperdynamic precordium without gallop, murmur, or rub. The abdomen was soft and nontender, with normoactive bowel sounds and no hepatosplenomegaly. Except for the patient's generalized weakness, neurologic findings were unremarkable.

White blood cell count was 3,000/µL (55% neutrophils, 33% bands, 10% lymphocytes, and 2% monocytes); hemoglobin, 11.7 g/dL; platelet count, 177,000/µL; albumin, 1.6 g/dL, total bilirubin, 1.9 mg/dL; and direct bilirubin, 1 mg/dL. Elevated levels were found for alkaline phosphatase (ALP), lactate dehydrogenase (LDH), aspartate transferase (AST), alanine transferase (ALT), amylase, and blood urea nitrogen; creatinine measured 2.8 mg/dL (normal range, 0.5 to 1.2 mg/dL) and lipase was 281 U/L (normal range, 30 to 90 U/L). Prothrombin time was 12 seconds and activated partial thromboplastin time, 28.8 seconds. Blood gas showed pH of 7.54; Pco2, 21 mm Hg; Po2, 93 mm Hg; and O2 saturation, 98%.

FANA titer was 1:320 with speckled pattern; anti-dsDNA antibody index, 2.9; SS-A index, 5.1; SS-B index, 11; anti-Sm index, 10.5 (an index over 1.1 is positive). C3 level was 41 mg/dL (normal range, 65 to 170 mg/dL) and C4 level was 20 mg/dL (normal range, 15 to 45 mg/dL). Urinary protein exceeded 300 mg/dL without cells or casts. A chest film demonstrated bibasilar opacities greater on the left side; heart size was normal.
Ventilation quantitation imaging technique showed matched defects in the left lower lobe. Doppler ultrasonography of the legs revealed no deep venous thrombosis (DVT). Transthoracic echocardiographic examination, which had been normal a month earlier, now showed a small pericardial effusion, left ventricular systolic dysfunction, and a possible apical thrombus. The patient was hydrated with intravenous fluids and fully heparinized for possible pulmonary embolism; prednisone therapy was continued at the same dosage. Over the next few days, liver function test results worsened: AST, ALT, LDH, ALP, and total and direct bilirubin levels increased dramatically. Levels of creatinine rose to 3.6 mg/dL and lipase to 1,556 U/L; p-amylase measured 328 U/L (normal range, 5 to 45 U/L). A hepatitis profile was negative. Abdominal ultrasonography demonstrated bilateral pleural effusions; an enlarged liver with a thickened, nondistended gallbladder with sludge and no calculi; markedly enlarged echogenic kidneys; and a mildly enlarged pancreas with a mild decrease in echogenicity. Total parenteral nutrition through a central venous line was initiated because of acute pancreatitis. Intravenous administration of methylprednisolone also was begun.

Blood cultures grew Staphylococcus aureus, and vancomycin was given. Pancytopenia developed; the patient was transfused with red blood cells. In addition, he received ventilatory assistance after acute respiratory distress syndrome developed. A second abdominal ultrasound scan showed multiple hypoechoic zones, which ranged from 1.9 cm to 6.2 cm in diameter, in the liver and spleen that suggested infarcts, abscesses, metastases, or vasculitis. Abdominal CT scan revealed bilateral pleural effusions, ascites, multiple hypodense areas in the liver and spleen, and multiple distended small bowel loops (A). Ultrasound-guided biopsy of the hypodense liver lesions showed ischemic infarctions; no blood vessels were seen (B). Assessments for anticardiolipin antibodies and lupus anticoagulants were negative; tests for hypercoagulable states detected no abnormalities. Drs Nasef, Stone, and Kaplan note that the few reported cases of hepatic infarction in SLE were associated with antiphospholipid antibodies. In this patient, who had no such antibodies, vasculitis appears to have caused the hepatic and splenic infarcts.

Liver disease in SLE is usually chronic, mild, and not related to a hypercoagulable state. Studies indicate that 25% to 30% of patients with lupus anticoagulant have associated thromboembolic phenomena, including DVT; pulmonary embolism; retinal vein thrombosis; cerebrovascular accidents; myocardial, splenic, or pancreatic infarctions; Budd-Chiari syndrome; and hepatic veno-occlusive disease.

The doctors believe that hepatic and splenic infarction in this patient was caused by arterial occlusion complicated by low blood pressure. Vasculitis and/or intravascular clotting may have accounted for the arterial occlusion. Hypoxia may be attributed to occlusion of pulmonary vessels, and occluded renal vessels can explain the worsening renal function. Similarly, pancreatic vessel occlusion could account for the acute pancreatitis. The patient's good response to high-dose corticosteroid therapy supports the conclusion that blood vessel inflammation was responsible for the arterial occlusions. This patient's health improved slowly. Transesophageal echocardiography showed improved systolic function and failed to verify the apical thrombus; heparin therapy was discontinued. Liver and kidney functions and blood studies gradually recovered. After 3 weeks of hospitalization, the patient was discharged and instructed to take prednisone, 60 mg/d. Several weeks after discharge, a CT scan of the abdomen showed a reduction in the size of the hepatic and splenic lesions.

Source URL:
http://www.rheumatologynetwork.com/printpdf/hepatic-and-splenic-infarction-systemic-lupus-erythematosus/page/0/1

Links: