A 41-year-old man was evaluated for an upper gastrointestinal bleed. He was stabilized and an upper esophagogastroduodenoscopy revealed gastritis and esophageal varices. A computed tomography (CT) scan of the abdomen showed hepatosplenomegaly and moderate ascites. Analysis of the ascitic fluid was consistent with portal hypertension. A liver biopsy demonstrated noncaseating granuloma consistent with sarcoidosis, there was no evidence of liver cirrhosis on pathology. Portal hypertension associated with sarcoidosis is rare; after the first reported case in 1949, 35 other cases have been reported in the English literature, with only 16 patients presenting with portal hypertension without evidence of liver cirrhosis. Our patient is among the small group reported to have sarcoidosis-related portal hypertension without evidence of liver cirrhosis. The present case illustrates the importance of recognizing an uncommon manifestation of sarcoidosis.

CASE PRESENTATION

A 41-year-old man presented to the emergency department complaining of palpitations and dizziness with exertion for one month. He reported passing dark stools for three weeks and noted decreased appetite. He denied chest pain, cough, dyspnea, orthopnea, paroxysmal nocturnal dyspnea, abdominal pain, nausea, vomiting, diarrhea or weight loss. His medical history included sarcoidosis and steroid-induced diabetes mellitus while on prednisone, glipizide and insulin. The patient migrated from Nigeria 20 years earlier, and denied toxic exposures, use of alcohol, nicotine or recreational drugs.

On examination, he was afebrile with a blood pressure of 121/61 mmHg, a pulse rate of 93 beats/min, a respiratory rate of 16 breaths/min and oxygen saturation of 98% on room air. He was anicteric with pale conjunctiva. Respiratory, cardiovascular and neurological examinations were normal. His abdomen was soft, with mild epigastric tenderness but no guarding or rebound tenderness. His liver span was 14 cm and his spleen extended 4 cm below the left costal margin. No masses were palpated and bowel sounds were normal. The rectal examination revealed dark stools.

Laboratory studies revealed a white blood cell count of 1.6×10^9/L, a hemoglobin level and hematocrit of 53 g/L and 16.7%, respectively, with a mean corpuscular volume of 68 fL, and a platelet count of 6.4×10^9/L. Electrolytes and coagulation studies were normal, with alanine aminotransferase, aspartate aminotransferase and alkaline phosphatase levels of 37 U/L, 27 U/L and 127 U/L, respectively. He was admitted for symptomatic anemia due to gastrointestinal blood loss and was transfused three units of packed red blood cells. Esophagogastroduodenoscopy revealed gastritis and grade 2, moderately sized esophageal varices that were not actively bleeding. A CT scan of the abdomen showed hepatosplenomegaly, ascites and thickening of the colonic mucosa with a patent portal system (Figure 1). Colonoscopic biopsy of the colonic mucosa was normal. An HIV 1/2 ELISA test was negative and hepatitis serology revealed previous exposure to hepatitis A and B viruses with no evidence of chronic infection. Work-up for collagen vascular diseases was negative, and his angiotensin converting enzyme level was within normal limits. Paracentesis revealed straw-coloured ascitic fluid with a serum ascitic albumin gradient of 1.8, consistent with portal hypertension. Liver biopsy showed no evidence of cirrhosis but demonstrated noncaseating granulomas consistent with sarcoidosis (Figure 2). He was started on prophylactic propranolol for recurrence of variceal bleeding. Furosemide and spironolactone were added for management of ascites. The patient declined splenectomy.

DISCUSSION

Sarcoidosis is a systemic disorder of unknown etiology characterized by noncaseating granulomas with a propensity for several body systems. Lung involvement is most common but liver involvement occurs in approximately 70% of cases. Patients with hepatic sarcoidosis are typically asymptomatic with normal liver enzyme levels. Hepatic granulomas are found on CT imaging in <5% of patients and are typically...
between 0.5 cm to 0.8 cm in size. Clinical manifestations identified include jaundice, chronic cholestasis, portal hypertension, Budd-Chiari syndrome, and intrahepatic cholestasis resembling primary biliary or sclerosing cholangitis (1).

Portal hypertension is defined by a portal pressure gradient >11 mmHg or the presence of esophageal varices, first reported in association with sarcoidosis by Mino et al (2) in 1949. Between 1949 and 2001, an additional 35 cases were reported in the English literature (16 had portal hypertension without evidence of cirrhosis). Several mechanisms have been postulated to explain the pathophysiology of portal hypertension related to sarcoidosis. Maddrey et al (3) proposed that arterial-venous shunts within granulomas in the liver and spleen cause elevated portal blood flow resulting in a compensatory increase in intrahepatic resistance. The resistance in the intrahepatic sinusoids may also increase due to obstruction from confluent sarcoid granulomas. Another proposed mechanism is that presinusoidal obstruction by granulomas in the portal vein cause an increase in pressure and restrict flow (4,5). A third theory is that granulomatous phlebitis in portal and hepatic veins leads to cirrhosis and focal fibrosis, which then increases pre- and postsinusoidal resistance (6).

The present patient is among a small group reported to have sarcoidosis-related portal hypertension. His portal hypertension resulted in variceal bleeding that presented as symptomatic anemia. Hypersplenism was likely a consequence of his portal hypertension and may have been responsible for his leukopenia and thrombocytopenia. Our patient's angiotensin-converting enzyme level remained normal throughout his hospital course. This is not unusual because angiotensin-converting enzyme levels are elevated in only 60% to 70% of patients with sarcoidosis and do not correlate well with disease activity. In patients with portal hypertension, splenectomy or insertion of a portocaval shunt has been shown to reduce portal pressures and offer symptomatic relief. Steroids are of no benefit in sarcoidosis-induced portal hypertension.

The present case illustrates the importance of recognizing an uncommon presentation of sarcoidosis.

REFERENCES
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