Hepatosplenic sarcoidosis

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ABSTRACT

Sarcoidosis is a systemic disorder characterized by development of granulomas within various organs in the body. It commonly affects people of African American and Scandinavian heritage between the ages of 20 and 40. The lungs are involved in 90% of patients. We report a case of sarcoidosis of the spleen and liver with minimal pulmonary involvement.

KEYWORDS Abdominal pain; elevated liver function tests; extrapulmonary sarcoidosis; hepatosplenic sarcoidosis; pruritus

Sarcoidosis is a multisystem inflammatory disease characterized by noncaseating granulomas.¹ It affects approximately 34 in 100,000 African Americans. Approximately 90% of primary cases of sarcoidosis present in the pulmonary system, with only 10% of cases presenting with primary extrapulmonary involvement.^{2,3} We report a case of sarcoidosis of the spleen and liver without pulmonary involvement, presenting as chronic abdominal pain and pruritus with elevated liver function tests.

CASE REPORT

A 34-year-old black man presented for diffuse abdominal pain and pruritus of 6 months' duration. He had diabetes mellitus type 1, essential hypertension, and a seizure disorder. His medications included amitriptyline, insulin lispro, insulin glargine, and lamotrigine. His vital signs were within normal limits. Physical exam revealed mild hepatosplenomegaly and signs of excoriation over multiple areas of the skin. Initial workup revealed elevated liver function tests (*Table 1*). He had a normal coagulation profile (prothrombin time 13.6 s, international normalized ratio 1.2). Hepatitis and HIV screening was negative, as was testing for cytoplasmic anti-neutrophil cytoplasmic antibodies, perinuclear anti-neutrophil cytoplasmic antibodies, and anti-nuclear, anti-mitochondrial, and anti-smooth muscle antibodies. Magnetic resonance imaging (MRI) of the abdomen showed hepatomegaly, splenomegaly, and diffuse punctate hypoattenuating nodules throughout both the liver and spleen

with reactive lymph nodes in the retroperitoneum (*Figure 1a*). Chest computed tomography (CT) showed enlarged mediastinal lymph nodes (*Figure 1b*) without any evidence of interstitial lung disease or parenchymal involvement (*Figure 1c*).

Esophagogastroduodenoscopy and a colonoscopy with random biopsies were performed. Macroscopically, gastroduodenal and colonic mucosa were normal and no varices or ulcers were noticed; histopathology findings were not significant. An endoscopic retrograde cholangiogram showed a normal bile duct with no biliary dilation, strictures, or obstructing stones or masses and no obvious lesions in the

Table 1. Patient laboratory values at baseline and after 6 months of prednisone

Variable	Baseline	After 6 months of prednisone
Alkaline phosphatase (IU/L)	1178	759
Aspartate aminotransferase (IU/L)	241	69
Alanine aminotransferase (IU/L)	269	79
Gamma-glutamyl transferase (IU/L)	1410	577
Total bilirubin (mg/dL)	2.1	0.9
Direct bilirubin (mg/dL)	1.3	0.3
Angiotensin-converting enzyme (U/L)	267	Not performed

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Received December 9, 2019; Revised February 20, 2020; Accepted February 27, 2020.

Figure 1. Patient imaging. (a) Axial postcontrast T1 fat-saturated MRI shows diffuse heterogeneous enhancement of the liver and spleen due to multiple granulomas. (b) Coronal contrast-enhanced CT image through the chest shows enlarged mediastinal (paratracheal and subcarinal) lymph nodes (arrows). Also seen are liver and spleen changes. (c) Axial CT of the chest in the lung window setting shows no parenchymal or interstitial changes.

Figure 2. Multiple noncaseating granulomas (arrowheads) within the spleen. Hematoxylin and eosin, 20× magnification.

liver. Endoscopic ultrasound showed hypoechoic lesions in the spleen. When biopsied, these lesions showed multiple nonnecrotizing granulomas (Figure 2). Gram stains and other special stains were negative. Cultures for acid-fast bacilli, anaerobic organisms, and fungal organisms were negative. Given these findings, the patient was diagnosed with hepatosplenic sarcoidosis with minimal pulmonary involvement. He was started on oral prednisone. Gradually his abdominal pain and pruritus improved and his liver enzyme levels decreased (Table 1).

DISCUSSION

Sarcoidosis can affect different organs and tissues, but more than 90% of patients have pulmonary¹⁻⁴ and mediastinal lymph node involvement at onset, while isolated extrapulmonary localization is rare (<10%).⁵⁻⁹ Although the liver is the third most affected organ, most patients are symptomatic and have mild nonspecific symptoms such as nausea, vomiting, weight loss, and abdominal discomfort.⁶⁻⁹ Laboratory findings are nonspecific as well. Serum markers such as angiotensin-converting enzyme, adenosine deaminase, and soluble interleukin-2 may be helpful in diagnosis.¹⁰ The diagnosis of sarcoidosis depends on clinical and

radiologic findings, histopathologic evidence of noncaseating granulomas, and exclusion of infectious or malignancy-related etiologies.¹¹

The differential diagnosis of multiple hepatic and splenic lesions includes metastases, lymphoma, and fungal infections. Clinicians should be aware of hepatosplenic sarcoidosis as a potential cause for abdominal pain, pruritus, and elevated liver function tests in adults.

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