Case of isolated hepatic sarcoidosis

Dear Editor,

This letter is in regard to the article "Atypical sarcoidosis: case reports and review of the literature" published in European Review for Medical and Pharmacological Sciences by Giovinale et al.1 We share our experience managing a patient with hepatic sarcoidosis.

A 35-year-old African-American male was admitted to the psychiatry ward due to suicidal ideation. He also complained of epigastric pain, associated with nausea, vomiting and weight loss. During admission it was noted that he had abnormal liver enzymes: alanine transaminase 119 U/L, aspartate transaminase 146 U/L and alkaline phosphatase 270 U/L. Laboratory tests for ceruloplasmin, alpha antitrypsin and calcium were within the normal range. Serological tests for hepatitis A, B and C virus were negative. Autoantibody screens, including antinuclear antibody, antimitochondrial antibody and smooth-muscle antibody, were also within the normal range. A computed tomography scan of the chest and magnetic resonance imaging of the abdomen were not suggestive of lymphoma or any solid mass. A liver biopsy showed chronic hepatitis with moderate activity, mild portal fibrosis and small non-caseating granulomas (Figure 1). Special stains were negative for both acid fast and fungal organisms. A rhodanine stain did not show evidence of copper accumulation making a chronic cholestatic process, such as primary biliary cirrhosis, less likely. His electrocardiogram, Mantoux test and chest x-ray were normal. The liver biopsy findings and the increased activity of the serum angiotensin converting enzyme (92 U/L) indicated hepatic sarcoidosis. As our patient had only minimal complaints and his liver enzymes were not significantly abnormal, we chose not to treat him with steroids. His liver functions have improved, and the patient is doing well after two years of follow up.

Sarcoidosis is a multisystem granulomatous inflammatory disease characterized by the accumulation of mononuclear inflammatory cells followed by the formation of non-caseating epithelioid granulomas at the site of involvement. The diagnosis of hepatic sarcoidosis is suggested by liver biopsy. However, liver biopsy alone is not sufficient to establish the diagnosis. Ruling out infections, particularly tuberculosis, and an elevated serum angiotensin converting enzyme level are also important steps in diagnosis. Most patients are asymptomatic and have only biochemical abnormalities, especially elevated alkaline phosphatase. Rarely do patients have cirrhosis, cholestatic liver disease and hepatic vein thrombosis.

The treatment of hepatic sarcoidosis depends on the clinical manifestations of the disease. As the majority of patients are asymptomatic and the only manifestation is non-caseating granulomas, no treatment is needed when there is no evidence of signs and symptoms of liver dysfunction. Whether corticosteroids can prevent hepatic disease progression or the development of complications is unproven and thus not recommended for hepatic disease alone.2 A trial of corticosteroid therapy may be of benefit in patients with constitutional symptoms, bile ductal involvement or portal hypertension by sarcoidosis.2-3. When a patient develops hepatic sarcoidosis, with portal hypertension, splenectomy with or without portacaval shunts should be considered to reduce the risk of varices bleeding and to prolong survival.

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Figure 1. High magnification view of the granuloma composed of lymphocytes and epithelioid cells. No evidence of necrosis is seen.

References

