

Introduction

Primary biliary cholangitis (PBC) is a disease characterized by immune-mediated destruction of intrahepatic bile ducts resulting in cholestatic liver disease. Sarcoidosis is a multisystem, chronic granulomatous disease of unknown etiology with hepatic involvement seen in majority of the patients. Very rarely, hepatic sarcoidosis can present as diffuse intrahepatic biliary inflammation, destruction and cholestatic liver disease, thereby resembling PBC. We present a rare case of hepatic sarcoidosis with PBC and non-alcoholic fatty liver disease (NAFLD).

Case Description

A 47 year old female with history of asymptomatic pulmonary sarcoidosis, type 2 diabetes mellitus, morbid obesity underwent liver biopsy for evaluation of fatty liver and abnormal liver enzymes (AST 41 U/L, ALT 55 U/L, alkaline phosphatase 238 U/L, Total bilirubin 0.5 mg/dL). Biopsy showed steatohepatitis with non-necrotizing granulomas consistent with sarcoidosis. Patient was asymptomatic and not on treatment for sarcoidosis or any other hepatotoxic medications. Social history was pertinent for no significant alcohol intake. Patient continued to have persistently abnormal liver enzymes and was further evaluated with a comprehensive chronic liver disease work up. This came back positive for Anti-mitochondrial antibody (AMA), raising suspicion for underlying PBC. Patient underwent repeat liver biopsy which showed similar findings of steatohepatitis (Figure 1A) and non-necrotizing granulomas (Figure 1B) along with focally identified biliary inflammation (Figure 1B) suggestive of underlying chronic biliary disease such as PBC given positive serologic testing for AMA. Iron and copper stains were negative. Patient was subsequently started on therapy with Ursodiol.

Histopathology

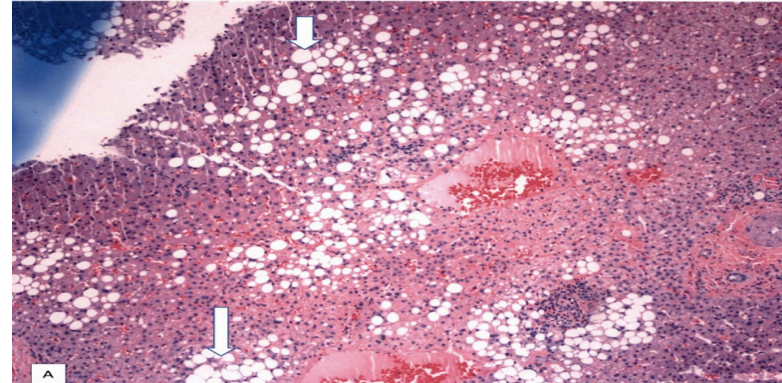


Figure 1A: Moderate large-droplet steatosis (white arrow) with mild to moderate lobular inflammation is seen.

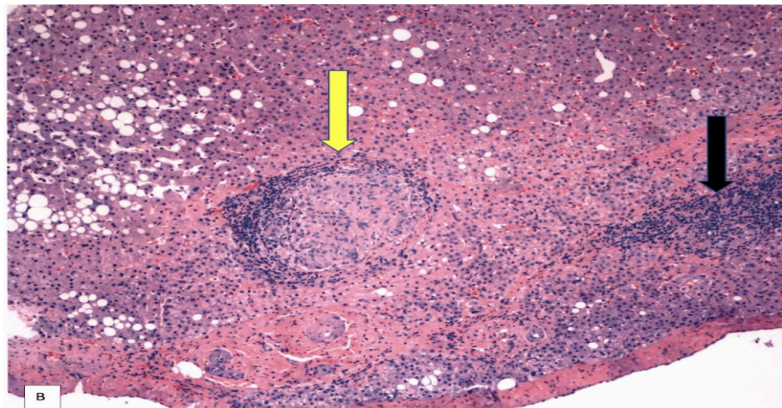


Figure 1B: Scattered non-necrotizing granulomas (yellow arrow) with focal portal inflammation (black arrow) is seen.

Discussion

Hepatic sarcoidosis most commonly manifests as non-necrotizing granulomas as seen on liver biopsy. However, in this case the liver biopsy showed evidence of three pathological processes- PBC, sarcoidosis and NAFLD. Patients with hepatic sarcoidosis very rarely develop an underlying co-existing cholestatic liver disease. The postulated underlying mechanism of chronic cholestasis in sarcoidosis involves confluent granulomas causing granulomatous cholangitis and ductopenia. It is prudent to identify this co-existing pathological process in order to initiate appropriate treatment and prevent further hepatic damage. Therefore, chronic cholestasis in hepatic sarcoidosis should prompt extensive evaluation for an alternative cause of chronic liver disease.

References

1. Alempijević T, Sokić-Milutinović A. *et al.* Primary biliary cirrhosis and hepatic sarcoidosis--a case report. *Vojnosanit Pregl.* 2014 Jan.
2. Keeffe EB. Sarcoidosis and primary biliary cirrhosis. Literature review and illustrative case. *Am J Med.* 1987 Nov.
3. Akshatha Manohar *et al.*, An Overlap of Sarcoidosis and PBC. *Journal of Clinical and Diagnostic Research.* 2021 Jul.