

Isolated Hepatic Sarcoidosis Presenting As Non-cirrhotic Portal Hypertension

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Isolated Hepatic Sarcoidosis Presenting As Non-cirrhotic Portal Hypertension

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Introduction

Sarcoidosis is a systemic inflammatory disease characterized by the formation of noncaseating granulomas. Most commonly it manifests as diseases of the pulmonary system, although it can involve any organ in the body. Hepatic involvement is well known and can present as abdominal pain, pruritus, hepatomegaly, and jaundice. Rarely does it progress to portal hypertension or cirrhosis. Hepatic sarcoidosis in the absence of pulmonary involvement is seen in less than 10% of cases. Here we highlight a case of isolated hepatic sarcoidosis discovered incidentally presenting as portal hypertension.

Case

A 47-year-old Caucasian male with past medical history of type 2 diabetes mellitus, essential hypertension, and alcohol abuse presented to the Emergency Department after a motor vehicle accident. Trauma workup including CT scan of the chest, abdomen, and pelvis incidentally showed scattered small pulmonary nodules, fatty liver, and splenomegaly with splenic varices. Subsequent

outpatient MRI showed hepatosplenomegaly and portal hypertension. The patient was referred to gastroenterology for further evaluation. Abdominal ultrasound with doppler showed patent hepatic vessels and normal bile ducts. Initially, it was thought that the patient's liver findings were secondary to fatty liver disease given risk factors of metabolic syndrome and daily alcohol use of 6-8 beers per day. Labs were significant for thrombocytopenia with a platelet count of 129 thou/cmm, normal albumin of 3.6 g/dL, normal liver function panel including total bilirubin 1.0 mg/dL, AST 22 U/L, ALT 29 U/L, and AP 108 U/L, and normal INR of 1.1. Serologic testing for chronic liver disease was negative except for an elevated smooth muscle antibody 1:160 and serum immunoglobulin G 1827 g/L which were suggestive of autoimmune hepatitis. An EGD showed small esophageal varices and isolated gastric varices in the fundus. He ultimately underwent a percutaneous liver biopsy that showed noncaseating granulomas without significant fibrosis. There was no steatosis or features of autoimmune hepatitis. Stains for

acid-fast bacilli and fungi were also negative. The patient was diagnosed with hepatic sarcoidosis presenting as non-cirrhotic portal hypertension. Other organ involvement was not found. He was started on nadolol for prophylaxis of gastroesophageal bleeding. Treatment for sarcoidosis was not initiated given the lack of clinical symptoms and normal liver enzymes.

Discussion

Our case demonstrates the importance of considering hepatic sarcoidosis as a cause of portal hypertension. Liver involvement is common in sarcoidosis, up to 70% of cases, but significant complications including cirrhosis and portal hypertension are rare, about 3%. Non-cirrhotic portal hypertension as seen in our case is pre-sinusoidal and thought to be secondary to granuloma compression of the portal vein. Treatment is reserved for those with clinical symptoms with first line therapy being glucocorticoids. Alternatives include ursodeoxycholic acid, immunosuppressants, such as methotrexate and chlorambucil, and rarely liver transplant.

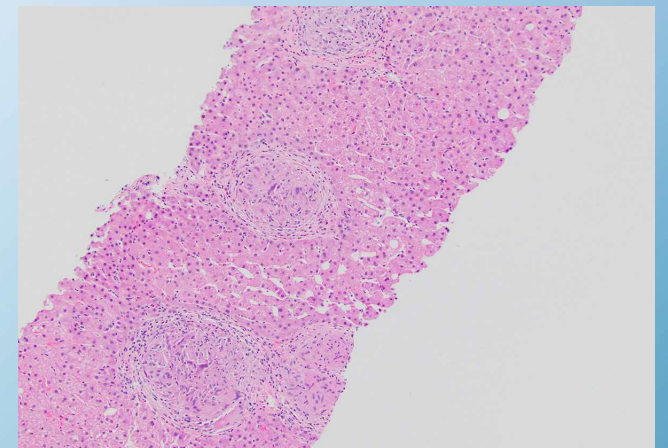


Image 1. Histological slide showing liver specimen overview (100x).

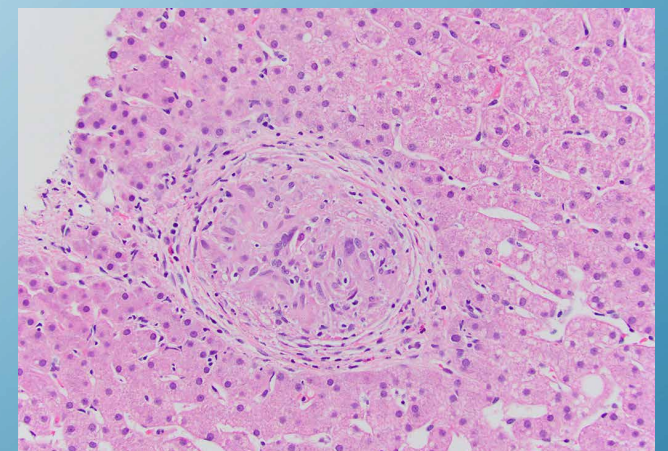


Image 2. Histological slide showing liver granuloma (200x).