Hepatic Sarcoidosis Presenting as Decompensated Liver Cirrhosis

Bishal Paudel, MD; Emily S. Edwards, MD, MS; Eric S. Sobel, MD, PhD; Amy J. Sheer, MD, MPH
University of Florida College of Medicine, Gainesville, FL

Learning Objectives

- Demonstrate clinical findings of hepatic sarcoidosis and recognize it as one of the rare causes of liver dysfunction

Background

- Sarcoidosis is a disorder characterized by the formation of non-caseating granulomas in multiple organ systems.
- While lungs are the most common site of disease activity, hepatic involvement can be seen in many patients.
- Most patients with sarcoid involvement of the liver are asymptomatic; however, some can present with symptoms of liver injury.[1]

Case Description

- 36-year-old female with past medical history of recently diagnosed cryptogenic liver cirrhosis presented to the emergency department with complaints of generalized abdominal pain and left flank pain.
- Patient was hemodynamically stable on presentation. She was found to be hypercalcemic at 13 mg/dl. Pertinent labs from admission are summarized in Table 1. MELD-Na score was 11. Her physical exam was notable for hepatomegaly, abdominal distension and right upper quadrant tenderness.
- Eight months prior to presentation, the patient was admitted to an outside hospital due to hematemesis. Esophagogastroduodenoscopy with variceal banding was performed, and she was diagnosed with cryptogenic liver cirrhosis. CT Abdomen/Pelvis performed during the hospitalization demonstrated marked heterogeneity of the liver.
- Follow-up MRI performed outpatient showed mild hepatosplenomegaly and “diffuse small areas of altered signal intensity throughout the liver suspicious for diffuse hepatic metastatic disease” (Figure A).
- Upon admission to our service, her hypercalcemia was treated with IV fluids. Infectious workup including hepatitis panel came back negative. Autoimmune labs including antinuclear antibody, antimitochondrial antibody, anti-smooth muscle antibody and anti-liver-kidney microsome antibody were also negative.

Pertinent Labs	Result	Reference Range
Calcium	13 mg/dl	8.4-10.2 mg/dl
Ionized Calcium	1.62 mmol/l	1-1.5 mmol/l
Parathyroid Hormone	8 pg/ml	12-88 pg/ml
1,25-Dihydroxyvitamin D	96.4 pg/ml	19.9-79.3 pg/ml
Alkaline Phosphatase	192 IU/L	33-133 IU/L
Aspartate Aminotransferase	51 IU/L	0-37 IU/L
Alanine Aminotransferase	47 IU/L	0-35 IU/L
Sodium	131 mmol/l	136-145 mmol/l
International Normalized Ratio	1.4	0.8-1.1

Table 1: Pertinent labs from the day of admission

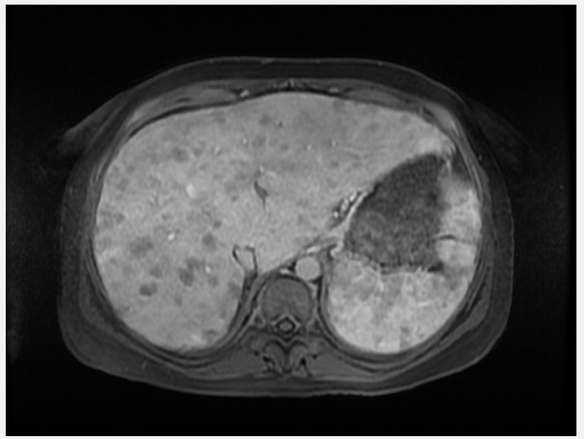

Figure A: Hypointense liver lesions on MRI, initially presumed suspicious for diffuse hepatic metastatic disease, later confirmed to be sarcoid lesions

- Tumor markers including CA 19-9, CA 27-29 and CA 125 were normal.
- Liver biopsy performed showed hepatic parenchyma with numerous non-necrotizing granulomas and associated fibrosis. Serum soluble interleukin-2 receptor level was elevated at 2916.1 pg/mL (175.3-858.2 pg/ml).
- Patient was then started on 20 mg prednisone daily for treatment of hepatic sarcoidosis.

Discussion

- Hepatic sarcoidosis can have a wide spectrum of clinical presentation, ranging from incidental finding to end-stage liver disease. It is very important to keep hepatic sarcoidosis on the differential when a patient is diagnosed with a new liver disease.
- The presentation of hepatic sarcoidosis can be puzzling and may resemble many other disease processes including malignancy, infection and autoimmune liver diseases. Since definite diagnostic criteria have not yet been formalized for hepatic sarcoidosis, it can often be challenging for providers to make a prompt diagnosis resulting in patient distress and clinical decompensation.
- A detailed clinical history along with pertinent labs/imaging is warranted if hepatic sarcoidosis is suspected. Liver biopsy is usually required to confirm the diagnosis.[1] In our case presentation, hypercalcemia, elevated serum soluble interleukin-2 receptor and liver biopsy finding of non-necrotizing granulomas helped establish the diagnosis of hepatic sarcoidosis.
- In a patient diagnosed with hepatic sarcoidosis, a thorough workup ruling out other possible liver disease is also critical as hepatic involvement of sarcoidosis may not exclude other underlying pathology.
- For patients with hepatic sarcoidosis, assessing symptoms of liver involvement and biochemical evidence of cholestasis is crucial. While observation is indicated for asymptomatic liver disease, corticosteroids and/or ursodeoxycholic acid are the first-line agents for symptomatic disease. Duration of treatment depends on treatment response.[1,2,3]
- Different immunosuppressive agents can be used as second-line treatment options. However, risk-to-benefit profile should be strongly considered before starting these agents. Severe cases of hepatic sarcoidosis require liver transplantation.[1,2,]

References

1. Tadros, Micheal et al. “Hepatic Sarcoidosis.” *Journal of clinical and translational hepatology* vol. 1,2 (2013): 87-93. doi:10.14218/JCTH.2013.00016
2. Shah, Neil, and Arnab Mitra. “Gastrointestinal and Hepatic Sarcoidosis: A Review Article.” *Clinical liver disease* vol. 17,4 301-307. 1 May. 2021, doi:10.1002/cld.1055
3. Sedki, Mai et al. “Hepatic Sarcoidosis: Natural History and Management Implications.” *Frontiers in medicine* vol. 6 232. 30 Oct. 2019, doi:10.3389/fmed.2019.00232