Abdominal sarcoidosis including hepatic and portal lymphadenopathy as a primary manifestation in a case of systemic sarcoidosis

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Introduction

- Sarcoidosis is a rare systemic inflammatory disease characterized by non-caseating granulomas.
- Most patients are asymptomatic but hepatic sarcoidosis can progress to portal hypertension which can be cirrhotic or non-cirrhotic. Definitive diagnosis is made by liver biopsy. [1]
- This is a case of hepatic sarcoidosis presenting with elevation in alkaline phosphatase (ALP) which improved with treatment.

Figure 1. Ultrasound of the abdomen showing hepatic steatosis.

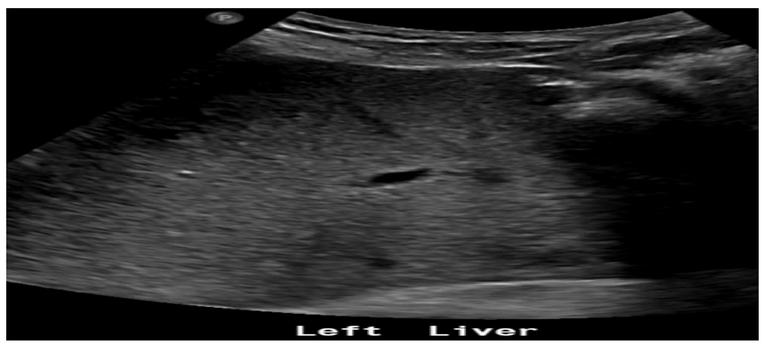
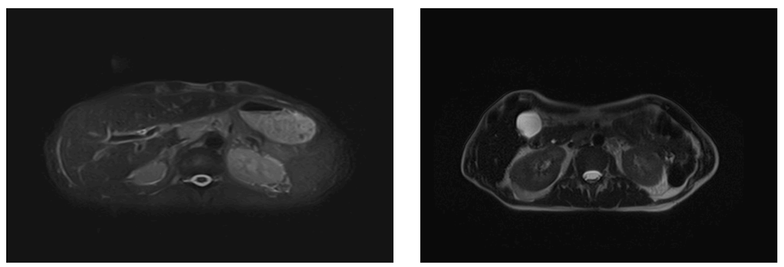


Figure 2. Magnetic resonance cholangiopancreatography showing periportal lymphadenopathy.



Case

- A 50 year-old man presented with three months of worsening dyspnea on exertion and dysphagia to liquids and solids. On arrival, the patient was tachycardic, tachypneic and hypoxemic. Laboratory values during hospital stay are shown in Table 1.
- Computed tomography of the chest showed a small apical pneumothorax and reticular opacities with bronchiectasis. Figured 1 and 2 depict findings of ultrasound and MRCP respectively. Work-up for fungal, autoimmune and infectious etiologies was negative except for an anti-mitochondrial antibody of 29 units and anti-smooth muscle antibody of 30 units.
- Liver biopsy revealed granulomatous portal and lobular inflammation, portal lymphocytic inflammation with ductitis, focal interface hepatitis and focal bridging fibrosis (Figure 3 and 4). Lung biopsy showed non-necrotizing granulomas that initially was thought to be due to atypical mycobacteria and not sarcoidosis. The patient was not immediately started on steroids thereafter. Ursodiol was initiated to help with cholestasis. Biopsy of portal lymph nodes confirmed non-caseating granulomas.
- The patient was then diagnosed with systemic sarcoidosis with hepatic and pulmonary involvement. He was started on steroids in the outpatient setting in conjunction with ursodiol. Liver enzymes subsequently improved at the end of hospital stay as shown in Table 1.

Table1. Progression of Liver function tests during course of illness.

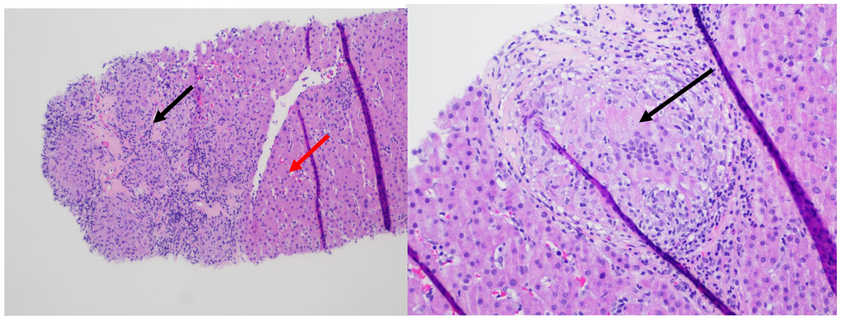
Lab value	Day 1	Day 15	Day 30
Alkaline phosphatase	458	232	188
Alanine transaminase	113	40	59
Aspartate transaminase	110	56	82
Total bilirubin	0.6	0.6	0.7
INR	1	1	1

Discussion

- Sarcoidosis should be among one of the differentials in patients presenting with predominant ALP elevation in the absence of bilirubin elevation and concomitant lung manifestations.
- A liver biopsy is essential for diagnosing hepatic sarcoidosis and to rule out other similar pathologies like primary biliary cholangitis and primary sclerosing cholangitis.[2]
- Ursodiol in conjunction with steroids can help improve liver enzyme elevations seen in hepatic sarcoidosis.[1,2]

Figure 3. H &E stained images of a large lobular focus of non-caseating granulomatous inflammation with interdigitating hyaline fibrosis in 200x magnification (black arrow). Hepatic parenchyma without steatosis and a small focus of lymphocytic inflammation (red arrow)

Figure 4. H&E stain of portal tract expanded by a poorly formed granuloma with a multinucleated giant cell in 400x magnification (black arrow). The remaining portal tract shows a lymphohistiocytic infiltrate with infiltration and damage of the bile duct by lymphocytes.



References

1. Ryland K. L. (2020). Hepatic Sarcoidosis: Incidence, Monitoring, and Treatment. *Clinical liver disease*, *16*(5), 208–211. https://doi.org/10.1002/cld.1002
2. Tadros M, Forouhar F, Wu GY. Hepatic Sarcoidosis. *J Clin Transl Hepatol*. 2013 Dec;1(2):87-93. doi: 10.14218/JCTH.2013.00016. Epub 2013 Dec 15. PMID: 26357609; PMCID: PMC4521279.