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Unraveling Extrapulmonary Sarcoidosis in a Patient With Unexplained Cholestatic Liver Injury

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Introduction

- Sarcoidosis is a rare autoimmune condition of unknown etiology characterized by non-caseating granulomas most commonly the lungs and hilar lymph nodes.
- We report a case of sarcoidosis that presented as hepatomegaly and recurrent cholestatic liver disease.

Case Summary

- 46-year-old F with ESRD, Type 2 DM, latent TB and daily consumption of herbal teas such as Manzanilla and RUDA leaves daily presenting with RUQ pain, pulsatile abdomen, jaundice, and jugular venous distention.
- During each (of 3) admissions, she was found to have significantly elevated ALP to a peak of 2003 U/L and Tbili peak of 37.4 mg/dL, with ALT & AST to 40 U/L and 60 U/L respectively. Viral panels and tumor negative.
- On her second admission, she had rapid clinical improvement after initiating steroids for Bell's palsy, otherwise, on each other admission she was given ursodeoxycholic acid which led to transient improvement.
- Initial U/S abdomen, MRCP and CT liver triphasic showed hepatomegaly, otherwise unremarkable.
- Two liver biopsies revealed sinusoidal congestion and dilation, hepatocellular and sinusoidal fibrosis consistent with chronic venous outflow obstruction and few non-necrotizing granulomas.
- TTE found to be normal twice however the third TTE showed mild tricuspid and pulmonary insufficiency and borderline pulmonary artery systolic pressure. Right heart catheterization showed borderline mild pulmonary hypertension and hemodynamics not suggestive of a cardiac etiology.
- In a last effort to attempt to locate tissue for biopsy, a CT C/A/P showed an area of enhancement at the junction of segment 5 and 6 of the liver that is nonspecific but could relate to hepatic sarcoid.
- She was started on Prednisone 40mg for presumed hepatic sarcoid and Rifampin 600mg for latent TB.
- She will subsequently undergo bronchoscopy w/ BAL and biopsy as well as renal biopsy to further establish a unifying diagnosis.

Discussion

- This patient had several potential etiologies for liver injury including untreated TB and daily consumption of herbal teas.
- However, after an extensive workup that was all found to be negative, infiltrative disease, specifically sarcoidosis, became the top differential given non-caseating granulomas on liver biopsy, facial nerve palsy, peripheral neuropathy, renal disease, mild pulmonary hypertension and CT findings.
- Lack of pulmonary involvement is an uncommon presentation in sarcoidosis, occurring in 5-9% of cases overall¹.
- Non-pulmonary sarcoidosis has been considered an under-recognized entity wherein the time to diagnosis is delayed compared to pulmonary sarcoidosis with extra-pulmonary involvement.
- Hepatic sarcoidosis occurs in 11-80% of cases and is mostly asymptomatic¹.
- Only 5-30% of patients present with symptoms of jaundice, nausea, vomiting, abdominal pain, and hepatosplenomegaly¹.

References

1. Ryland, K. L. (2020). Hepatic sarcoidosis: Incidence, monitoring, and treatment. *Clinical Liver Disease*, 16(5), 208