A Case of Seronegative Autoimmune Hepatitis During Pregnancy

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Introduction

Autoimmune hepatitis (AIH) is a rare disease that usually affects women at reproductive age and is associated with an increased risk of adverse maternal and fetal outcomes. Diagnosis of AIH during pregnancy, particularly in absence of autoantibodies, and considering the need to exclude unique liver diseases in pregnancy, poses a challenge for clinicians. We present the case of a patient with new-onset seronegative AIH during pregnancy.

Case Report

A 29-year-old Hispanic female, currently G2P101 at 19 weeks and 5 days presents to ED complaining of yellowing of her skin and eyes for 1 week, associated with nocturnal pruritus. She denies any nausea or vomiting for the last 3 weeks, abdominal pain, diarrhea, constipation, alcohol use, sick contacts or recent travel. She had a history or pre-term delivery at 32 weeks, 6 years prior. Physical exam was significant for generalized jaundice, scleral icterus and mild conjunctival pallor. Laboratory investigations showed normocytic anemia, significantly elevated AST (452 U/L), ALT (165 U/L), alkaline phosphatase (182 U/L), hyperbilirubinemia (total bilirubin 24mg/dL, direct bilirubin of >15mg/dL), elevated bile acids (total bile acid 270.6 ummol/L, cholic acid 87.7 ummol/L, chenodeoxycolic acid 182.9 ummol/L) and elevated INR (1.4). Hepatitis panel and autoimmune work up, which was repeated twice, were negative. Abdominal ultrasound was unremarkable. MRCP showed nonspecific hepatic parenchyma heterogeneity on T2 signal suggestive of infiltrative process.

A diagnosis of intrahepatic cholestasis of pregnancy (IHCP) with atypical presentation was made, and patient was started on ursodeoxycholic acid, which improved her symptoms. At day 15 of hospital course, she developed lower abdominal and vaginal bleeding, caused by placental abruption. The fetus was deemed non-viable fetus and an emergent C-section was performed. Patient was eventually discharged with outpatient GI follow up. At clinic follow up, recurrent rise in liver function test was noted, and repeat autoimmune work up was significant for elevated IgG (3600). Liver biopsy was performed, which showed active chronic inflammation with lymphoplasmacytic infiltrates, eosinophils, and neutrophils, moderate interface hepatitis with ballooning degeneration and emperipolesis and moderate portal fibrosis with bridging fibrosis, suggestive of autoimmune hepatitis. Patient was subsequently started on steroid therapy, with appropriate response as symptoms progressively resolved and liver function tests normalized.

Discussion

AIH is a rare condition that can manifest with antepartum and postpartum flares, having negative consequences to fetal and maternal outcomes, hence the importance of controlling AIH activity prior to and throughout pregnancy, and in the early postpartum period. Liver biopsy should be considered in cases of antibody negative AIH, however there is paucity of data regarding its safety in pregnancy. Our patient had the rare presentation of an initially seronegative AIH flare during pregnancy, which could be attributed to the potential physiologic effects of pregnancy in the regulation of immune system mechanisms.

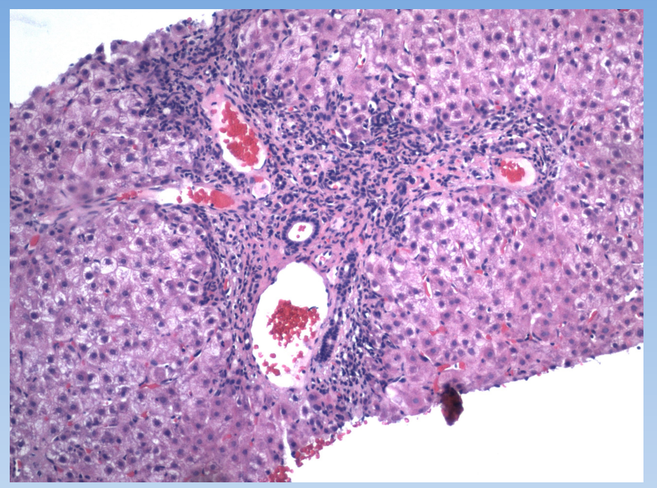

Fig 1. Portal inflammation

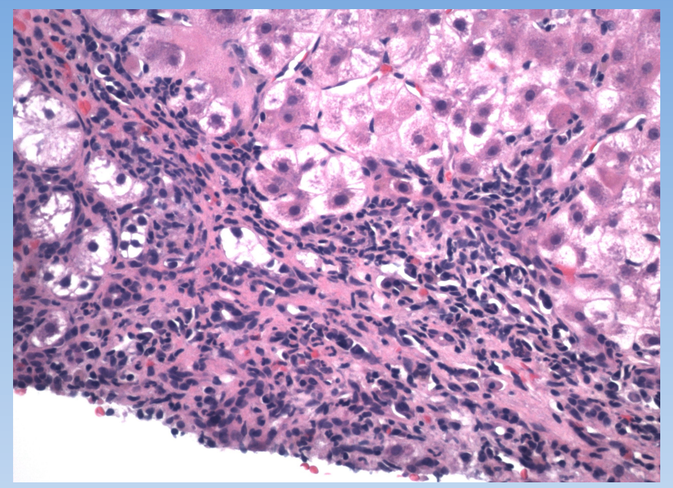

Fig 2. Interface hepatitis

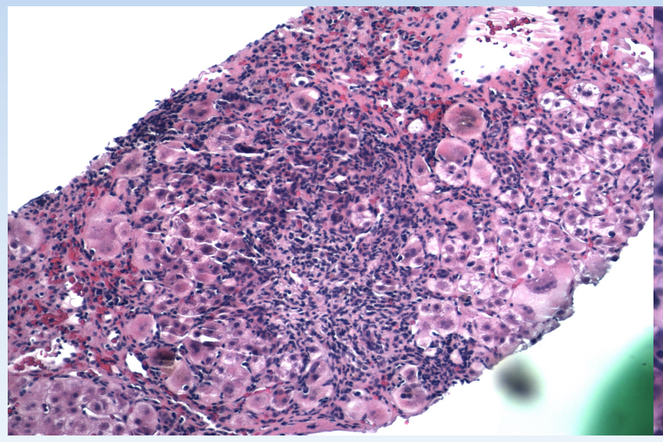

Fig 3. Hepatitis with hepatocyte degeneration.

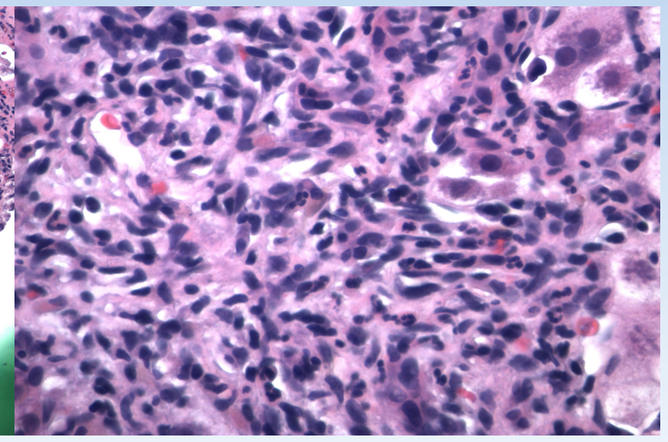

Fig 4. Aggregates of plasma cells and neutrophils.

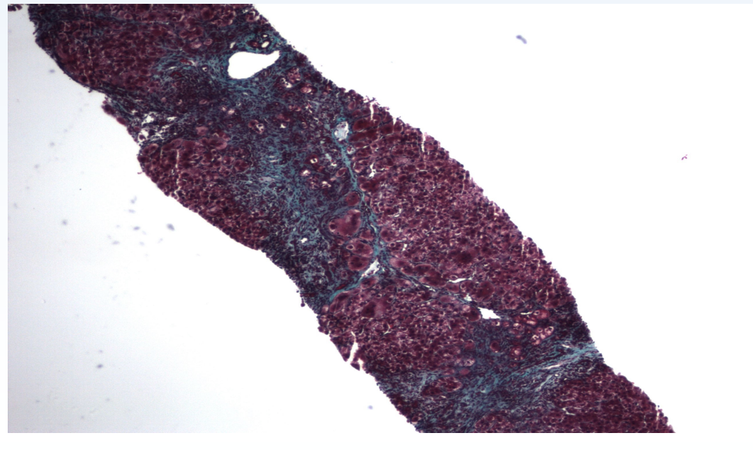

Fig 5. Trichrome stains showing portal fibrosis and bridging fibrosis.

References

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