A Fatal Case of Acute Liver Failure secondary to Autoimmune Hepatitis

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Introduction

- Autoimmune hepatitis (AIH) – a inflammatory condition characterized by the circulation of autoantibodies that damage the liver [1].
- Diagnosis can be difficult due to a variety of presenting symptoms ranging from asymptomatic to acute liver failure. [1]
- Responsiveness to immunosuppressive therapy may support the diagnosis [1]
- A retrospective analysis of 115 patients with AIH revealed that approximately 9% of them presented with acute hepatitis. [2]
- Patients should be considered for urgent liver transplantation. [1]

Case Presentation

HPI :
• 66-year-old male presented with the complaints of upper abdominal pain, fatigue, bloating, constipation, dizziness, pruritus, anorexia, dark colored urine, light colored stools, and 10 lbs of weight loss over the past weeks.
• **I**nitially was hospitalized due to elevated liver enzymes 5 months ago – followed by hyperbilirubinemia and jaundice
PE :
• Icteric sclera, jaundiced skin, and mildly encephalopathic
Labs :
• AST 2100 IU/L, ALT 1900 IU/L, INR 2.3, total bilirubin 24 mg/dL, direct bilirubin of 18 mg/dL
• Creatinine of 1.24mg/dL
• Immunoglobulin level 7.2 g/L, total protein of 10 g/L, albumin of 2 g/L, and 4+ urinary bile
• Anti-smooth muscle antibody with a titer of 1:80.
Imaging :
• CT of the abdomen and pelvis with contrast = heterogeneous hepatic parenchymal enhancement with mild periportal edema, and gastric wall thickening *(figure 1)*
• MRCP w/ and w/o contrast = moderate diffuse heterogeneous arterial phase enhancement with trace central periportal edema., mild splenomegaly, mild varices, and trace abdominal ascites

Biopsy :
• Transjugular liver biopsy = scattered foci of residual liver parenchyma with scattered apoptotic hepatocytes and lymphocytes, fibrotic liver tissue, dense lymphoplasmacytic infiltrate (60% plasma cells)
• Iron stain did not show any iron deposition in the liver parenchyma and copper stain was negative.

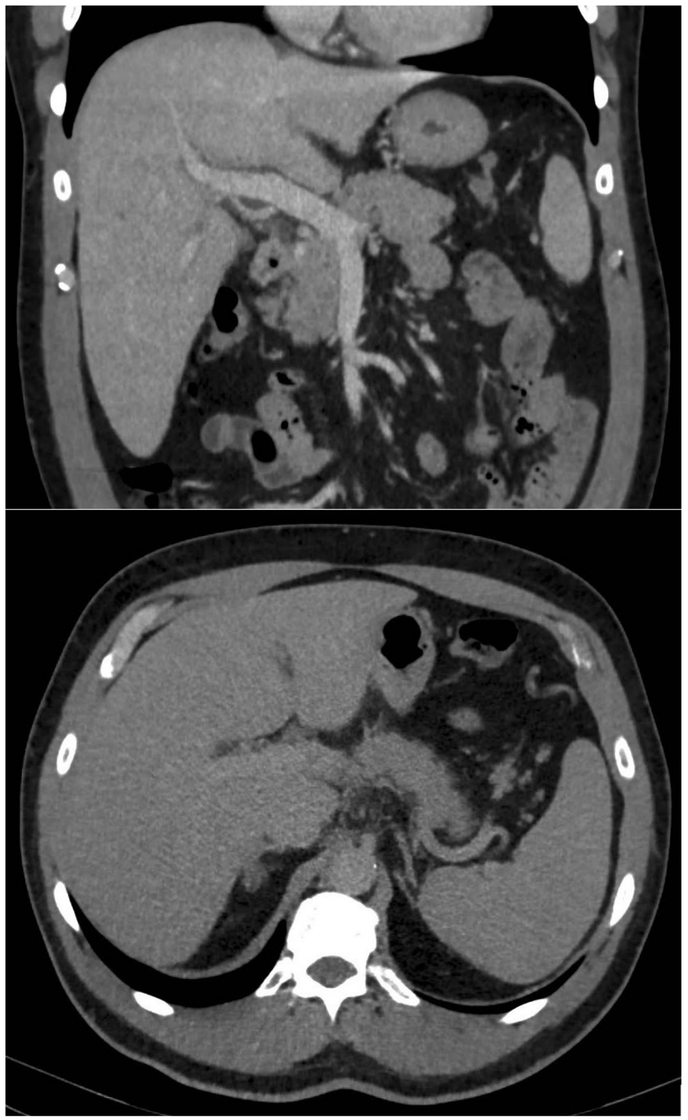

Figure 1:
Top: Coronal view showing heterogenous hepatic parenchymal enhancement with mild periportal edema

Bottom: Transverse view showing hepatomegaly, parenchymal enhancement

References

1. Rahim MN, Miquel R, Heneghan MA. Approach to the patient with acute severe autoimmune hepatitis. JHEP Rep. 2020;2(6):100149. Published 2020 Jul 21. doi:10.1016/j.jhepr.2020.100149
2. Kessler WR, Cummings OW, Eckert G, Chalasani N, Lumeng L, Kwo PY. Fulminant hepatic failure as the initial presentation of acute autoimmune hepatitis. Clin Gastroenterol Hepatol. 2004;2(7):625-631. doi:10.1016/s1542-3565(04)00246-0
3. Montrief T, Koyfman A, Long B. Acute liver failure: A review for emergency physicians. Am J Emerg Med. 2019;37(2):329-337. doi:10.1016/j.ajem.2018.10.032
4. Ganger DR, Rule J, Rakela J, et al. Acute Liver Failure of Indeterminate Etiology: A Comprehensive Systematic Approach by An Expert Committee to Establish Causality. Am J Gastroenterol. 2018;113(9):1319. doi:10.1038/s41395-018-0160-2

Patient Course

• Extensive work ruled out drug-induced, viral, and alcoholic causes of acute liver failure
• Autoimmune hepatitis was suspected as a possible etiology and supported by positive anti-smooth muscle antibody
• Biopsy was consistent with acute hepatitis with extensive liver cell necrosis (80%) and the presence of several plasma cells - highly indicative of autoimmune hepatitis.
• The patient was started on high dose prednisone and liver transplant evaluation was promptly initiated due the rapid deterioration of his liver function
• On the tenth day of his hospital stay, his condition rapidly declined – he became hypotensive, hypoxic, tachycardic, and tachypneic
• Laboratory testing revealed a lactate level of 14.6, consistent with severe acidosis.
• He was admitted to the intensive care unit, intubated, started on continuous renal replacement therapy, and multiple vasopressors. Despite aggressive treatment, the patient unfortunately expired on the eleventh day of his hospital stay.

Discussion

- This case demonstrates the importance of considering autoimmune hepatitis as a possible etiology of acute liver failure.
- Despite being mainly thought of as a chronic condition, autoimmune hepatitis can manifest in an acute manner as demonstrated by our case. Overlooking autoimmune hepatitis as a cause of acute liver failure may cause delays in treatment and obtaining a liver transplant.
- In one study, of 142 cases of acute liver failure that were originally labeled as “acute liver failure of indeterminate etiology”, 34 of them were found to be caused by AIH upon further investigation. [4]
- This highlights the importance of keeping AIH high on the list of possible etiologies for cases of ALF which may be labeled as idiopathic or caused by an indeterminate etiology.
- Although the pathogenesis of autoimmune hepatitis is complex and not fully understood, it is thought that certain environmental triggers may play a role in inciting hepatic dysfunction in patients with underlying autoimmune hepatitis. [1]
- One clue that may help in identifying AIH early in the initial phases of the work up is to ask regarding potential exposures to environmental triggers such as hepatotoxic drugs, recent infections, and recent immunizations.