Seronegative Autoimmune Hepatitis: An Unusual Presentation

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Introduction

Seronegative autoimmune hepatitis (AIH) presenting as acute liver failure is uncommon. Prompt recognition and treatment of AIH can significantly alter morbidity and mortality.

We present the case of a patient with moderate dementia, who was admitted for transaminitis and jaundice. A diagnosis of AIH was made later in her hospital course.

Case Presentation

A 74-year-old woman with past medical history of GERD and moderate Alzheimer's dementia was noted to have transaminitis on routine outpatient labs (ALT 272, AST 183), which did not resolve over four months.

Her only medications were calcium and famotidine. She took no other supplements or substances. She presented to our hospital after recent repeat outpatient chemistries showed worsening transaminitis.

Family noted she was unable to care for herself as usual and was less alert than her baseline.

In the ED, vital signs were normal. Initial labs revealed elevated total bilirubin 14.8 mg/dL, direct bilirubin 10.0 mg/dL, INR 1.5, AST/ALT 1428 and 1224 IU/L, Alk Phos 168 IU/L, Albumin 3.4 g/dL.

Upon admission, she appeared fatigued and jaundiced. Abdominal exam was benign.

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Hospital Course

Initial imaging in the ED included CTAP, which noted a small lesion in the pancreas, with pancreatic and periportal adenopathy. It also noted an enlarged gallbladder. She was admitted to medical oncology with a presumed diagnosis of pancreatic cancer.

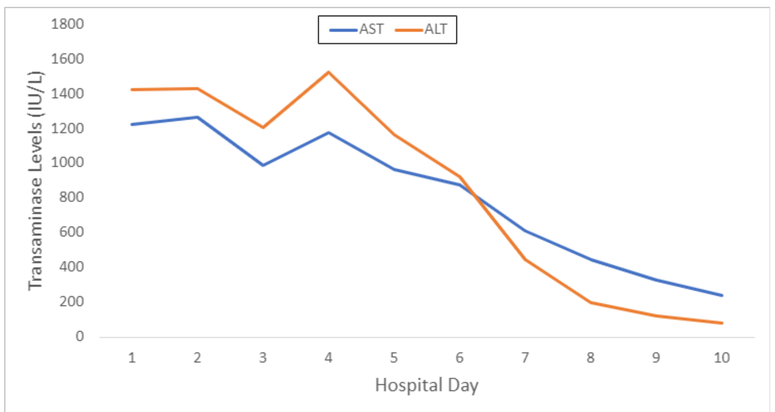
Further tests, including immunologic serologies (ANA screen, anti-smooth muscle antibodies (ASMA), LMK-1 antibodies) were negative. EBV, CMV and hepatitis panel were negative. Immunoglobulin G was noted to be elevated (2700).

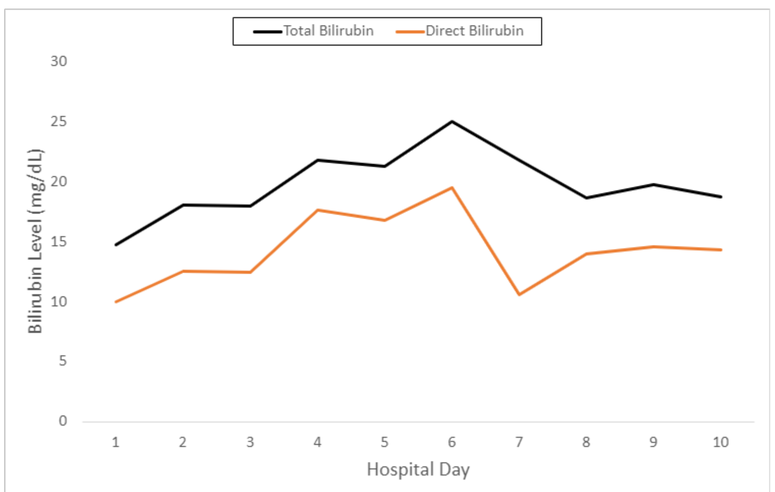
She could not complete MRCP and underwent endoscopic ultrasound (EUS) on hospital day 4, which revealed normal pancreas and bile ducts. EUS-guided biopsy of the liver was then done.

On hospital day 6, she developed acute liver failure, with obtundation and increase in INR to 2.1. Methylprednisolone and N-acetylcysteine were started. Due to dementia, she was ineligible for liver transplant.

On hospital day 8, pathology confirmed autoimmune hepatitis with bridging fibrosis. By hospital day 10, liver function was improving (Figures 1-2); however, she developed fever and, despite broad-spectrum antibiotics, suffered from septic shock and expired on day 11.

Figures





Discussion

Fulminant-onset autoimmune hepatitis (AIH) with hepatic encephalopathy is rare, occurring in <6% of patients with AIH.

The frequency of seronegative autoimmune hepatitis in acute and severe AIH is further estimated to be ≤7%.

This case illustrates the challenge in diagnosis, particularly when confounding factors are present.

Our patient suffered from dementia and aphasia, which masked developing hepatic encephalopathy.

Additionally, pancreatic cancer was initially suspected as the cause of her painless jaundice, which further delayed steroid treatment.

A high index of suspicion for AIH is needed in patients with liver failure of uncertain etiology, as early treatment with steroids may improve the clinical outcome.

References

- Sherigar JM, Yavgeniy A, Guss D, Ngo N, Mohanty S. Seronegative Autoimmune Hepatitis A Clinically Challenging Difficult Diagnosis. Case Rep Med. 2017;2017:3516234. doi: 10.1155/2017/3516234. Epub 2017 Jul 6. PMID: 28761444; PMCID: PMC5518508.
- Francque S, Vonghia L, Ramon A, Michielsen P. Epidemiology and treatment of autoimmune hepatitis. Hepat Med. 2012 Mar 16;4:1-10. doi: 10.2147/HMER.S16321. PMID: 24367228; PMCID: PMC3846915.
- 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 Jul;2(7):625-31. doi: 10.1016/s1542-3565(04)00246-0. PMID: 15224287.