Primary Hepatic Amyloidosis as a Mimic of the Rapid Development of Decompensated Cirrhosis

Debra W. Yen, MD[1], Divya Sharma, MD[2], Marshall Weesner, MD[3], Michael Schoech, MD[1]
[1]Division of Digestive Diseases, University of Cincinnati, [2]Department of Pathology, University of Cincinnati, [3]Division of Gastroenterology, Cincinnati VA Medical Center

INTRODUCTION

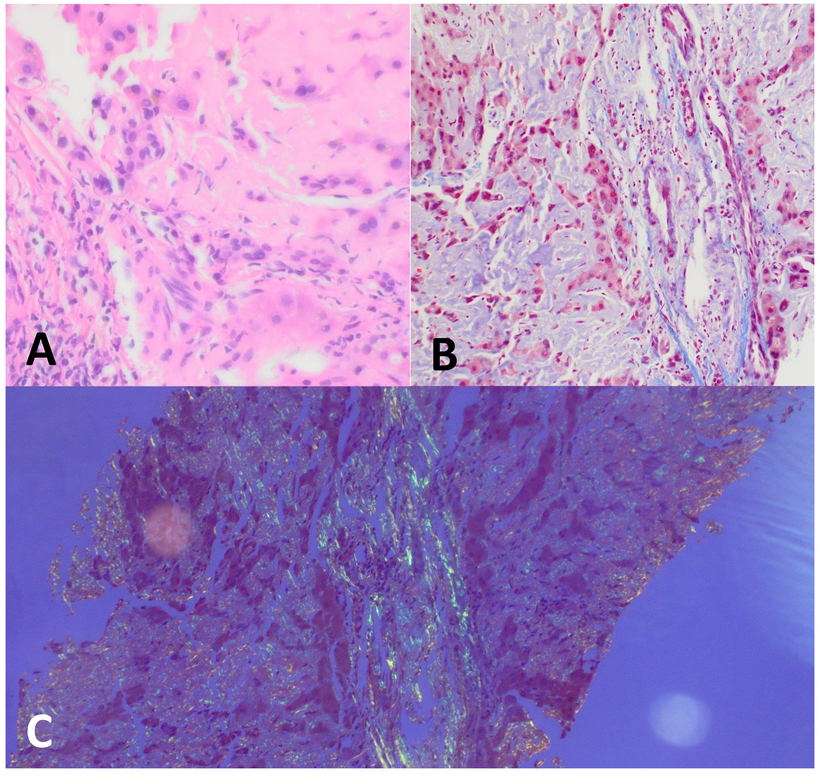
- Primary AL amyloidosis is a systemic disease characterized by the deposition of insoluble fibrils from immunoglobulin light chains.
- Hepatic involvement is common, though clinical manifestations are rare.
- This is a case of primary hepatic amyloidosis that mimicked the rapid development of decompensated cirrhosis.

CASE DESCRIPTION

- 63 yo M with h/o remotely treated HCV presented with new onset ascites for 1 month.
- He also presented with AKI (Cr 3.1) and nephrotic range proteinuria.
- Of note, liver biopsy 5 years ago had only portal fibrosis (F1 fibrosis).
- Initial workup
  - Labs: AST 201, ALT 64, TBili 1.3, ALP 582 alb 2.5, plt 105, INR 1.5
  - Paracentesis: 4L removed. SAAG > 1.1, protein < 2.5, consistent with ascites from portal hypertension
  - Serologic testing for other etiologies of chronic liver disease was negative.
- Further testing
  - Urine immunofixation: monoclonal free kappa light chains
  - K/L ratio: 8.35 (ref range 0.26-1.65)
- Liver and kidney biopsies
  - Amyloid, confirmed by Congo red stain
- Diagnosis: AL amyloidosis
- Unfortunately, he died from VF arrest 1 week later.

PATHOLOGY

Liver biopsy (2022)



Liver biopsy (2017)

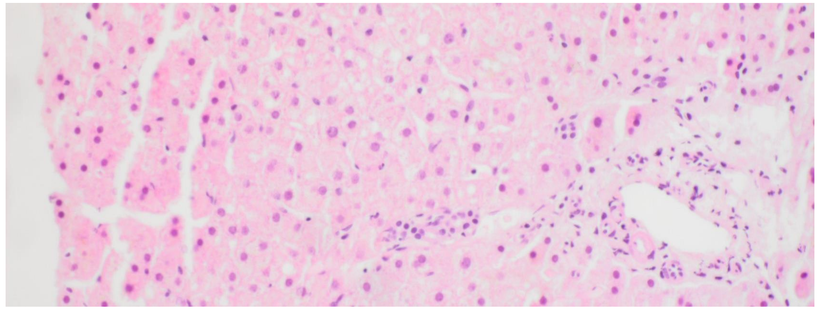


Fig 2. H&E stain 20X (2017): Normal hepatic parenchymal architecture. No significant fibrosis, inflammation or amyloid deposition.

Fig 1. A. H&E stain 20X (2022): Deposition of abundant amorphous, acellular, eosinophilic amyloid deposits within portal vessels and hepatic sinusoids leading to hepatocyte atrophy.
B. Trichrome stain 20X (2022): gray blue amyloid deposits within sinusoidal spaces and portal tracts. Bright blue collagen deposition is minimal.
C. Congo red stain 10X (2022): Apple-green birefringence under polarized light within sinusoids and portal tracts.

DISCUSSION

- This patient had a recent unremarkable liver biopsy but seemed to have progressed to decompensated cirrhosis in a short period of time.
- Ultimately, his entire presentation was attributable to primary hepatic amyloidosis.
- In a patient without known liver disease who rapidly develops signs of portal hypertension and systemic features of other end-organ involvement (i.e. proteinuria, cardiomyopathy), amyloidosis is important to include on the differential.

LEARNING POINTS

- Most frequent findings of hepatic amyloidosis are hepatomegaly and elevated alkaline phosphatase.
- Sinusoidal portal hypertension may occur and manifest as ascites, splenomegaly, and bleeding esophageal varices.
- Portal hypertension is thought to develop from the decreased vascular space of hepatic sinusoids from massive perisinusoidal amyloid deposits.
- Diagnosis involves biopsy of other organs and/or the liver. Bleeding risk is elevated for liver biopsy.
- Prognosis is poor (median survival is 9 months).