New Diagnosis of Systemic Amyloidosis in a Patient with Chronic Hepatitis B and Elevated Liver Stiffness Score on MR Elastography

Cristina I. Batarseh[1], Atoussa Golder-Najafi[2], Ali Khader[3], Carmi S. Punzalan[4,5]

[1] Department of Internal Medicine [2] Department of Anatomic Pathology [3] Department of Diagnostic Radiology[4] Department of Gastroenterology [5] Division of Transplant and Hepatobiliary Diseases, Lahey Clinic

Beth Israel Lahey Health
Lahey Hospital & Medical Center

Introduction

- Amyloidosis is a disorder of abnormal deposition of protein into bodily tissue.
- This case describes a patient with chronic hepatitis B (HBV) who was found to have systemic amyloidosis after a magnetic resonance elastography (MRE) was consistent with advanced fibrosis.
- This case highlights the importance of confirming the stage of fibrosis with liver biopsy, especially if the clinical picture is not consistent with LS measurements.

Case Description

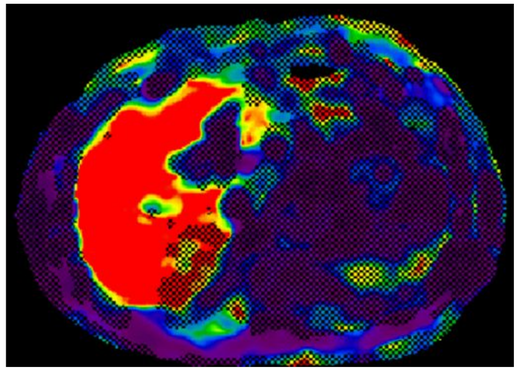
- A 58-year-old male initially evaluated for chronic HBV on Tenofovir Disoproxil Fumarate since 2009.
- On evaluation he reported short-term memory deficit, impaired concentration, weight loss, and anorexia for 2 years.
- Physical examination was unremarkable. Labs included normal CBC, LFTs, and INR, baseline creatinine, and undetectable HBV DNA. Shear wave ultrasound elastography (SWE) revealed increased echogenicity and mild steatosis, but liver stiffness (LS) measurements could not be obtained.
- After options for further testing were discussed, the patient elected to have a MRE (Fig1 A/B). He was found to have a LS measurement of 11.75 kPa consistent with cirrhosis. Liver appeared normal on T2 without steatosis. No stigmata of chronic liver disease or contour nodularity was observed. He ultimately had a non-focal liver biopsy which revealed diffuse deposition of amorphous congophilic material consistent with amyloid involving sinusoids and portal tracts, mild increased portal and periportal fibrosis (stage I-II/IV), and mild chronic portal inflammation.
- Findings were consistent with amyloidosis (Fig2 A/B). Amyloidosis was typed by laser capture liquid chromatography and tandem mass spectrometry as AL–lambda.
- Further work up ruled out renal and cardiac involvement. Bone marrow biopsy revealed lambda-typic plasma cell dyscrasia. The patient being treated with daratumumab and cyclophosphamide + bortezomib+dexamethasone.

Discussion

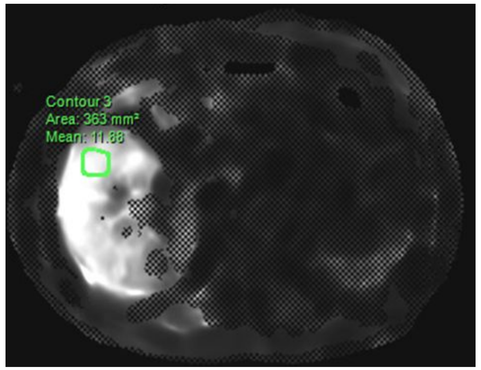
- SWE and MRE provide noninvasive information about fibrosis in patients with chronic liver disease.
- In this case, the LS measurement was elevated due to hepatic amyloidosis rather than cirrhosis . Amyloidosis has a wide variety of presentations. The most common findings in hepatic amyloidosis are hepatomegaly and elevated alkaline phosphatase, none of which was present in our case.
- Gastrointestinal and hepatic involvement are usually associated with worse outcomes, thus early diagnosis is essential.

Figure 1

A



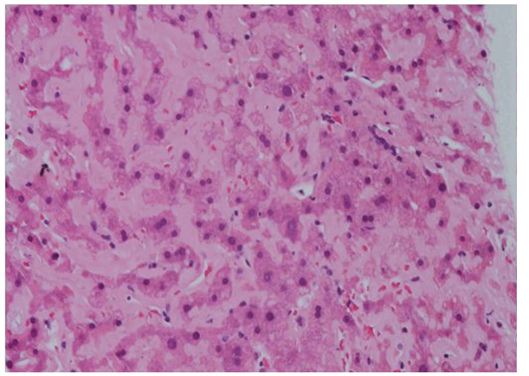
B



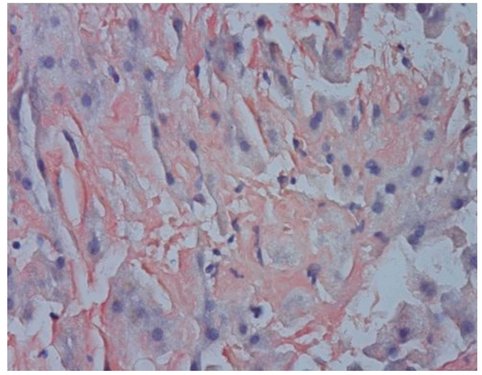
- (A & B) MR elastography with (A) showing the sampleable portion of liver parenchyma demonstrating elevated stiffness as colored red. (B) is one of four liver samples used to calculate liver stiffness measurement.
- Increased liver stiffness, in the appropriate clinical setting, is compatible with liver fibrosis. <2.5 kPa = Normal, 2.5 to 3.0 kPa = Normal or Inflammation, 3.0 to 3.5 kPa = Stage 1-2 fibrosis, 3.5 to 4 KPa = Stage 2-3 fibrosis, 4.0 to 5.0 KPa = Stage 3-4 fibrosis, >5.0 KPa = Stage 4 fibrosis or Cirrhosis.

Figure 2

A



B



- (A & B) Non-focal liver biopsy 20X H&E and 40X congo red stain respectively, showing deposition of amorphous congophilic material in sinusoidal tracts.