Out of the woods and into the jungle;
A case of an uncommon problem presenting commonly.

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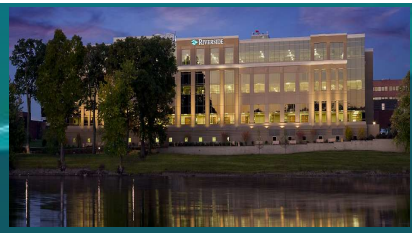
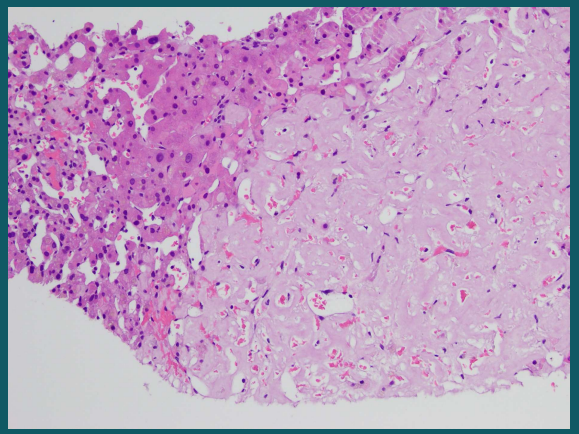


Background

Amyloidosis is an emerging diagnosis due to increase in availability of screening and more awareness in the medical community to the broad and unique presentation of the disease. We present a case of a patient previously believed to have cirrhosis of the liver due to alcohol, that was eventually determined to be pseudocirrhosis from amyloidosis and cardiac disease.

Case Presentation

A 71-year-old male with a history of chronic heart failure and recently diagnosed MET-ALD cirrhosis presented with abdominal distension. Abdominal ultrasound revealed ascites, and paracentesis with fluid analysis demonstrated a serum-ascites albumin gradient (SAAG) of 1.9g/dL and a peritoneal fluid protein concentration of 3.4 g/dL. Given the elevated SAAG (>1.1g/dL) and peritoneal fluid protein (>2.5g/dL), the differential diagnosis included heart failure, Budd-Chiari syndrome, and veno-occlusive disease; cirrhosis was considered less likely due to the high fluid protein. Further investigation included a Doppler ultrasound, which excluded Budd-Chiari syndrome, and an echocardiogram, which revealed a normal ejection fraction with no wall motion abnormalities or signs of acute heart failure. Notably, the echocardiogram showed an apical sparing strain pattern, raising suspicion for amyloidosis. Serum protein electrophoresis (SPEP), urine protein electrophoresis (UPEP), immunofixation, and technetium-99m pyrophosphate (PYP) scan were ordered to investigate this further. In the meantime, the patient was started on diuretics, with initial improvement in ascites. However, the ascites rapidly reaccumulated. With the cause still undetermined, a liver biopsy was performed along with measurement of hepatic vein pressures. The hepatic vein pressure gradient was found to be zero, though the liver biopsy revealed extensive sinusoidal amyloid deposition, confirming the diagnosis of hepatic amyloidosis. The patient underwent an additional therapeutic paracentesis and was subsequently discharged with referrals to hematology and hepatology for further management.



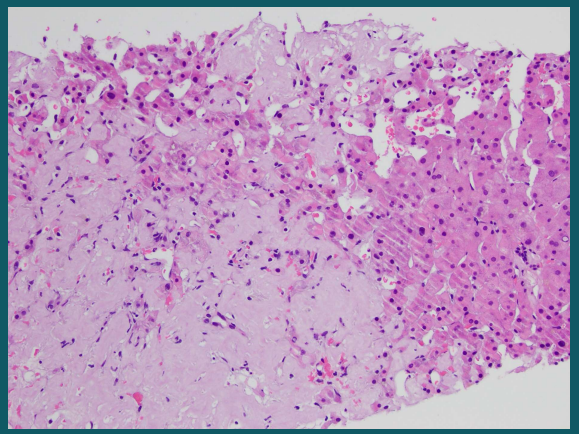


Figure A (above) and B (below): 100x Liver biopsy demonstrating amyloidosis in a core tissue sample.

Discussion

Amyloidosis is a condition of abnormal plasma cell function, creating abnormally constructed light chain antibodies. The most common form of amyloidosis is AL amyloid, which is found deposited in the extracellular matrix of organs, (1). Often the affected organs of amyloid include the heart (50-60% of cases). Regarding gastrointestinal (GI) manifestations of amyloid include deposition into the luminal structures of the stomach, esophagus, large intestine and, with the greatest frequency, the small intestine. Hepatic involvement often appears to be hepatomegaly and elevated alkaline phosphatase (3). The most common form of amyloidosis is AL amyloidosis that is associated with monoclonal light chains in urine and serum. AA amyloidosis is most common in patient who suffer from GI manifestations. In rare cases, one can display symptoms of severe cholestasis and intractable ascites; which can cause confusion in diagnosis as they are also common symptoms of cirrhosis (1,2).

As it is increasingly rare for fulminant hepatic failure, nodular appearance of the liver, and recurrent ascites; one can reasonably conclude that the patient above was notably suffering from a restrictive cardiomyopathy increasing portal pressures (4).

Accurate diagnosis of amyloidosis is essential to treatment for patients. A diagnosis can be confirmed on biopsy with polarized light eliciting the characteristic apple green coloration and red in normal light. This is accomplished through Congo Red Staining of the tissue sample, (1,3). As was noted in our case, Trichrome stain was negative for fibrosis, directly contradicting the diagnosis of cirrhosis. Instead, establishing an even stronger case for amyloidosis being the primary diagnosis.

This case report serves to remind clinicians that one should consider a differential diagnosis of amyloidosis when more than one organ system is affected, (1). This can be accomplished through careful chart review to evaluate the involvement of other organ systems to ensure accurate diagnosis be achieved at earliest interval.

Sources

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