An unusual cause of portal hypertension

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Introduction

A 66-year-old male with a medical history of coronary artery disease presented with nonspecific symptoms of anorexia, lethargy and an increase in abdominal girth. He was diagnosed 3 months ago with chronic liver disease of undetermined aetiology. He did not smoke and was a social drinker till 4 months ago. He was taking Furosemide, Aldactone, Pantoprazole, Acetylcysteine and Atorvastatin. He had no personal or familial history of liver diseases.

Physical examination revealed a tender but distended abdomen and large ascites was present. The laboratory tests showed progressive cholestatic liver enzymes (total bilirubin 1,09 mg/dL, AST 130 U/L, ALT 129 U/L, GGT 517 U/L and ALP 641 U/L) and a severe hypoalbuminemia. Laboratory workup made a metabolic, autoimmune, and viral aetiology of the suspected cirrhosis less probable. Urinalysis showed a rapidly progressive proteinuria in the nephrotic range accompanied by the presence of serum spike of monoclonal lambda light

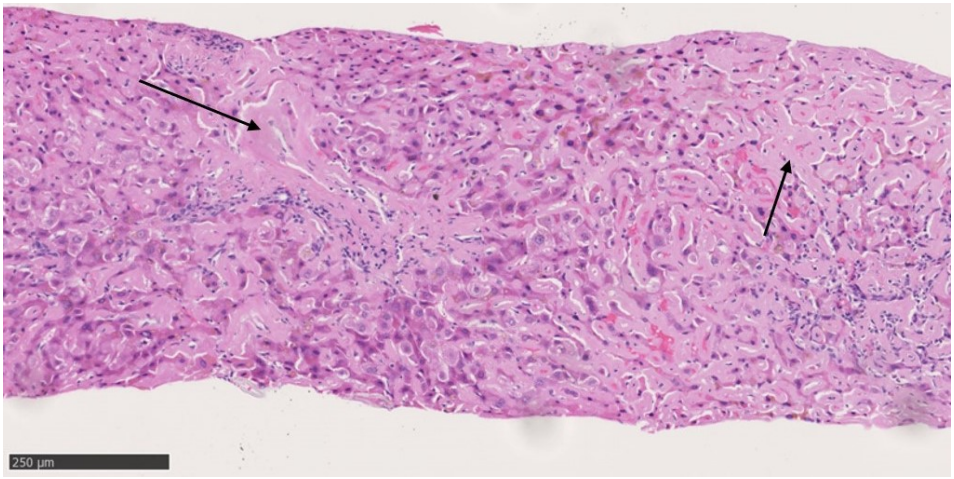

Figure 1a. — HE-staining; extensive space of Disse deposition of amyloid (seen as smudgy eosinophilic linear deposits perisinusoidal – see arrows). Linear sinusoidal pattern of deposition producing compression atrophy of liver-cell plates.

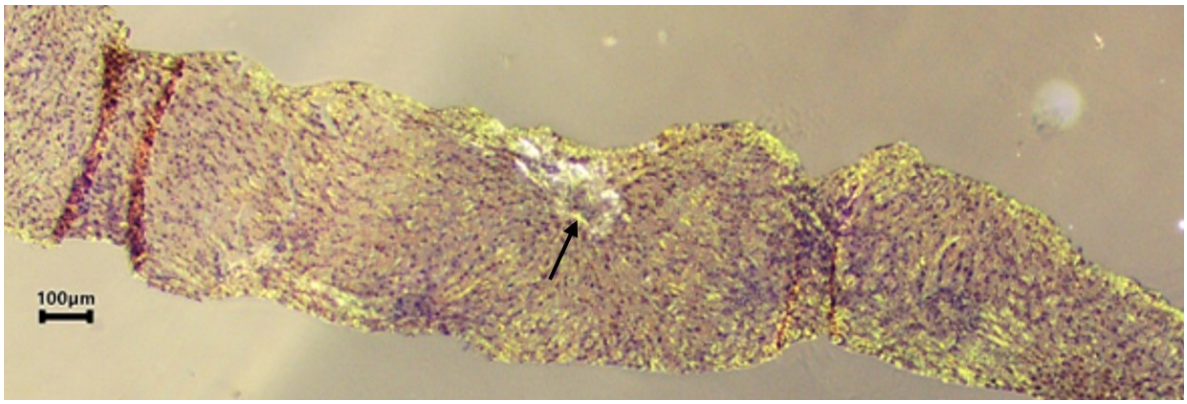

Figure 1b. — Congo red-stained amyloid shows green birefringence (see arrow) in polarization microscopy.

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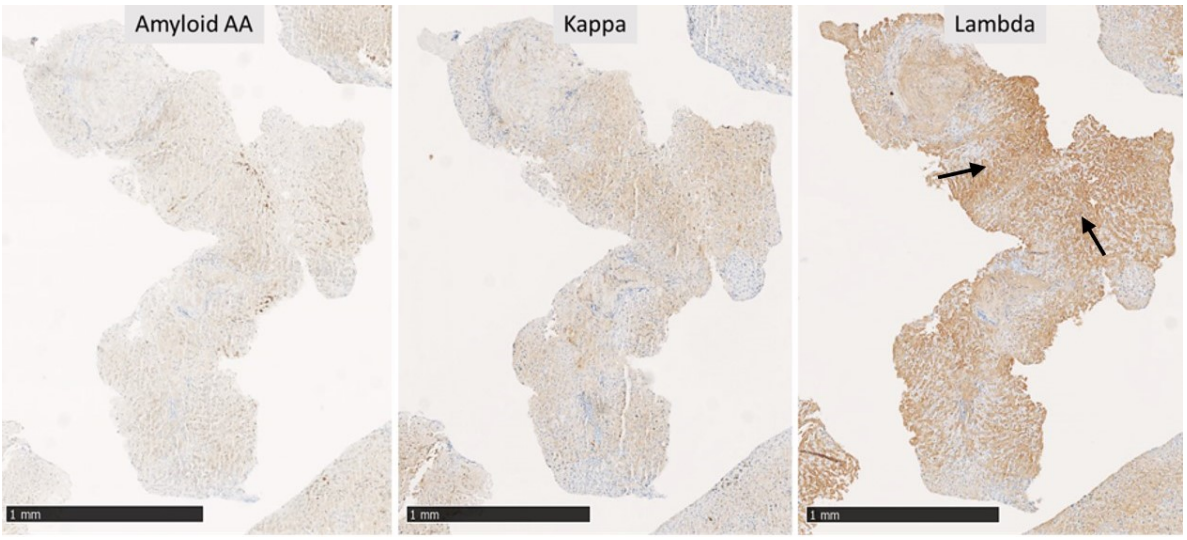

Figure 2. — Immunohistochemistry for AA amyloid and kappa and lambda light chain are suggestive of lambda light chain amyloid (see arrows).

chain protein on electrophoresis with immunofixation. Doppler echocardiography revealed a hypertrophic cardiomyopathy based on diffuse left and right ventricular hypertrophy with relative apical sparing. A diagnostic paracentesis was undertaken and the results were consistent with portal hypertension (SAAG 1,6 g/dL).

Considering the concomitant presence of hypertrophic cardiomyopathy, nephrotic syndrome and a presentation of portal hypertension of undetermined aetiology in presence of a monoclonal IgG lambda light chain spike, a transjugular liver biopsy (figure 1 and 2) was performed with HVPG measurement consistent with significant portal hypertension (15 mmHg).

Question

What is the most likely diagnosis based on clinical presentation, laboratory and pathology results?

Answer

Microscopy showed sinusoidal deposition of amorphous eosinophilic material causing atrophy of liver cell plates with Congo red stain and immunohistochemistry positive for lambda amyloid deposits. His final diagnosis was established as AL amyloidosis with the involvement of heart, liver and kidneys. The patient had a symptomatic hepatic involvement due to portal hypertension in this advanced case of amyloid infiltration. He was started on cyclophosphamide, bortezomib and dexamethasone but unfortunately succumbed a few days later.

The highlights of this case are to consider infiltrative diseases such as amyloidosis in a patient presenting with cholestatic hepatitis. A liver biopsy is essential for the diagnosis (2). Hepatic involvement in amyloidosis is common, although symptomatic presentations due to portal hypertension are rather exceptional (1,2). Recognition of this process is critical because it identifies patients with widespread organ involvement and portends a poor prognosis (1,2).

Keywords: hepatic amyloidosis, portal hypertension', infiltrative liver diseases.

Conflict of interest

The authors have no potential conflict of interest relevant to this clinical image to be reported.

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