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Case Report

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Hepatic Amyloid

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Introduction

The term 'amyloidosis' refers to the extracellular deposition of low molecular weight fibrillar proteins (amyloid) that stain positively with Congo red and exhibit apple-green birefringence under polarized light. The disease leads to a wide range of clinical manifestations depending on its type, location, and amount deposited. Primary amyloidosis is the most prevalent type of amyloidosis, and the presence of hepatic amyloid has been described in up to 90% of patients.

Case Report

A 73-year-old woman with the following relevant personal history: heart failure, permanent atrial fibrillation, IgG-kappa multiple myeloma in complete remission, and lymph node tuberculosis. She presents to the Emergency Department due to asthenia, marked weight loss, and jaundice evolving over a couple of months. She had recently started taking paroxetine and digoxin, both prescribed by a private cardiologist. On examination, hepatomegaly of 3 fingerbreadths and bilateral ankle edema with pitting were noted. Analytically, there was predominant hyperbilirubinemia of 8.95, mainly direct, and a slight elevation of transaminases. Abdominal ultrasound did not reveal any hepatic findings, so admission for further evaluation was decided.

During this admission, an abdominal MRI was performed, showing heterogeneous liver parenchyma with a mosaic appearance, as well as dilation of the inferior vena cava and suprahepatic veins (all radiological signs of passive hepatic congestion). Subsequently, suprahepatic vein manometry was performed, revealing a portal pressure gradient of 4 mmHg, along with a transjugular liver biopsy. The biopsy showed massive infiltration by amyloid material with immunoreactivity for Kappa light chain. The patient was diagnosed with systemic AL amyloidosis with hepatic involvement (confirmed by biopsy) and possibly cardiac involvement.

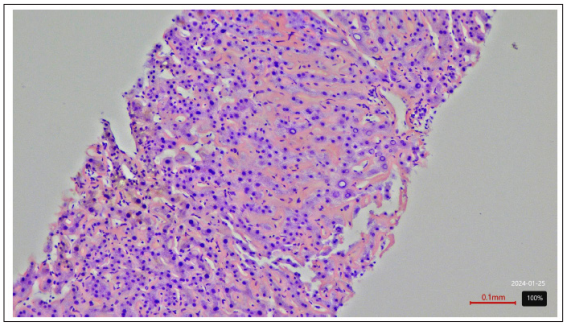

Figure 1: Congo red Staining

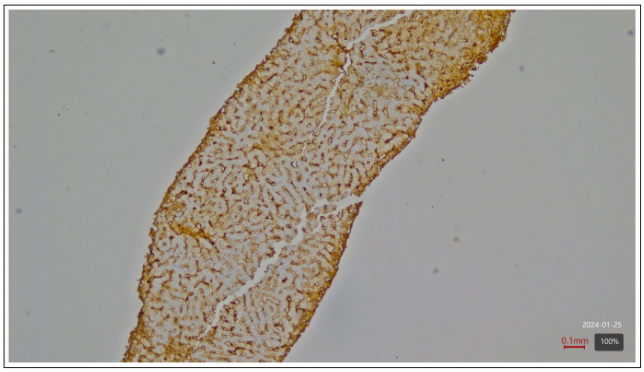

Figure 2: Immunofluorescence: Expression of Kappa Light Chains

Discussion

The most frequent clinical findings of hepatic amyloidosis are constitutional symptoms and hepatomegaly. Although patients may present with associated ascites, most likely due to concurrent heart failure or hypoalbuminemia. Jaundice is rare but constitutes a sign of poor prognosis. The most common analytical finding is elevated alkaline phosphatase. Radiological and endoscopic findings of amyloidosis are nonspecific. A liver biopsy is needed to establish the diagnosis of amyloid and rule out other infiltrative liver diseases, such as sarcoidosis, tuberculosis, neoplasms, and glycogen storage diseases. Regarding treatment, in addition to symptomatic therapy, it is based on treating the underlying disease [1-5].

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