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DOI: 10.17235/reed.2023.9598/2023

Link: [PubMed \(Epub ahead of print\)](#)

Please cite this article as:

Fernandes Daniel Alvarenga , Assis-Mendoza Guilherme Rossi, Costa Larissa Bastos Eloy da , Freitas Leandro Luiz Lopes de , Boin Ilka de Fátima Ferreira Santana , Reis Fabiano. Amyloidosis: a rare cause of severe acute liver failure. Rev Esp Enferm Dig 2023. doi: 10.17235/reed.2023.9598/2023.

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Amyloidosis: a rare cause of severe acute liver failure

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Conflict of interest: the authors declare no conflict of interest.

Keywords: Amyloidosis. Hepatic amyloidosis. Multiple myeloma. Liver failure. Multidetector computed tomography.

Dear Editor,

Gastrointestinal amyloidosis can be primary, more associated with monoclonal plasma cell dyscrasia, or secondary, usually secondary to a tissue-destructive, chronic inflammatory process (such as inflammatory bowel disease, for example) and long-term dialysis. The rare presentation of severe acute liver failure in systemic amyloidosis can make the diagnosis and management more difficult. Hepatomegaly with signs of diffuse infiltrative disease and periportal involvement associated with thoracic and other abdominal radiological findings in the appropriate clinical context may constitute a diagnostic imaging clue in this challenge.

Case report

A 61-year-old male patient with severe acute liver failure of unknown etiology, without a history of alcohol consumption, presented with hypovolemic shock. He was confused, with encephalopathy, blood pressure (BP) 90/60 mmHg, heart rate (HR) 77, oxygen saturation level (SpO₂) 99, Glasgow 13, with noradrenaline via central venous access. TGO was 4,414 and TGP 895. Computed tomography (CT) of the abdomen and thorax was performed, showing a diffuse alveolar septal pattern (Fig. 1A). He rapidly evolved with hemodynamic instability and subsequently died. Hepatic anatomopathological evaluation (Fig. 1B-D) followed by Congo red investigation was compatible with amyloid deposits (Fig. 1E and F).

The patient had hypercellular bone marrow with plasma cell proliferation. Ultimately, the patient was diagnosed with systemic primary (AL) amyloidosis associated with multiple myeloma (kappa-light chain restriction) involving the liver, spleen, lung, adrenals and kidneys.

Discussion

Acute liver failure (AHF), defined as the onset of hepatic encephalopathy within four weeks of developing jaundice in a patient with no previous history of chronic liver disease, is a rare presentation in amyloidosis (1,2). Brain magnetic resonance imaging (MRI) with bilateral and symmetrical hyperintensity on T1 weighted images (due to manganese deposits) in the globus pallidus and subthalamic regions may corroborate the diagnosis of hepatic encephalopathy (3).

Abdominal involvement can occur in primary and secondary forms of systemic amyloidosis. In cases of hepatic involvement, hepatomegaly with decreased hepatic attenuation on CT may be observed (1,4). The three most important CT and MRI findings of hepatic involvement in systemic amyloidosis are hepatomegaly, the heterogeneous aspect of the liver and especially periportal involvement. Periportal involvement is seen as low attenuation on CT and low signal intensity on T1-weighted and T2-weighted images on MRI (4). Hepatic amyloidosis can be differentiated by imaging from cirrhosis due to the absence of segment four atrophy. Occasionally, the radiologic findings may precede the clinical findings. Although the prognosis of systemic amyloidosis is still reserved, an earlier diagnosis may contribute to a better

survival (4,5).

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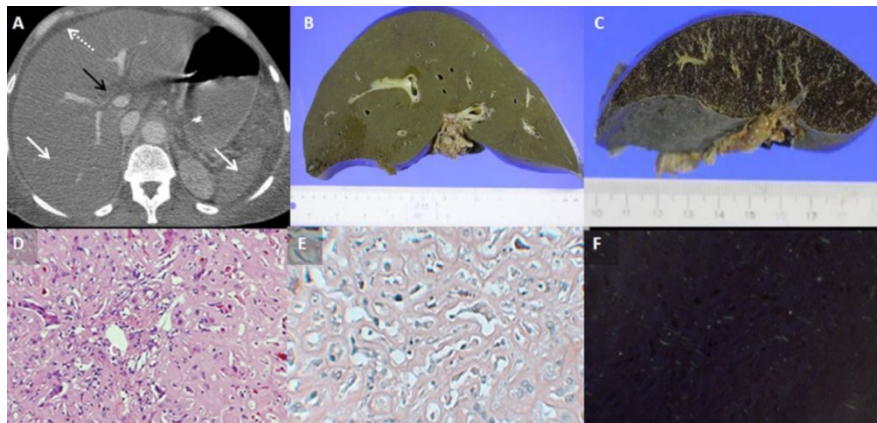


Fig. 1. A. Contrast-enhanced CT of the abdomen (portal phase, axial) revealed an enlarged liver, with regular contours and hypo-perfusion throughout the parenchyma (white arrows), periportal involvement (black arrows) and free perihepatic fluid (dashed white arrows). The same pattern with hypo-perfusion throughout the parenchyma in the arterial, portal and equilibrium phases after contrast medium was observed in the liver and spleen due to reduced blood content caused by the accumulation of amyloid material. B and C. Anatomopathological evaluation. B. Enlarged liver, with pallor. C. Macroscopic examination of the spleen after formalin fixation. D-F. Microscopic examination of the liver using hematoxylin-eosin (HE x200). D. Massive intrasinusoidal deposition of homogeneous and acellular eosinophilic material accompanied by a reduction in the number of viable hepatocytes, which were more frequently found in the 1-periportal zone. E and F. Congo red (400x) staining with regular light microscopy and polarized microscopy (400x) compatible with amyloid deposits.