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PICTURES IN DIGESTIVE PATHOLOGY

Massive hepatic amyloidosis with fatal hepatic failure

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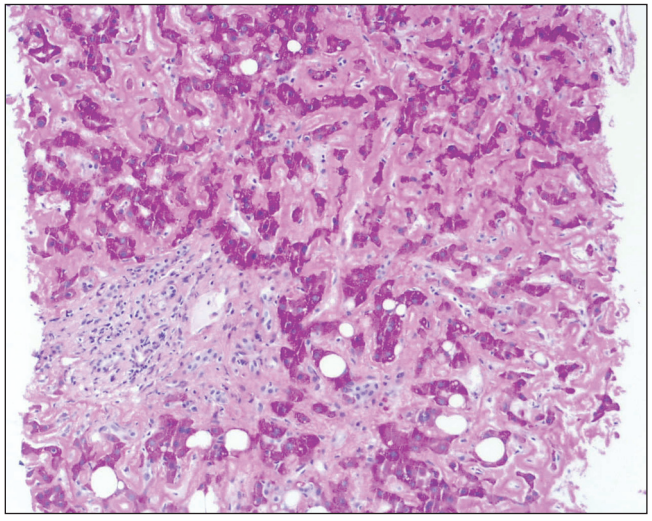

Fig. 1.- Liver biopsy: acellular hyalin material in sinusoids with compression of hepatic trabeculae.
Biopsia hepática: material hialino acelular en sinusoides con comprensión de las trabéculas hepatocitarias.

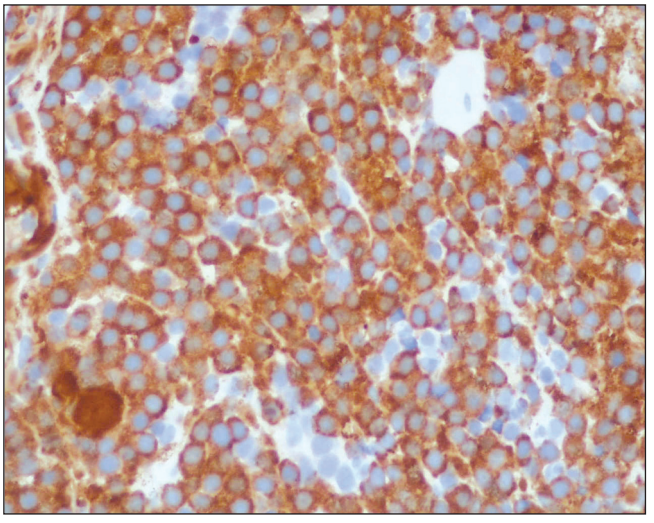

Fig. 2.- Liver biopsy: positive Congo red staining with typical birefringence.
Biopsia hepática: tinción positiva al rojo congo con birrefringencia típica.

A 55-year-old woman diagnosed with high blood pressure, hypothyroidism, and various vertebral fractures attributed to severe osteopenia was admitted to the hospital with a gastrointestinal bleeding secondary to a benign gastric ulcer. During the examination, attention was drawn to an indolent smooth-surface, hard hepatomegaly associated with severely tender spinous processes in lumbar vertebrae. Laboratory parameters showed normocytic anemia (Hb 8.7 g/dl, VCM 84 fl) with a prothrombin activity of 35% and changes in the liver tests (GGT 588 U/L, AP 231 U/L, normal transaminases and bilirubin). In view of these findings an abdominal ultrasound was obtained, which showed liver enlargement without signs of portal hypertension, but evidences of chronic liver disease with no definite findings. Because of the presence of severe coagulation problems, a transjugular liver biopsy was carried out, and the diagnosis of amyloidosis was reached; later, a bone marrow puncture was performed before the advent of vertebral fractures in the lumbar region. After the diagnosis of primary amyloidosis and plasmocytoma, treatment was begun with high-dose of dexamethasone; however, liver and renal function impaired progressively, the patient developed jaundice and died.

Liver involvement is frequent in systemic amyloidosis, and varies according to different series from 56 to 90% if we exclude familial amyloidosis (1,2). The onset of the disease usually involves a presentation with symptoms secondary to liver dysfunction, including abdominal distension and hepatomegaly. Liver function tests may be abnormal and usually include elevated alkaline phosphatase and GGT. The presence of jaundice and particularly serum bilirubin levels above 5 mg/dl is exceptional and commonly associated with a poor outcome (2). The presence of severe intrahepatic cholestasis at onset is exceptional; thus, Goenka et al. (2) described the existence of only 27 cases, of which 22 died before the sixth month after development of jaundice. The presence of high alkaline phosphatase levels up to 4 times the normal value results in poor prognosis, with a mean survival of 3.3 months since jaundice development (3). Treatment with melphalan and prednisone, together with the assessment of potential liver transplantation are the only therapeutic options.

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