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Portal hypertension and intrahepatic cholestasis in primary amyloidosis

The liver is a common site of amyloid deposition in primary systemic amyloidosis. Here, we report the case of a 59-year-old male patient who presented with hepatomegaly, ascites and jaundice.

A 59-year-old man was admitted to a hospital on March 2010 because of abdominal distension and fatigue that had lasted three months. The patient who had been recommended surgical therapy upon detection of an antral polyp of 3 cm size during the upper gastrointestinal endoscopic examination. In the patient, for whom a gastric operation had been decided due to this lesion, ascites and cirrhotic appearance of the liver had been observed during his operation, and a biopsy had been taken. The patient was admitted to the hepatology clinic after two weeks operation due to complaints of increasing abdominal distention and jaundice. On examination he was found to be deeply jaundiced with a 5 cm smooth non-tender hepatomegaly, splenomegaly and ascites. Blood counts were as follows; white blood counts, 12.300/mm^3; hemoglobin, 11.6 gr/dL; and platelet count, 180.000/mm^3. Serum chemistry values included alkaline phosphatase 630 IU/L (normal <120), aspartate aminotransferase 108 IU/L (normal 0 to 40 IU/L), alanine aminotransferase 51 IU/L (normal, 0 to 45 IU/L), total bilirubin 6.7 mg/dL (normal, 0.2-1.2 mg/dL), direct biluribin 5.4 mg/dL (normal, 0-0.2 mg/dL) and prothrombin time 13.8 sec (normal, 12.5 sec). Serum albumin level was 2.4 gr/dL (normal 3.5-4.5 gr/dL). Urinalysis revealed a specific gravity of 1025, albumin 3+. Paracentesis showed an asitic fluid albumin content 0.8 gr/dL, lactate dehydrogenase and amylase values were normal. Serum samples did not contain hepatitis B surface antigen, HCV antibody or anti-hepatitis A IgM. On admission, magnetic resonance imaging (MR) demonstrated ascites and enlarged liver (Figure 1). No diagnostic feature was detected in the liver biopsy taken during the operation of the patient. Intraperitoneal hemorrhage from an unknown origin occurred 12 days after admission. We decided to re-examine the patient's liver biopsy, which had been previously examined by the external center. The patient died two weeks later after admission. During the microscopic re-examination of the biopsy, diffuse amyloid deposition was noted in the Disse spaces and the portal area (Figure 2). Immunohistochemically, lambda light chain stained very weakly whereas kappa light chain was diffusely and strongly positive.

Amyloidosis is characterized by extracellular deposition of abnormal protein. Hepatic involvement is common, but the clinical manifestations are usually mild with hepatomegaly and an elevated alkaline phosphatase level.

Liver biopsy may be helpful diagnostically in patients with hepatic amyloidosis. Atrophic compression of hepatocytes by amyloid infiltration in the space of Disse is a well-known and striking histopathologic feature of hepatic amyloidosis. The risk of bleeding with liver biopsies is controversial. Some investigators report an increased risk of hemorrhage (1) while others claim that liver biopsies are safe (2). Coagulopathy is frequently seen in patients with AL, and several factors may contribute to bleeding complications such as deficiency of factor X, a decrease of vitamin K-dependent clotting factors, increased antithrombin activity and fibrinolysis, and intravascular coagulation (3). Bleeding tendencies can also be explained by amyloid infiltration in vessels, which, once lacerated, may not constrict or clot normally. Splenomegaly and functional hyposplenism correlate poorly with degree of splenic involvement with amyloid, although hyposplenism has been shown to be a poor prognostic factor in amyloidosis. Functional asplenia is related with related neutrophilic leukocytosis. Functional asplenia or hyposplenism may predispose patients to spontanous splenic rupture (4). The mechanism of rupture is associated with the distension of a

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poorly extensible organ capsule due to tissue infiltration, while at the same time the capsuleis infiltrated also, thus becoming inelastic. In our patients, the paracentesis performed revealed hemorrhagic ascites which might be associated with splenic rupture.

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