Letters to the editor

To the Editor,

Amyloidosis is a disease characterized by the deposition of altered proteins in tissues. Amyloidosis is classified as primary or secondary disease. Hepatic involvement, secondary amyloidosis is common, but primary systemic amyloidosis (amyloid light [AL] chain) is an uncommon entity (1). It may result in massive hepatomegaly, elevated serum alkaline phosphatase (ALP) levels and rapidly progressive liver failure (2).

A 51-year-old male presented with nausea, vomiting and jaundice. Hepatomegaly and ascites were noted on physical examination. Laboratory tests revealed the following values; WBC: 13200/mm³, Hct: %36, Plt: 487000/mm³, PT: 16.7 sn, INR: 1.48, APTT: 40.7 sn, ALT: 45 U/L, AST: 69 U/L, ALP: 235 U/L, GGT: 255 U/L, LDH: 186 U/L, T. Bilirubin: 24 mg/dL, D. Bilirubin: 17 mg/dL, Albumin: 2.3 g/dL, Ca: 12 mg/ dL. Protein electrophoresis revealed polyclonal hyper-gammopathy. Serum and urine kappa and lambda light chain levels were elevated. Roller formation on peripheral smear was observed and plasma cells were increased (6%). Markers for hepatitis and autoimmunity were negative. Abdominal tomography showed hepatomegaly, blunt contours and homogeneous parenchyma on liver. Endoscopic retrograde cholangiopancreatography (ERCP) revealed normal gall tracts. Examination of abdominal ascites showed characteristics of transudate. Space of Disse, pa-

REFERENCES


Lee La JANG, Kyu-Jong KIM, Hee Kyung CHANG, Mi Jung PARK, Jong Bin KIM, Jun Sik LEE, Seong Joo KANG, Hee Sang TAG
Departments of Internal Medicine and Pathology, Kosin University College of Medicine, Busan, Korea

A rare case of primary systemic amyloidosis presenting with hepatic failure
Hepatik yetmezilik ile gelen primer sistemik amiloidoz

To the Editor,

Amyloidosis is a disease characterized by the deposition of altered proteins in tissues. Amyloidosis is classified as primary or secondary disease. Hepatic involvement, secondary amyloidosis is common, but primary systemic amyloidosis (amyloid light [AL] chain) is an uncommon entity (1). It may result in massive hepatomegaly, elevated serum alkaline phosphatase (ALP) levels and rapidly progressive liver failure (2).

A 51-year-old male presented with nausea, vomiting and jaundice. Hepatomegaly and ascites were noted on physical examination. Laboratory tests revealed the following values; WBC: 13200/mm³, Hct: %36, Plt: 487000/mm³, PT: 16.7 sn, INR: 1.48, APTT: 40.7 sn, ALT: 45 U/L, AST: 69 U/L, ALP: 235 U/L, GGT: 255 U/L, LDH: 186 U/L, T. Bilirubin: 24 mg/dL, D. Bilirubin: 17 mg/dL, Albumin: 2.3 g/dL, Ca: 12 mg/dL. Protein electrophoresis revealed polyclonal hyper-gammopathy. Serum and urine kappa and lambda light chain levels were elevated. Roller formation on peripheral smear was observed and plasma cells were increased (6%). Markers for hepatitis and autoimmunity were negative. Abdominal tomography showed hepatomegaly, blunt contours and homogeneous parenchyma on liver. Endoscopic retrograde cholangiopancreatography (ERCP) revealed normal gall tracts. Examination of abdominal ascites showed characteristics of transudate. Space of Disse, pa-

Figure 1. The pathologic specimen revealing: liver biopsy staining with Congo red and non-staining with potassium permanganate.

Address for correspondence: Bennur Esen GULLU
Bursa Yuksek Ihtisas Training and Educational Hospital, Department of Nephrology, Bursa, Turkey
Phone: + 90 224 369 50 50 • E-mail: bennuresen@yahoo.com

Manuscript received: 25.02.2010 Accepted: 26.04.2010
renchyma and portal space were stained with Congo red during hepatic biopsy, and this continued after potassium permanganate. It was not immunohistochemically stained with AA antiserum. Biopsy showed diffuse amyloid infiltration, which was consistent with AL type amyloidosis (Figure 1). Kappa and lambda light chains were significantly increased on bone marrow biopsy. Congo-stained amyloid accumulation without potassium permanganate on the vessel wall in the adjacent tissue during bone marrow biopsy was considered as systemic involvement. Factor deficiency was determined, which is consistent with the literature (3). Electromyography (EMG) examination revealed sensory neuropathy, which might have been related to muscle involvement of the primary amyloidosis (4). His echocardiography showed increased diameters of the ventricular septum, left ventricular posterior wall and left atrium, with decreased left ventricular ejection fraction. All these findings were considered as amyloid involvement of the heart.

Prognosis is poor in patients with hepatic amyloidosis. The mean life span for these patients is nine months. Some patients benefit from systemic chemotherapy; however, multiorgan failure is a contraindication for chemotherapy (5).

Melphalan and prednisone therapy was offered, but the patient declined treatment and was discharged home with medical optimization of his hypercalcemia. He died 20 days after discharge.

In conclusion, clinicians should consider the diagnosis of primary hepatic amyloidosis in patients who present with involuntary weight loss or hepatomegaly. Other clues to the diagnosis include an unexplained elevated serum ALP level and proteinuria. Primary systemic amyloidosis should be kept in mind within the differential diagnosis of patients with unexplained cholestatic jaundice.

REFERENCES