REFERENCES

1. Williams GT, Bussey HJ, Morson BC. Inflammatory 'cap' polyps of the large intestine. Br J Surg 1985; 72: S133.
2. Ng KH, Mathur P, Kumarasinghe MP, et al. Cap polyposis: further experience and review. Dis Colon Rectum 2004; 47: 1208-15.
3. Bookman ID, Redston MS, Greenberg GR. Successful treatment of cap polyposis with infliximab. Gastroenterology 2004; 126: 1868-71.
4. Tomiyama R, Kinjo F, Kinjo N, et al. Gastrointestinal: cap polyposis. J Gastroenterol Hepatol 2003; 18: 741.
5. Maunoury V, Breisse M, Desreumaux P, et al. Infliximab failure in cap polyposis. Gut 2005; 54: 313-4.
6. Konishi T, Watanabe T, Takei Y, et al. Cap polyposis: an inflammatory disorder or a spectrum of mucosal prolapse syndrome? Gut 2005; 54: 1342-3.
7. Oshitani N, Moriyama Y, Matsumoto T, et al. Protein-losing enteropathy from cap polyposis. Lancet 1995; 346: 1567.

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 yetmezlik 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/mm^3, Hct: %36, Plt: 487000/mm^3, 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. Biluribin: 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-

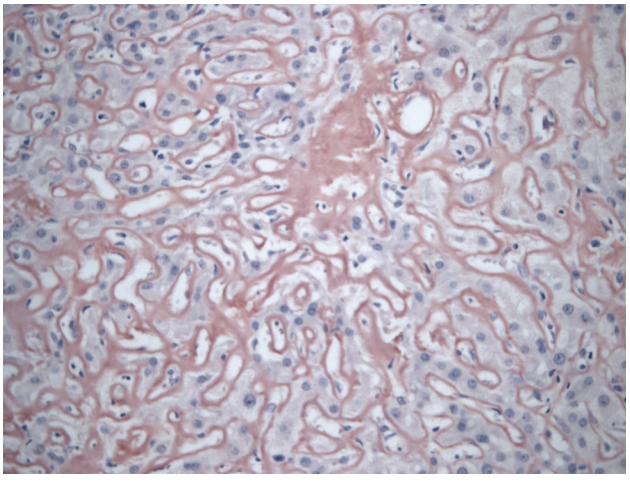

Figure 1. The pathologic specimen revealing: liver biopsy staining with Congo red and non-staining with potassium permanganate.

Address for correspondence: Bennur Esen GÜLLÜ
Bursa Yüksek İhtisas Training and Educational Hospital, Department of Nephrology, Bursa, Turkey
Phone: + 90 224 360 50 50 • E-mail: bennuresen@yahoo.com

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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 anticore. 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

1. Buck FS, Koss MN. Hepatic amyloidosis: morphologic differences between systemic AL and AA types. Hum Pathol 1991; 22: 904-7.
2. Bandyopadhyay SK, Bandyopadhyay R, Dutta A. Primary amyloidosis presenting as intrahepatic cholestasis. Indian J Pathol Microbiol 2006; 49: 557.
3. Dong MK, Cui Y, Xia H, et al. A case of primary hepatic amyloidosis with factor X deficiency. Zhonghua Gan Zang Bing Za Zhi 2006; 14: 684.
4. Hull KM, Griffith L, Kuncl RW, Wigley FM. A deceptive case of amyloid myopathy. Clinical and magnetic resonance imaging features. Arthritis Rheum 2001; 44: 1954-8.
5. Gertz MA, Kyle RA. Hepatic amyloidosis (primary [AL], immunoglobulin light chain): the natural history in 80 patients. Am J Med 1988; 85: 73-80.

Bennur Esen GÜLLÜ[1], İbrahim HATEMİ[2], Gülşen ÖZBAY[3], Nükhet TÜZÜNER[3], Abdullah SONSUZ[2]

Department of, [1]Nephrology, Bursa Yüksek İhtisas Training and Educational Hospital, Bursa
Departments of, [2]Gastroenterology, [3]Pathology, İstanbul University, Cerrahpaşa School of Medicine, İstanbul