Primary hepatic amyloidosis: A Case Report
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INTRODUCTION
Primary systemic amyloidosis AL is an uncommon disease characterized by the extracellular deposition of insoluble fibrils derived from immunoglobulin light chains. Widespread deposition of amyloid fibrils causes multisystem organ dysfunction. The heart, kidney, and peripheral nerves are most commonly affected by primary systemic amyloidosis. The liver is also a common site of amyloid deposition. The clinical manifestations are often and nonspecific and diagnosis of primary systemic amyloidosis may be overlooked.

We report a case of 68-year-old man who had biopsy-proven liver involvement, and describe his clinical presentation and contrast-enhanced computed tomographic findings, which was investigated in few studies 1,2.

CASE REPORT
In November 2005, a 68-year old man presented with mild right epigastric pain and abdominal fullness for 2 months was admitted to our department. He also had anorexia, pedal edema, and fatigue. On admission his blood pressure was 120/80 mmHg, heart rate was 75 beats/min, and temperature was 37°C. The physical examination showed the liver to be firm and enlarged accompanied by pedal edema, the spleen was not palpable.

On admission, laboratory data were as follows: red blood cell count, 5.3x10^9 cells/L; hemoglobin, 96 g/L; platelet count, 127x10^9 cells/L; serum urea, 8.2 mmol/L; serum creatinine, 98μmol/L;

Liver tests produced the following results: total bilirubin, 20 μmol/L; conjugated bilirubin, 11μmol/L; aspartate aminotransferase, 46U/L; alanine aminotransferase, 75 U/L; lactate dehydrogenase, 209 U/L; alkaline phosphatase, 396 U/L; l-glutamyl transpeptidase (GT), 310 U/L; Cholesterol, 3.2 mmol/L; prothrombin time, 17.8 s; partial thromboplastin time, 48.9 s; total protein, 65 g/L; and albumin, 31 g/L. Hepatitis B surface antigen and antihepatitis C virus antibodies were absent, as were antinuclear, antimitochondrial, and anti-smooth muscle antibodies, and tumor markers.

Urinalysis showed 2.24g of proteinuria every 24 h. Serum and urinary immunoelectrophoreses showed increase of monoclonal light-chain. In the serum, the immunoglobulin light-chain concentration was 239 g/L (normal, 0–18.5) and the immunoglobulin light-chain concentration was 184 g/L (normal, 0–50). And in the urine, the immunoglobulin light-chain concentration was 17.2 g/L (normal, 6.29–13.5) and the immunoglobulin light-chain concentration was 10.5 g/L (normal, 3.13–7.23).

The ECG on admission revealed left anterior bundle branch block and low ECG voltages 0.5mV mean ORS in limb leads I, II, III, aVL and aVF. Echocardiogram showed hypertrophy of left ventricular wall and an interventricular septum thickness of 12cm normal, 8-10.

Ultrasound examination of the upper abdomen showed a diffusely enlarged liver with heterogeneous echogenicity and no definitive evidence of focal lesions Fig. 1.
Abdominal enhanced-contrast CT scans revealed presence of a markedly acute left lobe hepatic margin and asymmetric hepatomegaly of triangular shape with an apex at the falciform ligament and heterogeneous attenuation, splenomegaly with diffuse hypoperfusion.

**Figure 2**
Figure 2: Pre-contrast CT scan showed asymmetric and triangular hepatomegaly with the apex at the falciform ligament (black arrow), mild atrophic change of the lateral border of both hepatic lobes (white arrow), splenomegaly with diffuse low density.

**Figure 3**
Figure 3: Arterial phases CT scan shows the lack of parenchymal enhancement in the liver and spleen.

**Figure 4**
Figure 4: Portal phases CT scan shows heterogeneous enhancement in the liver and spleen.
Hepatic needle biopsy specimen revealed an irregular expanded portal triad infiltrated with an amorphous eosinophilic material that stained with Congo red. Amyloid deposit along the sinusoidal wall and marked atrophy of the liver cell cord were observed. Bone marrow aspirate showed a small increase in the percentage of plasma cells 8%. These examinations confirmed the diagnosis of AL, which was classified as primary systemic AL.

DISCUSSION

Primary amyloidosis often involves the liver. Hepatomegaly and weight loss are the most common clinical manifestations in patients with hepatic amyloidosis. Previous study had reported that 27% of patients with primary systemic amyloidosis present with hepatomegaly, and that 81% of patients with primary amyloidosis with liver involvement have hepatomegaly while jaundice and portal hypertension.
Diagnosis of amyloidosis mainly relies on tissue confirmation by biopsy of affected organ. Diffuse infiltration of liver and spleen with amyloid materials will increase fragility of texture, and spontaneous rupture and massive hemorrhage may occur. So hepatic needle biopsy should be performed carefully in order to avoid possibility of serious complication. And alternative sites such as abdominal fat aspirate and rectal and labial salivary gland biopsies with a total sensitive of 80% have been recommended.

For primary amyloidosis, the mainstay of therapy has been chemotherapy, traditionally with melphalan and prednisone. But this therapy has only about a 30% response rate. High-dose chemotherapy followed by autologous peripheral blood stem cell transplantation ASCT has recently been deemed the treatment of choice, with a response rate from 50% to 60%. However, ASCT is associated to a relatively high treatment related mortality 13% to 14%. Patients with primary systemic amyloidosis who have biopsy-proven liver involvement (primary hepatic amyloidosis) have poor prognoses, the median survival of patients has been reported as 8.5 months.

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References
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