Hepatic amyloidosis

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INTRODUCTION

Hepatic amyloidosis is a rare disease that presents as an infiltrative disease involving liver. Amyloidosis is a systemic disease characterized by the extracellular deposition of amyloid protein in many organs.1-4 Progressive organ involvement leads to organ malfunction and death usually resulting from renal and/or cardiac involvement. Liver and spleen are major sites of involvement.1-5 The wide range of presenting symptoms encountered makes rapid clinical diagnosis difficult. None of the present imaging techniques is capable of specifically demonstrating the presence of amyloid. Even when suspected clinically and radiologically, the diagnosis of amyloidosis depends on a tissue biopsy to confirm the presence of amyloid deposits.5

To provide more detailed understanding of hepatic amyloidosis, here a case of amyloidosis involving liver is presented with radiological imaging findings including computed tomography (CT).

CASE

A 45-year-old female was admitted with the complaint of generalized edema. She got about 6 kg of weight gain during recent one week. She had dizziness, dyspnea, anorexia, nausea and epigastric discomfort. She presented physical examination findings including neck vein engorgement, abdominal distension, hepatomegaly and pitting edema. She had history of syncope about one month ago, but brain magnetic resonance imaging (MRI) showed no abnormal finding. The laboratory findings showed leukocyte count of 11,500/mm³, hemoglobin level of 9.1 g/dL and platelet count of 284,000/mm³. Biochemical tests showed that the serum level of alanine aminotransferase was 30 IU/L, aspartate aminotransferase was 55 IU/L, and alkaline phosphatase elevated up to 398 IU/L. Serum albumin was 3.7 g/dL, total bilirubin was 1.2 mg/dL, and creatinine was 2.3 mg/dL. Alpha fetoprotein was less than 1 ng/mL. The results for hepatitis B surface antigen and anti-HBs were negative. Urine analysis showed albumin positive, and otherwise negative.

Renal disease was suspected and abdominal ultrasonography was done. Liver showed diffuse coarse echo and hypoechogenic pattern with hepatomegaly (Fig. 1A). Both kidneys showed also diffuse increased cortical echo suggesting renal parenchymal disease (Fig. 1B).

Dynamic contrast enhanced CT was taken because of abnormal liver echo and size. On arterial phase, there was no enhancing lesion in liver (Fig. 2). On portal phase, size of liver increased, and especially left lobe showed bulging contoured enlargement and ill-defined hypoattenuation (Fig. 3A, 3B). There were multiple ill-defined hypoattenuated areas in right lobe. Spleen showed normal enhancement, but size increased. Both kidneys also showed multiple irregular decreased attenuation lesions (Fig. 3C). Because liver showed decreased echo on ultrasonography, it was assumed that it was not fatty infiltration. These CT findings suggested diffuse
infiltrative disease, so liver biopsy was done. Percutaneous fine-needle-aspiration biopsy was performed under ultrasound guidance, and a histological examination of the biopsy specimen showed amorphous material stained by Congo red stain. The deposit demonstrated characteristic positive birefringence with polarized light. Biopsy results led to a final diagnosis of extensive amyloidosis.

Additional examinations were done to evaluate the extent of amyloidosis involvement. 2D Doppler echocardiography showed enlarged both atrium and thickened left ventricle wall with normal ventricular contractility, which suggested infiltrative disease such as amyloidosis. Bone marrow aspiration and colonic mucosa biopsy showed also amyloidosis. Urine Immunoelectrophoresis showed abnormal zone of restriction in kappa light chain, suggesting Bence-Jones protein, free kappa type.

She was diagnosed as systemic amyloidosis, and chemotherapy was done. However, hepatic failure with hepatic encephalopathy was aggravated and she got discharged in a moribund state.

**DISCUSSION**

Amyloid is defined as a substance which: (1) stains positively with Congo red, (2) exhibits apple green birefringence by polarization microscopy, (3) shows aggregations of approximately 10 nm wide fibrils on electron microscopy, (4) exhibits a β-pleated sheet configuration, and (5) shows resistance to proteases other than pronase. Progressive deposition of amyloid compresses and replaces normal tissue, and this leads to organ dysfunction and a wide variety of clinical syndromes, some of which have severe pathophysiological consequences.

Amyloidosis is usually seen in a systemic form, but 10-20% of cases can be localized. Men are affected more than women, and the mean age of presentation is 55-60 years. Some causes of secondary amyloidosis are multiple myeloma (10-15%), rheumatoid arthritis (20-25%), tuberculosis (50%), or familial Mediterranean fever (26-40%). Approximately one-third of patients with primary amyloidosis develop congestive heart failure. It is the leading cause of death in patients with primary and myeloma-related amyloidosis.

In case of hepatic amyloidosis, amyloid is deposited in the parenchyma, along the sinusoids within the space of Disse, or in blood vessel walls. Hepatocytes are severely compressed by extensive accumulation of amyloid and they may atrophy or nearly disappear. In advanced cases with massive infiltration, the liver is enlarged with rubbery elastic consistency, and it