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CASE REPORT

CHOLESTASIS AND RENAL FAILURE IN A PATIENT WITH SECONDARY AMYLOIDOSIS

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ABSTRACT

Amyloid, was first described in the 19th century by Virchow, which means starch or cellulose. Extracellular deposition of this unique protein fibrils in tissues, often leading organ dysfunction is known as amyloidosis. In systemic amyloidosis, amyloid fibrils may deposit in different organs including kidneys, heart and liver. Although liver is effected frequently, clinical liver disease is rare. There are a few cases that secondary amyloidosis presented with hepatic involvement and cholestasis which seems to be limited to primary amyloidosis. Hepatic amyloidosis with cholestasis may be a predictor of involvement of other organs and thus of poor prognosis. We report a case of a 67 year old man with renal failure and prominent cholestasis due to AA amyloid deposition in the liver.

Key Words: Cholestasis; Renal failure; Amyloidosis

CASE REPORT

A 67-year-old man was admitted because of jaundice noticed one week before admission and oliguria. High blood pressure and abnormal renal function tests were found one month ago. At that time he had become oliguric and needed hemodialysis several times. The appearance of jaundice

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was one week before admission. He has no history of tobacco or alcohol use. In family history his brother has died due to chronic liver disease.

In physical examination he was icteric and vital signs were within normal limits. Hepatosplenomegaly and ascites were not found. Rare rhonchus was noticed in respiratory system and neurologic examination was normal.

In laboratory examination; Hemoglobin 9.3 g/dL, hematocrit 26.6%, white blood cells 13,400/mm^3 Blood urea nitrogen 44 mg/dL, creatinine 5.1 mg/dL, glucose 43 mg/dL, aspartate aminotransferase 54 IU/L, alanine aminotransferase 52 IU/L, total bilirubin 13.8 mg/dL, direct bilirubin 8.2 mg/dL, alkaline phosphatase 883 IU/L, gamma-glutamyl transferase 107 IU/L, albumin 1.8 g/dL and total protein 3.9 g/dL. Prothrombin time was 23.1 s (normal; 13 s). Daily proteinuria was 0.5 g. Antibody for hepatitis B surface antigen was positive. Cardiomegaly was noticed in telecardiogram. Electrocardiographic examination was revealed low voltage in all derivations. Computed tomography of the abdomen revealed bilateral pleural effusion, atelectasis and minimal ascites. Kidneys were bilaterally small and nonfunctioning. Liver, spleen and pancreas were normal. At follow up period his mental status worsened and he had became comatose. Hypotension and bradycardia developed. Despite of dextrose infusion serum glucose level spontaneously decreased to 40 mg/dL. Urinary output was about 400 mL per day. Clinical diagnosis was chronic renal failure and hepatic precoma with unknown etiology. The patient died, on the 9th hospital day in the hospital. Postmortem liver and myocardium biopsies revealed AA amyloid deposition.

DISCUSSION

Amyloidosis is described as extracellular deposition of amyloid fibrils in tissues. Any organ may be involved, leading a variety of clinical symptoms. Sometimes local depositions may be without any symptoms. Systemic amyloidosis is classified into primary and secondary forms. The classifications on the basis of the nature of the precursor plasma proteins that form the fibril deposits can be made.[6] Amyloid fibrils consist of lambda and kappa light chains in primary amyloidosis and there is no underlying disease. Secondary amyloidosis is commonly related to chronic inflammatory states like rheumatoid arthritis, inflammatory bowel disease and familial Mediterranean fever or unknown etiology. Amyloids are formed from serum amyloid A. In renal involvement nonselective proteinuria, nephrotic syndrome and renal failure are frequently seen. In 40–60% of cases with amyloidosis the cause of death is end stage renal failure. It is unusual for patients with either primary or secondary amyloidosis to manifest clinical liver disease or cholestasis. Amyloid deposition can be detected in 60–100% of liver specimens in

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patients with amyloidosis. Cardiac involvement is a common presentation of primary amyloidosis whereas rarely seen in secondary form. The deposition of amyloid in the myocardium can cause cardiomegaly or in severe cases congestive heart failure may be preceded with predominantly right-sided heart physical signs.[3,6] Respiratory system, skin, joints, central nervous system, endocrine system and hematopoietic system can be the sites for amyloid deposition.

We did not initially consider diagnosis of amyloidosis in our patient who has multiple problems like chronic renal failure, hepatic precoma, hypoglycemia and cholestasis. The short history of a decrease in urinary output may reflect the progressive renal failure. Although, in patients with hepatic failure having cholestasis, systemic amyloidosis may not be initially considered as a diagnosis, elevation of alkaline phosphatase level out of proportion to others and the pattern of hepatic infiltration may suggest it. Hepatic involvement of systemic amyloidosis rarely causes apparent liver disease clinically.[5] There are many case reports diagnosed as hepatic involvement of primary amyloidosis that has elevated alkaline phosphatase levels and cholestasis.[1,2,4,7] Hypoglycemia observed in the reported patient may be considered as a clue to the involvement of surrenal gland with amyloidosis or due to end stage hepatic failure. Cardiac involvement in systemic amyloidosis may cause cardiomegaly and congestive heart failure. In this case, cardiomegaly in telecardiogram and low voltage on ECG can be a clue of cardiac amyloidosis. Hypotension during the clinical course may be related to cardiac amyloidosis or surrenal insufficiency. Postmortem biopsy -revealed cardiac amyloidosis.

Hepatic involvement in amyloidosis is not infrequent but liver disease or cholestasis is rarely seen clinically. There are a few cases reported, which have hepatic involvement with AA Amyloidosis that it seems to be unique for the AL Amyloidosis.[5,6,7] The ability of diagnosis and treatment of disease are delayed because of progressive nature and rarity.[3] Hepatic involvement in systemic amyloidosis developing cholestasis may be a sign of involvement of other organs like heart or kidney and of poor prognosis.

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