A RARE CAUSE OF ACUTE LIVER FAILURE

BHARATH KUMAR AYAPATI ATYAGI

Department of Medical Gastroenterology,
CHRISTIAN MEDICAL COLLEGE

Abstract:
Hepatic involvement in systemic amyloidosis is common but is rarely symptomatic. It is usually in the form of hepatomegaly and asymptomatic elevation in serum alkaline phosphatase. Hyperbilirubinemia is rare and signifies worse prognosis. Hepatic failure due to systemic amyloidosis is very rare and it can manifest as acute or subacute liver failure. We report a case of a 55 year old man presenting with progressive jaundice followed within a month by ascites and encephalopathy. He was diagnosed as acute hepatic failure and had a rapidly worsening clinical course with fatal outcome. The diagnosis of amyloidosis was made on postmortem liver biopsy.

Keyword: Systemic Amyloidosis, Acute liver failure, Jaundice, Myeloma, Encephalopathy, Congo red staining

INTRODUCTION
The term ‘Amyloid’ was described first by Virchow in 1854 in a liver autopsy specimen. Amyloidosis is characterized by deposition of an abnormal proteinacious material, usually in the form of fibrils, in extracellular space. This leads to organ dysfunction. Heart and kidneys are the most common organs affected in amyloidosis.2 Liver and gastrointestinal tract involvement is less common and usually has mild symptoms.3 Hepatic involvement occurs in primary and secondary forms of amyloidosis4 and there are few case reports in literature of cases presenting as subacute, acute or fulminant liver failure, particularly in primary amyloidosis(AL) in the setting of myeloma.5–8

Herein, we present a rare case of primary amyloidosis (AL) presenting as acute liver failure.

CASE REPORT:
A 55 year old man from Assam presented with progressive shortness of breath of five months duration without associated orthopnea or paroxysmal nocturnal dyspnea. Three months later, he noticed progressively worsening jaundice without cholestatic symptoms, abdominal pain or fever. After 3 weeks of onset of jaundice, he developed worsening abdominal
One day prior to presentation, he developed drowsiness. He received native medications for few weeks after onset of jaundice. He had diabetes of two years duration and was on oral antidiabetic drugs. There was no history of alcohol intake. On physical examination, he was conscious and oriented. He was pale, icteric and had bilateral pitting pedal edema. He had tachycardia with a blood pressure of 90/60 mm of Hg at admission. There was free fluid in the abdomen with shifting dullness. Heart sounds were muffled and he had asterixis. Laboratory tests showed a haemoglobin of 8.7 gm/dL, total leucocyte count of 21,300/cu.mm and a platelet count of 2,58,000/cu.mm. Serum bilirubin was 21.3 mg/dL with a conjugated bilirubin of 17.5 mg/dL. Serum total protein and albumin were 5.8 gm/dL and 2.8 gm/dL respectively. SGOT and SGPT were 80 U/L and 14 U/L. Alkaline phosphatase was 453 U/L and GGT was 145 U/L. Serum creatinine was 3.9 mg/dL with a serum potassium of 6 mg/dL. Ascitic fluid evaluation showed a total leucocyte count of 310 cells/cu.mm with 96% lymphocytes. Ascitic fluid protein and albumin were 2.2 gm/dL and 1.4 gm/dL with a serum ascitic fluid albumin gradient(SAAG) of 1.4. Serology for HIV, Hepatitis B,C,A and E viruses were negative. Serological tests for autoimmune hepatitis (ANA, ASMA, Anti LKM, Anti SLA/P) were negative. Urine for Bence Jones protein was positive and serum protein electrophoresis showed a faint peak in gamma region. Bone marrow biopsy showed mild hypercellular marrow with 12% plasmacytosis including atypical forms. Cultures for mycobacteria and fungi were negative. Upper GI endoscopy showed features of severe portal hypertensive gastropathy.

Chest radiograph showed cardiomegaly and bilateral pleural effusion. ECG showed trifascicular block and 2D Echo showed moderate pericardial effusion with no tamponade and mild diastolic dysfunction. There was hepatosplenomegaly, ascites and grade I renal parenchymal changes on ultrasonography. He had rapidly worsening sensorium and hypotension and required inotropic support and mechanical ventilation. He had intermittent bradycardia for which a temporary pacemaker was inserted. The relatives opted for withdrawal of supports. A post mortem liver biopsy was performed and it showed deposition of pink amorphous material with congo red birefringence and a diagnosis of acute liver failure due to systemic amyloidosis with cardiac and renal involvement was made. The histological findings are shown in figures 1 and 2.
Liver- Hematoxylin and Eosin staining showing deposition of pink amorphous material (Amyloid) in the extracellular spaces and atrophy of hepatocytes

DISCUSSION:
Amyloidosis is caused by extracellular deposition of fibrils composed of low molecular weight subunits (most of which are in molecular weight range of 5-25 kD) of a variety of proteins, many of which circulate as constituents of plasma. Amyloidosis is currently classified based on the chemical characterization of precursor protein and around 30 protein folding diseases exist, all characterized by a specific soluble precursor protein that aggregates in the form of insoluble fibrils. The most common among them are AL and AA types. Myeloma associated (AL) (primary) amyloidosis is the commonest and is associated with plasma cell dyscrasias and malignant B-cell lymphomas. It is characterized by deposition of the variable region of the immunoglobulin kappa or lambda light chains. The second most common is Amyloid-associated (AA) (secondary or reactive) amyloidosis, which is seen in chronic infectious and inflammatory conditions, Hodgkin’s lymphoma and other malignancies and characterized by deposition of amyloid A fibrils, which are derived from serum AA precursor protein. In a Mayo clinic series, 474 patients with AL amyloidosis were reviewed. The most common symptoms were weakness, fatigue and weight loss. The most common physical findings was hepatomegaly (24%). Elevated serum alkaline phosphatase was noted in 25% of patients but hyperbilirubinemia was rarely seen and carried a bad prognosis. Hepatic failure is a rare consequence of hepatic amyloidosis and cases of fatal subacute and acute hepatic failure were previously reported in association with both types of amyloidosis. The treatment options of AL amyloidosis include melphalan, prednisolone along with consideration of liver transplantation in cases of liver failure. In conclusion, though amyloidosis rarely presents with hepatic failure, it should be considered in patients with hepatic decompensation especially if cardiac and renal involvement are present.

BIBLIOGRAPHY:


