Primary Biliary Cirrhosis — A Case Report

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INTRODUCTION

Primary Biliary Cirrhosis (PBC), a disease of intrahepatic bile ducts, is a relatively uncommon disease even in the West. In India only a few cases have been reported so far1,2. Intense pruritus and increasing skin pigmentation are the usual presenting features with development of jaundice at a later stage3. In this report, we describe a patient with primary biliary cirrhosis, who presented with recurrent bleeding from oesophageal varices.

CASE REPORT

A 55 year old housewife from Bihar presented with recurrent episodes of upper gastrointestinal bleeding for two years. There was no history of jaundice, pruritus, pedal oedema, abdominal distension, abdominal pain or joint pain. Physical examination revealed mild pallor and a firm hepatomegaly. There was no evidence of ascites or peripheral signs of liver cell failure. Her initial investigations were as follows: Total serum bilirubin 0.6mg%, direct 0.2mg%, alkaline phosphate 1634 μ/1, SGOT 126 μ/1, SGPT 72 μ/1, Serum total protein 9gm%, Albumin 2.9 gm% Globulin 6.1gm%, HBsAg negative, Serum cholesterol 219 mg%, Serum triglycerides 75mg%. Her serum was positive for antinuclear antibody and rheumatoid factor, but was negative for LE cells. Gastroscopy revealed varices in the oesophagus and gastric fundus. After initial assessment, she underwent a percutaneous needle biopsy of the liver, the features of which are described later. Following the biopsy report, she was investigated further and found to be positive for antimitochondrial antibody. Her serum immunoglobulins were also markedly elevated, with an IgG of 3.4gm%, IgA of 566 mgm% and IgM of 650 mgm%.

Pathology

Microscopically, the needle biopsy sample of the liver revealed features of primary biliary cirrhosis in stage 2-3, characterized by a dense chronic inflammatory cell infiltrate in the portal tracts consisting predominantly of lymphocytes and with complete destruction of bile ducts (Fig. 1 & 2). There was extension of the inflammation into the periportal parenchyma, forming linkages between the adjacent portal tracts. Bile ductular proliferation was noted at the periphery of some of the portal tracts. Epithelioid cell granulomas were not present in serial sections. There was no evidence of cirrhosis. Bile stasis was not conspicuous. There was no significant abnormality in the rest of the liver parenchyma except for focal liver cell necrosis and mild Kupffer cell

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hyperplasia. Accumulation of copper and copper associated protein was seen in several periportal hepatocytes (Fig. 3).

DISCUSSION

The diagnosis of PBC was made in this case on the basis of clinical biochemical and histological features. PBC occurs mostly in middle aged women. Although pruritus and skin pigmentation are the usual presenting features, in some patients the initial presentation may be bleeding from oesophageal varices.\(^4,5\)

On the other hand, some patients can remain asymptomatic for many years and the incidental finding of hepatomegaly, a raised serum alkaline phosphatase or antimitochondrial antibody may result in recognition of the disease.\(^3\) Skin xanthomas can occur in patients with hypercholesterolaemia. PBC can be associated with other immunological abnormalities.

The most helpful diagnostic test is the demonstration of serum antimitochondrial antibody which is present in more than 95\% of cases. Serum immunoglobulins of all types are raised but elevation of IgM is most consistent.\(^3\) Histologically, the evolution of the disease is characterised by increased periportal inflammation with fibroinflammatory septum formation, and eventual progression to cirrhosis which is predominantly micronodular.

The presence of periportal inflammation with bridging as seen in this case can lead to a mistaken diagnosis of chronic active
hepatitis (CAH). However, a diagnosis of PBC rather than CAH is indicated by the presence of bile duct epithelial damage with complete destruction of bile ducts, prominent lymphoid infiltrates or epithelioid cell granulomas in the portal tracts, bile ductular proliferation, absence of significant lobular changes and a variable pattern of the disease process within the liver.

Although a histological staging has been described in PBC, there is frequently a poor correlation between the clinical state and the histological staging. As the evolution of the disease is variable within the liver and there is individual variation in progression of the disease, a histological staging in a given needle biopsy specimen may not represent the entire spectrum of the disease. To date there have been only isolated reports of PBC from India\textsuperscript{1,2}. On the other hand, increasing awareness of this disease has resulted in recognition of a number of cases in recent years, in a country where this disease was previously considered uncommon\textsuperscript{3}.

The treatment of PBC is different from that of chronic active hepatitis, in particular steroids being contraindicated in the former disease\textsuperscript{4}. It is therefore important to consider the diagnosis of PBC in the appropriate clinical setting, when histological features are consistent and antimitochondrial antibody is present.

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Accumulation of copper associated protein shown as black granular deposits in periportal hepatocytes. (Shikata’s orcein x 315)

REFERENCES


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