Case Report

Its Congenital Hepatic Fibrosis; Not Cirrhosis At All

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ABSTRACT

Congenital hepatic fibrosis is a rare condition characterized by extensive fibrosis of liver but with preserved normal lobular architecture inherited as autosomal recessive trait. We report a 19 year old female admitted to Bangabandhu Sheikh Mujib Medical University with the complaints of lump in upper abdomen since last 13 years and episodes of fever and abdominal pain for same duration. She was diagnosed with hepatic TB on hepatic histology. Congenital hepatic fibrosis is a rare cause of portal hypertension that presents during childhood. Prognosis of congenital hepatic fibrosis is good. Life threatening events in these patients are related with variceal bleeding and episodes of cholangitis. Owing to relatively good liver function these patients tolerate portosystemic shunt surgeries quite well. Though rare, congenital hepatic fibrosis should be included in the differential diagnosis of portal hypertension in early life.

Key words: Fibrosis; Ductal plate malformation; Portal hypertension

INTRODUCTION

Congenital hepatic fibrosis is a rare condition characterized by extensive fibrosis of liver but with preserved normal lobular architecture, often occurring in association with cystic lesions of the kidney. It is inherited as autosomal recessive trait. The underlying abnormality lies in the development of biliary ductules, classically described as “Ductal Plate Malformation”.1,2 It commonly presents in childhood with features of portal hypertension nevertheless incidental diagnosis in adulthood or even autopsy diagnosis has been reported.1 Though frequently confused with liver cirrhosis2, congenital hepatic fibrosis do not present with feature of hepatocellular insufficiency.1 Absence of signs of liver failure may rather indicate possibility of congenital hepatic fibrosis in those who are mistakenly considered to have liver cirrhosis. Here we report a case of congenital hepatic fibrosis that was misdiagnosed and treated as a case of liver cirrhosis for 13 years.

CASE REPORT

A, 19 year patient was admitted to Bangabandhu Sheikh Mujib
Congenital hepatic fibrosis is a rare cause of portal hypertension that presents during childhood. Consequently, the manifestation is mainly that of portal hypertension and is devoid of features of hepato cellular failure. Primary defect lies in the involution of ductal plates that forms bile ducts resulting into persistence of the ductal plate with an increase in duct elements and an increase in portal fibrous tissue called as Ductal Plate Malformation. Though the underlying pathogenesis mainly involves ductal system, the portal fibrous tissue called as Ductal Plate Malformation. Of note, the fibrotic bands contain abnormal or ectatic bile ducts and numerous portal vessels commonly called as Von Meyer’s Complexes.²

Seventy seven percent of patients with congenital hepatic fibrosis have esophageal varices at presentation.³ Rupture of esophageal varices and subsequent bleeding is the commonest presentation. However other presentations like cholangitic and mixed cholangitic and portal hypertensive varieties also occur. Association with intra hepatic choleodochal cyst has been seen and such cases are termed as Caroli’s syndrome.³ Medullary sponge kidney and polycystic kidney disease are the most common associated abnormalities. Presentation in such association is further complicated by occurrence of hypertension, renal disease, pulmonary emphysema, cerebellar hemangioma and berry aneurysms of the arterial system of brain.¹

Congenital hepatic fibrosis can have a wide variety of manifestations and presentations.

Prognosis of congenital hepatic fibrosis is seen to be good.⁴ Life threatening events in these patients are related with variceal bleeding and episodes of cholangitis.⁵ Owing to relatively good liver function these patients tolerate portosystemic shunt surgeries quite well. But reports of development of nodular transformation later in life cannot be neglected.⁷ Other factors that may guard prognosis are associated renal disease and their complications.

In our case, the suspicion of congenital hepatic fibrosis was...
raised when polycystic kidneys were noted in association with long standing portal hypertension and relatively preserved liver function. Repeated episodes of abdominal pain and fever in our case were later related with cholangitis. The reason she did not bleed from varices may be that she was prescribed oral propranolol since detection of her varices and this might have masked the presentation. The case we report here was mixed cholangitic and portal hypertensive type. Her variceal size was medium sized and no associated red weal markings were noted. In background of preserved liver function her variceal characteristics appeared benign and were controlled by propranolol alone. ERCP to visualize biliary tree can sometimes detect biliary abnormalities that are inapparent on other imaging techniques. Cholangiogram however is also known to be an inciting event that may trigger overt manifestations and complications in other wise asymptomatic individuals. This fact together with lack of consent for the procedure, ERCP was not done in our case. Recurrent cholangitis in this patient was also benign and patient was doing well with oral antibiotics (ciprofloxacin) during such episodes. Repeated episodes of cholangitis and ectatic biliary tree may precipitate biliary stones. Chenodeoxycholate have been recommended for preventing biliary stones. Treatment with ursodeoxycholic acid (UDCA) for cholangitic variety of congenital hepatic fibrosis is also recommended by some authors. It may also improve bile salt dependent bile flow and prevention of biliary stone formation. Further management plans in this case include regular surveillance of varices and management of cholangitic episodes along with prophylaxis for biliary stone formation.

CONCLUSIONS

Though rare, congenital hepatic fibrosis should be included in the differential diagnosis of portal hypertension in childhood. Relatively preserved liver function and hard non-tender hepatomegaly, splenomegaly along with ballotable kidneys should further obviate the need to consider this diagnosis.

Acknowledgements: We thank patient party who gave approval for publication of the case.

REFERENCES