Case report
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First Living Donor Liver Transplantation For Congenital Hepatic Fibrosis In Azerbaijan.

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Congenital hepatic fibrosis (CHF) is a rare autosomal recessive disease. Cases have been reported from all over the world but the exact incidence of the disease is not known. The diagnosis sometimes is difficult to establish and one of the main diagnostic method is histological evaluation. The management and prognosis of CHF is dependent on alimentary bleeding secondary to portal hypertension. In late childhood abdominal pain, cholangitis and features of hypersplenism complicate the problem. Herein we present the case report of patient with CHF. Our choice of treatment was living donor liver transplantation. This procedure is a very difficult but only life – saving chance for patients with CHF.

Keywords: liver fibrosis, living donor liver transplantation, congenital hepatic fibrosis, chronic liver failure

Introduction

Congenital hepatic fibrosis (CHF) is an unusual condition in which portal hypertension (PH) occurs without significant hepatic or renal functional impairment and characterized histologically by defective remodeling of the ductal plate. CHF is a subtype of group of congenital disorders described as fibropolycystic disease with a wide clinical spectrum depending upon the time of presentation and degree of hepatic involvement. Herein we report a case of living donor liver transplantation for patient with CHF.

Case Report

An 21-year-old man was admitted with slowly progressive distension of abdomen and fullness in upper abdomen of 7 months duration, and history of 3 time hematemesis during last 7 months and 3 times of EVL. There was no history of pain abdomen, jaundice, or any skin bleeds or hyperpigmentation. On tracing the pedigree no other family member was known to be affected. The man weighed 62.7kg, with a height of 167 cm. Body mass index was 22.5. General examination revealed pallor and conjunctival xerosis without any signs of liver cell failure or icterus. Temperature, pulse and BP were normal. On abdominal examination spleen measured 9 cm below costal margin with tip below umbilicus without signs of hypersplenism, liver span was 6 cm with no evidence of free fluid in the abdomen. Kidneys were not palpable and other systemic examination was normal. On investigations, hemoglobin was 10.5 g/dl, total leukocyte count was 3060/mm3, platelet count was 106000/mm3, and peripheral blood smear revealed thrombocytopenia, leukopenia, normal erythrocytes and no malarial parasite (MP). Liver function tests revealed total bilirubin of 1.1 mg/dl and serum aspartate transaminase was 9.6 IU/ L, serum alanine transaminase was 10.5 IU/L and alkaline phosphatase was 75 IU/L.

Bone marrow aspiration showed erythroid hyperplasia with normoblastic reaction and no abnormal cells, LD bodies or MP. HBsAg, anti HCV, ANCA, ASMA and serol-

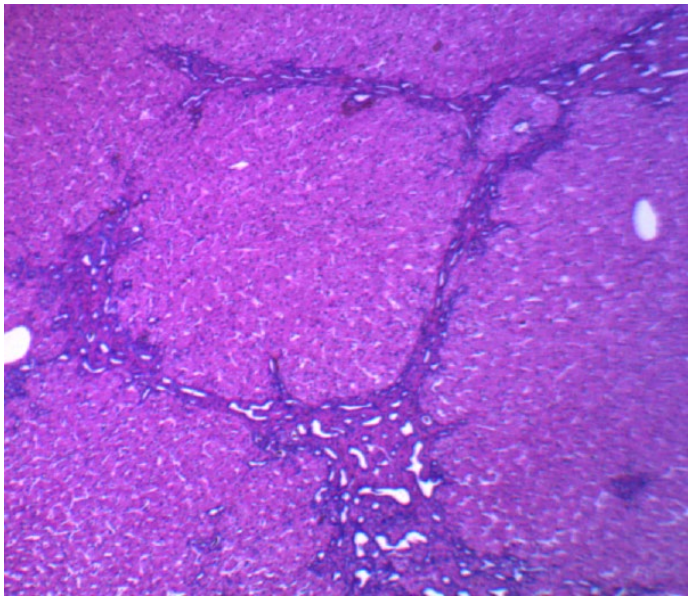

Figure 1. Histological examination. Liver tissue with distorted architecture composed of nodules of different sizes surrounded by fibrous septa.

ogy malaria were negative. Alpha 1 antitrypsin was 2.01 gr/L. Ultrasound of abdomen showed normal liver, kidneys and normal caliber of portal vein with no evidence of ascites. Upper GI endoscopy revealed grade III esophageal varices. Slit lamp examination of eyes was normal. Prothrombin time and INR were also within normal limits. Abdominal contrast CT shows the signs of chronic liver decease without any mass and splenomegaly, the dilated intrahepatic bile ducts particularly in right lobe and many portocaval shunts especially around spleen.

Liver biopsy showed liver tissue with distorted architecture composed of nodules of different sizes surrounded by fibrous septa. On fibrous septa dilated bile ducts and marked cholangiolar proliferation was seen. Inflammatory infiltration is minimally on fibrous septa. There was focal mild dilatation of interlobular ducts. These histological features confirmed the diagnosis of CHF and possibility of inactive cirrhosis. During the hospital stay, the patient remained asymptomatic and had no evidence of active bleeds.

This patient was treated by living donor liver transplantation. Procedure was performed without any deviations from standard technique.

Discussion

Congenital hepatic fibrosis is a rare autosomal recessive disease named by Kerr in 1961. Clinically reserved for a condition in which PH occurs without significant impairment of liver or kidney function [1]. Cases have been reported from all over the world [2-7] but the exact incidence of the disease is not known. CHF has usually autosomal recessive inheritance and initial presentation may be at around 3-6 months. The presentation ranges between 1.8-14 years [8], PH is a usual accompaniment and renal involvement is seen with < 10% tubules being affected. Classically affected patients are asymptomatic until the age of 5 or 7 years when manifestations of PH or cholangitis lead to the diagnosis. Several clinical forms are described which depend on the variable predominance of PH and cholangitis. Cholangitis form of CHF is more severe and usually occurs in late childhood and adult life [9]. Blyth and Ockenden(10) have divided their patients into 4 groups called perinatal, neonatal, infantile and juvenile in accordance with the age at clinical presentation. Renal involvement is maximal in perinatal group and minimal in juvenile group. Our patient had presented with PH, with no clinical or histological evidence of cholangitis and renal abnormalities.

The usual presentation of CHF is with abdominal distension(4), hematemesis or melena, failure to thrive, jaundice, anemia, hepatomegaly and splenomegaly [1,8]. The other features of CHF are abdominal pain (splenic infarction), fever (cholangitis in dilated ductules), ascites, etc. [2-5]. CHF is particularly associated with infantile polycystic kidney disease or intrahepatic bile duct dilatation (Caroli's disease) [1]. The diagnosis is based on liver functions which are well preserved, features of hypersplenism, elevation in levels of alkaline phosphatase and gamma glutamyl transferase [1, 5]. Other associated disorders with CHF are medullary sponge kidney, Ivemark's Familial Dysplasia, Meckels syndrome, vaginal atresia and rarely adult type polycystic kidney disease or nephronopthises, Jenunne's syndrome, tuberous sclerosis, etc. These conditions were ruled out in our case by absence of other clinical features/malformations associated with these conditions and relevant investigations. Biliary hamartomas (von Meyenberg complexes) are frequently associated with CHF and are detected on histology and by imaging. Hallmark of diagnosis is liver biopsy which shows bands of fibrous tissues often containing linear or circular spaces lined by cuboidal epithelium. There is diffuse portal and perilobular fibrosis varying in thickness but it does not distort lobular structures. The limiting plate is intact and parenchyma is separated by islands of fibrosis. There are no inflammatory changes and regenerative nodules are absent or few [8]. The cholangitis form of CHF is difficult to differentiate from Caroli's disease characterized by nonobstructive dilatation of intrahepatic bile ducts occurring as an isolated abnormality without portal fibrosis. This suggests a spectrum of congenital biliary tree disease with portal fibrosis and normal caliber ducts at one end and multiple intrahepatic, even extrahepatic, dilatations without fibrosis at the other end. Overlapping of CHF and Caroli's disease has been confirmed by histological studies [9].

The management and prognosis of CHF is dependent on alimentary bleeding secondary to PH. In late childhood abdominal pain, cholangitis and features of hypersplenism complicate the problem. However, prognosis may be greatly improved by shunt surgery but survival in some patients may be limited by degree of renal failure (I). In our case choice of treatment was living donor liver transplantation.

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