CAROLI DISEASE AND CAROLI SYNDROME

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Introduction

Caroli syndrome consists of Caroli disease and congenital hepatic fibrosis [1]. It is an uncommon congenital disorder of the intrahepatic bile ducts. The disease is presumably of autosomal recessive hereditary character [2]. A classical case history of the Caroli syndrome is presented. Non surgical management with endoprosthesis is discussed.

Case Report

A 10-years-old boy was admitted with complaints of pain in the upper abdomen, fever, vomiting and lump abdomen of 2 1/2 months duration. There was no family history of hepatic disease. He was poorly developed. Weight was 20 kg and height was 128 cm. He was febrile (102 °F). No icterus was present. The abdomen was full in upper part. The liver was firm, nontender and palpable 7 cm below the right costal margin. Features of portal hypertension in the form of ascites, splenomegaly and distended abdominal wall veins were present. Laboratory investigations revealed Hb 6.2 gm%, TLC 12,200/mm³; Liver function tests: S.bilirubin 2.8 mg%, ALT 21 IU/L, AST 56 IU/L, ALP 89 IU/L, Total proteins 8.5 gm/dl, Albumin 3.0 gm/dl, Globulin 5.6 gm/dl and A:G Ratio 0.55:1. HBsAg was negative by RPHA. Serum Amylase was 92 Somogyi Units. Ultrasonography revealed multiple cystic dilatations in both lobes of liver and both kidneys (renal polycystosis). Computed tomography (Fig 1) and endoscopic retrograde cholangiography (Fig 2) confirmed the presence of multiple communicating dilatations of the bile ducts affecting the whole of the liver but mainly of right lobe of liver. Patient was managed with nasogastric aspiration, broad spectrum antibiotics, blood transfusions and other supportive measures. Endoscopic papillotomy and nasobiliary drainage (endoprosthesis) was carried out to decompress the biliary tree. The cholangitis resolved and patient is in the waiting list for liver transplantation.

Discussion

Suspicion of Caroli disease is delayed owing to its rarity. It is characterized by ectasia of the intrahepatic bile ducts without other abnormalities. In Caroli syndrome, congenital intrahepatic ductal dilatation is associated with features of congenital hepatic fibrosis

and polycystic kidney [2]. Caroli disease and Caroli syndrome are inherited in an autosomal recessive fashion. The aetiology of Caroli disease is unknown. The ductal plate malformation theory and infantile obstructive cholangiopathy theory have been postulated. Male and females are equally affected.

The clinical features reflect recurrent bouts of cholangitis due to bile stasis. The classical features of this entity are well exemplified by our case. The child had scattered intrahepatic dilated communicating cysts with features of congenital hepatic fibrosis in the form of ascites, splenomegaly and distended abdominal wall veins. Ultrasonography, computed tomography scan, percutaneous transhepatic cholangiogram or endoscopic retrograde cholangiographic patterns are usually diagnostic of Caroli syndrome by showing the dilatation of the first and second order branches of the intrahepatic biliary ducts.

Fig 1 : Computed tomography scan of liver demonstrating multiple cysts scattered through out the whole liver. A single large cyst is seen in the postero-inferior aspect of the right lobe of the liver.

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Fig. 2 : Endoscopic retrograde cholangiography showing multiple communicating dilated intrahepatic bile ducts mainly in the right lobe of the liver

Definitive therapy for this disease is limited. Large cysts may benefit from internal or external drainage

procedures. In this case we did endoscopic retrograde biliary drainage by keeping a nasobiliary stent in the right hepatic duct. Hepatic lobectomy is performed for isolated lobar disease. For panhepatic disease, orthotopic liver transplantation is the only therapeutic option.

Caroli syndrome should be included in the differential diagnosis of chronic cholestasis of unknown cause. If recurrent cholangitis is frequent inspite of medical therapy, the outlook is poor and a significant number of patients die 5 to 10 years after the appearance of cholangitis [3]. Approximately 7% frequency of cholangiocarcinoma has been reported complicating this condition [4].

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