

Case Reports

Forme Fruste Choledochal Cyst

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Two girls presenting with features of pancreatitis were diagnosed to have minimal dilatation of extra hepatic biliary duct (EHBD) associated with pancreatico-biliary maljunction (PBMJ). A high degree of suspicion is required to diagnose this condition that has been termed Forme fruste choledochal cyst (FFCC). Both did well with pancreatico-biliary disconnection procedure and reconstruction of biliary channel using enteric conduit.

Key words: Choledochal cyst, forme fruste choledochal cyst, long common pancreatico-biliary channel, pancreatico-biliary maljunction.

Lilly *et al*(1) 1985 described four patients having stenosis of the distal common bile duct, a 'long common channel' secondary to a proximal junction of the common bile and pancreatic ducts, cholecystitis and the classical pathological microscopic features of choledochal cyst in the wall of the common

bile duct and coined the term "forme fruste choledochal cyst" (FFCC). Okada, *et al.*(2), however, had probably described the same entity four years earlier and had termed it as the "common channel syndrome". The definition of FFCC has been refined in recent years and now it is known as a variant of a choledochal cyst that has minimal or no dilatation of the extrahepatic bile duct (EHBD) and is associated with pancreatico-biliary malunion (PBMU)(3-5). We report two young girls having FFCC who were successfully treated by excision of the diseased extra-hepatic biliary duct and hepaticoduchojejunostomy and review the literature briefly.

Case Report

Case 1

A six-year-old girl presented with recurrent episodes of upper abdominal pain, fever, vomiting since last three years. She never had clinical jaundice in any of these episodes. The symptoms would recur every three months or so and would be severe enough to necessitate hospitalization, administration of intravenous fluids and antibiotics. The liver enzymes were found only minimally raised, but this biochemical abnormality was consistently noted. She had undergone repeated abdominal ultrasonography, barium meal with follow through and even barium enema elsewhere for the same; no clear justification of the latter studies was forthcoming. One of the abdominal ultrasonography was reported as choledochal cyst, but no definite details or pictures were available. A clinical diagnosis of relapsing pancreatitis was made; the serum amylase during the present hospitalization was

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minimally raised (449 IU/L; normal values <130-400 IU/L).

Magnetic resonance cholangio-pancreaticography (MRCP) revealed a focal minimal dilatation of common bile duct just distal to confluence of cystic duct and hepatic duct, normal distal common bile duct (CBD), pancreatic duct (PD) and intrahepatic biliary radicals (IHBR). Endoscopic retrograde cholangio-pancreaticography (ERCP) performed in the quiescent phase revealed minimal dilatation of the common hepatic duct long common pancreatico-biliary channel; CBD, PD and IHBR were normal.

Diagnosis was revised to forme fruste choledochal cyst. Excision of extrahepatic biliary duct system and Roux-en-Y hepatico-jejunostomy was performed. The histopathology of the excised biliary duct showed dense connective tissue mild inflammatory infiltrate and patchy loss of the biliary epithelial lining. The child has been doing well three and half years post-operatively on follow up.

Case 2

A ten-year-old girl presented with recurrent episodes of epigastric pain, since last three years. There was no history of vomiting or fever, though on close probing, history of transient self-limiting jaundice three years ago could be elicited. The child was being treated for gastritis elsewhere Examination was unremarkable. LFT were normal. Abdominal ultrasonography showed dilated CBD with a normal liver. MRCP revealed minimal dilatation of EHBD and long common pancreatico-biliary channel.

A pre-operative diagnosis of forme fruste choledochal cyst was made. Excision of extrahepatic biliary duct system and Roux-en-Y hepatico-jejunostomy was performed. The

histopathology was consistent with the clinical diagnosis. The child is under close follow up and has been doing well one year post-operatively.

Discussion

FFCC represents 4-21% of all choledochal cysts(1-4). The presenting symptoms include recurrent abdominal pain, recurrent jaundice, fever and pancreatitis and these closely resemble those seen in patients with classical choledochal cyst(1-5). The differentiation of FFCC from choledochal cyst is essentially done on findings of CECP or intraoperative cholangiopancreaticogram. The role of MRCP has been not reported specifically for the diagnosis of FFCC hitherto, though its use in the diagnosis of choledochal cyst has been well established. There is scant literature available on normal dimensions of extra hepatic biliary duct (EHBD) and the common pancreaticobiliary channel in children. The available literature mentions the maximum normal diameter of EHBD in children to vary from 3 to 6 mm(6). The cut off diameter above which the diagnosis of FFCC is unacceptable has been arbitrarily decided as 10 mm previously(5). The maximum normal length of the common pancreaticobiliary channel in infants has been reported as 3 to 4 mm; it increases with age to a maximum of 5 mm in adolescents(7). The demonstration of a longer common pancreaticobiliary channel for a particular age with EHBD measuring less than 10 mm on ERCP or intraoperative cholangio-pancreaticogram clinches the diagnosis of FFCC(5).

Other subtle differences that have been noted between FFCC and choledochal cyst include higher incidences of dilatation of common pancreaticobiliary channel and presence of protein plugs or debris at the level of common channel in FFCC as compared to the classical choledochal cysts(3). Dilatation

of the common pancreaticobiliary channel that was seen in as many as two-thirds of the patients of FFCC in one series(3). A dilated common channel is believed to be related to the development of protein plugs and pancreatitis with pancreaticolithiasis(8).

The treatment of choice for FFCC in children is EHBD excision and hepatico-jejunostomy. Although open sphincteroplasty and endoscopic sphincterotomy have also been reported as treatment modalities for this condition(9), it is not favored because the anatomical abnormality allowing mixing of biliary and pancreatic secretions in the two ductal systems is not fully corrected unless a pancreaticobiliary disconnection is done. Similarly, a simple cholecystectomy as prescribed for PBMJ in adults is not justified in children(10).

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