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CASE REPORT

Forme Fruste Choledochal Cyst – a single centre study with review of literature

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Abstract:
Forme fruste choledochal cyst (FFCC) is a rare disease which needs high index of suspicion for diagnosis. We are presenting 3 cases of FFCC managed in our department in last 2 year. In one case FFCC was suspected on clinical evaluation and diagnosis was confirmed by magnetic resonance cholangiopancreatography and endoscopic retrograde cholangiopancreatography whereas in rest, diagnosis was made on clinical suspicion and confirmed by MRCP and peroperative cholangiogram. We are presenting series of three cases of FFCC, because it is a rare disease and need high index of suspicion for its diagnosis and once diagnosed it can be cured.

Keywords: *Choledochal cyst, pancreatitits, surgery*

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Introduction

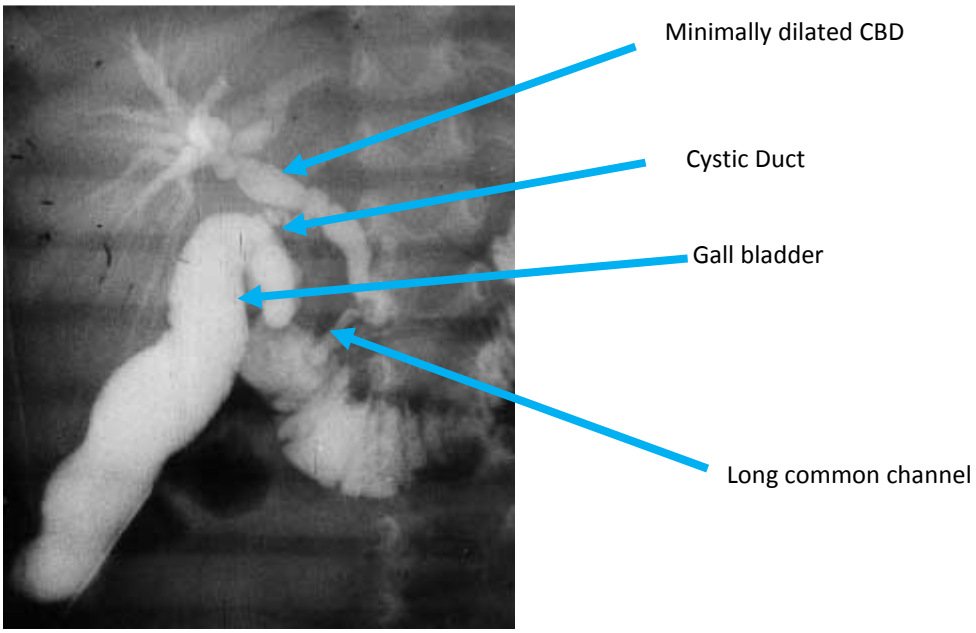
Choledochal cysts are rare disease of unknown cause and their presentation with the classical triad of pain, jaundice, and mass is rarely seen (1). They usually present with vague and nonspecific symptoms, impeding prompt diagnosis. Lilly *et al*(2), described four patients with stenosed distal common bile duct, with a 'long common channel' secondary to an abnormal junction of the common bile duct and pancreatic duct. These patients had classical pathological microscopic features of choledochal cyst in the wall of the common bile duct for which they coined the term "forme fruste choledochal cyst". The definition of "forme furste choledochal cyst" (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 extra-hepatic bile duct (EHBD) with associated pancreatico-biliary malunion (PBMU) (3-4). We have managed three cases in last 2 years and presenting them in view of clue for diagnosis and management in light of existing knowledge.

Case report

Case – 1: A four and half year old male child, presented with history of pain in upper abdomen, colicky in nature not associated with vomiting and occurs for 20 min every 3-4 hourly and subsides by its own. Abdominal examination showed mild hepatomegaly. Liver function revealed; total bilirubin of 3.2 gm%, direct 2.1 gm%, mild increase in SGOT and SGPT (88 and 76 U/I respectively) with raised alkaline phosphate and gama gutryl transaminase (437 and

130 U/L respectively). USG abdomen showed – mildly dilated common bile duct (CBD) with normal pancreas and no intra hepatic biliary radicles (IHBR) dilatation. Magnetic resonance cholangiopancreatography (MRCP) revealed dilated CBD measuring 6.8mm in its maximum diameter with prominent pancreatic duct. Based on the history and MRCP finding it was suspected as a case of FFCC. On exploration there was mildly dilated common bile duct and intra-operative cholangiogram showed minimally dilated common bile duct with long common channel pancreatico-biliary channel (figure -1). Resection of dilated CBD, pancreaticobiliary disconnection followed by Roux-en-Y hepaticojejunostomy was done. Patient is doing well after follow up of 18 months is doing well.

Figure-1: Peroperative cholangiogram showing mildly dilated Common Bile Duct with long common channel



Case – 2: A nine year old female child presented with complaint of recurrent peri-umbilical pain since 1 year. She had treated symptomatically for last 6 months and required hospitalization twice during this period in some private hostpial. General and systemic examination dose not revealed any significant findings. LFT revealed total bilirubin of 1.8gm%, SGPGT and SGOT mildly raised (59 & 31 respectively) ALP was 401 IU, GGT – 200 IU and serum amylase was 350 IU. USG abdomen showed mildly dilated CBD measuring 7.2mm with choledocholithiasis. MRCP showed bilateral IHBR dilatation with dilated right and left hepatic duct diameter of 7.6 mm (figure-2). Exploration showed minimally dilated CBD and intraoperative cholangiogram showed minimally dilated CBD with long common pancreaticobiliary confirming the diagnosis of FFCC. Resection of the CBD followed by Roux-en-Y hepaticojejunostomy done and patient is doing well after 18 month of follow up.

Case – 3: A twelve year old female presented with acute onset of upper abdominal pain of 2 days duration which was severe in intensity, located in upper abdomen, associated with vomiting and low grade fever. No history of jaundice, fever with chills & rigors and clay colored stools. LFT revealed total bilirubin of 3.2mg/dl, conjugated bilirubin 1.8mg/dl, SGOT/SGPT was 37/20 whereas ALP was 378 U/L and GGT was 170U/L. USG abdomen showed common bile duct dilated measuring 8.4 mm. ERCP showed long common Pancreaticobiliary channel (Figure – 3) Patient was posted for elective operation after 6 weeks of ERCP. On exploration the finding was consistent with the forme fruste choledochal cyst. Resection of the CBD followed by Roux-en-Y

hepaticojejunostomy done and postoperative event was unremarkable

Figure -2: MRCP showing mildly dilated Common Bile Duct with distended Gall bladder

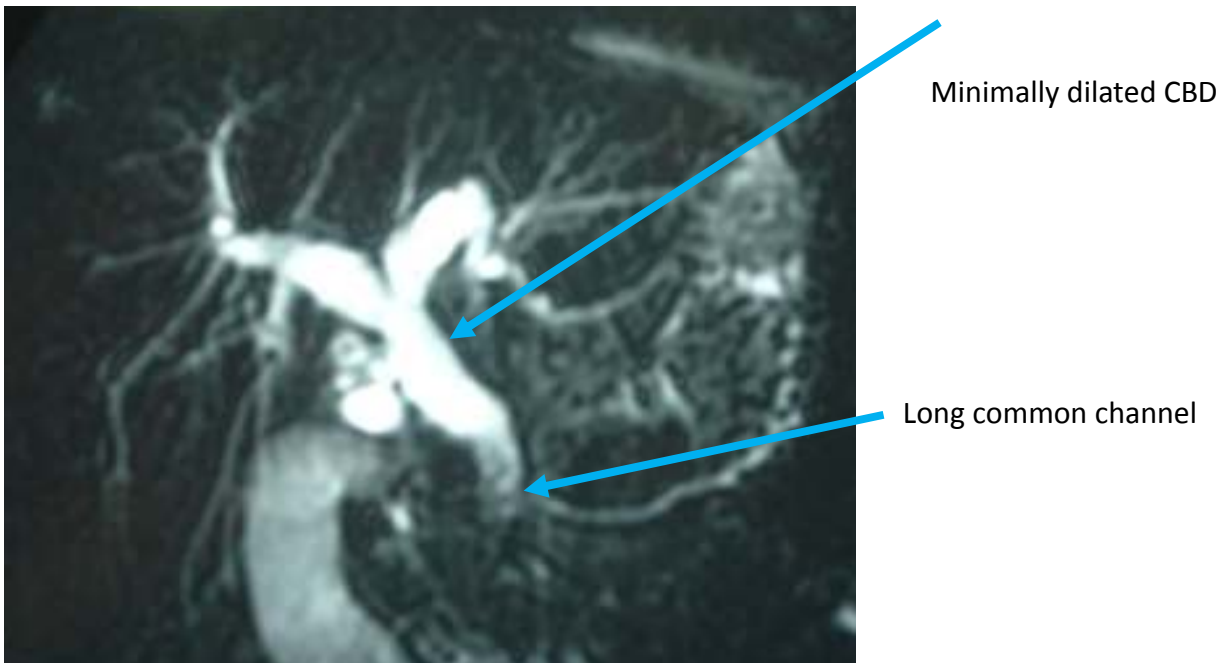
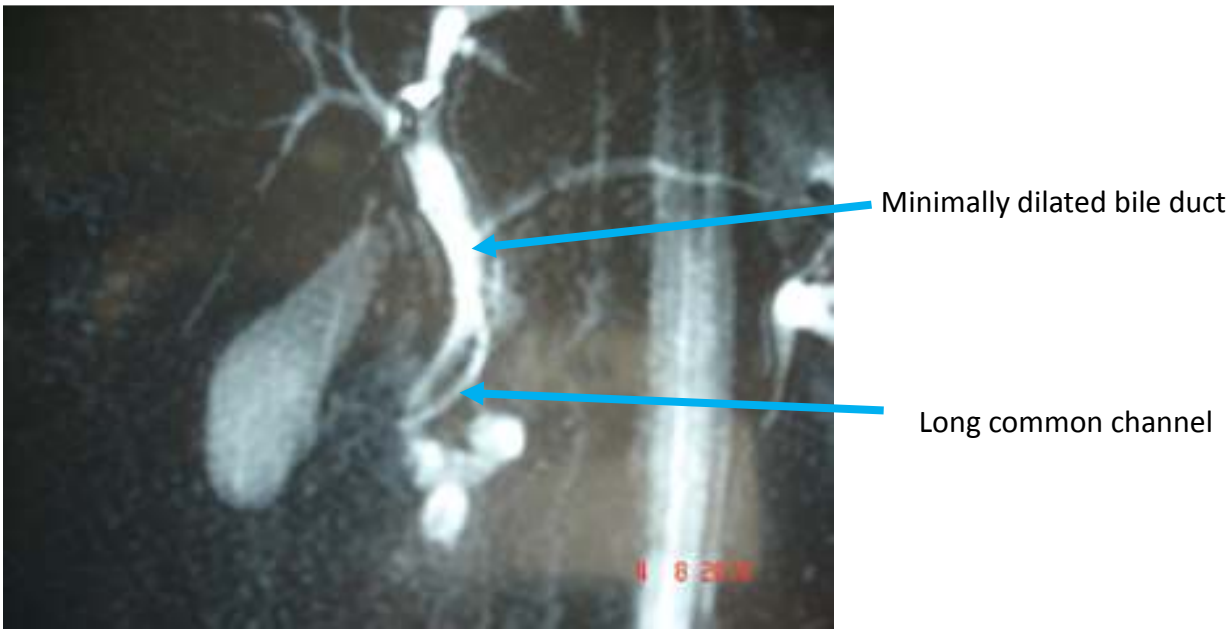


Figure-3: ERCP showing mildly dilated Common Bile Duct with long common channel



Discussion

Choledochal cysts are rare disease of unknown cause and Pancreaticobiliary mal-union is almost always associated with choledochal cyst (6). PBMU was first reported by Arnolds in 1906 (5) and defined it as anomalous junction between the pancreatic duct and the CBD that lies outside the duodenal wall and beyond the influence of the sphincter of Oddi (6,7,8). PBMU allows pancreatic secretions to constantly regurgitate into the biliary duct, where bile activates the pancreatic enzymes and these activated enzymes damages the biliary mucosa (9) making it vulnerable for malignancy.

In 1981, Okada et al.(11) were the first to report a case of choledochal cyst with minimal dilatation of the cyst and called it as " common channel syndrome " but later on Lilly et al in 1985 (2) coined this entity as FFCC which accounts for 4-21% of all choledochal cysts (1-4). The gross difference between choledochal cyst and FFCC is that; in choledochal cyst the CBD is severely dilated whereas in FFCC there is minimal dilatation of the CBD. The cut off diameter of CBD for considering it to FFCC is arbitrarily decided as 10 mm (12). There are two pre-requisite for diagnosis of FFCC; of them is demonstration of long common pancreatico-biliary channel for a particular age and another is dilated common bile duct with maximum diameter not exceeding 10 mm.(12).

The upper normal limit of common bile duct reported in literature for children aged below 3 months is less than 1.2 mm and less than 3.3 mm for children 1 to 13 years (13). In all cases of our series the maximum diameter of the CBD was ranged between 6.8 mm to 8.5 mm. Second important component for documentation of FFCC is demonstration of the long common pancreatico-biliary channel which can be demonstrated either by ERCP or peroperative cholangiogram. Available literature regarding the normal length of common pancreatico-biliary channel in children is very scanty, though the available literature search sowed that maximum normal length of common channel in neonate and infant is 3 mm which increase with age up to maximum of 5 mm in adolescent up to 15 years (14). The length of common pancreatico-biliary channel was more than 6 mm in all cases which was demonstrated on peroperative cholangiogram and ERCP.

Clinically these patients present with features of cholangitis, pancreatitis or non-specific pain in abdomen. Biochemical investigation may reveal conjugated hyperbilrubinemia with near normal SGOT/SGPT but there may be considerable elevation of ALP and GGT hence are not specific for diagnosis. It is observed that the hydrostatic pressure within the pancreatic duct is usually greater than that in the bile duct (10), but still bile may reflux into the pancreatic duct in presence of anomalous junction leading to acute pancreatitis which explains the frequent episodes of pancreatitis in such cases. So the patients presenting with recurrent episode of pancreatitis in absence of any predisposing factor (like alcohol intake, gall stone) or with recurrent cholangitis in absence of CBD calculi with minimal dilatation CBD should be investigated to rule out FFCC.

The mean age patients in present series was 8.1 years, all patients presented with pain in abdomen, one patient had history of symptoms suggestive of pancreatitis but none of our patients presented with features of cholangitis. All these patients showed prominent common bile duct with maximum diameter of CBD ranging between 6 mm to 8 mm which in presence of history of recurrent pain and or fever and non-clinical conjugated hyperbilrubenimia, elevated ALP and GGT suspected us to further investigate the patients to rule out FFCC. In hindsight, their clinical symptoms and laboratory data were most likely caused by stasis or obstruction at the level of the long common channel directly associated with the pathogenesis of PBMU. It is difficult to resolve these pathological symptoms without a diversion procedure, hence the optimal treatment is; resection of the CBD, pancreatico biliary disconnection and restoration of bilio-enteric continuity in form of Roux-en-Y hepaticojejunostomy (7,8) Few authors had suggested open sphincteroplasty (15) and endoscopic sphincteromomy (16) as a treatment modalities for FFCC but excision of dilated common bile duct with pancreatico bililary disconnection and bilioenteric anastomosis is considered as a treatment of choice(7,8). In past only cholecystectomy had been done for these cases but symptoms were not resolved and patients continued to have recurrent episode of pancreatitis and responded to cyst excision with hepaticojejunostomy (12). If PBMU is found accidentally in an asymptomatic child, close observation is required, and if any symptoms at all are noted, CBD excision with biliary reconstruction is recommended. However, close postoperative follow-up is required because postoperative stricture formation and recurrent cholangitis are problems associated with reconstruction of the non-dilated biliary tree but none of our patient developed any complication in follow up of 2 years.

Conclusion: Though "forme fruste choledochal cyst" is rare disease but it is easy to diagnose if the index of suspicion is high and proper diagnostic facility is available. The disease is totally curable. The FFCC should

be ruled out in patients presenting with recurrent attacks of cholangitis or pancreatitis especially if ultrasonography revealed a mildly dilated common bile duct.

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