Choledochal Cyst of Cystic Duct in Children a Rare Entity

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ABSTRACT

Choledochal cyst of the cystic duct is an uncommon entity which may occur as isolated disease or may be associated with choledochal cyst of the other part of the biliary tree. In most of the reported series it was diagnosed at time of operation but now days to better investigation modality it can be diagnosed preoperatively if the index of suspicion is high. We are presenting a series of such case in pediatric age group (which includes an infant) where the diagnosis was made during operation with a light on means and ways for suspecting this entity before operation and how to differentiate other lesion with this entity with relevant review of literature.

KEY WORDS

Choledochal cyst, Cystic duct, MRCP

INTRODUCTION

Choledochal cyst (CDC) defined as a cystic dilatation of the common bile duct is a rare abnormality. Traditionally, choledochal cysts have been classified into five main types as described by Todani et al. Choledochal cyst of cystic duct (CDC of CD) was not described by Todani et al. and now it is described as type VI choledochal cyst. Choledochal cyst of cystic duct is very rare and usually diagnosed at time of surgery in past.[1-3] Awareness of the disease and better modalities of investigation helped us to diagnose and manage these cases successfully. We are reporting three cases of choledochal cyst of cystic duct one of which was in infant and one was associated with forme fruste type choledochal cyst.

CASE SERIES

A 13 year old male presented with complaint of abdominal pain since 4 months with occasional vomiting. Pain was severe in nature and occurs once or twice in months with frequency gradually increased to one to twice in week with in two months. The pain usually responds to intravenous analgesics and heed need hospitalization twice during this period. Laboratory finding revealed mild elevation of alkaline phosphtase (ALP) and gamma gutryl transferase (GGT), with no rise in bilirubin and serum amylase level. Ultrasound was suggestive of cystic lesion at the site of insertion of cystic duct with mildly distended Gall bladder (GB). Magnetic resonance cholangiopancreatography (MRCP) demonstrated dilated cystic duct with mild dilatation of common bile duct at the site of insertion of the cystic duct (fig. 1) and rest of the common bile duct CBD was prominent with diameter of 8 mm (distal CBD) with long common channel with no evidence of calculus in CBD. In view of long common channel and mild dilatation of the CBD the diagnosis of Forme fruste choledochal cyst was suspected. We are not able to demonstrate wide opening of the cystic duct into prominent CBD, in view of diagnosis patient was planned for cyst excision with Roux

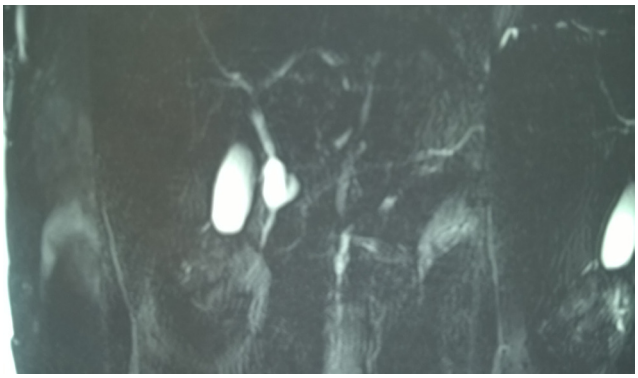

Figure 1. Showing mild dilatation of the CBD, mildly dilated gall bladder and dilated cystic duct entering into bile duct

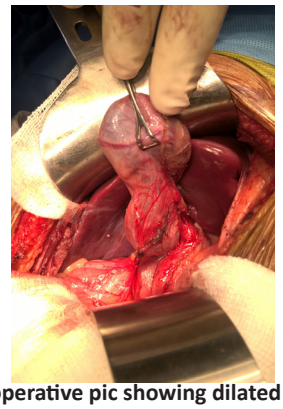

Figure 2. Intra-operative pic showing dilated cystic duct

-en-Y hepaticojejunostomy (HJ). Exploration revealed dilated cystic duct (fig. 2) with wide opening of cystic duct into CBD. Patient did well in post-operative period and in follow up of more than 6 month.

A 4 month infant presented with classical triad of choledochal cyst that is pain, jaundice and right hypochondrial lump. Laboratory parameter revealed increase in total leucocyte count, raised total and conjugated bilirubin, alanine aminotransferase (SGPT) aspartate aminotransferase (SGOT), alkaline phosphatase (ALP) andgamma-glutamyl transferase (GGT). Ultrasound showed fusiform dilatation of the CBD, with grossly distended GB and prominent cystic duct and bilateral intrahepatic biliary radicle dilatation (IHBRD). MRCP confirmed the finding of the ultrasound (fig. 3) and showed grossly dilated cystic duct, but not able to localize the wide opening of the cystic duct. Patient was treated conservatively with intravenous antibiotics and excision of the cyst with Roux-en-Y Hepaticojejunostomy was done after treating cholangitis. Exploration revealed very tortuous, long (4 cm) grossly dilated cystic duct.

Five year old male child presented with complaint of pain in abdomen with occasional vomiting since one year admitted to our ward with no history of jaundice, fever or weight loss. Blood investigation showed mild elevation of total leucocyte count, serum amylase, ALP and GGT. Ultrasound revealed distended gall bladder with type IVA choledochal cyst. MRCP showed grossly distended GB with grossly dilated cystic duct again the wide opening of cystic duct was not commented on MRCP. Exploration revealed

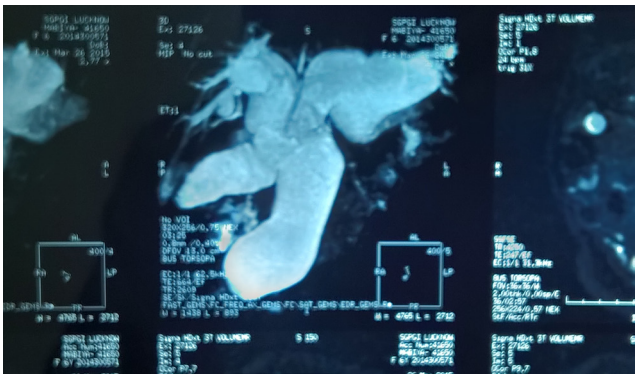

Figure 3. Grossly dilated bile duct, with grossly dilated left and right hepatic duct in 4 month old child with grossly distended gall bladder and dilated tortuous cystic duct

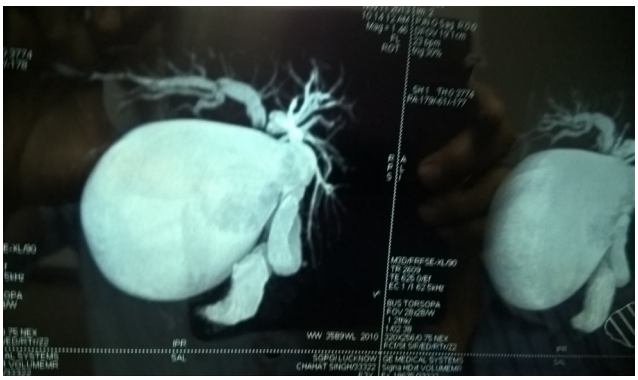

Figure 1. Off line- Saccular dilation of the bile duct with mildly distended gall bladder with near normal cystic duct – this will differentiate choledochal cyst of cystic duct: because though the obstruction is beyond the insertion of cystic duct even though it is not dilated

distended gall bladder with tortuous and dilated cystic duct with wide opening. Excision of the cyst with Roux-en-Y HJ done and patient did well in postoperative period.

DISCUSSION

Choledochal cyst is a congenital dilatation of the extrahepatic biliary tract with or without dilatation of the intrahepatic biliary tract. Type 1 choledochal cyst is the most common type (80-90%).[4-6] About two-thirds of patients are clinically symptomatic before the age of 10 years and rest remain asymptomatic till adulthood. Classical triad of abdominal pain, jaundice and a palpable right upper quadrant mass is present in 30-60% of patients presenting in the first decade of life and only in 20% of patients presenting later in life.

Choledochal cysts were originally classified into three types by Alonso-LEJ et al. in 1959.[5] Other variations were subsequently recognized and the above classification was modified by Todani-et-al, but does not include CDC of CD, inclusion of this variety in choledochal cyst classification

Cystic malformation of the CD is a less known entity and was firstly described by Bode and Aust in 1983. It had been described with different names as choledochal cyst of cystic duct, cystic duct cyst, type 6 choledochal cyst choledochocele of cystic duct and cystic malformation of CD

Table 1. **Profile of the cases of choledochal cyst of cystic duct in pediatric age group**

References	Presentation/no of cases	Associated disorder	Time of diagnosis	Management	Outcome
Champetier et al.[2]	Pain and Jaundice/one	Choledochal cyst of bile duct	Preoperative	Total excision of the bile duct with HJ	Good
Manickam et al.[15]	Pain /one	No details	Preoperative	Not done	
Yoon et al.[13]	Pain fever, chills/one	Fusiform dilatation of the bile duct along with fusiform dilatation of the cystic duct	Preoperative	Total excision of the cystic lesion of CD with excision of the part of the CBD with HJ	Good
Maheshwari et al.[26]	Pain in one, jaundice and pain in other no details of other cases/ 6 cases	Fusiform dilatation of the bile duct in two cases	Preoperative	Three of these were not operated and for those who had been operated not details are available	Not available
Shah et al.[20]	Pain/one	None	Preoperative	Total excision of the cystic lesion of CD with excision of the part of the CBD with HJ	Good
Goya et al.[6]	Pain/one	None	Preoperative	Not operated	
Sethi et al.[14]	Pain and Jaundice/one	Intra hepatic duct cyst	Preoperative	Total excision of the cystic lesion of CD with excision of the part of the CBD with HJ	Good
Present study	Pain, Classic triad in one	Two had fusiform dilatation of the CBD and one had? Forme Fruste Choledochal cyst	Intra-operative	Total excision of the cystic lesion of CD with excision of the part of the CBD with HJ	Good

have been used.[7-13] Extensive literature search on pubmed and google showed only 40 cases of choledochal cyst of CD in 20 studies. In most of the studies it has been described in adults.[3-5,7-11,14-18] Twelve such cases are reported in world literature in pediatric patients of this entity in pediatric population in seven studies (table-1).[2,6,13,15,19,20,26] We are reporting three cases in pediatric patients of which one was infant.

Most widely accepted theory for development of choledochal cyst is primary abnormality of pancreatic biliary ductal junction (anomalous pancreatic biliary ductal junction: APBDJ) with long common channel is the primary abnormality and this abnormality affects the sphincteric action of common channel.[7,20,21] Incidence of APBDJ had been reported in 33-90.2% of choledochal cyst and it explain the development of type I, III and IV CDC but does not explain the development of type II and V cysts (the cysts which have normal common bile duct). Other hypotheses are aganglionosis with proximal biliary dilatation these theories explain the pathogenesis of choledochal cyst of CBD but do not explain the pathogenesis of choledochal cyst of cystic duct.[22] The most acceptable explanation for development of CD cyst is that it may result from a combination of APBDJ, acute angulation, and wide orifice of CD hepatic duct junction.[23] In our cases wide opening of the cystic duct at junction with common hepatic duct was observed in all three cases where as APBDJ with long common channel was documented in only one case. None of our patients had stricture of aganglionosis of CBD.

The classical triad of pain, jaundice and right hypochondrial mass of choledochal cyst are seen approximately in 6-38% cases.[3,24] The main presenting symptoms in cases of choledochal cyst of CD is similar to other common variant of choledochal cyst. The most common presenting feature for choledochal cyst of CD is abdominal pain in upper abdomen which usually exaggerated by a fatty meal. In our series two patients presented with complaint of upper abdominal pain with no history of exaggeration of pain on taking fatty meals. Jaundice and abdominal lump was a presenting feature in one case (infant). Vomiting was seen in two cases. Many patients also had associated complicating biliary tract disease like; cholangitis, cholecystitis, cholelithiasis, gall bladder polyp, cystolithiasis. In our series only one patient (infant) presented with classical triad and rest two patients had no feature other than pain.

Cystic duct attaches the gallbladder to the extrahepatic bile duct; its point of insertion into the extrahepatic bile duct marks the division between the common hepatic duct and the common bile duct. The CD usually measures 2 to 4 cm in length and contains prominent concentric folds known as the spiral valves of Heister. The CD frequently exhibits a tortuous or serpentine course. The normal diameter of the cystic duct is variable, ranging from 1 to 5 mm.[25] Choledochal cyst of CD may present as isolated finding or with associated cyst of the common hepatic or bile duct. Extensive literature search showed that there are two anatomical pre-requisite for the diagnosis of this entity. Firstly the diameter of CD should be more than 5 mm and secondly the diameter of opening of the CD at common hepatic duct should be wide (no clear cut demarcation for leveling CD opening as wide opening is available in literature, though arbitrary we considered the opening as wide if we were able to negotiate K-90 tube (external diameter > 20 FR) through the CD opening in our study but it is not documented anywhere in the literature). Dilated

CD may be observed in cases of biliary obstruction if the obstruction of the bile duct is distal to the junction of the CD and though in such cases, dilatation of CD is proportionally much less in comparison to the dilatation of bile duct and opening is usually narrow. That's why for diagnosis choledochal cyst of CD these two prerequisite should be met (CD must be dilated adequately in proportion of bile duct with wider opening in bile duct). We observed dilated and tortuous cystic duct in two more cases but the dilatation of the cystic duct was much less than the dilatation of the CBD and on exploration the opening was narrow (able to admit only no 10 Fr Ryles tube) so was not included in our series. Because of extreme rarity of this disease, the diagnosis of this entity needs high degree of the suspicion and in most of the published series diagnosis was made at the time of operation as was in our study.[13,14] It is important to recognize this entity before avoid laparoscopic cholecystectomy because of its anatomical similarity with type-2 choledochal cysts in which management may vary.[3,7] Ultrasound is the first investigation used for biliary symptoms which can diagnose the choledochal cyst but it usually fails to demonstrate the communication of cystic duct with common hepatic duct so cannot comment on the wider opening of CD in common hepatic duct though it can measure the diameter of the CD and secondly it is operator dependent. Abdominal MRI with MRCP is preferred over CT scan for further evaluation of sonographic finding for better display of biliary anatomy and relationships. It can diagnose the origin of cyst from cystic duct as well as associated biliary abnormalities. It can accurately measure the diameter of the CD as well as the width of the opening of the CD into the common hepatic duct. Normal CD diameter has an upper limit of 5 mm in adults but the literature on the size of cystic duct in children is not available that's why in our series it was considered as 5 mm more for leveling the dilated CD as choledochal cyst of CD.[25,26] The cystic duct dilatation may be fusiform or saccular; with former variety more common than the latter, and two of our patient had fusiform dilatation. The width of opening of cyst into the CBD is important especially in cases of isolated cyst because it is difficult to differentiate it from Type-II choledochal cyst and the management of the two entities may differ. In most of the studies the diagnosis was made post-operatively only two studies it was diagnosed preoperatively in present series only in one case it was suspected before surgery and was diagnosed at time of operation but in rest suspected and diagnosed during operation.[15,17] Because of the rarity of the disease and scanty literature there are no uniform guidelines for the treatment of choledochal cysts of the CD. Some advocate that to treat isolated choledochal cysts of the cystic duct with a narrow opening in the CBD with simple cholecystectomy and cyst excision either by open surgery or laparoscopically, where as others prefer complete excision with bilio-enteric anastomosis.[2,9-10,12,15] We recommend complete excision with biliary drainage, in view of the increased risk of developing bile duct and gallbladder cancer in the presence of cystic biliary duct anomalies and APBJ especially in the cases of children owing to large life span ahead.

Cystic malformations of the cystic duct had a characteristic features which are common to other types of choledochal cysts in Todani's classification. Although rare, they need to be recognized. We recommend that these cases should be treated with complete excision cystic duct and common bile duct with Roux en Y bilio-enteric anastomosis to avoid any future complication owing to long life span ahead in cases of children. Cystic duct cyst is an extremely rare anomaly, and advanced examination is required for the diagnosis

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