Uncommon Site of Perforation of Biliary Tree in Choledochal Cyst Associated with Chronic Calcific Pancreatitis- A Case Report

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Abstract

Choledochal cyst presents with various clinical features and complications. Presentations and complications vary across the various age groups. Early identification and early surgical intervention have shown to reduce the complications related to the disease. The treatment of choice is Roux -en-Y hepaticojejunostomy. We present a case of 7 years-old boy who presented with biliary peritonitis due to choledochal cyst.

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Keywords

- ruptured choledochal cyst
- pancreatitis with choledochal cyst
- hepaticoduodenostomy
- biliary peritonitis in choledochal cyst
- unusual site of rupture of choledochal cyst

The perforation was in the proximal biliary tree near the confluence which is not a common site of perforation, either in the case of spontaneous biliary tree rupture or in the choledochal cyst. The patient also had chronic calcific pancreatitis. Such a combination is not mentioned in the literature. Unconventional surgical procedure (hepaticoduodenostomy) was performed as a part of treatment, reserving jejunum for future pancreatic drainage, if the need arises.

Introduction

Choledochal cyst (CDC) is a congenital dilatation of the extrahepatic biliary tree. It is one of the consistent components of a broader spectrum of malformations in the pancreaticobiliary junction. The disease is ten times more commonly seen in eastern hemisphere and often seen in infancy or early childhood.[1] CDC was initially classified by Alonso-Lej, modified by Todanifron as the types 1 to 5.[2] Pancreatitis associated with CDC is usually seen in the adult population. Chronic calcific pancreatitis in children is relatively rare. Biliary perforation and perforation of CDC are well documented entity, but biliary tree perforation proximal to CDC is also not clearly documented in the CDC literature. Biliary tree perforation can take place due to increased ductal pressure by an obstacle like a distal biliary stone, sphincter of Oddi dysfunction, malignancy, necrosis and ischemia due to thrombosis of the intramural vessels, pancreatic reflux and infection.[3] We present a case of biliary perforation of the dilated system complicated by chronic calcific pancreatitis in a young boy.

Case Presentation

A 7 years-old boy presented to emergency department with upper abdominal pain and nonbilious vomiting of 3 days duration. There was no associated fever and jaundice. He had history of previous nonspecific abdominal pain which had subsided with conservative management. He was afebrile, anicteric and dehydrated at presentation. Patient had normal blood pressure, tachycardia, epigastric tenderness

and guarding along with the abdominal distension suggesting peritonitis which was denoted by complete blood count (14,500/cumm). Chest and abdomen radiographs were normal. Abdominal ultrasonography showed dilated common bile duct and main pancreatic duct along with the ductal calculi. Serum amylase, lipase and liver function tests were within normal limits.

Computed tomography scan of the abdomen showed dilated extrahepatic biliary tree, choledocholithiasis with a 3 mm perforation in the common hepatic duct wall and chronic calcific atrophic pancreatitis along with the pancreatic calculi measuring 1X 1cm in the ampulla and multiple small calculi in main pancreatic duct. **(Figure 1)**

Emergency exploratory laparotomy was performed after initial resuscitation. Intra operative findings showed dilatation of common bile duct (CBD) suggestive of choledochal cyst (Type 1) and 3-4 mm perforation with biliary leakage just distal to confluence of common hepatic ducts. **(Figure 2)** The hepatic ducts were inflamed. There were no biliary stones. After peritoneal lavage, excision of choledochal cyst and hepatico-duodenostomy was performed as jejunum was reserved for pancreatic drainage if the need arises in future. Drain was placed in Morrison's pouch. He continued to have minimal bile leak post operatively which gradually decreased with conservative management and nutritional support and subhepatic drain was removed on post op day 14. Our patient improved and was discharged in the stable condition. The cyst fluid amylase was noted to be 395IU/ml. The patient has been symptom free during the follow up for 9 months.

Discussion

Biliary peritonitis secondary to the perforation of the extra hepatic biliary tree is a rare occurrence, with the common site being junction between cystic duct and common bile duct which is the weakest site of hepatobiliary tree.[4] The incidence of biliary peritonitis due to ruptured CDC is not more than 3.3 % in most series.[4-5] Our patient was presented with biliary tree perforation proximal to CDC, but just distal to the confluence of hepatic ducts. This was also associated with chronic calcific pancreatitis with dilated pancreatic ducts which is not a usual presentation. Such a combination is not mentioned in the english literature. Jaundice is the prominent feature

in infants, whereas cholangitis and pancreatitis are common in adults.[6] Perforations are usually noted in the anterior or posterior wall of the cyst. Presentation is in the form of ascites in neonates and acute abdomen in older children.[4] The cause of the rupture is thought to be due to the raised intraabdominal pressure (eg. vomiting), the long common channel theory leading to reflux of pancreatic secretion into the biliary tract as pancreatic ductal pressure is higher compared to biliary tract. Hypoperfusion of inflamed dilated duct is also a known etiology of rupture. Pregnancy is another risk factor CDC rupture.[3]

Pancreatitis is well associated with CDC due to abnormal pancreato-biliary-duodenal junction. There are case reports of calcific pancreatitis associated with CDC in adult patients. But there is no clear description of atrophic calcific pancreatitis in the pediatric age group. Whether pancreatitis causing distal biliary obstruction has led to the perforation of the biliary tree in our patient or CDC perse had led to this phenomenon in our case is unclear. Magnetic resonance cholangiopancreaticography has high sensitivity and specificity in the diagnosis of CDC and anomaly of pancreatic and biliary ducts, which has replaced percutaneous biliary dye study or ERCP in the work up of CDC. Role of ERCP is also widely discussed in the diagnosis and management of pediatric choledochal cyst. It defines the types of CDC. It is also helpful in initiating management of biliary peritonitis. Need for miniature instruments and pediatric gastroenterology expertise limit this option. Pancreatitis is one of the serious complications of ERCP, whereas it can as well be used as a treatment option in pancreatitis.[7] Surgical management in infancy renders CDC surgery easy and also can avoid spontaneous perforation in future if left untreated.[6] Differentiating spontaneous billiary perforation from the CDC perforation is necessary to choose the appropriate treatment. Dilatation due to CDC may appear similar to the dilatation caused by distal obstruction in children.[3] Nuclear scintigraphy may also be used as an effective tool in evaluating CDC. It can as well diagnose sealed off biliary collection form the active leak. Spontaneous biliary rupture is managed by drainage of the collection with or without the insertion of T-tube into the biliary tree, whereas treatment of ruptured CDC includes complete excision of the cyst with

restoration of biliary tract continuity. Residual cyst wall poses a significantly high risk for malignancy in the future. Initial biliary drainage of the ruptured cyst eases the subsequent definitive management of CDC.[8] Biliary peritonitis associated with biliary dilatation should raise the suspicion of following differential diagnosis in children; CDC, biliary hamartoma, biliary rhabdomyosarcoma, biliary stones, pancreatitis. Roux en Y hepaticojejunostomy is the treatment of choice in type 1 CDC.[7] Our patient underwent cyst excision and hepaticoduodenostomy. The pancreatitis was not addressed in the present procedure, but jejunum was reserved for future pancreatic drainage, if the need arises. Hepatico duodenostomy is comparable with hepaticojejunostomy, with higher rate of bile reflux issues.[9] Hence we should keep the options of unconventional therapy if special situation arises.[10]

Conclusion

Biliary peritonitis in choledochal cyst needs a thorough evaluation. The surgical procedure should be tailored depending on the prevailing situation. Unconventional procedure must also be kept in mind while treating if the need arises.

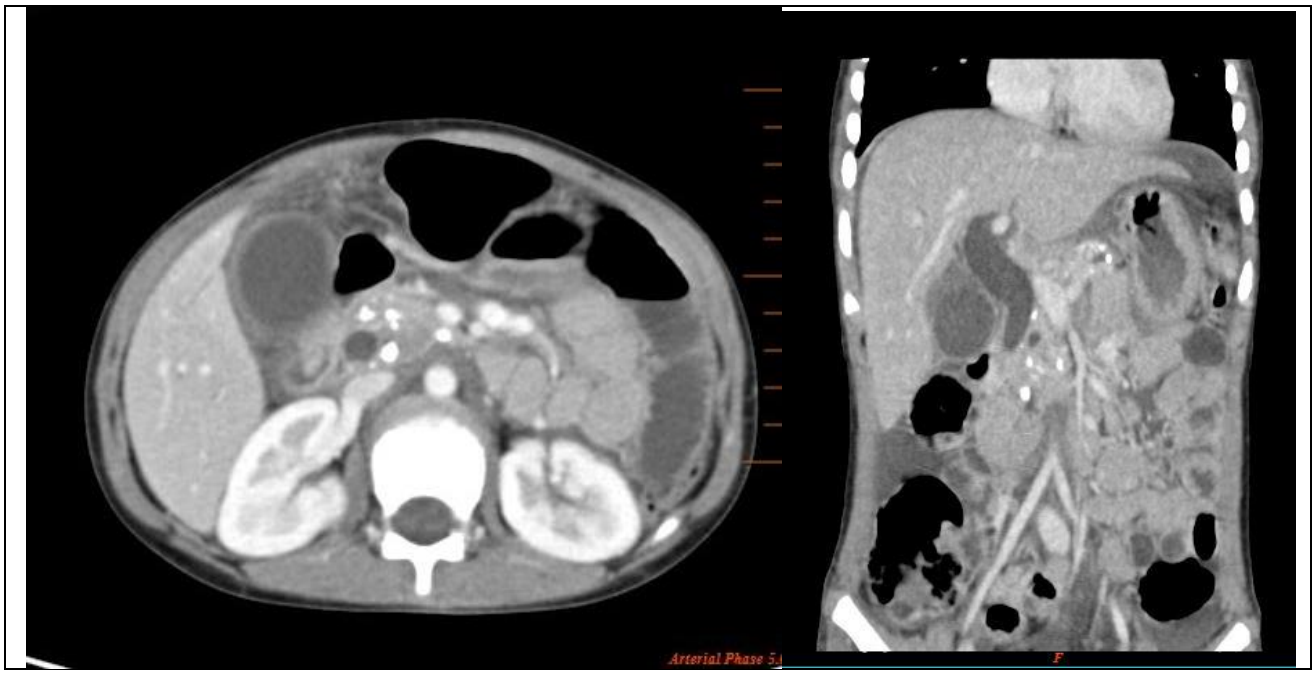

Figure 1: CT scan images of coronal and transverse view showing dilated common bile duct and perihilar collection of bile. Calcification in the pancreas can be noted in pancreatic parenchyma

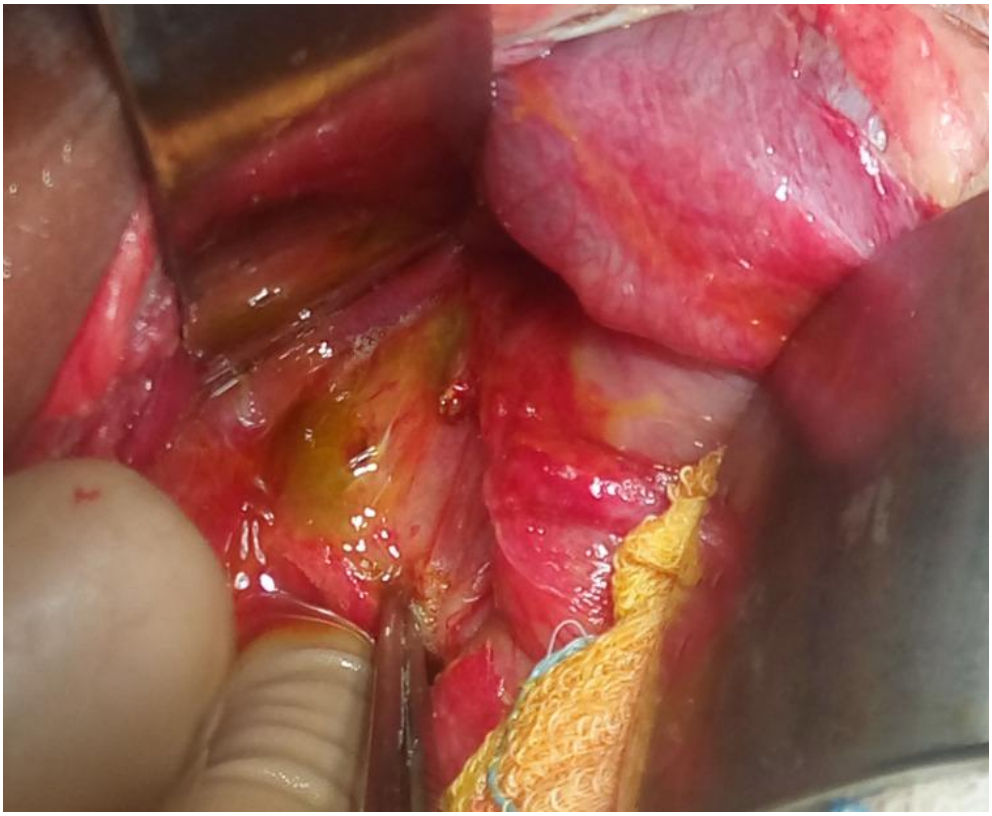

Figure 2: Intra-operative photograph showing perforation in the confluence of right and left hepatic duct

Ethical Consideration

Written consent for participation was obtained from the parent or guardian of the participant in the study**.** This study was approved by Shri Dharmasthala Manjunatheshwara University.

Conflict of interests

There is no conflict of interest

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