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Choledochal Cyst with Pancreas Divisum – Surgical Insight into A Rare Association

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ABSTRACT

Congenital pancreaticobiliary anomalies like pancreatic divisum (PD), choledochal cysts (CDC), anomalous pancreaticobiliary ductal union (APBDU) have been reported in 5.7% of patients undergoing magnetic resonance cholangiopancreatography. CDC's are characterized by abnormal dilatations of the intrahepatic and/or extrahepatic portion of the biliary tree and can be complicated by cystolithiasis, cholangitis, pancreatitis and malignant transformation necessitating surgical managemnet. While CDCs are commonly associated with APBDU, combination of CDC with PD is rare and a potential surgical challenge. We report a case of recurrent cholangitis in a patient with CDC and coincidental classic PD and illustrate how preoperative identification, fastidious dissection technique to safeguard both the pancreatic ducts and simple intraoperative preemptive strategies may decrease consequences of distal stump blowout.

PD, pancreatic divisum; CBD, common bile duct; CDC, choledochal cysts; APBDU, anomalous pancreaticobiliary ductal union; MRCP, magnetic resonance cholangiopancreatography; DOS, Duct of Santorini; DOW, Duct of Wirsung; PTBD, percutaneous transhepatic biliary drainage; RYHJ, roux en Y hepaticojejunostomy.

Introduction

Congenital pancreaticobiliary anomalies, namely pancreatic divisum (PD), choledochal cysts (CDC), and anomalous pancreaticobiliary ductal union (APBDU), arise from developmental errors during gestation [1]. PD is the most common and results from the failure of fusion of embryological ventral and dorsal buds, resulting in two main ducts (Duct of Santorini- DOS and Duct of Wirsung- DOW) draining the pancreatic parenchyma into the duodenum [2]. It has been reported in 5.7% of patients undergoing magnetic resonance cholangiopancreatography (MRCP) [3].

On the other hand, CDCs are rare congenital bile duct malformations characterized by abnormal dilatations of the intrahepatic and/or extrahepatic portion of the biliary tree and can be complicated by cystolithiasis, cholangitis, pancreatitis, and malignant transformation [4,5]. While CDCs are commonly

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associated with APBDU [5], the combination of CDC with PD is infrequent but a potential surgical challenge [6,7]. Treatment of CDCs entails complete excision of the extrahepatic biliary tree with restoration of bilioenteric continuity via a hepaticojejunostomy at the hilum [8,9]. While proximal mobilization of the CDC till the biliary confluence is straightforward unless the surgical dissection is made difficult by adhesion of the cyst to the hepatic artery and portal vein secondary to inflammation [10], excision of the distal segment of CDC necessitates dissection from pancreatic tissue in addition to the hepatic artery and portal vein. This might be particularly challenging in a large intrapancreatic portion of CDC, particularly if surrounding planes are inflamed and friable and or the main pancreatic duct is intimately related. In CDC with PD, the presence of two pancreatic ducts in the vicinity increases the risk of injury to either duct, with the potential for serious complications like postoperative pancreatitis, postoperative pancreatic fistula (POPF), intraabdominal collections, bleeding, stone formation [11].

Further, incomplete dissection with a large remnant intrapancreatic component may give rise to subsequent malignant transformation with an estimated risk of 0.7% to 6% [12]. The authors have increasingly appreciated the contribution of distal stump management in overall outcomes following CDC excision [13]. Liu et al. have even proposed a classification and management of CDCs in adults based on the relation between the intrapancreatic portion of CDC and the pancreatic duct [11] to provide a more targeted strategy for complete and safe cyst excision, protect the pancreatic ducts, and thus prevent life-threatening complications like POPF, collections, bleeds, and late malignant transformation.

Although there are reports of CDC with PD presenting with pancreatitis, and treated with minor duct sphincterotomy [6], there is scant literature on surgical challenges while excising such CDCs [14]. The authors report a case of recurrent cholangitis in a patient with CDC and coincidental classic PD and illustrate how preoperative identification, fastidious dissection technique to safeguard both the pancreatic ducts, and simple intraoperative preemptive strategies may decrease consequences of distal stump blowout.

Case Presentation

A 35-year-old lady presented to the emergency with features of cholangitis (fever, leucocytosis, bilirubin of 7.5mg%, AST of 215U/L, ALT of 356U/L, and alkaline phosphatase of 545U/L). She had two similar episodes in the past which resolved with oral antibiotics alone. Ultrasound abdomen revealed asymmetrical dilatation of CBD (58mm) suggesting CDC. MRCP (Figure 1) revealed dual congenital abnormality related to the ampulla of Vater, Todani's type 1 CDC, and PD with DOW opening near the termination of the CDC. The DOS was coursing in close proximity to the CDC. Since the current episode of cholangitis did not respond to antibiotics, endoscopic access was attempted for CBD stenting, but a tight stricture at the papilla prevented cannulation of distal CBD. Percutaneous transhepatic biliary drainage (PTBD) through segment III hepatic duct was done to drain the biliary system. Six weeks later, the patient underwent laparotomy and CDC excision with Roux en Y hepaticojejunostomy (RYHJ). The dissection of the intrapancreatic segment of choledochus was particularly difficult in view of the dense inflammatory adhesions. By maintaining the plane of dissection right on the cyst wall, vessels from the epicholedochal plexus were controlled with bipolar coagulation and the cyst was enucleated from the pancreatic head. Since it was known that the patient had PD, care was taken not to deviate into

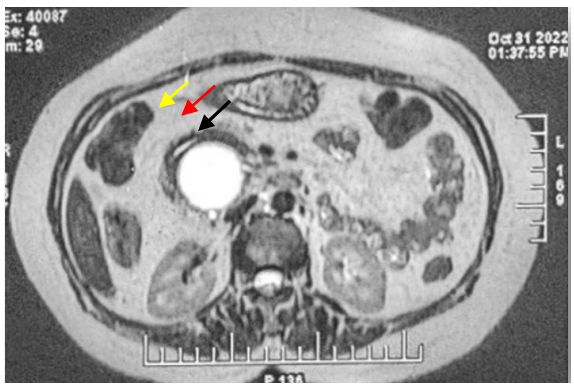

Fig. 1. Preoperative magnetic resonance cholangiopancreatography (MRCP)- Axial section: red arrow depicts the choledochal cyst; black arrow depicts the ventral pancreatic duct draining into the choledochal cyst; yellow arrow depicts dorsal pancreatic duct draining separately at minor papilla

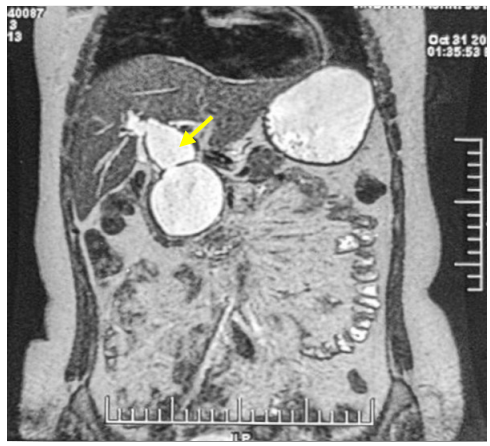

Fig. 1. Preoperative magnetic resonance cholangiopancreatography (MRCP) - coronal section showing the choledochal cyst marked by yellow arrow.

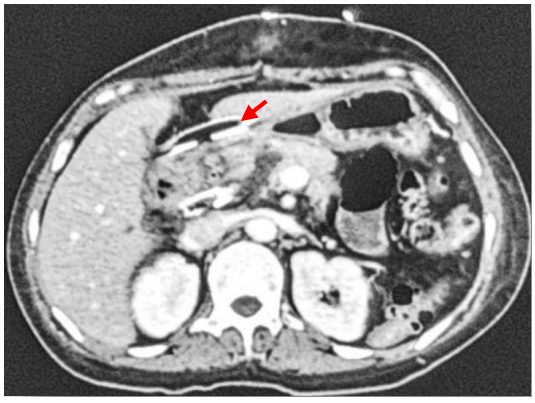

Fig. 3. Post operative computer tomography (CT): Black arrow represents normal pancreas and two drains can be seen in-situ near distal stump.

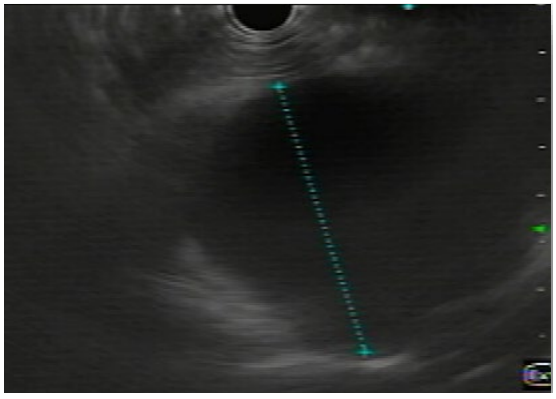

Fig. 4. Endoscopic Ultrasound (EUS) depicting fusiform dilatation of CBD with size 28 – 58mm.

the pancreatic parenchyma and injure DOS. The CDC was opened 5mm above its termination to identify the opening of the DOW, but the inflamed mucosa prevented visualization. The cyst was transected 5 mm above the commencement of the narrowed segment to avoid the DOW. The cut edge was secured with delayed absorbable monofilament polydioxanone suture. An RYHJ restored biliary drainage. Two wide bore latex drains were placed in the resultant cavity in the pancreatic head (Figure 2).

On postoperative day 3, drain fluid amylase of right and left drains were 442IU/L and 24IU/L respectively while serum amylase was 27IU/L suggesting postoperative pancreatic fistula (POPF), however, the patient was clinically stable and tolerating a full diet. These drains functioned well postoperatively and enabled effective drainage with subsequent discharge on the 7th postoperative day with drains in situ as controlled, low volume (daily output of 25 – 30 ml/day) external pancreatic fistula. She continued to thrive postoperatively, the drain output progressively decreased and both drains were removed 21 days after surgery, (Grade B POPF). At 6 months, she is well with normal liver function tests (McDonald criteria excellent outcome) with no features to suggest any clinical consequences of PD.

Discussion

Management of cholangitis in CDC ranges from antibiotics with or without drainage (endoscopic or PTBD) followed by surgical excision and RYHJ. An inflamed choledochus due to repeated previous episodes of cholangitis can increase chances of distal stump blowout and POPF [11]. Coexistence of a PD with CDC places two distinct pancreatic ducts (DOW and DOS) at risk of injury during mobilisation of the distal end. The authors' patient had dense pericholedochal inflammation following repeated episodes of cholangitis and PD with closely related DOS. To add to the challenge, DOW was opening into the narrowed lower end of CDC. Preoperative identification of these anatomical relations prompted the authors to maintain the plane of pericholedochal dissection on the cyst wall, avoiding breach of subjacent pancreatic tissue and the ducts. The pericholedochal plexus was painstakingly controlled using bipolar coagulation. The distal transection was done about 5 mm above the terminal end of the dilated part of choledochus after opening the choledochus and visualization to ensure near total removal of the distal end of CDC, without injuring the DOW opening [12]. The authors placed two drains in the resultant pancreatic bed which functioned well to create a controlled, external pancreatic fistula. CT scan on postoperative day 7 did not show any undrained collection or evidence of pancreatitis excluding potential inadvertent occlusion of either of the PDs. This enabled the authors to confidently discharge the patient within a week of surgery. The intraoperatively placed drains functioned effectively to create a controlled, low volume, external pancreatic fistula.

Reported incidence of POPF following CDC excision is 2 to 6% and can lead to life-threatening complications like intraabdominal collections and bleeds resulting in reinterventions and prolonged hospital stay [15]. Management options for distal POPF are conservative with or without additional PCDs, ERCP and PD stent, relaparotomy with oversewing of distal cyst stump and reinforcement with fibrin sealant [15]. Each of these additional interventions has a complication and failure rate. Correct interpretation of the MRCP guided the authors in distal transection without injuring the DOW. Meticulous dissection without straying into the pancreatic parenchyma [16] helped protect both the pancreatic ducts. Preemptively placing two wide bore drains in the resultant cavity

in the pancreatic head provided adequate egress to pancreatic juice when the inflamed distal stump failed to seal completely and enabled the authors to manage the patient without the need for additional intervention or prolonged hospital stay. Other authors have described more challenging strategies like CDC excision with papillectomy and reconstruction with a hepatico-jejunostomy and reinsertion of the uncinate pancreatic duct into the same jejunal loop [17], endoscopic minor papillary sphincterotomy followed by CDC excision and RYHJ [14].

A literature review reveals a more common association between PD and choledochocele compared to other types of CDC, 37% vs 10% [18]. MRCP is an elegant modality to diagnose anatomical variations related to the choledochus [3]. While choledochoceles are managed by endoscopic sphincterotomy [19], CDCs require surgical excision [5] within challenging surroundings in the presence of PD. The authors' case provides insight about the need to carefully study MRCP for the coexistence of CDC with PD, the need for meticulous dissection and opening of the distal end to safeguard the DOW before distal transection. Placing an additional drain in the resultant space in the pancreatic head may be sufficient for the management of POPF grade B.

Ethical Considerations

Compliance with ethical guidelines

There were no ethical considerations to be considered in this article.

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Conflict of Interests

The authors have no conflict of interest to declare.

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