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Complete Agenesis of Dorsal Pancreas with Type 1 Choledochal Cyst- A Rare Association and its Surgical Implications: A Case Report

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Abstract

Introduction: Choledochal cysts (CDC) are common congenital biliary anomalies frequently associated with anomalous pancreaticobiliary junctions. Complete agenesis of the dorsal pancreas (ADP), resulting from failed development of the dorsal pancreatic bud, is a rare developmental anomaly and its coexistence with CDC is exceptionally uncommon, with only two cases previously reported.

Case Presentation: An 11-year-old girl presented with one-week history of upper abdominal pain, jaundice and vomiting. Laboratory evaluation suggested obstructive jaundice. Magnetic resonance cholangiopancreatography demonstrated a type I choledochal cyst with a long common channel and complete dorsal pancreatic agenesis. Following treatment of cholangitis, she underwent complete cyst excision with Roux-en-Y hepaticojejunostomy. Histopathology showed inflammatory changes without dysplasia. Recovery was uneventful and at 18-month follow-up she remained asymptomatic with normal liver function tests and no clinical or biochemical evidence of endocrine or exocrine pancreatic insufficiency.

Conclusion: This is the third reported case of choledochal cyst associated with complete dorsal pancreatic agenesis and the first reported in a female child. Preoperative recognition of this rare association on MRCP is important because it facilitates meticulous surgical planning, enables safe intrapancreatic dissection and identifies patients who require long-term surveillance for pancreatic endocrine and exocrine dysfunction.

Keywords: Pancreatic agenesis; Obstructive jaundice; Anomalous pancreaticobiliary junction; Congenital anomaly

Introduction

Choledochal Cysts (CDC) are among the commonest congenital biliary anomalies encountered in children, frequently associated with an Anomalous Pancreaticobiliary Junction (APBJ) and other developmental anomalies of the pancreaticobiliary system [1]. Agenesis of Dorsal Pancreas (ADP) is a rare congenital anomaly resulting from failed development of the dorsal pancreatic bud, leading to partial or complete absence of the body and tail of the pancreas. The condition is often discovered incidentally on cross-sectional imaging but may be associated with abdominal pain, pancreatitis, diabetes mellitus or exocrine pancreatic insufficiency [2]. The co-existence of these two anomalies is exceptionally uncommon, with only two cases described in the literature to date [2,3] **(Table 1)**.

Table 1: Details of the cases of Agenesis of Dorsal Pancreas associated with Choledochal cyst described in literature. MRI- Magnetic resonance imaging, CBD- Common bile duct, PTBD- Percutaneous transhepatic biliary drainage. NA- Not available.

Study	Year of publication	Age/ Gender	Presenting complaint	Diagnostic modality	Associations	Treatment given	Intra-operative issues	Follow-up period	Post-op complications

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Oyachi et al [2]	2006	9 year/ Male	Jaundice and Abdominal discomfort with features of pancreatitis	MRI	Nil	Underwent PTBD followed by Roux-en-Y hepatico-jejunostomy after treatment of pancreatitis	NA	NA	None
Chapa et al [3]	2021	23 year/ Male	Intermittent abdominal pain	MRI	Hirschsprung's disease	Not mentioned	NA	NA	NA
Index case		11year/ Female	Upper abdominal pain with features of cholangitis	MRI	Nil	Conservative for cholangitis followed by Roux-en-Y hepatico-jejunostomy	None	18 Months	None

Recognition of this association is clinically important because the altered pancreatic anatomy may influence distal bile duct dissection during surgery and necessitates long-term surveillance for endocrine and exocrine pancreatic dysfunction. We report the first female child with a type I choledochal cyst associated with complete dorsal pancreatic agenesis and discuss its diagnostic and surgical implications.

Case Report

An 11-year-old girl presented with upper abdominal pain for one week's duration, associated with non-bilious vomiting, progressive jaundice and dark-coloured urine. There were no history of fever, pruritus or pale stools or similar complaints in the past. Her developmental and family history was unremarkable. On examination, she was afebrile with stable hemodynamics, icteric with tenderness in the right hypochondrium and epigastrium. There was no palpable mass, organomegaly or ascites.

Laboratory investigations demonstrated obstructive jaundice with a total bilirubin of 5.4 mg/dL, direct bilirubin of 2.1 mg/dL and elevated alkaline phosphatase of 439 IU/L. Complete blood count, blood glucose, serum amylase and lipase were within normal limits. Abdominal ultrasonography revealed a distended gallbladder with dilated Common Bile Duct (CBD) and mild intrahepatic biliary radicle dilatation. To further delineate the biliary and pancreatic anatomy, Magnetic Resonance Cholangiopancreatography (MRCP) was performed which revealed fusiform dilatation of the common bile duct with a maximum diameter of 14 mm consistent with a type I choledochal cyst with a long common channel of 16mm **(Figure 1a)**. In addition, the pancreatic body and tail were not visualized. The dependent stomach sign, with direct apposition of the stomach to the splenic vein confirmed complete agenesis of the dorsal pancreas **(Figure 1b)**.

The patient was managed initially for cholangitis with intravenous third generation cephalosporins for a week. Her clinical condition improved with normalization of inflammatory markers and gradual resolution of jaundice.

She subsequently underwent elective surgery two months after the initial presentation. Intraoperatively, a fusiform dilatation of the extrahepatic bile duct measuring 2 cm in diameter was identified extending from common hepatic duct to the intrapancreatic portion of the CBD **(Figure 1c)**.

Complete excision of the cyst was performed up to the pancreaticobiliary junction, followed by reconstruction with a Roux-en-Y hepaticojejunostomy. The postoperative recovery was uneventful. Oral feeds were initiated on Postoperative Day (POD) 4 and was discharged on POD 8 in good clinical condition. Histopathology revealed inflammatory changes without evidence of dysplasia or malignancy.

At 18-month follow-up, she remained asymptomatic with normal liver function tests and no clinical or biochemical evidence of exocrine or endocrine pancreatic insufficiency.

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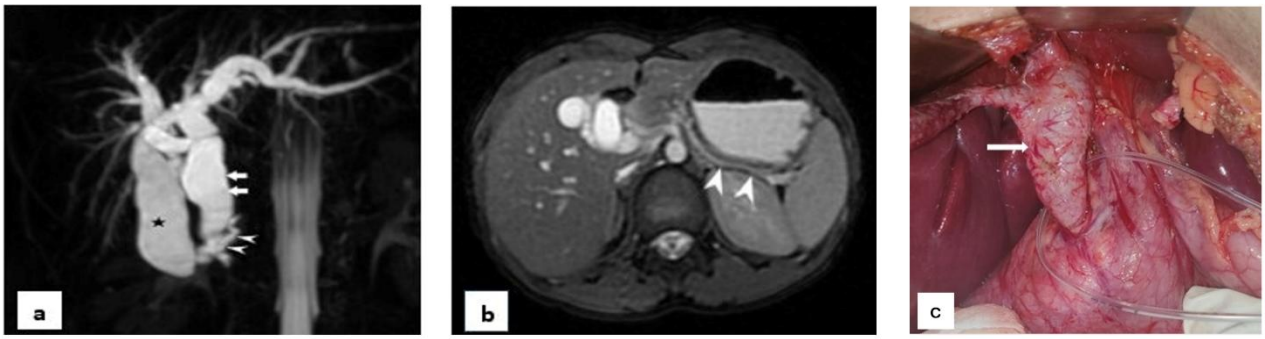

Figure: (a) Magnetic Resonance Cholangio Pancreatography (MRCP) showing fusiform dilatation of the entire Common Bile Duct (CBD) (arrows) with an Anomalous Pancreaticobiliary Junction (APBJ) (arrow heads) suggestive of choledochal cyst Type 1; (b) Axial image showing dependent stomach sign (arrowheads) suggestive of dorsal pancreatic agenesis; (c) Intra-operative image of the choledochal cyst after isolation from the porta (arrow).

Discussion

Choledochal cysts are congenital abnormal dilatation of the biliary tree with a higher incidence in Asian population and a female preponderance, as seen in our patient [4]. Commonly known association include an APBJ contributing to pathophysiology which may be seen in up-to 80% of cases [1]. Other associations include congenital cardiac anomalies and pancreas divisum, while an association with agenesis of the dorsal pancreas has only rarely been reported [2,3,5,6]. Notably, both previously reported patients were male in contrast to our female patient.

Embryologically, pancreas develops from a dorsal and a ventral bud arising from the foregut. The dorsal bud gives rise to body, tail and superior head with accessory duct of Santorini, whereas the ventral bud forms the inferior head and uncinate process bearing the main duct of Wirsung.

ADP is classified into three types: (I) total agenesis of the dorsal pancreas; (II) hypogenesis of the body and tail; (III) hypogenesis of the tail [7]. Our patient had complete (type I) dorsal agenesis. Because both anomalies arise from the foregut endoderm with an overlapping developmental window, a shared insult or disruption of common signalling pathways governing dorsal bud outgrowth and biliary morphogenesis may plausibly account for their coexistence. The clinical presentation is indistinguishable from that of isolated choledochal cysts and none of the reported cases have shown features specifically indicating ADP. Patients with ADP may develop endocrine insufficiency because most islet cells reside in the body and tail of the pancreas [8].

However, our patient showed no such features, most likely reflecting preserved endocrine reserve within the ventral pancreatic remnant. It is usually an incidental finding on imaging and MRCP is the modality of choice. An important imaging differential is pseudo-agenesis secondary to chronic pancreatitis or pancreatic lipomatosis.

The presence of the dependent stomach sign favours true dorsal pancreatic agenesis, as seen in our patient, whereas both pseudo-agenesis and pancreatic lipomatosis retain the pancreatic ductal anatomy; pancreatic lipomatosis additionally shows fatty replacement anterior to the splenic vein [9]. The treatment strategy mirrors that of an isolated choledochal cyst. Meticulous intra-pancreatic dissection to achieve complete cyst excision is preferred, both to minimize post-operative pancreatitis, which is seen in 4-5% of patients and to avoid malignant change in any residual cyst [10].

Apart from this, peri-operative monitoring for glycemic disturbance is advisable given the reduced pancreatic mass. Neither the previously reported cases nor our patient developed peri-operative complications attributable to ADP. Long-term clinical and biochemical surveillance is advisable to monitor for the development of exocrine or endocrine deficiencies and pancreatic cancer at a later life [9]. Our patient underwent successful complete cyst excision with Roux-en-Y hepaticojejunostomy and remains asymptomatic at 18 months of follow-up without evidence of pancreatic dysfunction. This case highlights the importance of systematically evaluating pancreatic morphology on preoperative MRCP in children with choledochal cysts, as recognition of associated anomalies may influence operative planning and postoperative surveillance. This case adds to the limited literature by highlighting the diagnostic and surgical implications of a rare association. However, as a single case with limited follow-up, the long-term pancreatic outcomes remain uncertain.

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Conclusion

Complete agenesis of the dorsal pancreas is a rare but clinically relevant anomaly that may coexist with choledochal cysts. Careful preoperative MRCP evaluation enables accurate diagnosis, facilitates meticulous surgical dissection and identifies patients who require long-term monitoring for endocrine and exocrine pancreatic dysfunction. Greater awareness of this association and additional reported cases is needed to better define its embryological basis and long-term outcomes.

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