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Case Report

Complex Pancreaticobiliary Maljunction with Pancreas Divisum and Obstructive Pseudocyst

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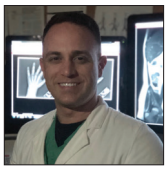

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ABSTRACT

Pancreaticobiliary maljunction (PBM) is the congenital junction of the bile and pancreatic ducts into a common channel located anatomically outside of the duodenal wall. The complex type (Type D) is the rarest type and occurs in conjunction with other pancreaticobiliary anomalies. We present a case of complex PBM with pancreas divisum presenting as acute pancreatitis and obstructive symptoms secondary to a pancreatic pseudocyst. Surgical management is discussed, as complex type PBM is most predisposed to post-operative complications.

Keywords: Pancreas divisum, Pancreatic pseudocyst, Pancreaticobiliary maljunction, Pancreatitis, Biliary cancer

INTRODUCTION

Pancreaticobiliary maljunction (PBM) is a rare congenital anomaly relating to the union of the pancreatic duct and common bile duct. Multiple theories regarding its embryological origin exist, though PBM likely arises from the abnormal fusion of the pancreatic duct and biliary tree which originate from different buds of the primitive foregut.[1] PBM is rare among non-Asian populations, reported at 1 per 100,000 individuals in the Western world. However, the incidence in East Asia is estimated to be 100–1000 times higher, with most cases reported in Japan.[2,3]

Differential considerations for PBM include choledochal cysts, annular pancreas, or pancreas divisum. These entities should be delineated using magnetic resonance cholangiopancreatography (MRCP). Ultrasound is often used in a screening capacity, but is alone an incomplete imaging workup for PBM. Endoscopic retrograde cholangiopancreatography (ERCP) is invasive, but should be used to diagnose equivocal cases.

In each of the four major PBM variations, designated as Types A-D, the common bile and main pancreatic ducts join early to form a long common pancreaticobiliary channel located anatomically outside of the duodenal wall.[4,5] PBM leads to regurgitation of pancreatic juice into the biliary system (pancreaticobiliary reflux) and bile into the pancreatic duct (biliopancreatic reflux) through the long common channel, predisposing patients to obstructing protein plugs, recurrent pancreatitis, biliary stones, and a 200 times greater lifetime risk for biliary cancer.[6] Early diagnosis and intervention are essential to mitigate these risks. Here, we discuss the presentation, diagnosis, and management of a complex (Type D) case of PBM – the rarest type of PBM which comprises <5% of cases.[5,7]

CASE REPORT

A 4-month-old female, born at term, was admitted with weight loss, fever (102 F), and progressively refractory non-bilious emesis. No congenital anomalies were detected in the patient's prenatal course. Physical examination was remarkable for a palpable epigastric abdominal mass. Ultrasound demonstrated a 6.4 cm pancreatic pseudocyst. Admission laboratories were significant for a markedly elevated serum lipase (2527 U/L) and leukocytosis (26,000/uL). The acute pancreatitis was managed non-operatively, and the child improved, feeding well enough to discharge from the hospital. However, over the next several weeks, she developed recurrent feeding intolerance as the pseudocyst appeared to enlarge and mature overtime.

Magnetic resonance imaging of the liver and pancreas was performed on a Siemens 3 Tesla scanner with 0.7 mL/kg intravenous gadoxetate disodium contrast demonstrating pancreas divisum [Figure 1] with the formation of a common channel between the distal common bile duct and the dorsal pancreatic duct of Santorini, which joined prematurely with the common bile duct into a 1.9 cm cystic malformation in the pancreatic head. The cystic malformation then connected with a complex network of ducts, including one thought to represent the duct of Wirsung and other smaller cysts that drained into the duodenum [Figure 2]. The distal common bile duct was normal in caliber without stricture or dilation. The common hepatic duct and cystic duct demonstrated normal morphology, but were slightly prominent.

A large, thick-walled pseudocyst in the anterior pancreatic body with an internal fluid-fluid level consistent with a pancreatic pseudocyst with internal debris, measuring up to 6.9 cm in length, exerted a mass effect on the pancreatic head and proximal pancreatic body. Ascites, thought to be secondary to prior pseudocyst rupture, was also demonstrated [Figure 3].

At the time of publication, the patient is clinically stable. The clinical team is allowing the patient time to grow before undergoing surgical intervention.

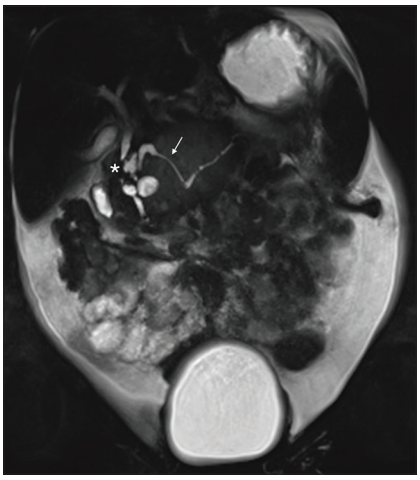

Figure 1: A 4-month-old female with emesis. Coronal T2 3D magnetic resonance cholangiopancreatography shows the pancreas divisum with the main pancreatic duct draining into the duct of Santorini (arrow). The common bile duct drains into a complex conglomeration of pancreatic head cysts (asterisk). The common bile duct is mildly prominent, and there is ascites.

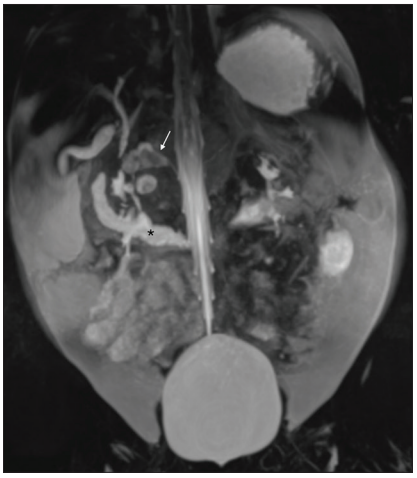

Figure 2: Same patient as above. Coronal T2 3D magnetic resonance cholangiopancreatography shows the 1.9 cm common cyst (arrow) that drains the duct of Santorini and the common bile duct. There are multiple cysts and ducts that extend throughout the pancreatic head with multiple connections to the duodenum (asterisk).

DISCUSSION

PBM is the anomalous congenital junction of the bile and pancreatic ducts into a common channel located anatomically outside of the duodenal wall.[4,5] The configuration of the common channel and presence of biliary tract dilation is variable in PBM, leading to various iterations of classification systems.[8] The 2015 committee on diagnostic criteria of the Japanese study group on PBM proposed a classification system of four types (A-D) which is simple and correlates with clinical features.[5] In stenotic type (Type A), a distal stenotic segment of common bile duct joins a non-dilated common channel. In non-stenotic type (Type B), the distal common bile duct and pancreatic duct taper gradually to form a common channel without focal stenosis or dilation. In dilated channel type (Type C), the narrowed distal segment of the common bile duct joins the common channel, and there is abrupt dilation of the common channel. Of the types of PBM, complex type (Type D) is the rarest, comprising <5% of cases of PBM.[5,7] The case presented is an example of complex type (Type D) PBM, wherein a complex network of interconnected cysts and channels with contributions from

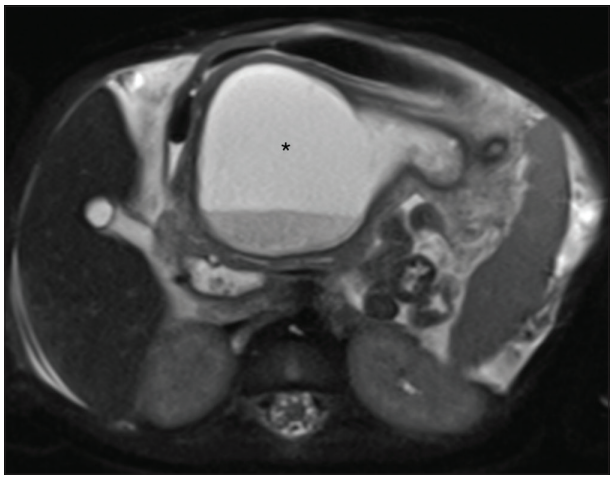

Figure 3: Same patient as above. Axial T2 fat-saturated image shows a large pancreatic pseudocyst (asterisk), with layering debris, exerting mass effect on the pancreatic head. There is ascites.

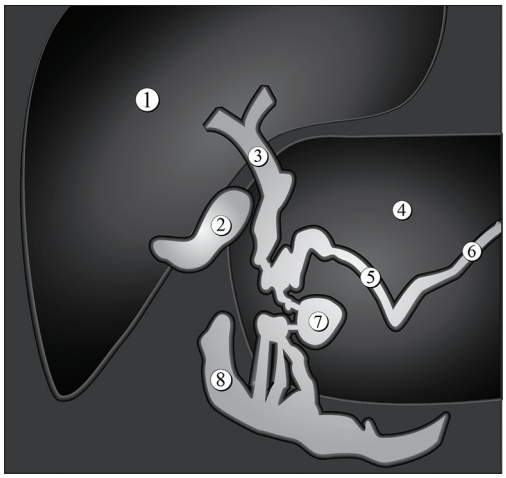

Figure 4: Diagram based on the same patient as above. (1) Liver, (2) gallbladder, (3) common bile duct, (4) pancreas, (5) duct of Santorini, (6) main pancreatic duct, (7) complex network of cysts and channels in the pancreatic head which extend to the duodenum, (8) duodenum. Of note, the pancreatic pseudocyst is not included in this diagram for simplicity and to focus on illustrating the congenital anomaly.

the pancreatic and biliary systems form outside the duodenal wall and drain to the duodenum [Figure 4].

In complex type (Type D), there is a complicated union of the pancreaticobiliary system in association with annular pancreas, pancreas divisum, or another complicated duct system. In the primitive foregut, the dorsal and ventral pancreatic buds normally fuse during embryonic week 7, resulting in a single drainage point of the main pancreatic duct to the duodenum at the sphincter of Oddi.[1,7] When the dorsal and ventral buds fail to fuse, there is separate or predominantly separate drainage through a minor papilla (dorsal duct) and major papilla (ventral duct), resulting in pancreas divisum. PBM and pancreas divisum are likely associated based on abnormal interaction of the embryological foregut elements – including the dorsal and ventral pancreatic buds with the hepatic diverticulum (which gives rise to the extrahepatic bile ducts) – between embryological weeks 4 and 7.[1,7,9] This anomalous configuration is illustrated in the case, which includes the elements of pancreas divisum and complex PBM.

Thorough evaluation of pancreaticobiliary anatomy is essential to make the correct diagnosis and guide patient care. MRCP is the preferred, non-invasive diagnostic imaging technique to assess pancreaticobiliary configuration and associated anomalies. Ultrasound may be used for screening when there is suspicion for pancreaticobiliary anomalies, but sonography alone offers an incomplete evaluation. In addition, invasive methods such as ERCP may be employed to map the pancreaticobiliary ductal system, especially in equivocal cases. ERCP may also contribute to patient management. Thorough understanding of the pancreaticobiliary system is essential for management and operative planning.[4] PBM causes various hepatobiliary and pancreatic disorders, including pancreatitis, biliary carcinoma, and cholangitis. There is also an association between Type I choledochal cysts and stenotic type PBM. As in our case, the patient presented with symptoms of bowel obstruction secondary to mass effect from a large pancreatic pseudocyst from chronic pancreatitis.

With the presence of a long common pancreaticobiliary channel, regurgitation of pancreatic juice into the biliary system (pancreaticobiliary reflux) and of bile into the pancreatic duct (biliopancreatic reflux) can occur.[10] This reciprocal reflux has been implicated in inhibiting excretion of bile and pancreatic juice, recurrent pancreatitis, and biliary cancer.[4,10] As such, patients with incidental or asymptomatic PBM should undergo prophylactic flow diversion surgery early to prevent malignant degeneration.

Standard management of PBM is the excision of the extrahepatic bile duct. However, in patients with complex type PBM, the concomitant congenital pancreatic anomalies, such as pancreas divisum as in our case, may predispose the patient to chronic pancreatitis even after excision of the extrahepatic bile duct. Additional strategies, such as sphincterotomy, may be considered in cases with pancreatic anomalies to lower chronic post-operative complications. The complex type PBM is most likely to have late post-operative complications, such as recurrent pancreatitis or protein plugs.[5]

CONCLUSION

PBM may present in unexpected ways based on the patient's associated anomalies or related complications. When biliary or pancreatic anomalies are suspected clinically, there should be thorough diagnostic imaging evaluation to assess for the

presence of PBM. Although rare, PBM should be managed surgically. Early intervention is a key to prevent chronic complications, including biliary malignancy.

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Declaration of patient consent

Patient's consent not required as patient's identity is not disclosed or compromised.

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Conflicts of interest

There are no conflicts of interest.

Disclaimer

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