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Case Report

Recurrent Acute Pancreatitis with Anomalous Pancreaticobiliary Ductal Union (Komi's Type IIIc3) with Santorinocele in a Child: A Rare Case Report

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Abstract

Anomalous pancreaticobiliary ductal union (APBDU) can cause recurrent acute pancreatitis. We describe the case of a 16-year-old boy with recurrent acute pancreatitis. The discussion provides a review of recent literature, supporting use of various diagnostic modalities and surgery as choice of treatment.

Keywords: *Anomalous pancreaticobiliary ductal union, choledochal cyst, Komi classification, recurrent acute pancreatitis, santorinocele*

Introduction

Recurrent acute pancreatitis is defined as more than two discrete episodes of diagnosed acute pancreatitis at least 2 months apart with asymptomatic interval period. The most common causes are alcohol abuse and gallstone disease. Anomalous pancreaticobiliary ductal union (APBDU) has been described as cause for idiopathic recurrent acute pancreatitis in both pediatric and adult population. This anomaly can cause acute pancreatitis, recurrent acute pancreatitis, and recurrent cholangitis.[1] It should be diagnosed and surgically treated before complications or carcinomas develop.[1,2]

We report a case of an APBDU with choledochal cyst and santorinocele (with calculi) associated with probable choledocholithiasis presented with recurrent acute pancreatitis and complicated by acute cholangitis.

Case Report

We admitted a 16-year-old boy with recurrent abdominal pain with a history of acute necrotizing pancreatitis (contrast-enhanced computed tomography [CECT] done elsewhere) 6 months before present admission. Etiological factors including trauma, drug, family history, and metabolic factors were excluded from the study. He was again admitted 2 months later with mild acute pancreatitis elsewhere.

On evaluation, magnetic resonance cholangiopancreatography (MRCP) [Figure 1 above] and CECT showed that common hepatic duct was dilated till terminal portion (choledochal cyst) with T2 hyperintense focus in terminal common bile duct (CBD) near ampulla suggestive of calculi, mildly prominent pancreatic duct draining separately into the second part of duodenum (like classic pancreas divisum) with moderate bilobar intrahepatic biliary dilatation with liver function test (LFT) of bilirubin total and direct of 1.2/0.4 mg/dl, aspartate aminotransferase (AST)/alanine aminotransferase (ALT) of 112/294 (<40 IU/L) IU/L, and alkaline phosphatase (ALP) of 610 U/L (<125 IU/L) and normal amylase and lipase. Physical examination was normal.

He developed high-grade fever with jaundice in hospital (bilirubin total and direct 3.6/2.6 mg/dl, AST/ALT 204/369 IU/l and ALP 902 IU/l). Endoscopic retrograde cholangiopancreatography (ERCP) with sphincterotomy with plastic stenting was done; no CBD calculi noted. His fever resolved and LFT improved, but abdominal pain was persisting. Repeat ERCP for CBD clearance and minor papillotomy was performed. Cholangiogram revealed complex connection of CBD, main pancreatic duct (MPD), and accessory pancreatic duct (APD) with cystic dilatation in APD [Figure 1 below]. Cholangiogram revealed no

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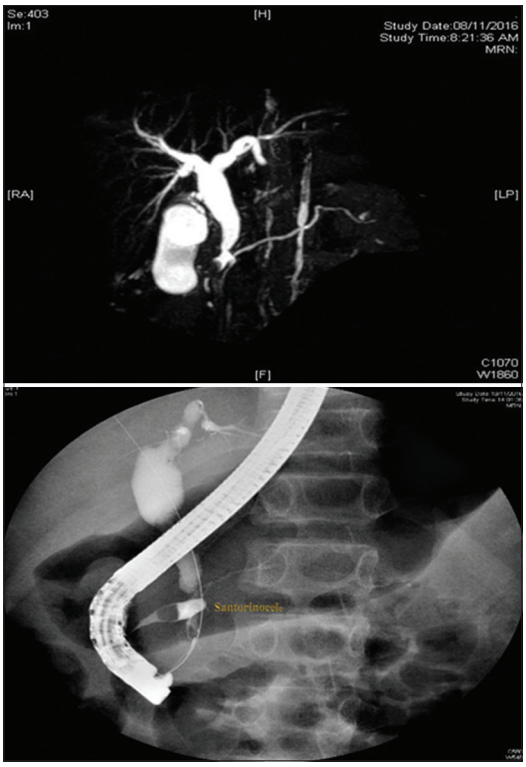

Figure 1: Preprocedure magnetic resonance cholangiopancreatography image (above) and endoscopic retrograde cholangiopancreatography image - santorinocele (below)

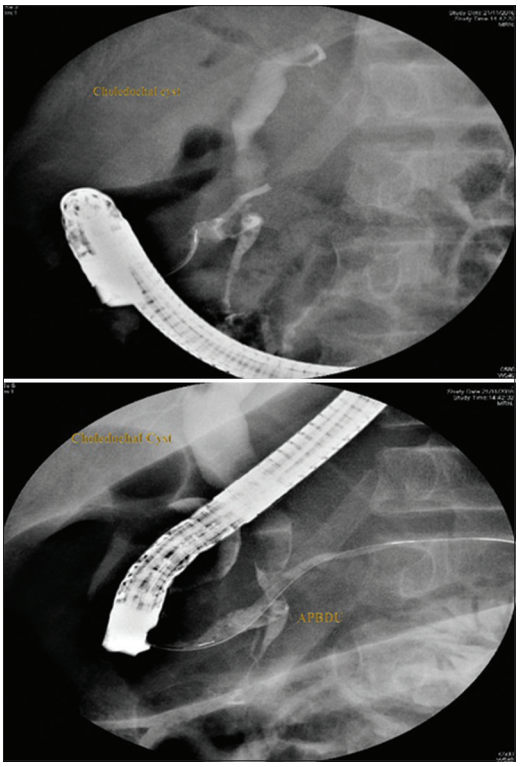

Figure 2: Choledochal cyst and anomalous pancreaticobiliary ductal union in endoscopic retrograde cholangiopancreatography images

CBD filling defect; contrast through major papilla also filled MPD and APD with a 7 mm filling defect in APD before its opening to minor papilla and pancreaticogram through minor papilla also filled MPD and CBD with both MPD and APD appeared to additionally communicate with CBD [Figure 2 above and below]. APD stenting was done following minor duct papillotomy and stent was removed 5 days later. The patient remained asymptomatic in terms of abdominal pain and there were no further episodes of fever or jaundice on 3-month follow-up. Need for cholecystectomy and pancreatectoduodenectomy was discussed with relatives.

Discussion

In APBDU, pancreatic and bile ducts join outside the duodenum and form a long common channel (usually >15 mm). The anomalous union is often associated with a choledochal cyst.[3] Association of APBDU with choledochal cyst may be present in up to 90% of cases.[4]

Gallbladder carcinomas develop in 15%–40%, with lesser frequency if associated with choledochal cyst.[2]

APBDU is a rare anomaly with frequency of 1.5%–3.2%.[4] Its diagnosis needs a high index of suspicion. Komi[5] proposed a new classification of APBDU [Figure 3], dividing it into three types. In Type III, APBDU is complicated with a patent APD with or without complicated network of ducts and is subclassified into Types IIIa, IIIb, and IIIc.[4] There are scarce data from India,[6,7] and none of them provides the prevalence of APBDU, in particular the new Komi Type III.

APBDU and choledochal cyst association[6] may be due to congenital weakness of CBD and reflux of the pancreatic juices in CBD. Subtype IIIC3 APBDU has been reported to be associated with ductal calculi in up to 43.8% of cases, but its mere presence causes symptoms is controversial.[8]

Santorinicele is a focal cystic dilatation of (which may contain protein plug or calculi) the termination of APD at the minor papilla.[9] This anomaly has been described in patients with pancreas divisum and recurrent acute pancreatitis.[9] Pancreas divisum as a cause for acute recurrent pancreatitis is still debatable and undiagnosed santorinocele with calculi/plug or APBUD may be playing a role in those cases.

Patients with pancreas divisum who may get benefit from endoscopic therapy is still debatable.[8] Santorinicele is due to stenotic accessory duct papilla or cause of relapsing dysfunction remains controversial. As per previous studies, the presence of santorinocele may help in selecting patients who might benefit from endoscopic treatment.[9]

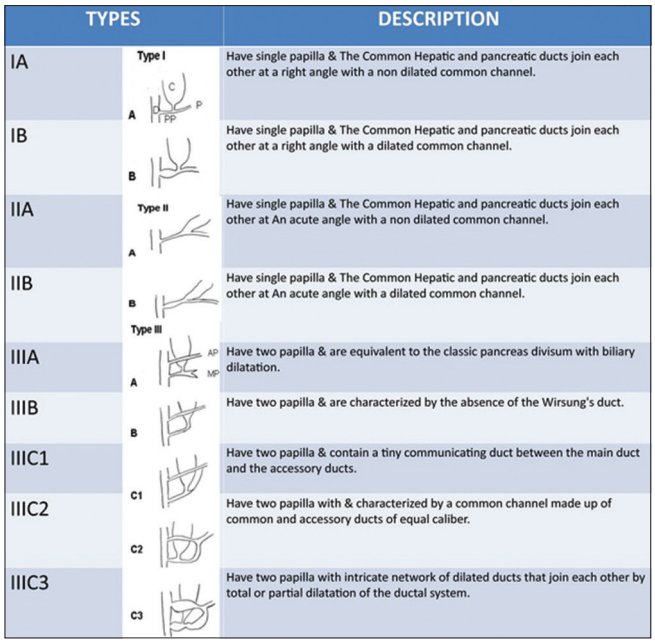

Figure 3: Komi's classification of anomalous pancreaticobiliary ductal union

ERCP is the investigation of choice;[7] MRCP has up to 82% detection rate. Endoscopic ultrasound has been reported as a good method for the detection of APDU.[10] Our patient had separate major and minor papillae, communicating with each other and with the dilated CBD and santorinocele (with probable calculi) without obvious CBD obstruction (choledochocele). After reviewing the literature, we found that this pattern was consistent with the Type IIIC3 anomaly.[5] Type IB, IIB, and IIIC3 APBDU are particularly associated with recurrent pancreatitis and are treated surgically, pylorus-preserving pancreatectoduodenectomy being the choice for Type IIIC3.[6] In APBDU patient without dilated CBD, cancer risk is low, and hence, cholecystectomy without duct resection is recommended in few studies.

Conclusion

In patients with idiopathic recurrent acute pancreatitis, particularly young patients, APBDU should be a differential. The complications and long-term sequelae of acute pancreatitis and risk of cancer in young patients warrant early and judicious management.[6,7]

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Conflicts of interest

There are no conflicts of interest.

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