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A RARE CASE OF CHOLEDOCHAL CYST WITH INCOMPLETE PANCREATIC DIVISUM AND ABNORMAL PANCREATICOBILIARY MALUNION

Gastroenterology

Bontha Vineesha — Senior resident, Department of Medical Gastroenterology, Sri Ramachandra Institute of Higher Education and Research.

Goutham Reddy Katkuri — Senior resident, Department of Medical Gastroenterology, Sri Ramachandra Institute of Higher Education and Research.

Shanmuganathan Subramanyam * — Professor, Department of Medical Gastroenterology, Sri Ramachandra Institute of Higher Education and Research. *Corresponding Author

Ganesh P — Professor and Head, Department of Medical Gastroenterology, Sri Ramachandra Institute of Higher Education and Research.

ABSTRACT

Abnormal pancreaticobiliary malunion (APBDU) is characterized by a junction of the bile duct and pancreatic duct outside the duodenal wall with a long common ductal channel leading to the duodenal lumen. APBDU may result from failure of the embryological ducts to migrate fully into the duodenum. Pancreas divisum, occurs from a fusion failure of the ventral and dorsal pancreatic buds, and characterized by a dominant santorine duct, is considered to be a predisposing factor to recurrent attacks of acute pancreatitis. In incomplete pancreas divisum, the ventral and dorsal pancreas are connected by a segmental branch. AUPBD frequently associated with choledochal cyst may have an implication not only as an etiological factor but as an associated disorder leading to a complicated clinical course. Our patient had a complex (type IV) type of APBDU which was associated with choledochal cyst and pancreas divisum. In children with type D, recurrent pancreatitis may occur even after excisional surgery due to associated pancreatic anomalies.

KEYWORDS

Choledochal cyst, pancreatic divisum, abnormal pancreatico biliary malunion.

CASE REORT:

A 16 year-old female child with no significant past illness presented to the outpatient department with 10-day history of abdominal pain and high colored urine. The pain was localized to the right upper quadrant, associated with nausea and vomiting and was intermittent, each episode lasting for 3-4 hours. The patient reported normal colored stools. Patient did not report fever, pale stools or pruritus during the course of illness. Examination was remarkable for scleral icterus and mild tenderness over the right hypochondrium. Blood investigations showed cholestatic jaundice (total bilirubin: 2.99 mg/dL, direct bilirubin: 1.63mg/dL, alkaline phosphatase: 319 IU/L, alanine transaminase: 204 IU/L, aspartate transaminase: 125 IU/L, serum albumin: 4.3 g/dL). Abdominal ultrasonography showed a normal liver size and echo texture, dilated common bile duct (1.8 cm) along with dilated intra hepatic biliary radicles. Multiple calculi were noted in the gallbladder, with wall thickening suggestive of calculous cholecystitis. An MRCP (Magnetic resonance cholangiopancreatography) was done (Figures 1-5). The findings were suggestive of a choledochal cyst (type IV a) along with a complex type of anomalous pancreato-biliary ductal union and a variant of pancreas divisum with choledocholithiasis. Patient was taken up for ERCP (Endoscopic retrograde cholangiopancreatography) under general anesthesia. Ampulla was noted deep in 2nd part of duodenum (Figure 6). Selective bile duct cannulation was done and guidewire was placed into the right intrahepatic duct. Cholangiogram (Figure 7) confirmed the findings of MRCP. Endoscopic biliary sphincterotomy was done. Balloon trawling was done to extract CBD stones. Multiple tiny calculi and sludge was extracted. Biliary stenting was done using a double pigtail stent of size 7Fr (Figure 8). Patient improved symptomatically and liver functions normalized over a week. Patient was referred to a specialized center for surgery.

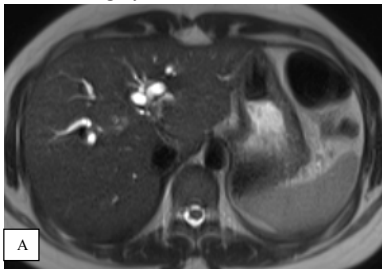

Figure 1: Axial T2W sequence shows mild to moderate dilatation of the intrahepatic biliary radicals (left arrow) involving the left lobe of the liver.

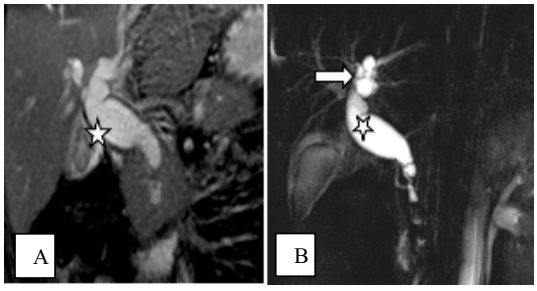

Figure 2A & 2B: Coronal T2W and reformatted 3D coronal MRCP shows cystic dilatation of the common bile duct (star) with segmental dilatation of the proximal intrahepatic biliary duct (right arrow) and abrupt narrowing of the distal end suggestive of choledochal cyst

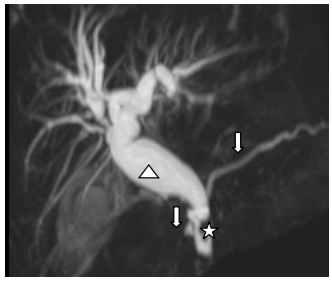

Figure 3: Reformatted 3D Coronal MRCP shows the abnormal course of the main pancreatic duct (down arrow) draining the body and tail of the pancreas which is seen to cross the CBD and finally unite at the stenotic CBD (star) distal the choledochal cyst (triangle).

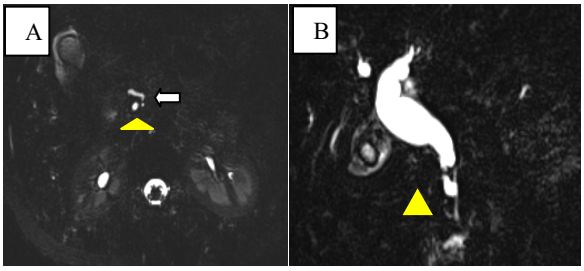

Figure 4a and 4b: Axial and Coronal 3D MRCP shows an abnormal

channel (yellow triangle) arising from the minor papillae which is seen to unite with main pancreatic duct (left arrow) proximal to the unification into the stenotic CBD.

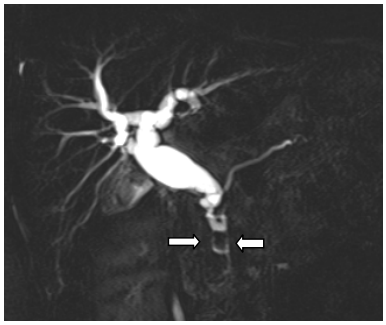

Figure 5: Reformatted Coronal 3D MRCP shows a minor pancreatic duct(left arrow) draining the head and uncinate process uniting with abnormal duct channel(right arrow) distal to the stenotic CBD and forms a common channel(star) draining into the third portion of the duodenum.

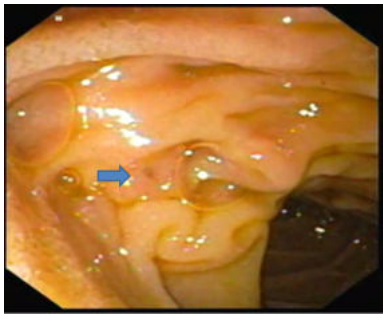

Figure 6: Duodenoscopy image of major papilla. Ampulla was normal (blue arrow), but was noted deep in 2nd part of duodenum

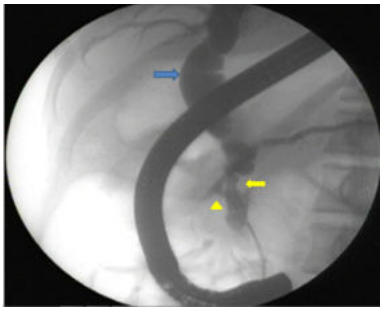

Figure 7 : Cholangiogram shows cystic dilatation of CBD (choledochal cyst)(blue arrow) along with long common channel (APBU)(yellow arrow) and a variant of pancreas divisum(yellow arrow head)

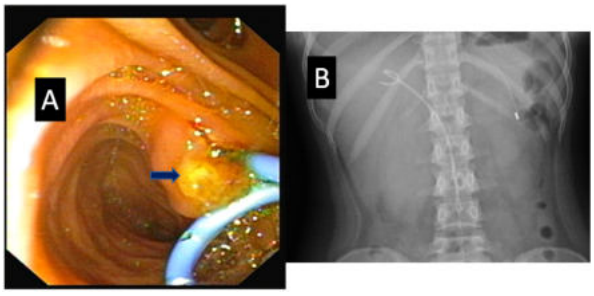

Figure 8a: Duodenoscopic image of post EST with CBD stent insitu with extracted sludge(blue arrow)
Figure 8b: Post procedure X-ray abdomen showing stent in situ

DISCUSSION:

Choledochal cyst is a spectrum of bile duct malformations characterized by dilatation of all or part of extrahepatic and/or intrahepatic bile ducts. Traditionally, choledochal cyst has been classified into 5 subtypes by Todani [1]. The incidence of choledochal cysts has wide geographical differences with Asian population having a higher incidence of 1:1000 as compared to 1:100,000 in the west [2-3]. One of hypothesis for pathogenesis of Todani types I and IV choledochal cyst has been suggested that the long common channel in abnormal pancreatobiliary ductal union (APBDU) induces a reflux of pancreatic juice into the bile duct (because of the higher pressure in the pancreatic duct compared with the bile duct), leading to inflammation, denudation, thinning of the ductal wall, and distal obstruction, resulting in cystic dilatation of the bile duct □.

APBDU is characterized by a junction of the bile duct and pancreatic duct outside the duodenal wall with a long common ductal channel leading to the duodenal lumen. APBDU may result from failure of the embryological ducts to migrate fully into the duodenum □. In our patient the papilla was noted deep in second part of duodenum. In a series published by Li L et al, in patients with APBDU, the distal displacement of the papilla was corresponding to the length of the common channel □.

ABPJ is present in 50 to 80 percent of patients with biliary cysts □⁻□. Complications or associated conditions with a choledochal cyst and APBDU are recurrent cholangitis, choledocholithiasis, bile duct and gallbladder cancer, and peritonitis caused by spontaneous perforation □, □⁻¹□. In one of the studies which compared AUPBD with and without choledochal cyst, there were no significant differences between the AUPBD-present group and the AUPBD-absent group in the incidence of gallstone disease, while the incidence of acute inflammation was 93% (26/28) in the AUPBD-absent group ($p < 0.01$). Carcinoma developed only in the AUPBD-present group (9/28, 32%) ($p < 0.05$). Pancreatic disorders (i.e. pancreatic stone, pancreatitis or pancreatic cancer) occurred in 12 of 28 cases in the AUPBD-present group (43%), while only in 1 of 16 cases in the AUPBD-absent group (6%) ($p < 0.05$). [11]

The Committee on Diagnostic Criteria of JSGPM has proposed a classification of APBDU into four types: (A) stenotic type, (B) non-stenotic type, (C) dilated channel type, and (D) complex type. [12]

Type A (stenotic type): The stenotic or narrow segment of the distal common bile duct joins the common channel, and dilatation of the common bile duct is seen.

Type B (non-stenotic type): The distal common bile duct without any stenotic or narrow segment smoothly joins the common channel. Localized dilatation of the common channel is not seen.

Type C (dilated channel type): The common channel is dilated. The narrow segment of the distal common bile duct joins the common channel, and abrupt dilatation of the common channel is seen.

Type D (complex type): Complicated union of the pancreaticobiliary ductal system as follows: PBM associated with annular pancreas, pancreas divisum, or other complicated duct systems.

Our patient had a complex (type IV) type of APBDU. In children with type D, recurrent pancreatitis may occur even after excisional surgery due to associated pancreatic anomalies.[12]

AUPBD frequently associated with choledochal cyst may have an implication not only as an etiological factor but as an associated disorder leading to a complicated clinical course. Therefore, we should make an effort to confirm the presence of AUPBD in patients with choledochal cyst. Adequate surgery may be required to prevent the occurrence of cancer.

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