A case series of choledochal cyst with pancreatic divisum: A rare association

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ABSTRACT
Choledochal cysts (CC) are congenital cystic dilations of the biliary tree usually associated with abnormal pancreaticobiliary ductal junction (APBDJ), but its association with pancreatic divisum has been rarely described. We encountered four cases of CC associated with pancreatic divisum (PD). Three had Type 3 PD and one had Type 1 PD. Two cases presented with pancreatic complications, with one case requiring preoperative minor papilla sphincterotomy for recurrent pancreatitis. The association of CC with PD is infrequent, and the variable presentation alters management strategy. PD may be one of the factors responsible for complications associated with CC.

Keywords: Choledochal cyst, pancreatic divisum, abnormal pancreaticobiliary junction

INTRODUCTION
Choledochal cysts (CC) are congenital cystic dilation of the biliary tree predominantly reported in the first decade of life (1). The pathogenesis of CC remains unclear. However, the most accepted theory is the presence of anomalous pancreaticobiliary duct junction (APBDJ) and long common channel (2). This is hypothesized to cause the reflux of pancreatic enzymes into the common bile duct leading to inflammation and duct ectasia, and reflux into the pancreatic duct causing pancreatitis (3,4). Pancreatic divisum (PD) is the most common congenital anomaly of the pancreas, which results when the embryological ventral and dorsal buds fail to fuse (complete PD) or fuse partially (incomplete PD). Although rare, PD can cause intraductal hypertension leading to abdominal pain and recurrent pancreatitis (5). The co-existence of CC and PD and the associated complications are rare, with less than ten documented cases in the literature (6). This report reviews our experience of four cases of CC with PD.

Case 1
A 20-year-old female patient presented with a history of epigastric pain for seven days. On clinical examination, the patient had tachycardia and epigastric tenderness. Her serum amylase and lipase levels were 900 IU/L and 726 IU/L, which was more than three times above the normal limits. Ultrasonography (USG) of the abdomen showed dilated common bile duct (CBD) of two centimeters with edematous pancreatitis which responded to conservative management. On MRCP, diagnoses of Todani Type IV A CC (7) with APBDJ and Type 3 pancreatic divisum were made. The patient underwent elective complete CC excision six weeks after recovery from pancreatitis. Postoperative course was uneventful. On follow-up for five years, the patient is doing well.

Case 2
A 28-year-old male patient presented with recurrent episodes of abdominal pain for 12 months. On clinical examination, the patient had tachycardia and epigastric tenderness. Her serum amylase and lipase levels were 900 IU/L and 726 IU/L, which was more than three times above the normal limits. Ultrasonography (USG) of the abdomen showed dilated common bile duct (CBD) of two centimeters with edematous pancreatitis which responded to conservative management. On MRCP, diagnoses of Todani Type IV A CC (7) with APBDJ and Type 3 pancreatic divisum were made. The patient underwent elective complete CC excision six weeks after recovery from pancreatitis. Postoperative course was uneventful. On follow-up for five years, the patient is doing well.
On postoperative day (POD) two, he developed acute pancreatitis with peripancreatic necrosis with a computed tomography severity index (CTSI) score of ten. On POD ten, the patient developed haemobilia with jaundice and melena, requiring two units of blood transfusion. Conventional angiogram did not reveal any active source of the bleed, and eventually, melena subsided with no drop in hemoglobin over the subsequent days. He developed sepsis with a total leukocyte count of 35,000 cells/mm$^3$. Necrosis was drained percutaneously. Drain fluid culture showed multi-drug resistant *Escherichia coli*. The patient did not respond to these interventions; a step-up approach was followed, and on POD 23 underwent surgical necrosectomy and drainage. Following surgery, the patient required prolonged ICU care with ventilator assistance. He eventually improved and was discharged on POD 44 in stable condition, but the patient was lost to follow up.

**Case 3**

A 12-year-old girl, with pain in the abdomen and vomiting ongoing for one month, was diagnosed with Type IV A CC of two centimeters with complete PD on MRCP and normal biochemical investigations. The patient underwent complete cyst excision with an uneventful postoperative course. She was discharged on POD six. The patient is currently doing well after five years of follow-up.

**Case 4**

A 35-year-old female patient presented with pain in the abdomen ongoing for 15 days. MRCP showed Type I CC with incomplete Type 3 pancreatic divisum (PD). The patient underwent complete excision of the cyst but developed a low output biliary leak, which was managed conservatively. The patient has been on regular follow-up and asymptomatic at five months of follow-up.

**DISCUSSION**

The presence of pancreatic divisum rules out APBDJ as the only cause of CC as most of the pancreatic juice drains through the dorsal duct into the minor papilla, which has no communication with the biliary system (8,9).

Although cases with PD rarely cause any symptoms, they have been implicated with acute pancreatitis in 25-38% of the patients (8). In addition, Cotton et al. have suggested the importance of the co-existence of stenotic accessory pancreatic duct along with PD in the causation of pancreatitis (10). The risk factors mentioned above for acute pancreatitis, although rare, also increase the risk of postoperative pancreatitis (11). In our study, one case with preoperative pancreatitis developed severe necrotizing pancreatitis in the postoperative period requiring operative intervention, which is a rare complication and has not been reported previously.

The incidence of acute pancreatitis with CC is 0% to 70.6% in children and 10% to 54.5% in adults (11). The incidence of acute pancreatitis in our series of adult CC was 3.7%. The causative factors described for acute pancreatitis with CC are fusiform dilatation with a non-stenotic distal bile duct above the common
Choledochal cyst with pancreatic divisum

channel, a dilated common channel, and protein plugs in the common channel, which leads to reflux of bile into the pancreatic duct, and therefore leading to increased incidence of acute pancreatitis (11–13).

As there is an uncertainty of PD being a causative factor in acute/chronic pancreatitis and the risks associated with the treatment options, the approach to the management of PD should be based on the clinical presentation. Treatment options available are endoscopic sphincterotomy or surgical sphincterotomy with sphincteroplasty.

A few authors have managed PD associated with Type 3 CC (choledochocele) with either endoscopic sphincteroplasty or sphincterotomy as therapeutic modality (14). Hackert et al. have reported a case treated with bile duct resection, papillotomy, hepaticejunostomy, and jejunal reinsertion of the uncinate pancreatic duct, which is an infrequently performed procedure (15). Other types of CC associated with PD could be managed with preoperative minor papilla sphincterotomy with or without stenting or trans-duodenal papillectomy followed by CC excision (6,9,16,17). In our study, two patients with CC and PD presented with acute pancreatitis. Only one patient required preoperative minor papilla sphincterotomy due to recurrent pancreatitis.

CONCLUSION

In conclusion, CC with PD is a rare congenital anomaly; PD may be an additional factor along with APBDJ for complications associated with CC, but larger sample size studies are required to prove this. Management strategy of these cases varies with the presence of complications.

REFERENCES


Pankreas divisum ile birlikte koledok kisti olgu serisi: Nadir bir birliktelik

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ÖZET


Anahtar Kelimeler: Koledokal kist, pankreas divisum, anormal pankreatikobiliyer bileşke

DOI: 10.47717/turkjsurg.2022.5609