CASE SERIES
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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 pain abdomen for 12 months. The patient had recurrent episodes of mild acute pancreatitis for which he was evaluated with MRCP and found to have Type I CC (two centimeters) with Type 1 PD and underwent minor papilla sphincterotomy elsewhere (Figure 1). Three months after acute pancreatitis attack, the patient underwent CC excision.

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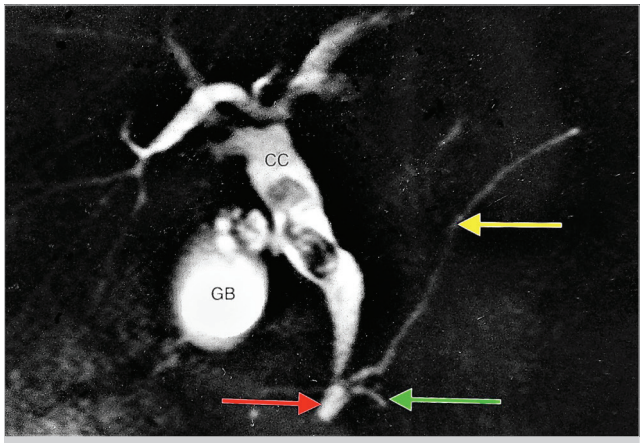

Figure 1. Magnetic resonance cholangiopancreatography (MRCP) of choledochal cyst (CC) with pancreatic divisum (PD). Yellow arrow; dorsal pancreatic duct draining separately into the minor papilla with no communication with the ventral duct (green arrow) suggestive of complete PD. Red arrow; long common channel formed by common bile duct and ventral duct draining into the major papilla.

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 (Figure 2). 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.

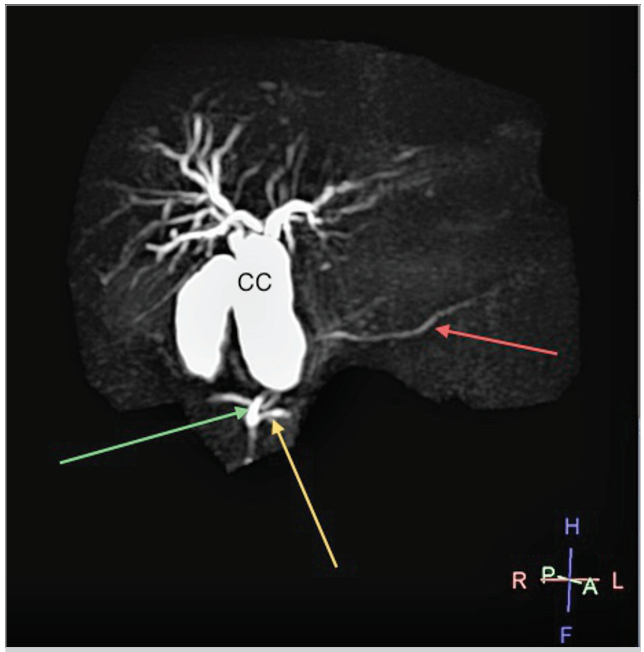

Figure 2. Magnetic resonance cholangiopancreatography (MRCP) choledochal cyst (CC) with incomplete (Type 3) pancreatic divisum (PD). Red arrow; dorsal pancreatic duct draining separately into the minor papilla with filamentous communication with the ventral duct (yellow arrow) suggestive of incomplete PD. Green arrow; long common channel formed by common bile duct and ventral duct draining into the major papilla.

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

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, papillectomy, hepaticojejunostomy, 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.

Ethics Committee Approval: This study was approved by Nizam's Institute of Medical Sciences Institutional Ethics Committee (Decision no: EC/NIMS/2940/2022, Date: 25.01.2022).

Informed Consent: Informed consent was obtained from the son of the patients.

Peer-review: Externally peer-reviewed.

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REFERENCES

1. *Urushihara N, Fukumoto K, Fukuzawa H, Mitsunaga M, Watanabe K, Aoba T, et al. Long-term outcomes after excision of choledochal cysts in a single institution: Operative procedures and late complications. J Pediatr Surg 2012; 47(12): 2169-74. https://doi.org/10.1016/j.jpedsurg.2012.09.001*
2. *Saluja SS, Nayeem M, Sharma BC, Bora G, Mishra PK. Management of choledochal cysts and their complications. Am Surg 2012; 78(3): 284-90. https://doi.org/10.1177/000313481207800334*
3. *Watanabe Y, Toki A, Todani T. Bile duct cancer developed after cyst excision for choledochal cyst. J Hepatobiliary Pancreat Surg 1999; 6(3): 207-12. https://doi.org/10.1007/s005340050108*
4. *Lal R, Agarwal S, Shivhare R, Kumar A, Sikora SS, Kapoor VK, et al. Management of complicated choledochal cysts. Dig Surg 2008; 24(6): 456-62. https://doi.org/10.1159/000111821*
5. *Ferri V, Vicente E, Quijano Y, Ielpo B, Duran H, Diaz E, et al. Diagnosis and treatment of pancreas divisum: A literature review. Hepatobiliary Pancreat Dis Int 2019; 18(4): 332-6. https://doi.org/10.1016/j.hbpd.2019.05.004*
6. *Ransom-Rodríguez A, Blachman-Braun R, Sánchez-García Ramos E, Varela-Prieto J, Rosas-Lezama E, Mercado MÁ. A rare case of choledochal cyst with pancreas divisum: Case presentation and literature review. Ann Hepatobiliary Pancreat Surg 2017; 21(1): 52-6. https://doi.org/10.14701/ahbps.2017.21.1.52*
7. *Todani T, Watanabe Y, Narusue M, Tabuchi K, Okajima K. Congenital bile duct cysts: Classification, operative procedures, and review of thirty-seven cases including cancer arising from choledochal cyst. Am J Surg 1977; 134(2): 263-9. https://doi.org/10.1016/0002-9610(77)90359-2*
8. *Kuzel AR, Lodhi MU, Rahim M. Pancreatic divisum: An unusual cause of chronic pancreatitis in a young patient. Cureus 2017 ; 9(11): e1856. https://doi.org/10.7759/cureus.1856*
9. *Arulprakash S, Balamurali R, Pugazhendhi T, Kumar SJ. Pancreas divisum and choledochal cyst. Indian J Med Sci 2009; 63(5): 198-201. https://doi.org/10.4103/0019-5359.53166*
10. *Cotton PB. Congenital anomaly of pancreas divisum as cause of obstructive pain and pancreatitis. Gut 1980; 21(2): 105-14. https://doi.org/10.1136/gut.21.2.105*
11. *Fujishiro J, Masumoto K, Urita Y, Shinkai T, Gotoh C. Pancreatic complications in pediatric choledochal cysts. J Pediatr Surg 2013; 48(9): 1897-902. https://doi.org/10.1016/j.jpedsurg.2012.12.038*
12. *Kaneko K, Ando H, Seo T, Ono Y, Tainaka T, Sumida W. Proteomic analysis of protein plugs: Causative agent of symptoms in patients with choledochal cyst. Dig Dis Sci 2007; 52(8): 1979-86. https://doi.org/10.1007/s10620-006-9398-4*
13. *Muthucumaru M, Ljuhar D, Panabokke G, Paul E, Nataraja R, Ferguson P, et al. Acute pancreatitis complicating choledochal cysts in children. J Paediatr Child Health 2017; 53(3): 291-4. https://doi.org/10.1111/jpc.13380*
14. *Sonoda M, Sato M, Miyauchi Y, Yazumi S, Nakamura M. A rare case of choledochocele associated with pancreas divisum. Pediatr Surg Int 2009; 25(11): 991-4. https://doi.org/10.1007/s00383-009-2460-5*
15. *Hackert T, Hartwig W, Werner J. Symptoms and surgical management of a distal choledochal cyst in a patient with pancreas divisum: Case report and review of the literature. Case Rep Gastroenterol 2007; 1(1): 90-5. https://doi.org/10.1159/000108636*
16. *Tuggle DW, Smith EI. Pancreas divisum, pancreatic pseudocyst, and choledochal cyst in an 8-year-old child. J Pediatr Surg 1989; 24(1): 52-3. https://doi.org/10.1016/S0022-3468(89)80301-X*
17. *Dalvi AN, Pramesh CS, Prasanna GS, Rege SA, Khare R, Ravikiran CS. Incomplete pancreas divisum with anomalous choledochopancreatic duct junction with choledochal cyst. Arch Surg 1999; 134(10): 1150-2. https://doi.org/10.1001/archsurg.134.10.1150*

OLGU SERİSİ-ÖZET
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Pankreas divisum ile birlikte koledok kisti olgu serisi: Nadir bir birliktelik

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ÖZET

Koledok kistleri (KK), genellikle anormal pankreatikobiliyer duktal bileşke (APBDB) ile ilişkili olan safra kanallarının konjenital kistik dilatasyonlarıdır ve pankreas divisum (PD) pankreas divisum ile ilgisi nadir bildirilmiştir. Serimizde PD ile ilişkili dört KK olgusu bulunmaktadır. Bu dört olgunun üçünde Tip 3 PD, birinde ise Tip 1 PD vardı. İki olgu pankreatik komplikasyonlar geliştirirken bunlardan biri rekürren pankreatit açısından preoperatif minör papilla sfinkterotomisi gerektirdi. KK ile PD ilişkisi nadir olmakla birlikte değişken başvuru şekilleri yönetim stratejisini değiştirmektedir. PD, KK ile ilişkili komplikasyonlardan sorumlu bir faktör olabilir.

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

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