Clinical Case Report

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Pediatric types I and VI choledochal cysts complicated with acute pancreatitis and spontaneous perforation

A case report and literature review

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Abstract

Rationale: Choledochal cysts are a congenital disorder of the common bile duct that can cause progressive biliary obstruction and biliary cirrhosis. They were classified by Todani into five types. Of these, type VI choledochal cysts are rarely reported in the literature.

Patient concerns: A 22-month-old girl presented with intermittent epigastralgia for approximately 10 days and fever for three days. Fasting and total parenteral nutrition were administered after admission. However, sudden onset of severe epigastric pain occurred. An abdominal sonogram showed turbid ascites and peritonitis was impressed.

Diagnoses: An emergent exploratory laparotomy was performed, and perforation of the posterior wall of types I and VI choledochal cysts was observed.

Interventions: Intraoperative cholangiography revealed concomitant types I and VI choledochal cysts with stricture of the distal common bile duct. Definite surgery for resection of the choledochal cysts and gallbladder was performed with Roux-en-Y choledochojejunostomy.

Outcomes: The patient had no evidence of ascending cholangitis at three years after the operation.

Lessons: Type VI choledochal cysts are rarely reported in the literature. To our knowledge, this is the first reported pediatric case of concomitant types I and VI choledochal cysts complicated with acute pancreatitis and spontaneous perforation.

Abbreviations: γ-GT = glutamyl transpeptidase, ALT = alanine aminotransferase, AST = aspartate aminotransferase, CBD = common bile duct, CT = computed tomography.

Keywords: acute pancreatitis, choledochal cyst, spontaneous perforation, type I, type VI

1. Introduction

Choledochal cysts are abnormal dilatations of the biliary tree. They are more common in some Asian countries (with an incidence of up to 1 per 1000 individuals).[1] The female-to-male ratio varies from 3:1 to 4:1.[1] The classic symptoms of such cysts in children are intermittent abdominal pain, jaundice, and the presence of a right upper quadrant abdominal mass. Cystic dilatation of the cystic duct, also called type VI choledochal cyst, is extremely rare.[2–17] We report the case of a 22-month-old girl with concomitant types I and VI choledochal cysts, complicated with acute pancreatitis and spontaneous perforation.

2. Case report

A 22-month-old girl presented with intermittent epigastralgia for approximately 10 days. The pain appeared to be cramping and intermittent, usually lasting for several minutes at a time, and was centered in the epigastric area. No radiating pain was felt. The severity of the pain could be reduced by resting but was aggravated by body movement and food intake. In addition, she had a fever (body temperature, approximately 38–39 °C) and demonstrated frequent postprandial vomiting for 3 days. Therefore, she was admitted to our hospital.

Physical examination revealed that the abdomen was soft and mildly distended. Percussion was tympanic. Bowel sound was slightly hyperactive. Tenderness was observed over the epigastric area, without rebound pain or muscle guarding. Laboratory data showed the following values: white blood cell count, 9.48×10^9 cells/L; hemoglobin, 20.3 mmol/L; mean corpuscular volume, 81.2 fL; platelets, 335×10^9 cells/L; neutrophils, 0.697; lymphocytes, 0.265; eosinophils, 0.0003; basophils, 0.0002; monocytes, 0.033; aspartate aminotransferase (AST), 275 U/L; alanine aminotransferase (ALT), 146 U/L; lipase, 7727 U/L; and gamma glutamyl transpeptidase (γ-GT), 691 U/L. Abdominal sonography revealed a cystic mass near the main bile duct and segmental dilatation of the

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CCT and PKH have contributed equally to this work.

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The authors report no conflicts of interest.

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main bile duct. Abdominal computed tomography (CT) revealed a complicated cystic dilatation of the biliary tree (Fig. 1). Relative heterogeneous enhancement of the pancreatic head was also observed. Fasting was ordered initially after admission. The patient's amylase/lipase level on the fourth day after admission was 428/1006 U/L. Her abdominal pain subsided on the fifth day after admission, and feeding was attempted; however, vomiting and severe abdominal pain developed again on the following day. On the sixth day after admission, the patient's amylase/lipase, ALT, and γ-GT levels were 1227/2873, 359, and 1303 U/L, respectively. Fasting was ordered again. Total parenteral nutrition was administered beginning on the seventh day after admission. However, sudden onset of severe epigastric pain occurred, and her fever flared up on the evening of the eighth day after admission, with amylase/lipase and γ-GT levels of 720/2669 and 875 U/L, respectively. An abdominal sonogram revealed turbid ascites within the peritoneal cavity and impressed peritonitis. Emergent exploratory laparotomy was performed, during which approximately 500 mL of bile-stained ascites was evacuated. Additionally, a distended gallbladder and bile-stained choledochal cyst were found, as well as a perforation of the posterior wall of the type I choledochal cyst. Intraoperative cholangiography revealed concomitant types VI and I choledochal cysts with stricture of the distal common bile duct (CBD) and a dilated common channel (Fig. 2). Definite surgery for resection of the choledochal cysts and gallbladder was performed with Roux-en-Y choledochojejunostomy. The resected specimen is shown in Fig. 3A, and the illustration is shown in Fig. 3B. The pathological findings are shown in Fig. 4. The patient was discharged uneventfully following surgery, and showed no evidence of ascending cholangitis at the 3-year follow-up performed in our outpatient department.

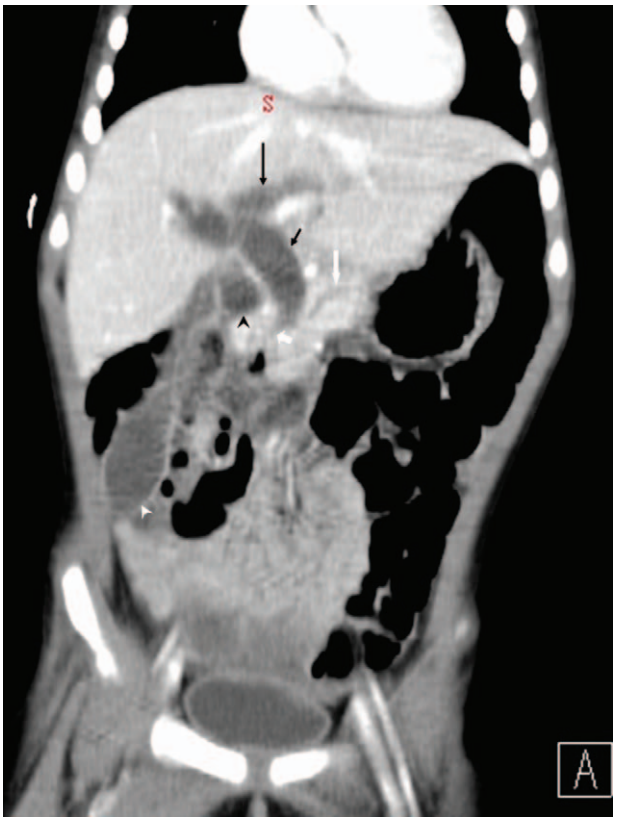


Figure 1. Abdominal contrast-enhanced computed tomography image in the coronal view showing that the intrahepatic duct (long black arrow) and common bile duct (CBD; short black arrow) were dilated with segmental stricture (short white arrow) of the distal CBD. The gallbladder (white arrowhead) is also dilated, and a cyst (black arrowhead) can be observed between the gallbladder and CBD. The pancreatic duct (long white arrow) is mildly dilated with relative heterogeneous enhancement of the pancreatic head.

3. Discussion

The primary classification of choledochal cyst was first described by Alonso-Lej et al,[18] and was later modified by Todani et al[19]

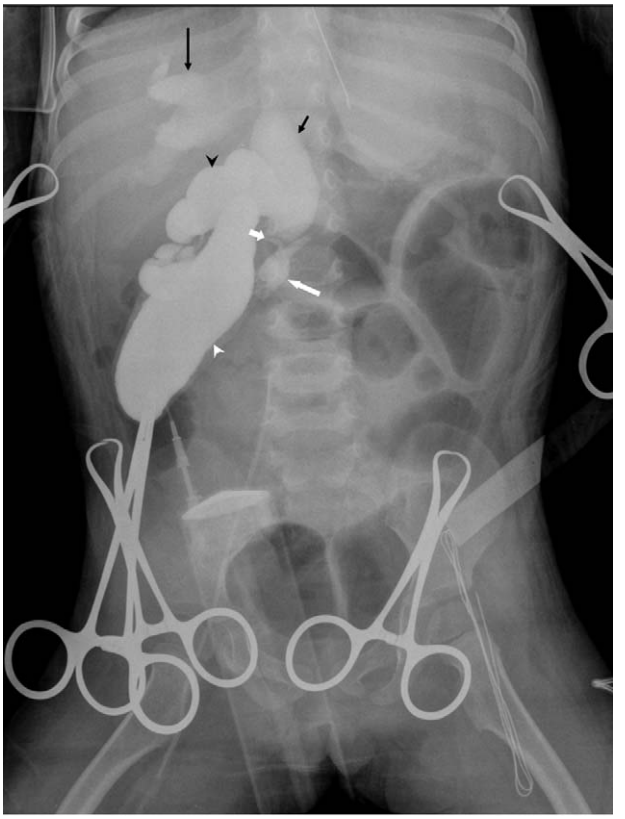
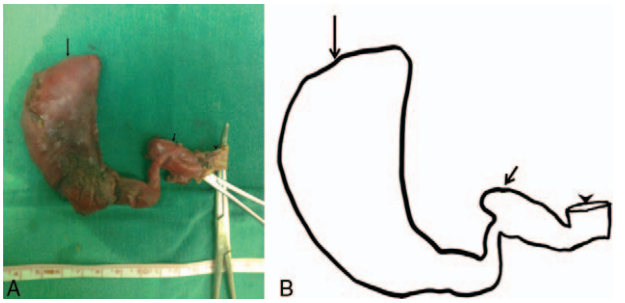


Figure 2. Intraoperative cholangiography image showing dilatation of the right intrahepatic bile ducts (long black arrow) and common bile duct (CBD; short black arrow), dilatation of the distal part of the cystic duct (black arrowhead), and segmental stricture of the distal CBD (short white arrow), as well as a dilated common channel (long white arrow) and hypertrophy of the gallbladder (white arrowhead).



Figures 3. A, B The resected specimen shows a pigeon-egg-sized choledochal cyst (short arrow) at the distal cystic duct. The gallbladder (long arrow) is markedly dilated, and its wall is hypertrophied with inflammatory changes. The arrowhead represents the common bile duct (A). The corresponding illustration is shown in (B).

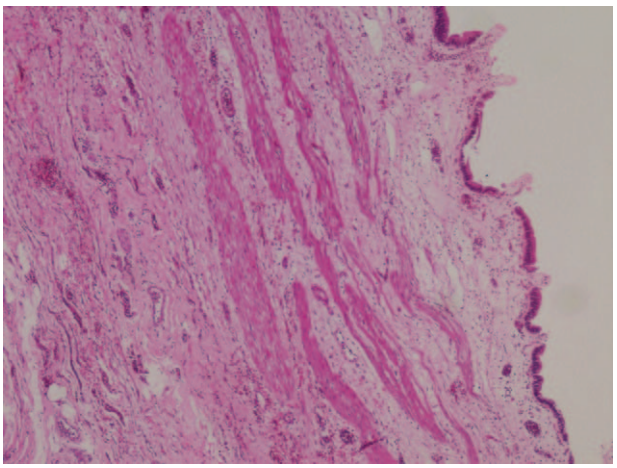


Figure 4. The histological pathology of choledochal cyst revealed columnar epithelium lining, smooth muscle layer, and adventitia which means this cyst originating in the bile duct (H&E stain, 40×).

in 1977 into 5 major types. The dilatation of the cystic duct, also known as type VI choledochal cyst, is rare in pediatric patients, with only 19 cases reported in the literature searching from a public domain database (PubMed) with the following keywords: choledochal cyst, type VI choledochal cyst, and cystic duct cyst with each possible combination for relevant articles (Table 1).[3,4,7–17] In this case, 1 choledochal cyst was located at the distal cystic duct entering the CBD, and another was located at the CBD. Furthermore, the cystic dilatation distal to the segmental stricture of the CBD was diagnosed to be an abnormal pancreaticobiliary common channel because intraoperative cholangiography (Fig. 2) revealed that the pancreatic duct entered this cystic dilatation. Based on the above-mentioned findings, the choledochal cysts in this case were a variant of concomitant types VI and I choledochal cysts. To our knowledge, the current patient is the youngest person with concomitant types VI and I choledochal cysts reported in the related literature. Preoperative diagnosis of concomitant types VI and I choledochal cysts is challenging, and intraoperative cholangiography is recommended in this unusual type of choledochal cyst.

Table 1

Literature review of published cases of type VI choledochal cyst in children.

Type	Illustration	Age (y)	Gender	Cystic duct abnormalities	Author
Type VI only		7	F	Inflamed 6 cm cyst confined to the cystic duct	Bode et al[3]
		18	F	Cystic dilatation	Serrudel et al[4]
		18	F	Dilated cystic duct with opening into CBD	De et al[7]
		0	F	Mild fusiform	Maheshwari[8]
		0.3	M	Mild fusiform	Maheshwari[8]
		4	M	Fusiform	Maheshwari[8]
		7	F	Fusiform	Maheshwari[8]
		15	F	Fusiform dilatation with wide opening	Yoon[10]
		10	F	Spherical cystic lesion	Shah et al[11]
		6	F	Fusiform	Goya et al[13]
		10	F	Dilatation of the cystic duct	Kaselas et al[15]
		0.3	M	Fusiform	Youn et al[17]
Type VI + Type III + IVb		16	F	Cystic duct enter in one of the extrahepatic cysts	Mishra et al[12]
Type VI + Type IVa		13	M	Cystic dilatation	Mishra et al[12]
Type VI + Type V		16	M	Fusiform	Sethi et al[14]
Type VI + Type I		3	F	Fusiform	Maheshwari[8]
		4	F	Fusiform	Maheshwari[8]
		15	M	Not available	Champetier et al[9]
		14	M	Cystic dilatation	Bhoil et al[16]
		1	F	Dilatation of distal part of cystic duct	This article

Choledochal cysts are related to significant complications, such as pancreatitis, cholangitis, secondary biliary cirrhosis, and spontaneous rupture of the cyst or cholangiocarcinoma. Choledochal cysts generally rupture spontaneously in children aged <4 years.[20,21] To our knowledge, this is the first reported case of spontaneous perforation of concomitant types VI and I choledochal cysts. Pancreatitis, proteinaceous plugs, or stones within a dilated common channel may be related to pancreaticobiliary malunion.[22–24] Komuro et al[24] reported that acute pancreatitis and protein plug formation were found more frequently in children between ages 1 and 15 years than in adults with choledochal cysts.

To date, few studies have discussed the optimal timing of surgery for choledochal cysts complicated with acute pancreatitis in children.[25] Cho et al[25] divided patients aged ≤15 years into 3 groups according to timing of surgery after diagnosis: ≤7 days (group A), 8 to 30 days (group B), and ≥31 days (group C). Length of operation, days of hospitalization, time to commencement of diet after operation, and change in hemoglobin concentration were similar in the 3 groups. The authors suggested that early cyst excision after the development of acute pancreatitis in patients with choledochal cysts reduced patients' symptoms, especially in instances of pseudopancreatitis.[25] However, at the time of operation in their study, even the highest amylase or lipase level of the patients in the 3 groups did not exceed 309 U/L. In addition, determining the timing of surgery in choledochal cyst complicated with acute pancreatitis and spontaneous perforation is challenging. Here, we report our experience in treating spontaneous perforation of concomitant types VI and I choledochal cysts with pancreatitis.

4. Conclusion

Choledochal cysts are congenital disorders of the CBD that can cause progressive biliary obstruction and biliary cirrhosis. Type VI choledochal cysts are rarely reported in the literature. To our knowledge, we present herein the first reported case of concomitant types I and VI choledochal cysts complicated with acute pancreatitis and spontaneous perforation.

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