Pediatr Surg Int (1990) 5: 22–26

Pediatric Surgery International

Choledochocele: an unusual form of choledochal cyst

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Abstract. Choledochocele, or type III choledochal cyst, is a rare anomaly. Two children with choledochocele, both younger than any previously reported patient, were recently cared for at the University of Michigan Medical Center and prompted a literature review of this subject. Since 1974, 40 cases of choledochocele have been reported with enough clinical information for critical review. Ten of these patients were 21 years of age or younger. Presenting symptoms were not specific for choledochocele; they were generally interpreted to result from other biliary or gastrointestinal disorders that are more common for each age group. In pediatric patients the most frequent signs and symptoms of choledochocele were abdominal pain (70%), nausea and/or vomiting (60%), jaundice (30%), and acute pancreatitis (30%). While two-thirds of adult patients with choledochocele had undergone prior cholecystectomy (with stones rarely found), this was observed only once in children. Obstructive symptoms led to evaluation of the stomach and duodenum with either barium upper GI series or endoscopy in all children. These demonstrated an extrinsic mass effect in 90% of the patients. Endoscopic retrograde cholangiopancreatography identified a choledochocele in all cases in which the study was successfully executed. Intravenous cholangiography was sensitive in children, but less so in the adult patients reviewed. Other imaging efforts (computerized tomography, ultrasound, radionuclide scanning) were less dependable. Transduodenal marsupialization is the treatment of choice for patients of all ages and was provided in both of these newly reported children.

Key words: Choledochocele – Choledochal cyst (type III) – Duodenal duplication

Introduction

A choledochocele is a cystic dilation of the distal, intramural portion of the common bile duct, typically protruding into the descending duodenum. Classification as a type III choledochal cyst is based upon the system originally presented by Alonso-Lej et al. [1] in 1959 for cystic abnormalities of the extrahepatic biliary tree (Fig. 1). Choledochoceles are a rare form of choledochal cyst in all reports. In Flanigan's 1975 review of the world's literature, 43 patients with choledochocele were described among a study population of 955 patients [4]. Yamaguchi's 1978 review of 1433 choledochal cysts in the Japanese literature cited only 12 occurrences of choledochocele [18].

Two children with choledochocele are presented here, representing the youngest 2 patients ever reported. In addition, a review of choledochocele in the pediatric age group is provided [2, 3, 5, 8, 10, 11, 13, 16].

Case reports

Case 1. A 2 year-10-month-old boy was referred for evaluation of recurrent episodes of abdominal pain and sepsis. The child had a history of 12 hospitalizations or emergency department evaluations for this problem during the preceding 18 months. Episodes occurred at 2–3-month intervals, each lasting 3–4 days. During an episode the child complained of severe epigastric cramping and vomiting. There were three instances of

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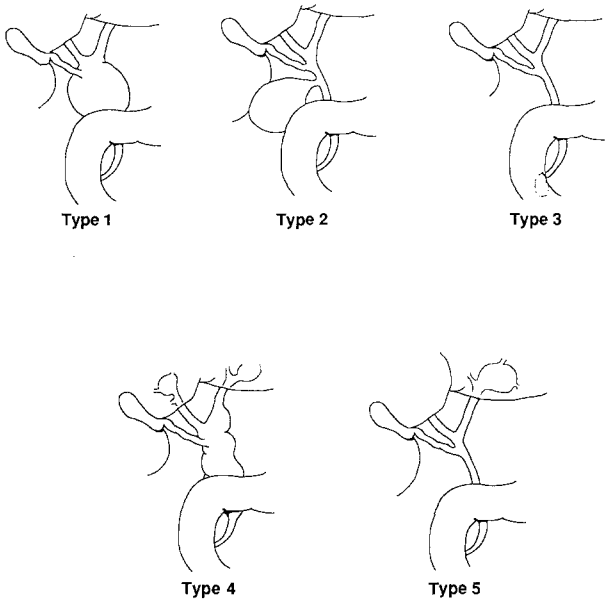

Fig. 1. Classification of choledochal cysts [1]

bacteremia with enteric organisms documented; two resulted in overt sepsis with hypotension. A barium upper GI study had suggested a filling defect in the second portion of the duodenum on two occasions (Fig. 2), however the endoscopic examination had been interpreted as normal.

Upon transfer, an abdominal ultrasound study revealed a cystic mass in the right upper quadrant that was thought to be a duodenal duplication. At laparotomy, the patient was found to have an annular pancreas and a palpable mass within the lumen of the duodenum just distal to the pancreatic annulus. Lateral duodenotomy revealed a 2-cm cystic mass covered with normal duodenal mucosa along the medial duodenal wall. No ampulla of Vater could be identified despite the presence of bile within the duodenal lumen. A small incision was made in the most dependent portion of the cyst, demonstrating that the cavity was bile-filled. Two orifices were seen in the cephalad aspect of the choledochocele and a contrast injection demonstrated these to be the common bile duct and pancreatic duct (Fig. 3). Marsupialization of the cyst was performed. In addition, duodenoduodenostomy was performed in order to correct the obstruction related to the annular pancreas. Microscopic examination of the cyst wall revealed duodenal mucosa. The patient was discharged home on the 6th postoperative day and has subsequently been asymptomatic.

Case 2. A 10-week-old term infant developed nonprojectile vomiting at 8 weeks of age followed 2 days later by the onset of acholic stools and jaundice. The total serum bilirubin level was 6.4 mg% with a direct fraction of 5 mg%. An abdominal ultrasound study revealed common bile duct dilation, but no choledochal cyst was seen. A radionuclide scan did not visualize the biliary tree. At laparotomy, a cholangiogram obtained through the gall bladder revealed an abrupt obstruction to bile flow at the level of the distal common bile duct. A 1.0 to 1.5-cm intraluminal mass was palpable in the second portion of the duodenum. A longitudinal, lateral duodenotomy revealed that the papillary mass was the distorted ampulla of Vater. A 2-mm posterolateral ampullotomy was performed and biliary concretions ("sand") filling the ampulla were extracted. A 2-mm probe was passed easily into the terminal common bile duct with prompt flow of clear bile. Transduodenal marsupialization of the choledochocele was performed. In this case, a cholecystoduodenostomy was additionally provided due to the extremely small size of the distal common bile duct and the uncertain location of the pancreatic duct. The patient recovered uneventfully and remains well 1 year postoperatively.

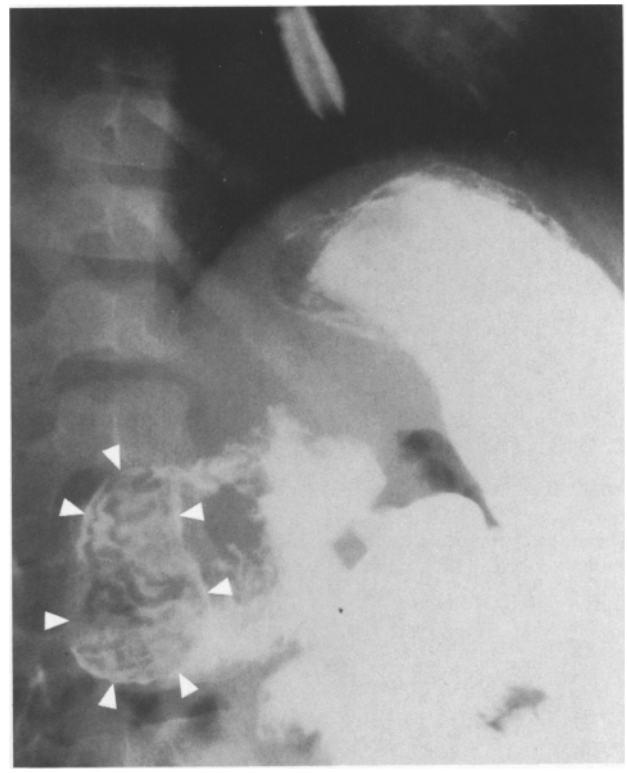

Fig. 2. Barium upper GI study, case 1. *Arrows* show filling defect secondary to the choledochocele

Literature review

Forty cases of choledochocele (including the 2 presented here) have been reported in the world literature since 1974 with enough data for review. Ten patients were 21 years of age or less and their clinical features are summarized below.

Diagnosis

Symptoms or signs related to a choledochocele in a child are typically present for some time prior to correct diagnosis, with a mean duration of 1.3 years between the onset of symptoms and treatment (range: 1 week to 4 years). Even longer delays were typical of the adult cases reviewed. Abdominal pain (typically epigastric or right upper quadrant), nausea or vomiting, jaundice, and pancreatitis were seen most frequently (Table 1).

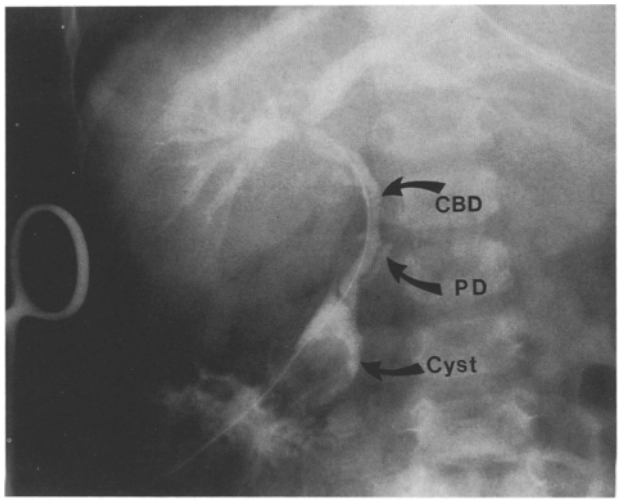

Fig. 3. Intraoperative cholangiogram, case 1. *Arrows* show the choledochocele (*cyst*), common bile duct (*CBD*), and pancreatic duct (*PD*)

The predictable absence of a right upper quadrant mass differentiates this entity from the much more common type I choledochal cyst.

A variety of diagnostic studies were employed in the evaluation of these children and adolescents with a choledochocele (Table 2). The most frequently obtained studies were those that evaluated the upper GI tract or the biliary tree. Scholz et al. [8] emphasized the radiological criteria for the diagnosis of choledochocele: a club-shaped dilation of the terminal common bile duct bulging into the duodenal lumen can usually be demonstrated as a non-filling mass on barium upper GI studies. A cystic dilation of the distal common bile duct may also be demonstrable on cholangiography. In this pediatric review, either a barium upper GI series or gastroduodenoscopy was always obtained and demonstrated a mass effect in 90% of the children. Endoscopic retrograde cholangiopancreatography (ERCP) correctly identified the choledochocele in all 3 children in whom it was employed. Intravenous cholangiography demonstrated the anomaly in four of five studies in children, although it was far less sensitive in the adult cases reviewed. Intraoperative cholangiograms demonstrated the choledochocele in 3 of the 4 patients in which it was obtained. Ultrasound was also helpful: three of five studies demonstrated extrahepatic biliary dilation and the choledochocele, yet two were nonspecific. Through the combined use of these imaging studies, a correct diagnosis was reached in 60% of cases prior to surgery for definitive treatment.

Additional anomalies were occasionally encountered: in case 1 an annular pancreas was found in conjunction with the choledochocele. One patient had an intrahepatic biliary cyst associated with the choledochocele. Intracystic biliary stones were reported in 2 of the 10 children.

Table 1. Symptoms and signs associated with choledochocele in children ($n = 10$)

	%
Abdominal pain	70
Nausea/vomiting	60
Jaundice	30
Pancreatitis	30
Sepsis	10
Malaise	10
Prior appendectomy	10
Prior cholecystectomy	10

Table 2. Diagnostic studies performed to evaluate pediatric patients with choledochocele

Study	No. Performed	% Positive	% Normal	% Nonspecific
Barium upper GI series	6	100	0	0
Gastroduodenoscopy	4	75	25	0
Ultrasound	5	60	0	40
Intravenous cholangiogram	5	80	20	0
Intraoperative cholangiogram	4	75	0	25
ERCP	3	100	0	0
Hydroxy imino diacetic acid	2	0	50	50
Liver/spleen scan	1	0	100	0
Liver biopsy	1	0	0	100

Treatment

Definitive treatment of the choledochocele was carried out operatively in 8 of 10 children and by endoscopic sphincterotomy in 1. In 1 child the parents refused treatment and the patient was lost to further follow-up. Transduodenal marsupialization of the choledochocele was the operative technique employed exclusively in pediatric patients although other procedures have been reported in adults. Postoperative complications were rare. One case of mild pancreatitis followed surgical intervention. Persistent elevation of serum transaminases occurred in 1 patient, and 1 instance of postoperative pneumonia was reported.

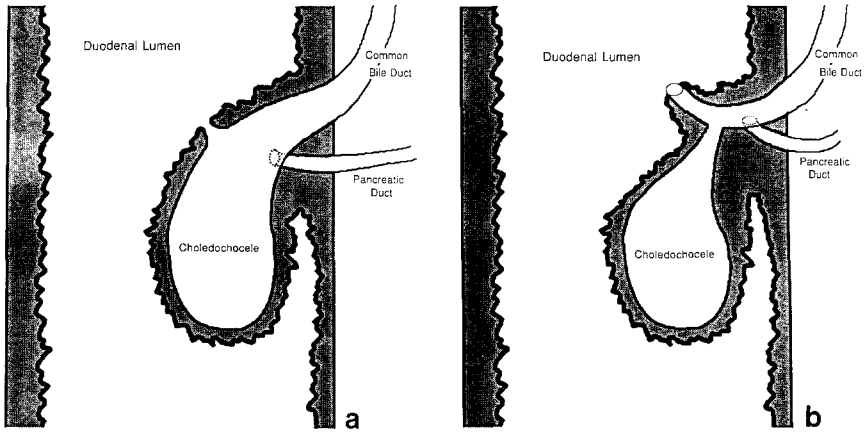

Fig. 4. Two observed anatomic variations with choledochocele: **a** choledochocele emptying into duodenum; **b** choledochocele emptying into distal common bile duct

From operative descriptions of the relationships of the biliary and pancreatic ducts with the choledochocele, it appears that two anatomic variations of choledochocele exist (Fig. 4). In the first, the choledochocele itself drains into the duodenum with the pancreatic and common bile ducts draining into it. In the second form the cyst appears to be a diverticulum of the distal common bile duct with the pancratic duct entering the bile duct proximally. These anatomic relationships are not described precisely enough in the reports available to determine the relative frequency of the variants. However, other authors have suggested that the former, both bile duct and pancreatic duct draining into the choledochocele, is seen more often [14]. Marsupialization is suitable for both variants.

Histologic evaluation of the choledochocele wall was reported in 6 of the pediatric cases; the cyst was lined with duodenal mucosa in all of them. In adult cases reviewed, both duodenal and biliary mucosa have been reported with equal frequency, and adenocarcinoma has been noted once.

The results of treatment were uniformly successful at relieving preoperative complaints, however follow-up was reported in only 6 cases and was generally of short duration (average = 1.3 years).

Discussion

Delay in arriving at the diagnosis of choledochocele appears to be related to the rarity of the lesion and failure to consider choledochal cyst in the initial differential diagnosis. Choledochoceles do not necessarily exhibit features typical of the more common type I choledochal cyst. Signs and symptoms are often nonspecific. Evaluation for jaundice and pancreatitis in a young patient should lead one to entertain this diagnosis.

Although classified as a choledochal cyst, the choledochocele has characteristics that some authors have suggested are more consistent with a duodenal duplication [6, 13, 17]. Classical duodenal duplications, like choledochocele, are most frequently located on the posterior-medial aspect of the second portion of the duodenum [9]. They are usually asymptomatic. When symptomatic, duodenal duplications may produce obstructive effects on the duodenum with vomiting, abdominal pain, and a palpable mass rather than biliary obstruction. Microscopic evaluation of the cyst wall reveals duodenal mucosa rather than biliary epithelium in all pediatric patients, perhaps supporting the concept of a closer relationship to duodenal duplications. In addition, the typical extramural union between the pancratic and biliary ducts seen with type I choledochal cysts is not a feature of choledochoceles [7]. At present, it must be acknowledged that the pathogenetic mechanisms that lead to the development of choledochoceles are not known. The classification with choledochal cysts is established in the literature and will no doubt continue to be used.

The risk of adenocarcinoma developing in type I choledochal cysts is reported to be approximately 10% [15], however only a single case of adenocarcinoma associated with a choledochocele has been reported [12]. Whether the rarity of underlying lesion accounts for this or whether different pathogenetic mechanisms are involved is not clear.

Summary

Our recent experience with two cases of childhood choledochocele prompted a review of the

relevant literature. Nearly half of the world's experience has been reported in the last 13 years, suggesting that the diagnosis is being made with increasing frequency. Improved physician education and wide application of new imaging techniques are certainly significant contributors to this increase. Presenting symptoms are nonspecific; errors in diagnosis and delays in therapy are common. Despite the nonspecific nature of presentation, a correct preoperative diagnosis was achieved in 60% of the current patients using a combination of imaging techniques. Transduodenal marsupialization of the choledochocele is the treatment of choice, with few serious complications reported. Demonstration of the variable periampullary pancreatic and biliary ductal anatomy is critical. In small infants, such as the patient presented in case 2, additional biliary tract drainage procedures may be advisable because of the small size of the common bile duct. This rare problem can be reliably and successfully managed given the correct diagnosis and attention to the technical precautions presented.

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Accepted June 14, 1989