The Ochsner Journal 7:131–132, 2007

Guess the Case from the Ochsner Archives

Jason A. Breaux, MD, William S. Richardson, MD, Vincent Adolph, MD

Ochsner Clinic Foundation, New Orleans, LA

INTRODUCTION

A 15-year-old Asian female with a past history of omphalocele and meningomyelocele at birth, both repaired in the neonatal period, presented to her pediatrician complaining of vague, intermittent, epigastric abdominal pain for approximately 10 years. These episodes occurred every 2–4 weeks, but prior to presentation had worsened and become more frequent. She described no exacerbating factors. Occasionally, the pain radiated to the left upper quadrant.

Proton-pump inhibitor therapy was initiated with no relief. There was no history of jaundice. She had no nausea or emesis, no fevers or rigors. Her past medical history was otherwise unremarkable except for a well-functioning ventriculoperitoneal shunt. There was no significant family history.

She had experienced normal growth and development after her initial problems at birth. On examination, she was afebrile and hemodynamically normal. Her examination was remarkable for nonicteric sclerae, a soft, nontender abdomen with a well-healed midline incision and no palpable abdominal masses. Her aspartate aminotransferase level was elevated at 101 u/L; the results of all other laboratory studies were normal. Her pediatrician ordered an abdomen and pelvis CT scan followed by endoscopic retrograde cholangiopancreatography and referred her to a pediatric surgeon.

QUESTION: What is the diagnosis and what treatment would you recommend?

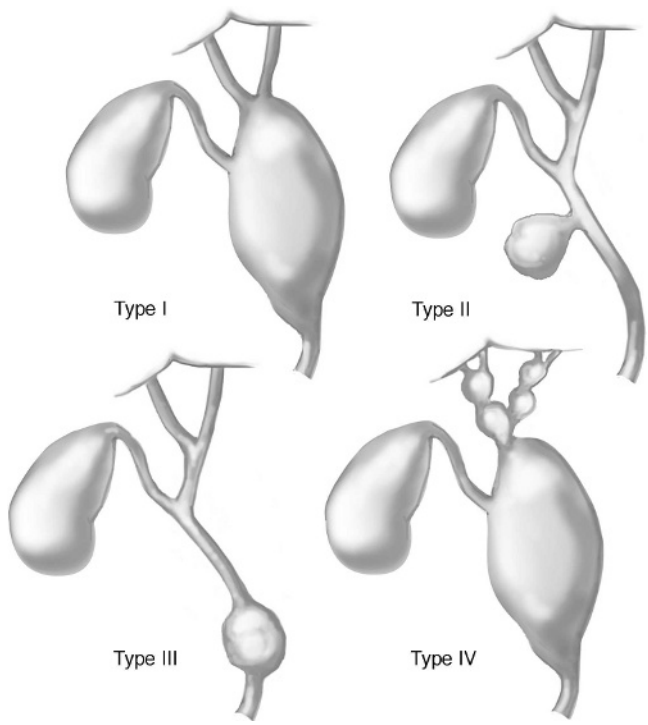
DIAGNOSIS AND TREATMENT

This patient had a Type I choledochal cyst with choledochocystolithiasis. She underwent an excision of the cyst from the liver hilum to the intrapancreatic bile duct, where the ductal caliber became normal, and a cholecystectomy. A Roux-en-Y hepaticojejunostomy reconstruction was performed using a 45–50 cm Roux limb. The patient did well and was discharged home on postoperative day 5.

CHOLEDOCHAL CYST

Choledochal cysts have an incidence of 1:13,000 in the United States. They are more common in females and in Asian countries, especially in Japan, where the incidence is as high as 1:1,000 (1,2,3). They are most commonly classified as Type I–V based on the classification scheme of Alonso-Lej as modified by Todani (4) (Fig. 1). A Type I cyst is a fusiform dilatation of the extrahepatic biliary tree. A Type II cyst is a saccular diverticulum of the common bile duct. A Type III cyst, also known as a choledochocele, is a dilatation of the distal common bile duct within the duodenal wall. Type IV cysts are multiple and involve both intra- and extra-hepatic bile ducts. Type V

Figure 1: Extrahepatic bile duct anatomy for Types I-IV choledochal cyst as classified by Todani. Type V (Caroli's Disease), with only intrahepatic dilatation not shown.



Address correspondence to:
William Richardson, MD
Department of Surgery
Ochsner Clinic Foundation
1514 Jefferson Highway
New Orleans, LA 70121
Tel: (504) 842-4070
Fax: (504) 842-3100
Email: wrichardson@ochsner.org

Image 1: Computed tomography image of upper abdomen revealing dilated extrahepatic bile duct with choledocholithiasis (arrow).

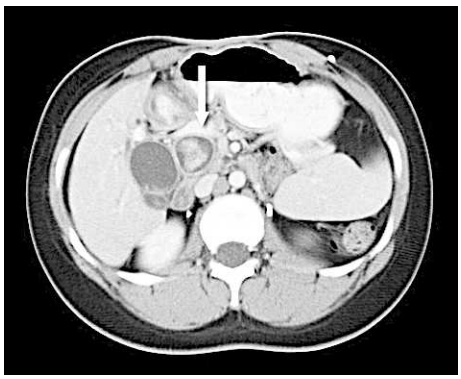
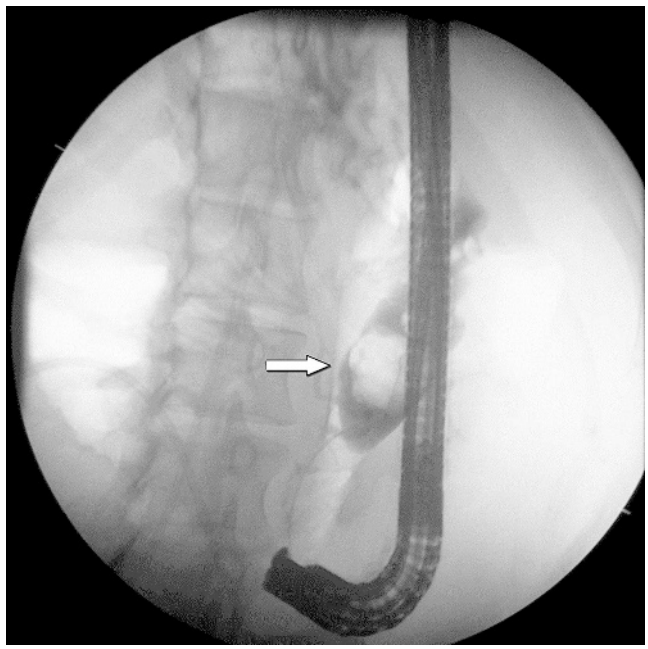


Image 2: Fluoroscopic image during endoscopic retrograde cholangiography revealing Type I choledochal cyst containing stone (arrow).



(Caroli's disease) consists of multiple cysts involving only the intrahepatic ducts. Type I and IV make up the majority (>95%) of choledochal cysts (1–5). The most common presenting symptom is abdominal pain. The classic triad of pain, jaundice, and an abdominal mass is present in less than 30% of patients (1–5).

In most patients (>90%) with choledochal cyst, a long common channel exists due to a more proximal convergence of the common bile duct and pancreatic duct in relation to the ampulla. This leads to reflux of pancreatic secretions into the biliary tree, causing chronic inflammation and dilatation of the bile ducts (2,6,7). This repeated injury can lead to stasis, inflammation, metaplasia, dysplasia, and ultimately carcinoma (8). Patients with choledochal cyst, especially Types I and IV, are at a 30- to 100-fold increased risk of developing cholangiocarcinoma compared to normal subjects. This risk increases linearly with age. Type III choledochal cysts are unique in that they are lined with duodenal mucosa and have a very low rate of malignant transformation (9).

The treatment employed for most choledochal cysts is excision and Roux-en-Y reconstruction. Internal drainage procedures, leaving the cyst in place, have been abandoned (10). There remains a very small risk (<1%) of cholangiocarcinoma even after excision, and whether leaving some remnant cyst wall increases this risk is controversial (11,12). Type II cysts are usually amputated at their neck and simple duct closure is performed. Asymptomatic Type III cysts may be observed; symptomatic cases require transduodenal marsupialization, either operatively or endoscopically. Type V cysts (Caroli's disease) may be resected if isolated to one hepatic lobe or may require transplantation for treatment (1–4,10).

REFERENCES

1. Besner GE, Paddock HN, Nguyen LT, et al. Choledochal cyst: surgical perspective. eMedicine, August 28, 2006. www.emedicine.com/ped/topic2932.htm. Accessed June 2007.
2. Cheney M, Rustad DG, Lilly JR. Choledochal cyst. World J Surg 1985;9:244–249.
3. O'Neill JA. Choledochal cyst. Curr Probl Surg 1992;29:361–410.
4. Todani T, Watanabe Y, Narusue M, et al. Congenital bile duct cysts: classification, operative procedures, and review of thirty-seven cases including cancer arising from choledochal cyst. Am J Surg 1977;134:263–269.
5. Babbitt DP. Congenital choledochal cyst: new etiological concept based on anomalous relationships of common bile duct and pancreatic bulb. Ann Radiol (Paris) 1969;12:231–240.
6. Okada A, Nakamura T, Higaki J, et al. Congenital dilatation of the bile duct in 100 instances and its relationship with anomalous junction. Surg Gynecol Obstet 1990;171:291–298.
7. Jordan PH, Goss JA, Rosenberg WR, et al. Some considerations for management of choledochal cysts. Am J Surg 2004;187:790–795.
8. Imazu M, Iwai N, Tokiwa K, et al. Factors of biliary carcinogenesis in choledochal cysts. Eur J Pediatr Surg 2001;11:24–27.
9. Benjamin IS. Biliary cystic disease: the risk of cancer. J Hepatobiliary Pancreat Surg 2003;10:335–339.
10. Fu M, Wang Y, Zhang J. Evolution in the treatment of choledochus cyst. J Pediatr Surg 2000;35:1344–1347.
11. Ishibashi T, Kasahara K, Yasuda Y, et al. Malignant change in the biliary tract after excision of choledochal cyst. Br J Surg 1997;84:1687–1691.
12. Watanabe Y, Toki A, Todani T. Bile duct cancer developed after cyst excision for choledochal cyst. J Hepatobiliary Pancreat Surg 1999;6:207–212.