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Type VI Choledochal Cyst—An Unusual Presentation of Jaundice

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Abstract

Choledochal cysts involving the cystic duct are extremely rare, and are usually associated with cystic dilations of the extrahepatic biliary tract. We describe a patient who presented with jaundice and was found to have a dilatation of the common bile duct on computed tomographic imaging, consistent with a choledochal cyst. He underwent a laparoscopic-converted-to-open cholecystectomy with excision of the choledochal cyst which was found to involve the cystic duct. Choledochal cysts involving the cystic duct are notably missing from the Todani classification. Although exceedingly rare, new cases of these types of cysts are being reported, in part due to advancement of diagnostic imaging modalities. We discuss the current classification scheme for choledochal cysts and we propose an expansion of this scheme.

Keywords

- ▶ choledochal cyst
- ▶ hepatobiliary
- ▶ jaundice
- ▶ cystic duct
- ▶ cholecystectomy
- ▶ Todani classification

Choledochal cysts involving the cystic duct are extremely rare, and are usually associated with cystic dilations of the extrahepatic biliary tract. Their incidence ranges from 1/10,000 to 1/100,000 in North America and Europe.^{1,2} The rate is remarkably higher in Asian populations with a reported incidence of 1 in 1,000, with about two-thirds of cases occurring in Japan.³ Choledochal cysts have a female-to-male ratio of 3:1 to 4:1,⁴ and although most common in the pediatric population, 20% of cases occur in adults.¹

Case Report

Our patient is a 27-year-old obese African American male who initially presented with jaundice. His medical history, review of systems, and abdominal examination were unremarkable. The patient was evaluated and found to have a dilatation of the common bile duct (CBD) on computed tomographic (CT) imaging, consistent with a choledochal

cyst. Endoscopic retrograde cholangiopancreatography (ERCP) and sphincterotomy without stenting led to resolution of jaundice but onset of abdominal pain within 24 hours. He was admitted for a laparoscopic cholecystectomy that was converted to an open cholecystectomy with excision of the choledochal cyst and construction of a hepaticojejunostomy.

Intraoperatively, the gallbladder, cystic duct, and distal portion of the CBD extending into the head of the pancreas were found to be dilated. The surgical dissection began at the porta hepatis. Identification of the cystic duct and artery were difficult due to the distorted anatomy. The gallbladder was taken down starting at its fundus, requiring incision, and resection of a bridge of liver to identify a plane of dissection. The distal CBD was divided at the head of the pancreas where it was normal in caliber. No stones were found within the duct. The proximal common hepatic duct was divided. The gallbladder, cystic, and CBD were resected and removed en bloc. Pathologic evaluation of the margins was negative for

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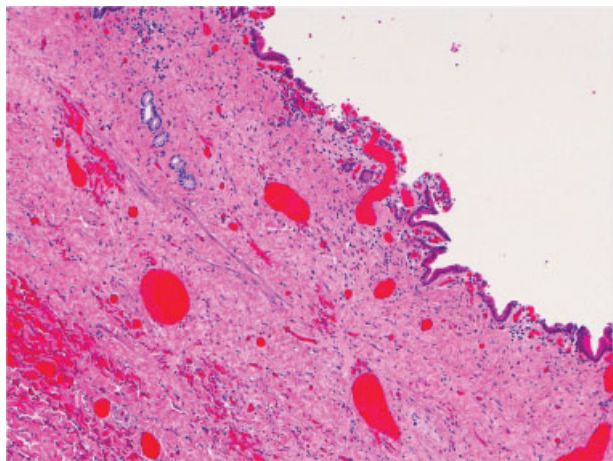


Fig. 1 Histologic image of choledochal cyst, showing no evidence of dysplasia.

tumor. The distal bile duct was oversewn. A Roux-en-Y hepaticojejunostomy was constructed. The patient recovered uneventfully and was subsequently discharged home.

Surgical pathology report was consistent with a choledochal cyst with no evidence of dysplasia (→ **Fig. 1**). The proximal common hepatic duct and distal CBD resection margins were unremarkable. The gallbladder was attached to a cyst measuring 3.8 × 2.5 cm (→ **Fig. 2**).

Discussion

Choledochal cysts are associated with an anomalous union of the pancreaticobiliary duct (AUPBD) in 33 to 90% of cases.² They present with the triad of jaundice, abdominal pain, and a palpable right upper quadrant mass 6 to 38% of the time.^{2,5} However, adults most commonly present with pain, fever, and nausea.¹ Approximately, 3 to 39% of patients develop cholangiocarcinoma.² This increased risk is believed to be due to chronic inflammation and ulceration of the cyst epithelium. The risk of malignancy (adenocarcinoma, anaplastic carcinoma, undifferentiated cancer, squamous cell carcinoma, and unspecified others) increases with age, with a

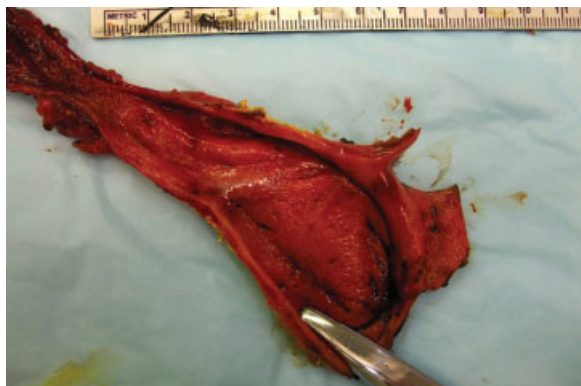


Fig. 2 Gross image of a choledochal cyst involving the cystic duct.

suspected cancer risk of 10% in the third decade.¹ Malignancies can develop in the extrahepatic bile ducts, gallbladder, intrahepatic bile ducts, liver, and pancreas.¹ Other complications of choledochal cysts include cholelithiasis, cholangitis, pancreatitis, portal hypertension, liver fibrosis, and cirrhosis.^{1,2}

Choledochal cysts are treated by complete excision of the extrahepatic duct, cholecystectomy, and Roux-en-Y hepaticojejunostomy.^{1,2,6} These procedures are preferred to internal drainage or laparoscopic cholecystectomy due to the possibility of malignant transformation.² Following treatment, these patients must be carefully monitored postoperatively with laboratory investigations of liver and biliary function as well as imaging modalities to detect the development of biliary strictures and/or carcinoma.^{1,2}

Important laboratory investigations include alanine aminotransferase, aspartate aminotransferase, bilirubin, alkaline phosphatase, gamma-glutamyl transferase, and amylase; although these results may be nonspecific. Radiological investigations include abdominal ultrasound, ERCP, magnetic resonance cholangiopancreatography (MRCP), percutaneous transhepatic cholangiography, intraoperative cholangiography, technetium-99 hepatobiliary iminodiacetic acid (HIDA) scan, and abdominal CT.^{1,7,8}

Todani et al⁹ developed a classification system for choledochal cysts based on location, shape, and number.^{2,4}

Type I: 50 to 80% of Cases

Type I cysts involve only the extrahepatic biliary system. Subclassifications include IA (involve the entire extrahepatic bile duct), IB (segmental involvement of the extrahepatic bile duct), and IC (diffuse, fusiform, or cylindrical involvement of the extrahepatic bile duct with AUPBD). Types IA and IB do not include AUPBD.^{1,2,4}

Type II: 2% of Cases

Type II cysts are true diverticula of the extrahepatic bile duct.^{1,2,4}

Type III: 4 to 4.5% of Cases

Type III cysts, also known as choledochoceles, are in the extrahepatic bile duct within the duodenal wall.^{1,2,4}

Type IV: 15 to 35% of Cases

Type IV cysts can include both the extrahepatic and intrahepatic biliary system. Type IVA can result in a primary ductal stricture involving the hepatic hilum. Type IVB involves multiple cystic dilations involving only the extrahepatic duct.^{1,2,4}

Type V: 20% of Cases

Type V cysts only involve the intrahepatic bile duct. It is believed to be due to a genetic predisposition to intrahepatic gallstones and autosomal recessive polycystic kidney disease.^{1,2,4}

To the best of our knowledge, choledochal cysts like the one described here involving the cystic duct are not included in this classification.

To be Determined: Type VI

Cysts involving the cystic duct are notably missing from the Todani classification. Although they are exceedingly rare, cases have been reported in the literature.^{2,7,8} With the advancement of diagnostic imaging technologies such as MRCP, EUS, and ERCP, we are seeing the emergence of new cases of these types of cysts. We recommend that the current classification scheme be expanded to include choledochal cysts involving the cystic duct.

Conclusion

We present a case in which a 27-year-old man with jaundice was found intraoperatively to have a choledochal cyst involving the cystic duct, an unusual variant which is not included in the current classification scheme, and which was not considered before the surgical operation. We believe that it is important to recognize this variant as a distinct pathologic entity so that the appropriate diagnostic workup and preoperative planning can be undertaken.

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