



A Rare Case of Choledochal Cyst Connecting Intra- and Extra-Hepatic Duct

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Abstract

Choledochal cysts are rare congenital dilatations of the extra and/or intrahepatic bile ducts. This disease often presents non-specific symptoms that perplex early diagnosis. We report a rare anatomical location of a choledochal cyst in a 5-year-old female patient, and it is a variant of the classical types of the hepatic duct cysts. Awareness of this type of variation in choledochal cysts would aid preoperative diagnosis, more accurate interpretation of imaging results and surgical planning.

Keywords: Choledochal cyst; Hepatic duct; Laparoscopic cyst

Introduction

Choledochal cysts are rare congenital dilatations of the extra and/or intrahepatic bile ducts found primarily in children and estimated of much higher incidence in Asia, where it reaches approximated 1:1000, as compared to Western population [1,2]. A choledochal cyst increases the risk of malignant transformation up to 10% and patients may still be exposed at higher risk for biliary malignancies even after surgical resection [1,3]. The most commonly used classification system updated by Todani et al. [4] described 5 broad types of choledochal cysts. The most commonly reported is type I, which includes cystic, focal or fusiform dilatations of the common bile duct. The clinical presentations are often non-specific, such as nausea, vomiting, abdominal pain and jaundice. Nevertheless, image techniques are essential to aid the diagnosis [5]. Laparoscopic cyst excision and hepaticojejunostomy for children have become a popular treatment protocol [6].

Case Presentation

A 5-year-old girl presented several episodes of right upper quadrant abdominal pain after an upper airway infection. She denied any fever, nausea/vomiting, abdominal cramping pain, tea-color urine or diarrhea during these episodes. Initially, she received survey at another hospital, where choledochal cyst was diagnosed. Medical staff there suggested further Magnetic Resonance Cholangiopancreatography (MRCP) and surgical survey. Her parents hesitated and came to our hospital for the second opinion. At presentation to our pediatric ward, her red blood cell count was $5.31 \times 10^6/\mu\text{L}$ (normal 4.28 to $5.05 \times 10^6/\mu\text{L}$), hemoglobin 10.6 g/dL (normal 11.6 g/dL to 13.7g/dL), Mean Cell Volume (MCV) 64.2 fL (normal 74.9 fL to 84.6 fL), and activated partial thromboplastin time (APTT) 35.1 sec (normal 24.3 to 32.7 sec). Her white blood cell count, total bilirubin, direct bilirubin, Alanine Transaminase (ALT), Alanine Aspartate Phosphatase (AST), Gamma-Glutamyl Transpeptidase (γ -GT), amylase and creatinine were all within normal limits.

Pre-operative abdominal ultrasound and MRCP (Figure 1) showed a cystic lesion near the common bile duct, 1.45 cm \times 1.29 cm in size, and Todani type 1 or 2 choledochal cyst was considered. During surgical exploration, a cyst lesion was found at the junction of intra- and extra-hepatic duct (Figure 2), which was confined within the common hepatic segment. This finding was confirmed by intraoperative cholangiography (Figure 3). The dilatation part of the cyst included part of left and right hepatic duct, as well as the common hepatic duct (Figure 4A). After cholecystectomy, the cyst was excised at about 1 cm below bifurcation point (Figure 4B). Reconstruction was carried out with a Roux-en-Y end-to-side hepaticojejunostomy. Seven days after surgery, the girl was discharged and followed up with normal daily activities 3 months later. Histological analysis confirmed that

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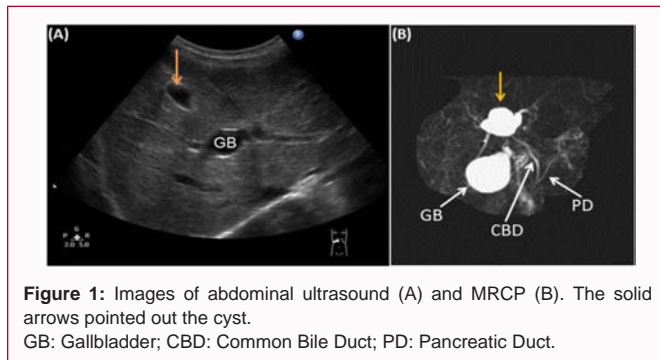


Figure 1: Images of abdominal ultrasound (A) and MRCP (B). The solid arrows pointed out the cyst.
GB: Gallbladder; CBD: Common Bile Duct; PD: Pancreatic Duct.

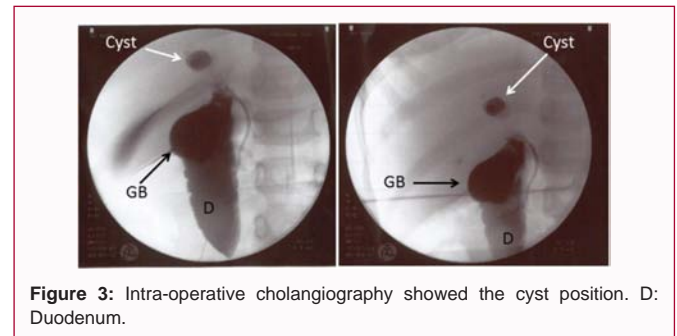


Figure 3: Intra-operative cholangiography showed the cyst position. D: Duodenum.

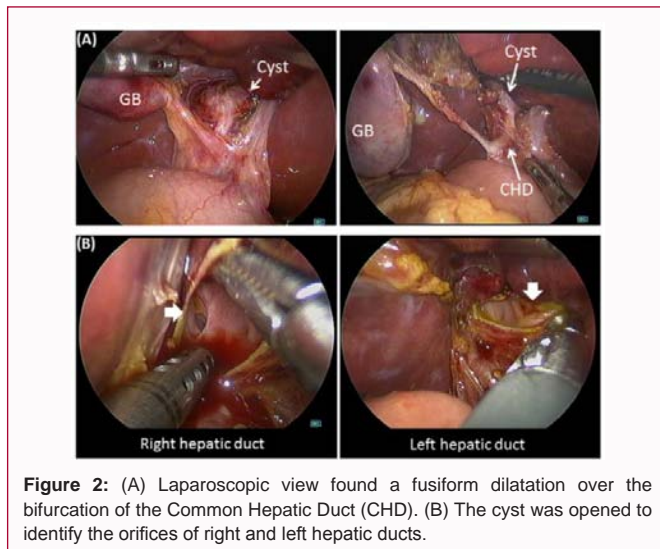


Figure 2: (A) Laparoscopic view found a fusiform dilatation over the bifurcation of the Common Hepatic Duct (CHD). (B) The cyst was opened to identify the orifices of right and left hepatic ducts.

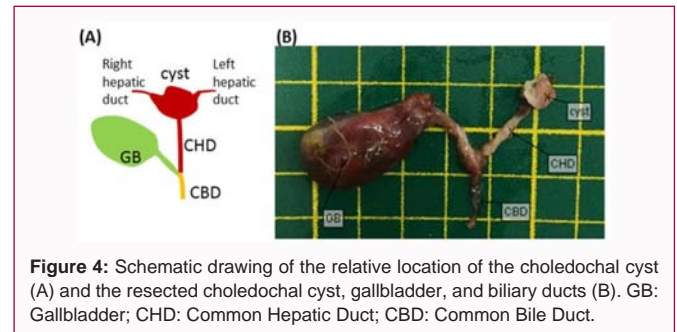


Figure 4: Schematic drawing of the relative location of the choledochal cyst (A) and the resected choledochal cyst, gallbladder, and biliary ducts (B). GB: Gallbladder; CHD: Common Hepatic Duct; CBD: Common Bile Duct.

the resected specimen contain thickened fibrotic wall with chronic inflammation and intact overlying biliary type epithelium. The cyst contained no stones, and the cyst wall was continuous with the common hepatic duct.

Discussion

More severe complications of choledochal cyst include pancreatitis, cholecystitis, liver cirrhosis and cholangiocarcinoma [7]. Surgical treatment and reconstruction is varied according to different types of choledochal cyst. In 2008, Calvo-Ponce et al. [8] introduced an extra variant type in addition to Todani's modified classification of biliary duct cysts according to their findings in a fifty year-old female patient, which is similar to this pediatric case. This is an extremely rare anatomical location for a choledochal cyst, which could be the cause of the unique clinical manifestation of our patient. The bile flow may be disturbed by the altered route, which leads to intermittent right upper quadrant abdominal pain without any fever or nausea. The gross examination and imaging of the cyst is quite misleading as that of type II choledochal cyst. However, under laparoscopic view, there is no saccular diverticulum from the common bile duct, but a fusiform dilatation sitting at the convergence of intra- and extra-hepatic ducts. Awareness of this variant type in choledochal cysts would aid pre-operative diagnosis, more accurate interpretation of imaging results and surgical planning.

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