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Cystic duct anomaly and pancreaticobiliary maljunction mimicking choledochal cyst

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ABSTRACT

A choledochal cyst (CC) is an abnormal congenital dilatation of the extrahepatic or intrahepatic biliary tree and is usually diagnosed during childhood. While a magnetic resonance cholangiopancreatography (MRCP) may be helpful for the diagnosis, laparoscopy and intraoperative cholangiography might be the final tool for the diagnosis and evaluation of anatomical variant and an effective surgical treatment. Herein we report a case of a 12-year-old male whose MRCP was suggestive choledochal cyst with gallbladder agenesis. He underwent an elective laparoscopic evaluation, and the intraoperative cholangiography and findings revealed that the patient was not a choledochal cyst but that the infundibulum of the huge gallbladder directly entering the main right hepatic duct with pancreaticobiliary maljunction. This case highlights that these rare entities can mimic each other on imaging; however, a laparoscopic approach and intraoperative cholangiography serves the dual purpose of diagnosing and treating this rare pathoanatomical entity.

1. Introduction

Choledochal cysts which are characterized of dilation of various ducts of the biliary tree, are relatively rare in the Western world but more common in Asia, in addition, it is well known that there are many aberrant bile duct variations, and to identify correct anatomy and to avoid intraoperative bile duct injury which could cause severe complication including the intraoperative bile duct injury, the bile leakage and the stenosis, surgeons should have many kinds of knowledge about possible variation of biliary trees [1–6].

Herein, we report a rare case of 12 years old male presenting the right hepatic duct entering the cystic duct with pancreaticobiliary maljunction and we would like to highlight the importance of the intraoperative evaluation of the biliary duct.

2. Case

A previously healthy 12 years old male presented with complaints of intermittent abdominal pain associated with nausea for a year, but no history of jaundice and fever, and admitted to our institution. Laboratory evaluation did not show any elevation of C-reactive protein level, white blood cell count, liver, renal and hematological dysfunction.

Ultrasound of the abdomen showed the dilation of the right hepatic duct with 10 mm diameter and the cystic lesion of 83 mm × 63 mm which are considered the gallbladder and contained no sludge or stones.

Magnetic resonance cholangiopancreatography (MRCP) showed pancreaticobiliary maljunction (PBM), dilated the right hepatic duct and the huge cystic lesion which are considered as a choledochal cyst (Fig. 1). We speculated that this patient had a choledochal cyst with PBM and the gallbladder agenesis [7], and decided to schedule an elective laparoscopic surgery.

Under general anesthesia, laparoscopic exploration started. The huge gallbladder was observed (Fig. 2a) and the exposure of the cystic duct and the cystic artery around Calot's triangle was continued. The cystic artery was identified around infundibulum of the gallbladder while the identification of the cystic duct was difficult due to severe inflammatory swelling and scarring of the gallbladder, the gallbladder was incised at the level of Hartmann's pouch (Fig. 2b), and the intra-

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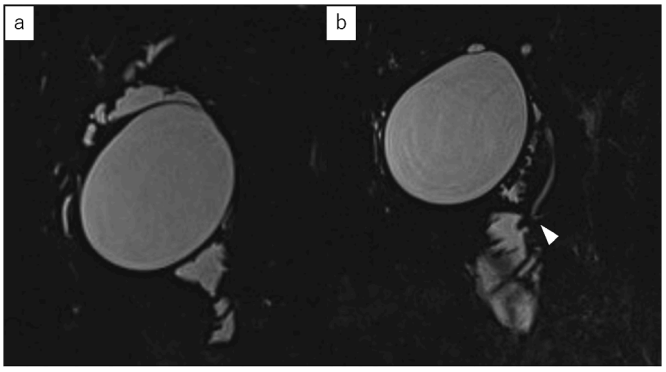

Fig. 1. Magnetic resonance cholangiopancreatography showing a cystic lesion at the porta closely abutting the neck of the gallbladder, cystic duct and right posterior hepatic duct with faint suspicious communication of the lesion with cystic duct. C: Cyst. GB: Gallbladder. CBD: Common bile duct. RHD: Right hepatic duct.

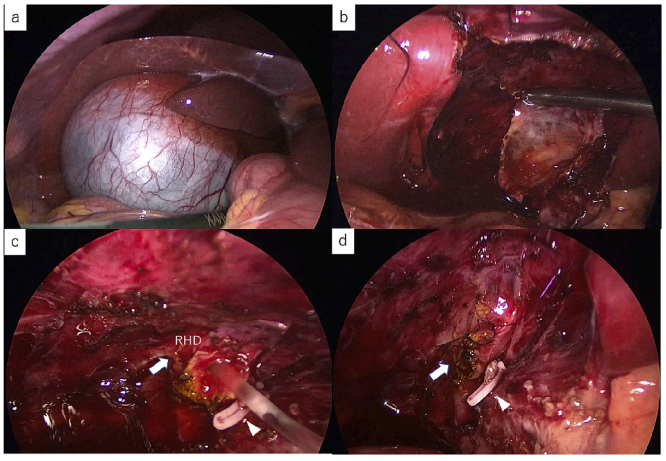

Fig. 2. Intraoperative findings. (a) A huge gallbladder was observed. (b) The gallbladder was incised at the level of Hartmann's pouch. (c) The gallbladder directly entered to the right hepatic duct (RHD, white arrow), the cystic artery was resected (arrow head). (d) The direct closure of the orifice of the gallbladder neck was performed carefully not to make a stricture of the right main hepatic duct (white arrow). The clip for resection of the cystic artery (arrow head).

operative cholangiography (IOC) revealed the infundibulum of the gallbladder directly enter the main right hepatic duct (Figs. 2c and 3). After identification of the anatomy of the biliary tree, the resection of a cystic artery and the direct closure of the orifice of the gallbladder neck was performed carefully not to make a stricture of the right main hepatic duct (Fig. 2d).

The postoperative course was uneventful, and the patient was discharged on postoperative day 7. At 1 year after surgery, the patient was on a normal diet and was asymptomatic.

3. Discussion

Biliary tree anomalies are quite common. Approximately 42% of the general population has some anatomical variation in the biliary tree [1,4,8–10]. This anatomical variation was not appreciated in both preoperative imaging studies and a retrospective evaluation in a postoperative period. In general, the biliary system is routinely examined using preoperative imaging modalities (MRCP or DICCT) in patients who are scheduled to undergo elective surgeries. However, the imaging data are sometimes inconclusive.

Based on our numerous experiences of choledochal cyst repair [2,3,6], we believe that IOC and 'direct vision' is one of the most important intraoperative evaluation tools for the identification of real anatomical variation of the biliary tree. In this case, we suspected our patient as a 'choledochal cyst with agenesis of gallbladder' preoperatively [7], because the common bile duct appeared to be continuous to the cystic duct (Fig. 1a), however, IOC revealed that the gallbladder directly enters to the right hepatic duct with PBM (Fig. 3) [11,12].

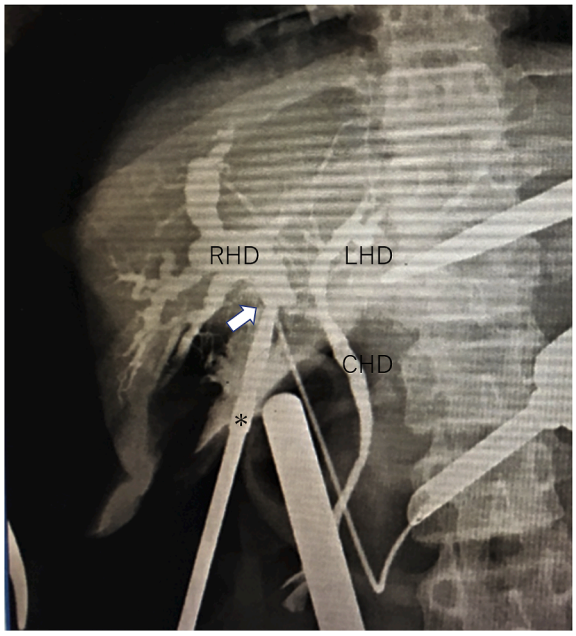

Fig. 3. Intraoperative cholangiography.
Gallbladder entered directly to the right hepatic duct (RHD, white arrow). (LHD: left hepatic duct, CHD: common hepatic duct, *:the forceps for pointing out of the orifice of the gallbladder entering to RHD.

Our experience may be a good example of reminding pediatric surgeons that biliary surgery should be based on objective indices, not based on preoperative evaluations or our assumptions.

4. Conclusion

We report a rare case of the gallbladder directly enter to the main right hepatic duct. Preoperative evaluation and intraoperative evaluation of the biliary duct, awareness of the variations of the aberrant biliary duct, especially intraoperative cholangiography and meticulous dissection are important in preventing bile duct injury.

Patient consent

Consent to publish the case report was not obtained. This report does not contain any personal information that could lead to the identification of the patient.

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Authorship

All authors attest that they meet the current ICMJE criteria for Authorship.

Declaration of competing interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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