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Open surgical treatment of choledochocele in a two years old child

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ABSTRACT

Choledochocele is a cystic dilation of the distal segment of the common bile duct protruding into the duodenal lumen. Which is an extremely rare congenital lesion. Here we report the case of a 2-year-old girl with a choledochocele complicated with choledocho lithiasis and acute pancreatitis who was treated by transduodenal cyst resection with sphincteroplasty. The patient had an uneventful clinical course for one year after the procedure. Very few reports have been published on this condition in infants.

1. Introduction

Choledochocele (CC) is a cystic dilation of the distal common bile duct protruding into the duodenal lumen and the third subtype of biliary cysts -according to Todani's classification [1] Which is a rare abnormality seen in 1.4–4.5% of the common bile duct (CBD) in the largest case series [2]. The choledochocele is most often diagnosed in adulthood. However, twenty percent of CC are seen in children, at an average age of 5 year-old [3]. Their diagnosis is mainly based on endoscopic cholangiopancreatography (ERCP). The treatment of choice is still controversial. However, the cyst size (i.e., > -3 - cm in diameter), the histology of the cyst (duodenal mucosa) and the classification (-type A3) of the choledochocele, may be the cut-off value, above which transduodenal cyst resection should be performed, otherwise ERCP is recognized as a feasible alternative treatment with satisfactory results [4]. Here we report the case of a 2-year-old girl with a Choledochocele complicated with choledocho lithiasis and acute pancreatitis who underwent a transduodenal cyst excision with sphincteroplasty. Very few reports have been published on this rare condition in infants.

2. Case report

A two-year-old girl presented to of our pediatric surgery department emergency room, with abdominal pain, fever and jaundice. The physical examination found the girl in poor general condition with an epigastric tenderness and without dynamic instability. Serum chemistry revealed: an increased levels of amylase (3156 UI/L), and lipase (1312 UI/L), that led to a diagnosis of acute pancreatitis. Bilirubin level was also elevated (total bilirubin: 129-mg/dL, direct bilirubin: 96-mg/dL) confirming the cholestasis. -Complete blood count showed hyper leukocytosis (white blood cells: 18.47 × 10*3) cells/mL and anemia (hemoglobin: 8.90 g/dL), and elevated C reactive protein (35.8 mg/L). An initial computed tomographic cholangiography (CTC) assessment (Fig. 1), showed intrahepatic and extra hepatic bile duct dilatation with a cystic formation projecting in front of the pancreas head, this cyst contained stones and also revealed an acute pancreatitis classified as Balthazar A, (managed conservatively). To confirm the diagnosis a magnetic resonance cholangiopancreatography (MRCP) (Fig. 2) revealed a small cyst (20 mm in diameter) in the distal common bile duct consistent with a type A2 choledochocele (separate openings of the pancreas duct and common bile duct into the cyst.

Pediatric (ERCP) was not available in our department to confirm the diagnosis and eventually as a therapeutic investigation with

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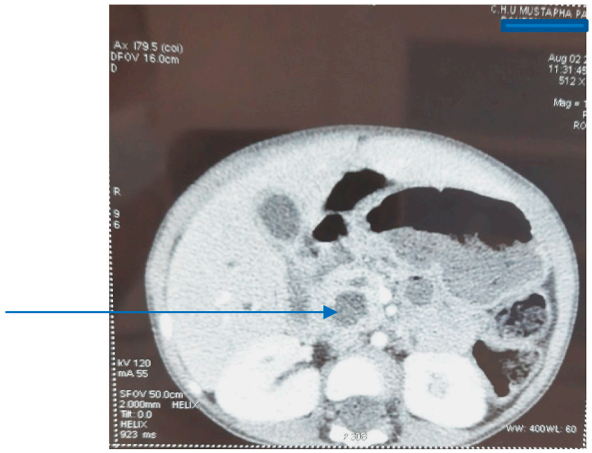

Fig. 1. Computed tomographic cholangiography (CTC) showed a cystic intraluminal lesion in the second part of the duodenum (arrow).

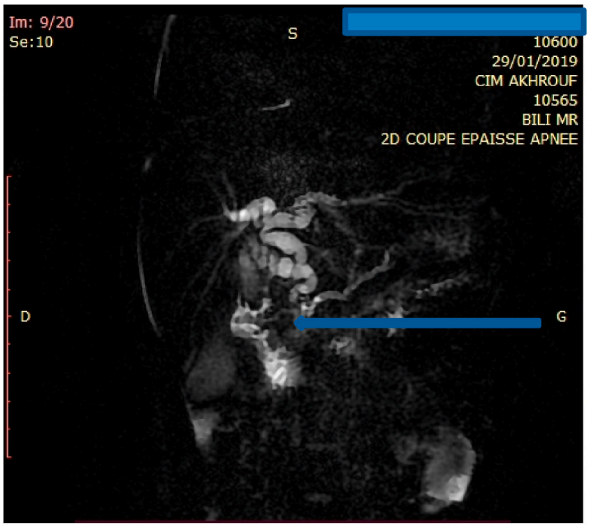

Fig. 2. Magnetic resonance cholangiopancreatography (MRCP) showed separate openings of the pancreas duct and common bile duct into the cyst (arrow).

the technical feasibility of endoscopic sphincterotomy. Hence, after biological markers normalization we admitted the infant to the operating room, where she underwent a laparotomy. At the exploration we noticed a dilation of the gallbladder and a tortuous cystic duct. We performed a cholecystectomy with cystic duct cannulation which enabled us to perform an intraoperative cholangiogram (Fig. 3) that showed that the distal CBD was dilated and protruded into the duodenal lumen, consistent with a choledochocele.

The anti-mesenteric wall of the second duodenum was opened (Fig. 4) and the cystic mass was dissected, the papilla was washed out by injection of physiological saline through the cystic stent, allowing the evacuation of cholestones. A sphincterotomy was performed at twelve and three o'clock to achieve pancreaticobiliary drainage and allowed stones to pass into the duodenum. The cystic stent was left, and the abdomen was closed without drainage.

The immediate postoperative course was uneventful. The jaundice regressed rapidly and the bile duct drainage dried up after the second day. A cholangiogram through the cystic stent was performed on the 6th postoperative day showing a normal outflow of pancreatic and biliary secretions (Fig. 5), then the biliary stent was removed. The patient was discharged on the 8th day after surgery. A pathological examination of the surgical resection found an intestinal mucosa. There were no abnormal findings on an abdominal ultrasound (Fig. 6) performed 6 months after the surgery. The patient had an uneventful clinical course one year after the procedure.

3. Discussion

Choledochocoele is a very rare pathological entity characterized by a dilatation of the common bile duct in its most distal segment as it enters the duodenal wall. In the classification of choledochal cysts by Todani et al., choledochocele corresponds to type III, representing a small percentage of all biliary tree cysts [5]. However, both Ziegler et al. [6] and Dong et al. [7] considered that the choledochal cysts should not include the choledochocele because patients with choledochocele differ from those with choledochal cyst

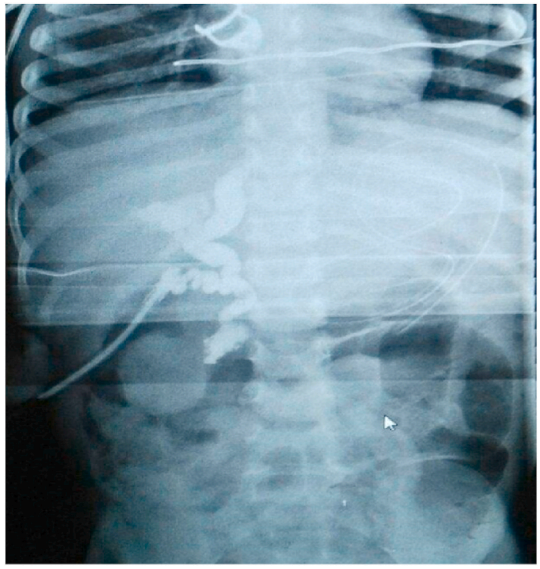

Fig. 3. Perioperative cholangiography.

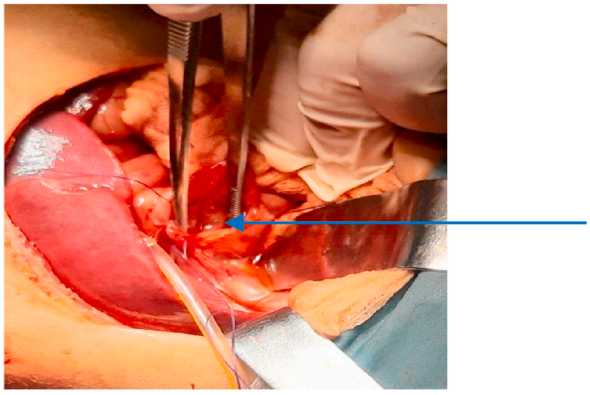

Fig. 4. Macroscopic appearance of the choledochocele through the duodenostomy. Arrow indicates the choledochocele.

in age, sex, presentation, pancreatic ductal anatomy, and in their management. The precise pathogenesis of the choledochocele remains unclear, Tanaka et al. suggested that the mechanism for formation was failed regression of a bile duct during embryogenesis [8]. Symptoms of choledochocele are nausea, vomiting, and abdominal pain. Cholestasis is the only manifestation in newborns; described in few cases reported [9]. Symptoms of choledochocele are related to biliary tract obstruction in 10–34% and pancreatic symptoms in 30–38% [10] as it was the case of our patient who had cholestasis and acute pancreatitis. Choledochocele can be associated with serious complications including cholangitis, pancreatitis, cholethiasis and malignant transformation. However the potential for malignancy (2.5%) is reported to be significantly lower than in other types of choledochal cysts due to the presence of duodenal epithelium lining the choledochocele and the lack of a massive reflux of pancreatic secretions into the biliary tree [11,12]. The preoperative diagnosis and understanding of the distal biliary tract anatomy in relationship to the choledochocele is essential in guiding the surgical treatment. This appears to be ideally made by ERCP, which provides the advantage of possible concomitant treatment. Nevertheless, pediatric duodenoscope was no available in our department, therefore a CT scan and MRCP were performed to confirm the diagnosis of CC and classify our choledochocele as type A2 according to the anatomical classification of Sarris Tsang that distinct choledochocele into type A (further sub classified as A1, A2, and A3) and type B [13]. Type A1 has a common opening of the pancreatic duct (PD) and CBD into the cyst. In type A2, there are separate openings of the PD and CBD. Type A3 is similar to type A2, but the cyst is small and intramural. In type B the ampulla empties directly into the duodenum and the cyst represents a diverticulum of the distal CBD protruding into the duodenal lumen. Surgical excision is recommended for the subtype A1, A2 and type B [14].

Established management of choledochocele when the cyst is less than 3 cm in diameter, with intestinal mucosa at biopsy of cystic wall [15], and type A3 in Sarris Tsang classification, include a sphincterotomy by ERCP [16], above which a transduodenal cyst resection should be performed. In our case since the size of the cyst is 2 cm and CC classified as type A2, we went for an open procedure. We performed a transduodenal cyst resection with sphincteroplasty. Choledochoceles lined with biliary mucosa should undergo

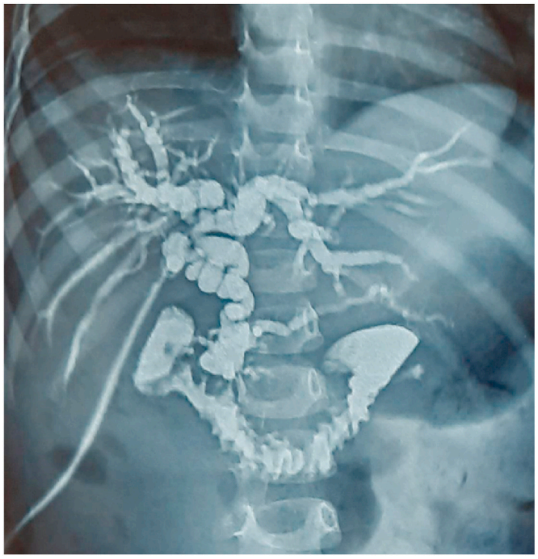

Fig. 5. Cholangiogram through the cyst stent post-operative period.

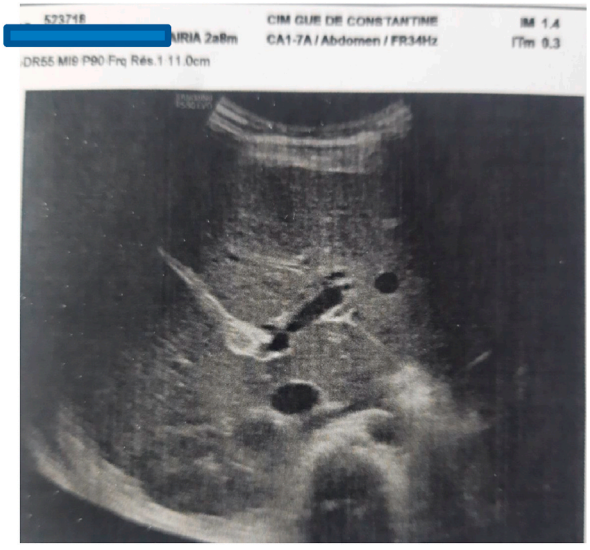

Fig. 6. Post-operative abdominal ultrasound. There were no abnormal findings.

indefinite surveillance to monitor the risk of malignant transformation [17]. Histopathological examination of the inner wall of the choledochocele of our case revealed the presence of duodenal mucosa. Consequently our patient will need limited monitoring.

4. Conclusion

Choledochocele is a very rare disease in children less than 5 years old and the treatment is still controversial. The choice of the treatment management depends on the cyst size, the type of mucosa lining the cyst, the classification of the choledochocele, and the facilities available. Cystoenterostomy with sphincteroplasty in infant is a safe procedure as shown in our case with successful outcome.

Ethics declarations

Ethics approval and consent to participate this study was conducted in accordance with the Declaration of Helsinki and approved by the research and ethics committee of Algeria; reference number is not applicable. This study does not contain any personal information that could lead to the identification of the patient.

Consent for publication

Written informed consent to publish this information was obtained from.
The parent of the study participant.

Source of funding

None.

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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