



Case report

Choledochal cyst: A challenging diagnostic and therapeutic entity in low-resource settings

Turyalai Hakimi^a, Habib Ahmad Esmat^{b,*}, Ramazan Karimi^a

^a Department of Pediatric Surgery, Kabul University of Medical Science, Kabul, Afghanistan

^b Department of Radiology, Kabul University of Medical Sciences, Kabul, Afghanistan

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ABSTRACT

Background: Choledochal cyst is an uncommon congenital biliary tract abnormality of unknown etiology. Its classical symptoms are jaundice, abdominal pain, and right upper quadrant mass. However, the disease may present with a vague and non-specific chronic abdominal discomfort. Delay in diagnosis and management may increase the risk of complications particularly the malignancy, which can directly affect the prognosis and outcome. Complete excision of the extra-hepatic duct, cholecystectomy, and Roux-en-Y hepaticojejunostomy through the open surgical intervention or laparoscopic procedure is the mainstay of treatment.

Case Presentation: A 14-year-old male was presented to our hospital complaining of vague abdominal pain for 5 years. The radiologic imaging showed the features of a type IVa choledochal cyst. He underwent complete excision of the extra-hepatic duct, cholecystectomy, and Roux-en-Y hepaticojejunostomy. Although the bile leakage occurred as a complication of the procedure, our team best managed the patient in the post-operative period until full recovery.

Discussion: The Choledochal cyst was first described by Vater, which is a congenital anomaly that sometimes may remain asymptomatic till adulthood. Surgical intervention is the mainstay of treatment. However, postoperative complications including bile leakage need a close follow-up of the patient.

Conclusion: Diagnostic delay (antenatal and postnatal) and non-specific symptoms will mask the real feature of the disease, especially in low-resource settings. Technical facilities and professional care of the patient may preclude complications.

1. Introduction

A choledochal cyst is a congenital dilatation along the biliary tract, commonly of the main part with an incidence of 1/100000–150,000 live births. Clinically it has five types with several sub-types based on the anomalous union (Komi classification) and anatomical location (Todani classification) [1–3].

The etiology is unknown, but one hypothesis says that pancreaticobiliary ductal congenital mal-union proximal to the sphincter within the duodenal may lead to reflux of pancreatic juice into the bile duct, contributing to the inflammation and dilatation of the biliary tract [4,5].

Ultrasonography (US) is the initial diagnostic modality of choice followed by computed tomography scan (CT-Scan), which may help diagnosis the continuity of the cyst within the biliary tract, relation to

surrounding organs, and staging of any possible malignant changes as biliary tract malignancy due choledochal cyst increases with age [6–8].

In the past, the treatment for choledochal cyst was the drainage procedure, but now the definitive treatment is the complete excision of the extra-hepatic duct, cholecystectomy, and Roux-en-Y hepaticojejunostomy [9].

Choledochal cyst generally has an excellent prognosis after complete resection. However, long-term surveillance is necessary if there is biliary stone or progressive dilatation of the intrahepatic bile ducts [10].

Herein, the authors present a case of a Choledochal cyst in a 14-year-old male, which presented with a chronic abdominal pain and successfully managed by an open surgical intervention with a favorable outcome.

This work has been reported in line with the SCARE 2020 criteria [11].

Abbreviations: US, Ultrasonography; CT, Computed Tomography; CBD, Common Bile Duct; ERCP, Endoscopic Retrograde Cholangiopancreatography.

* Corresponding author.

E-mail address: habib.smt@gmail.com (H.A. Esmat).

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2. Case presentation

2.1. Patient information

A 14-year-old male presented with an occasional vague abdominal pain more predominant in the right upper quadrant associated with anorexia for 5 years. He was born of a non-consanguineous couple and the first child of the family. This couple had given birth to 7 full-term children through normal delivery with the last child born on the 7th gestational month. Family history was negative for the mentioned problem and other anomalies. The physical exam was unremarkable. This family was living in the capital outskirts and according to the patient's father, during the pain episodes he has taken his child to the local clinic several times, but due to lack of health care facilities the general practitioner or sometimes physician assistant prescribed him some symptomatic relief medications.

2.2. Laboratory and radiological findings

His routine blood and urine biochemical examinations were within normal limits except for the amylase and lipase which were two times greater than normal. On abdominal ultrasound exam, cystic dilatation of the biliary tract was observed. On abdominal CT images, a prominent cystic dilatation of the common bile duct (CBD) and mildly dilated intrahepatic ducts consistent with a type IVa choledochal cyst was noted (Fig. 1a and b).

2.3. Therapeutic intervention

The patient was taken up for complete excision of the extra-hepatic duct, cholecystectomy, and Roux-en-Y hepaticojejunostomy through an open surgical intervention by a qualified surgery team in the pediatrics surgery unite. During the operation, the posterior hepatic duct wall was torn inadvertently as we used vicryl suture materials with the cut needle, so initially, we repaired the duct with 5.0 vicryl round needle followed by completion of the Roux-en-Y procedure (Figs. 2, 3). We put two drains; one in the sub-hepatic area and the other in the Douglas pouch. The muscle layers of the abdominal wall were closed according to the anatomic plan and the patient was taken to the intensive care unit as extubated postoperatively. 36 hrs following surgery, about 800 ml/day of bile drainage, was observed through the abdominal-hepatic drain, which is considered as postoperative bile leakage. The bile leakage gradually increased to 1200 ml per day till postoperative day 8th and then steadily decreased to 400 ml/day by day 15th of his

hospital stay. The patient was discharged from the hospital by post-operative day 18th, removing the sub-hepatic drain but leaving the drain in the Douglas pouch. We advised him regular wound dressing and keep phone contact with us for possible consultation if necessary. On day 4th of the homestay, due to carelessness (improper and careless dressing in local clinic), the drain was displaced, leading to stop the bile drainage through the catheter. On post-discharge day 7th, the patient father contacted us that his child complained of high-grade fever, tender abdomen, nausea, and vomiting, so we re-admitted the patient and started antibiotic therapy.

Based on the ultrasonography result of biloma, the emergency laparotomy was planned that we found the patient to have grade III biloma of 16–20 cm size, encapsulated in the sub-diaphragmatic area. The accumulated bile was suctioned, the area was washed with 2000 ml normal saline, and a drain was placed in the sub-diaphragmatic region. The operation was terminated and the muscle layers of the abdominal wall were closed according to the anatomic plan. After full recovery, he was discharged from the hospital with a satisfactory outcome and during the six months follow up the patient had a normal life.

3. Discussion

A choledochal cyst is a congenital anomaly of the biliary tracts, which is more common in the Asian population and frequently seen in females. It was first described by Vater. However, the first authenticated case was reported by Doughlas in 1852 [12].

Its classical symptoms are jaundice, abdominal pain, and right upper quadrant mass. There are several types of choledochal cyst based on the 2003 guidelines. Type I, which is most common (82% of cases) characterized by cystic dilations of CBD. Type II (3% of cases) are true diverticula of CBD. Type III (5% of cases) are cystic dilations of the CBD called choledochal. Type IV (9% of cases) are multiple cysts of intra-or extrahepatic ducts or both. Type V (1% of cases) is characterized by single or multiple intrahepatic cystic dilations [13].

Diagnosis is made based on clinical suspicion. However, abdominal ultrasound is the most suitable imaging modality which is inexpensive, non-invasive, but its results are observer-dependent [14,15]. An abdominal CT-Scan scan may accurately define the size, location, and continuity of the anomaly. Endoscopic retrograde cholangiopancreatography (ERCP) can help diagnosis the lower ductal anomalies, particularly, the abnormal joining of bile duct and pancreatic duct [14]. Magnetic cholangiopancreatography (MRCP) is now substituted ERCP since it is non-invasive and highly accurate [16,17]. In our case, the patient was a male child who presented with a

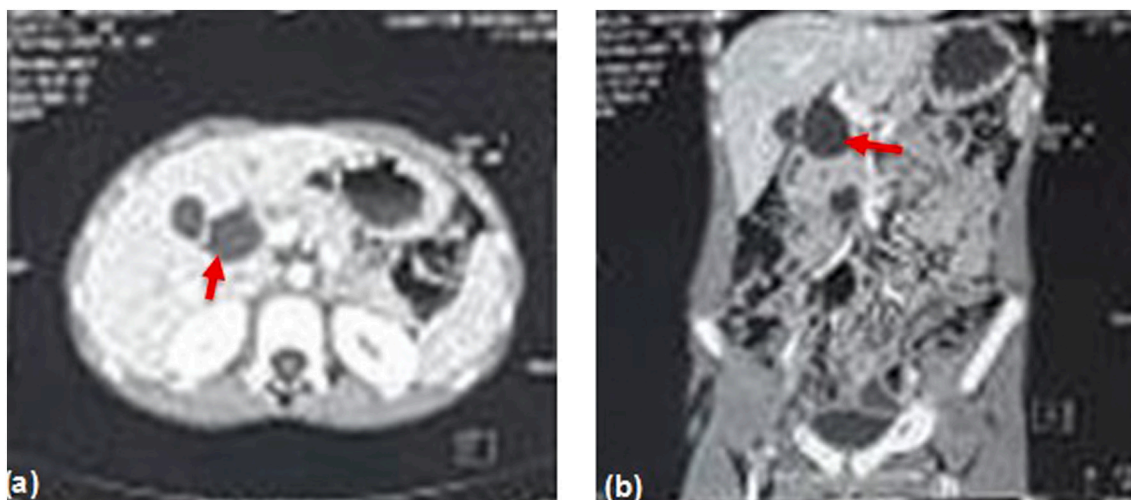


Fig. 1. Axial(a) and coronal (b) abdominal CT-images, showing a prominent cystic dilatation of common bile duct (red arrows) consistent with a Choledochal cyst.

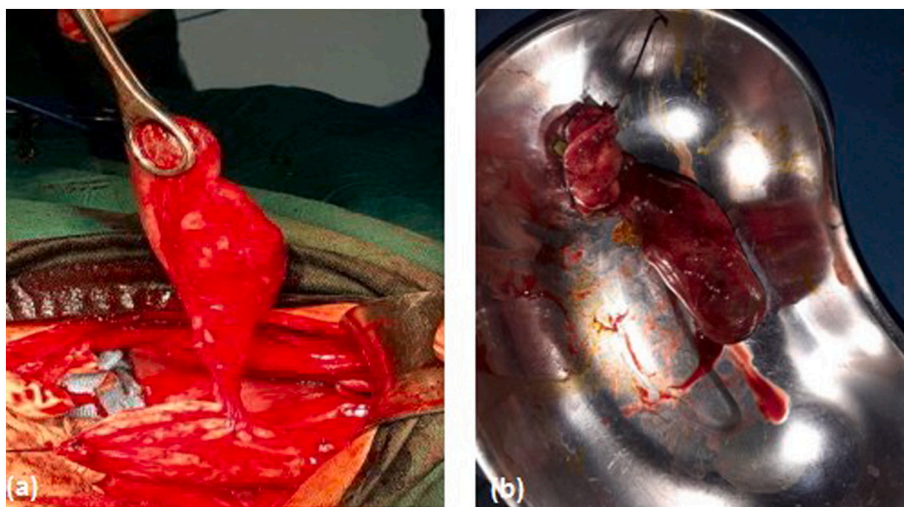


Fig. 2. Intra-operative (a) and (b) views, showing the resected choledochal cyst.

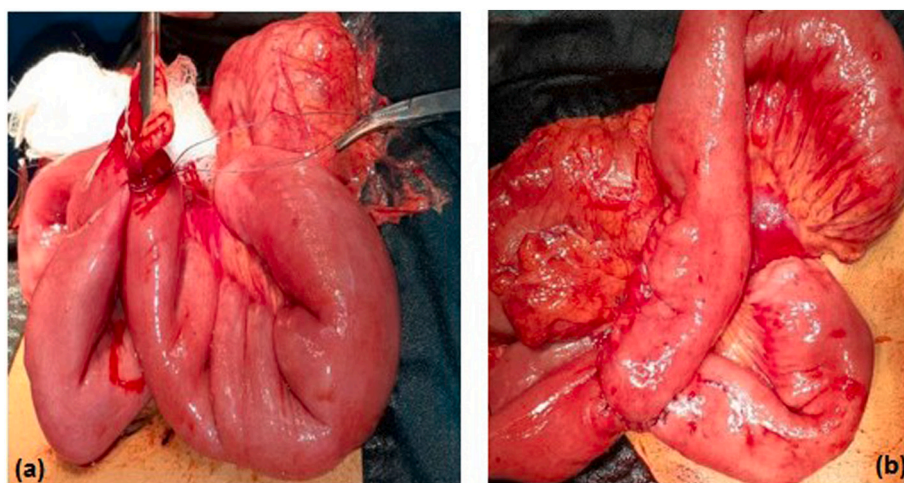


Fig. 3. Intra-operative (a) and (b) images, showing the sequential views of the Roux-en Y procedure.

longstanding vague abdominal pain. The abdominal CT images revealed a prominent cystic dilatation of the common bile duct (CBD) and mildly dilated intra-hepatic ducts consistent with a type IVa choledochal cyst.

The standard treatment of choledochal cyst consists of complete excision of the cyst and gallbladder with bile duct reconstruction through a Roux-en-Y hepaticojejunostomy [18,19]. It can be performed through the traditional open surgical approach or a minimally invasive surgical procedure using video laparoscopy, which is a very successful technique [6].

The surgical outcome is favorable in (85–90%) of cases. However, 6–10% of cases will need re-operation due to postoperative complications including bile leakage, anastomotic stricture, intrahepatic cholelithiasis, recurrent pancreatitis, adhesive bowel obstruction, and malignancy [20–22]. In our case according to our low-resource settings, we performed the open surgical intervention through a right subcostal (Kocher) incision. The cyst was successfully resected, but the bile leakage occurred as a postoperative complication. However, we managed the patient with re-operation through a midline incision for the evacuation of the biloma. Finally, he was discharged from the hospital with full recovery and a favorable outcome.

Although this is one of the documented cases, proved with imaging studies and managed by surgical intervention, the lack of the longtime follow-up may be the only limitation for this case.

4. Conclusions

Choledochal cyst is a rare congenital anomaly of the pediatrics age group. However, it may remain asymptomatic till adulthood. Low-resource settings, parents' literacy level, and less symptomatic cases may be the contributing factors in the diagnostic delay. Lack of the technical facilities and inadequate post-operative care (especially those families who wish their child to be discharged as early as possible and with no attention to the patient) can play an important role in post-operative complications.

CRediT authorship contribution statement

Concept – TH and RK; Design – HAE; Supervision – HAE; Resources and data Collection – TH; Literature Search – TH; Writing Manuscript – TH; Critical Review – HAE. All authors have read and approved the final manuscript.

Declaration of competing interest

The authors have no potential conflicts of interest to disclose.

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