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Choledochal cyst type I in infant: a case report

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ABSTRACT

Choledochal cysts are rare congenital anomalies characterized by dilatation of the bile ducts. The most frequent subtype of choledochal cyst is type I, which has fusiform dilatation of the common bile duct and poses significant challenges in diagnosis and management, particularly, in pediatric patients. We present a case report of a child diagnosed with a Type I choledochal cyst, detailing the clinical presentation, diagnostic workup, surgical intervention, and postoperative outcome. Early recognition and appropriate surgical management are crucial for optimal outcomes in children with this rare entity.

Keywords: choledochal cyst, children, congenital anomaly, surgical management

INTRODUCTION

Choledochal cysts are congenital malformations with dilatation of the bile ducts and are classified into several types based on various lengths and severity [1]. Choledochal cysts usually present with icterus, and abdominal enlargement, and resemble symptoms of biliary atresia or hepatitis [2]. Choledochal cysts can lead to progressive obstruction of the bile ducts and can eventually lead to biliary cirrhosis [3]. The incidence of choledochal cysts varies from 1:13.000 to 1:150.000 in children [4]. The incidence of choledochal cysts in Asia is high [5].

Choledochal cysts may occur in a portion of the bile duct within the liver (intrahepatic) or outside of the liver (extrahepatic). There are 5 types of choledochal cysts, the most common of which are type I choledochal cysts in the extrahepatic bile ducts, which account for 90% of all choledochal cysts, according to the Todani classification. Type II choledochal cysts are aberrant pockets or sacs within the duct. Type III cyst is located within the wall of the duodenum. Type-IV is divided into two subtypes: IVA cysts of intrahepatic and extrahepatic bile ducts (IVB), characterized by uncommon rare, numerous extrahepatic bile duct cysts. Type V, commonly known as Caroli's disease, combines intrahepatic cysts are combined with extrahepatic disease [3,6].

Type I choledochal cysts are rare but represent the most common subtype among pediatric patients and are characterized by fusiform dilatation of the common bile duct. The presentation of Type I choledochal cysts in children varies widely, ranging from asymptomatic cases incidentally discovered in imaging studies to symptomatic presentations such as abdominal pain, jaundice, and right upper quadrant mass remain diagnostic and management challenges, particularly in pediatric patients [3]. Diagnosis of choledochal cysts is usually done using multimodality imaging techniques and is mostly established with MRCP [6]. Early diagnosis and appropriate "surgical management are crucial to prevent complications such as cholangitis, pancreatitis, and malignancy [3]. Surgical treatment remains the cornerstone in managing type I choledochal cysts, aiming to resect the cystic segment and restore normal bil-

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iary anatomy. A case report of a child diagnosed with type I choledochal cyst is presented and focuses on the clinical presentation, diagnostic workup, and surgical intervention.

CASE REPORT

A 7-month-old child presented with jaundice for 1 week of age all over the body. Jaundice persisted and was followed by acholic stools and tea-colored urine (Figure 1). The patient was brought to the primary health facility at the age of 1.5 months because of persistent jaundice but was told to drink more and was told to be dried in the sun. The jaundice persisted at the age of 3 months. The patient suffered from progressive jaundice and an enlarged abdomen at 4 months of age. The patient was referred to a referral hospital for further examination.

There were no abnormalities during pregnancy. The patient was born by Caesarean section with an indication of premature rupture of membranes, full term, with a birth weight of 3700 grams, and immediately cried. The patient was breastfed until one month, and formula milk until now. There is no similar disease in the family.

The anthropometric examination showed severe malnutrition. The physical examination revealed vital signs were in the normal range. There was an icterus of the sclera, abdominal distended, hepatomegaly, splenomegaly, vein ectasis, and abdominal mass in the right upper quadrant. The patient's laboratory result was shown in Table 1, which showed anemia, cholestasis, hypoalbuminemia, and elevated liver enzymes.

FIGURE 1. Acholic stool

Abdominal ultrasonography showed a choledochal cyst Todani classification type Ia with gallbladder emptying disorder, no intrahepatic bile duct (IHBD) widening, and anechoic cystic lesion appeared, in extrahepatic bile duct (EHBD) to common bile duct (CBD). Magnetic resonance cholangiopancreatography (MRCP) revealed a cystic lesion with a fusiform shape size ±6 × 10.5 × 6.4 cm in the CBD which is hypointense on T1, hyperintense on T2, causing CHD dilatation size ± 1 cm, appears multiple cystic lesions in the left IHBD with the largest size ±0.8 × 0.7 cm with the conclusion of choledochal cyst type IVA (based on Todani classification) with sludge that causes CHD dilatation, hydrops gallbladder, and hepatosplenomegaly (Figure 2).

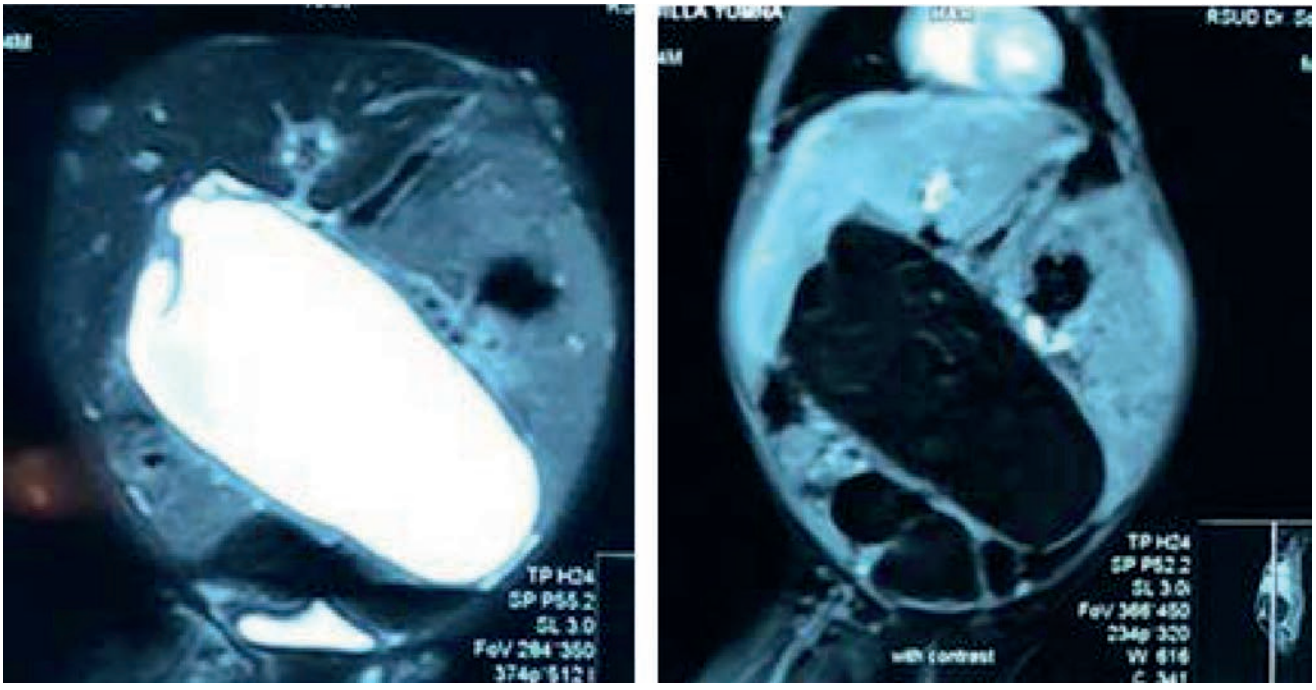

FIGURE 2. MRCP of the patient

TABLE 1. Laboratory results of the patient

Variable	Value
Hb/ Hct (g/dL/%)	9.7/ 28.5
WBC (103/µL)	11
Thrombocyte (103/µL)	153
Natrium (mmol/l)	132
Kalium (mmol/l)	3.2
Chloride (mmol/l)	106
PPT/ APTT (s)	12.8/ 35.1
Albumin (g/dL)	2.45
AST/ ALT (U/L)	68/ 41
Gamma GT (U/L)	339.1
Alkali Phosphatase (U/L)	240
Total Bilirubin/Direct Bilirubin (mg/dL)	8.3/ 5.9
BUN/ sCr (mg/dL)	8.4/ 0.3

Hb: Hemoglobin; Hct: Hematocrit; WBC: White Blood Cell; PPT: Plasma Prothrombin Time; APTT: Activated partial thromboplastin time; AST: Aspartat aminotransferase; ALT: Alanine aminotransferase; Gamma GT: Gamma-glutamyl transferase; BUN: Blood Urea Nitrogen; sCr: Serum Creatinine

The patient underwent Lily's procedure – hepaticojejunostomy Rouxen-Y. The cystic artery was ligated, followed by cholecystectomy, excision of CBD cyst, and the attached part of the cyst on the portal vein was performed Lily's procedure. A jejunum resection was performed 45 cm distal to the Treitz Ligament (Y lim) and an end-to-side jejunostomy suture was performed 75 cm distal to the treitz ligament (Roux limb 30 cm). Roux's limb was mobilized through the retrocolic space and continued with hepaticojejunostomy with bleeding ± 1400 cc during the operation and a drain was inserted (Figure 3). The patient underwent treatment in the Intensive Care Unit (ICU) for 4 days with intravenous analgesics, albumin transfusion, PRC transfusion during operation and post-operation, fresh frozen plasma transfusion, and empiric antibiotic injection.

DISCUSSION

Choledochal cysts are bile duct disease that can be either congenital or acquired. Choledochal cysts dilate the biliary tree, affecting extrahepatic and/or intrahepatic segments and eventually leading to cirrhosis. The precise etiology of choledochal cysts remains unclear although various theories have been proposed, including pancreaticobiliary maljunction, and abnormal pancreaticobiliary duct junction. Furthermore, genetic predisposition and environmental factors could affect to the formation of choledochal cysts [3].

The most prevalent symptoms in choledocal cysts were abdominal pain, followed by nausea/ vomiting, and jaundice. The classic triad of abdominal pain, jaundice, and right upper quadrant mass may also observed [7]. In this case, persistent jaundice, abdominal distension, hepatomegaly, and splenomegaly were obtained with laboratory tests showing cholestasis accompanied by impaired liver function such as hypoalbuminemia. Early diagnosis and treatment of choledochal cysts are essential for optimal outcomes. Low-resource settings, parental literacy, and less symptomatic cases may contribute to delayed diagnosis [8]. The patient suffered from persistent jaundice but was not adequately treated. The diagnosis of choledochal cysts is delayed, resulting in impairment of liver function. Studies showed that the mismanagement of neonatal jaundice among healthcare workers in terms of breastfeeding and sun exposure recommendations is still happening in cholestasis cases [9,10].

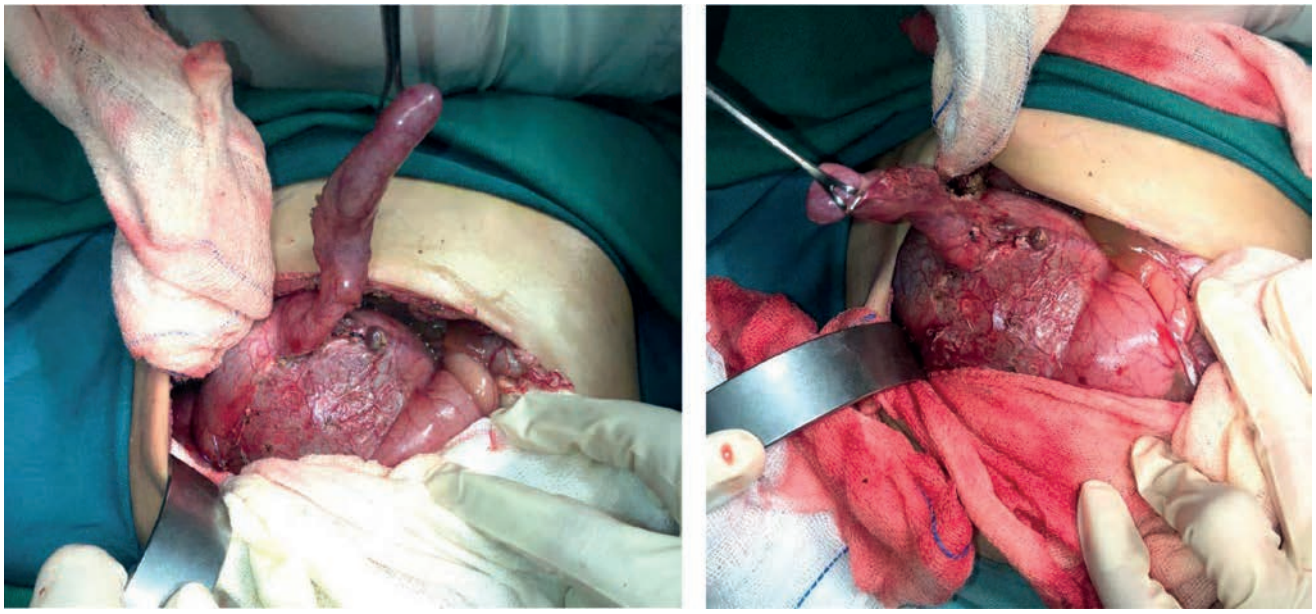

FIGURE 3. Choledochal Duct Cyst During Surgery

Choledochal cysts should be investigated if imaging shows dilatation of the bile duct or ampulla, particularly if there is no evidence of obstruction. In patients with ductal dilation, type I choledochal cysts should be considered in the differential diagnosis. The diagnosis of choledochal cysts is established by of abdominal ultrasound and computed tomography (CT). Ultrasonography is typically the initial imaging modality of choice due to its non-invasive method and accessibility [3]. Abdominal ultrasonography showed a type Ia choledochal cyst of Todani classification with impaired gallbladder emptying, and an anechoic cystic lesion from the extrahepatic bile duct (EHBD) to the common bile duct (CBD). Cysts are often suspected based on ultrasound findings when evaluating for jaundice, abdominal masses, or abdominal pain [11].

Infants presenting with early onset severe jaundice and cystic mass are most likely to be diagnosed with choledochal cysts [11]. Patients who are suspected of having bile cysts should undergo evaluation to establish the presence of the cysts and ascertain whether the cysts and the biliary tree are communicating. Additional studies may be needed to confirm the diagnosis. Magnetic resonance cholangiopancreatography (MRCP) or endoscopic retrograde cholangiopancreatography (ERCP) may be necessary to rule out the exclusion of biliary obstruction [12]. In this case, MRCP showed a cystic lesion with a fusiform shape causing a dilated CHD and multiple cystic lesions appeared in the left IHBD causing CHD dilatation, and hepatosplenomegaly.

Effective management of these children requires early cholangiography and surgical treatment [11]. Surgical intervention remains the cornerstone of treatment for choledochal cysts to prevent complications and alleviate symptoms. The surgical approach decision depends on various factors including the type and location of the cyst, the presence of associated anomalies, and surgeon's expertise. Patients with type I, type II, or type IV cysts are usually treated with cyst resection surgery due to the risk of malignancy. Patients with type III cysts have treatment if the cyst is symptomatic and manageable. Most typically, type V cysts are treated with supportive care to solve complications including sepsis and recurrent cholangitis. Type V cysts can be challenging to treat, and the majority of patients require a liver transplant. Hepaticojejunostomy (Roux-en-Y) and choledochal cyst excision combined with hepaticojejunostomy are common surgical technique. The primary goal of surgery is to achieve complete excision of the cyst with restoration of biliary-enteric continuity while minimizing the risk of postoperative complications such as anastomotic strictures and bile leakage [13].

Early surgical intervention is crucial to prevent complications associated with choledochal cysts, including cholangitis, pancreatitis, and biliary malignancy. Despite advancements in surgical techniques and perioperative care, choledochal cyst surgery may be associated with postoperative complications such as anastomotic strictures, bile leakage, and recurrent cyst formation. Long-term surveillance is essential to monitor for these complications and ensure optimal outcomes. Additionally, close follow-up is necessary to detect potential biliary malignancies, which have been reported as long-term complications of choledochal cysts [14,15].

CONCLUSION

Choledochal cysts, particularly Type I, represent a rare but significant congenital anomaly in children necessitating prompt surgical intervention to prevent complications and improve outcomes. Early detection is very important, especially in infants with persistent jaundice as choledochal cysts can lead to biliary cirrhosis. Long-term surveillance and close follow-up are essential to monitoring for potential complications such as biliary malignancy.

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