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Case Report

A choledochal cyst type IVa in a child treated with Roux-en-Y hepaticojejunostomy

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Abstract

A choledochal cyst is a bile duct anomaly that disrupts the transportation of bile from the liver to the gallbladder and small intestine. Choledochal cysts are rare, occurring in approximately one out of every 100,000 to 150,000 children in Western countries, with a girls-to-boys ratio of 4:1. Immediate surgery to excise the cyst and construct a biliary-enteric continuity is necessary to treat this condition. This case-report aimed to present a child with choledochal cyst type IVa who underwent a Roux-en-Y hepaticojejunostomy. A 3-year-11-month-old girl with an abdominal mass experienced jaundice, nausea, and vomiting over the past two years, which worsened in the last month. Abdominal ultrasonography indicated intrahepatic biliary dilatation. Abdominal computed tomography scan results confirmed a choledochal cyst type IVa, characterized by fusiform cyst dilatation at the bilateral intrahepatic bile duct, common hepatic duct, cystic duct, and common bile duct. The cyst exerted pressure on the pancreas and small intestine. Before the surgery, the patient was treated with ceftriaxone 100 mg/kg/day and gentamicin 5 mg/kg/day. Roux-en-Y hepaticojejunostomy was performed, involving the complete excision of the extrahepatic bile duct to reconstruct the biliary system. During the surgery, a retroperitoneal cyst measuring 20 cm x 10 cm with a volume of 200 ml was discovered. Following the surgery, the patient showed clinical improvement. Patient follow-ups indicated that no complications such as wound infection, acute pancreatitis, and the formation of pancreatic or biliary fistula occurred. This case highlights that Roux-en-Y hepaticojejunostomy proves to be an effective surgical approach for managing choledochal cyst type IVa in children, helping to prevent further complications.

Keywords: Choledochal cyst, hepaticojejunostomy, Roux-en-Y, congenital anomaly, bile duct

Introduction

A choledochal cyst is a dilatation of the biliary tree that affects either intrahepatic or extrahepatic segments or both. The most frequently used classification for these cysts is the Todani classification, which categorizes choledochal cysts into five main types [1]. Type I, the most common type, is a saccular or fusiform dilatation of the extrahepatic bile duct (50–90%) while type IV is the second most common (11–35%), involving the dilatation of extrahepatic or intrahepatic bile ducts [2]. Choledochal cysts can occur at any age. Clinical symptoms associated with choledochal cysts generally arise due to bile stasis, stone formation, recurring

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superinfection, and inflammation. Approximately two-thirds of patients exhibit clinical symptoms before the age of 10 years [3]. The classic triad of choledochal cyst symptoms includes abdominal pain, jaundice, and a palpable mass in the right upper quadrant of the abdomen [4,5]. However, only 20% of adults and 30–60% of patients under 10 years old have typical symptoms [6].

Ultrasonography (USG) is the initial imaging choice for determining the location and size of choledochal cysts. This method is noninvasive, fast, cost-effective, widely available, and has a sensitivity ranging from 70 to 97% [4,7]. Computed tomography (CT) scans, endoscopic retrograde cholangiopancreatography (ERCP), and magnetic resonance cholangio-pancreatography (MRCP) are used to determine the origin of the cyst and differentiate choledochal cysts from other intraabdominal cysts [4,5,7]. CT scans, being the most readily available and noninvasive imaging technique, are capable of revealing the cyst continuity with the bile duct or surrounding tissues as well as any associated malignancy [5,7].

The etiology of choledochal cysts is still a subject of debate. Some believe that choledochal cysts are congenital anomalies, given that most cases are diagnosed in infants and children. On the other hand, others believe that choledochal cysts are acquired because approximately 20% of patients receive a diagnosis in adulthood [7]. The prevent complications such as pancreatitis, cystolithiasis, and cholangitis associated with choledochal cysts, prompt diagnosis and treatment is key. The Roux-en-Y hepaticojejunostomy is the mainstay of treatment options with complete excision of the dilated bile duct. Choledochal cysts generally have an excellent prognosis after complete resection. The aim of this case report was to present the management of choledochal cyst type IVa in a child treated with Roux-en-Y hepaticojejunostomy, resulting in a good prognosis.

Case

A 3-year-11-month-old girl was referred from Ibnu Sina Sigli Hospital in Pidie Regency to Dr. Zainoel Abidin Hospital in the capital city of Aceh, Banda Aceh, Indonesia. The patient presented with symptoms including abdominal pain, hepatomegaly, recurrent bacterial infection, and secondary thrombocytosis. The patient's abdomen had enlarged over the past two years, with a recent worsening within the last month. This was accompanied by jaundice affecting the whole body. The patient had intermittent fever two weeks before admission. There was no history of tea-colored urine, clay-colored stools, diarrhea, or paleness.

The abdominal USG suggested an intrahepatic bile duct dilatation (**Figure 1**). The non-contrast abdominal CT scan revealed a cystic and fusiform dilatation of the intrahepatic bile duct, common hepatic duct, cystic duct, and common bile duct. This dilation exerted pressure on the pancreas and intestines (**Figure 2**). The patient was diagnosed with choledochal cyst type IVa and was scheduled for surgery with Roux-en-Y hepaticojejunostomy once stabilized.

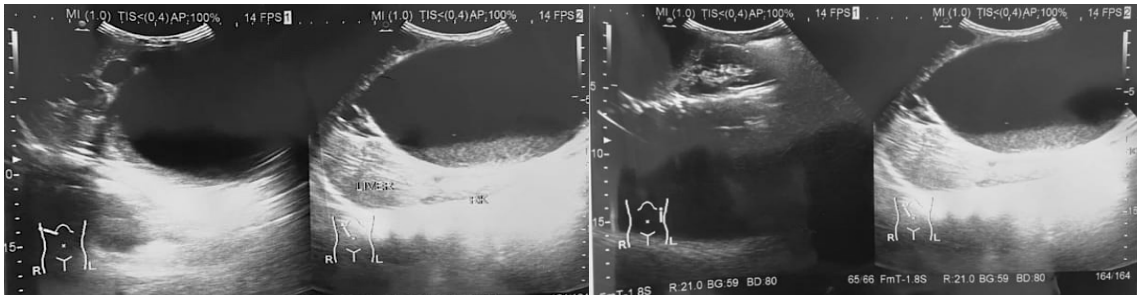

Figure 1. Abdominal ultrasonography (USG) shows a cystic lesion with intrahepatic bile duct dilatation.

The laboratory results revealed a hemoglobin level of 10.3 g/dL, leukocyte count of 16,400/mm^3, platelet count of 829,000/mm^3, serum glutamic oxaloacetic transaminase (SGOT) level of 162 U/L, serum glutamic pyruvic transaminase (SGPT) level of 143 U/L, total bilirubin level of 9.28 mg/dL, direct bilirubin level of 5.98 mg/dL, and indirect bilirubin level of 3.30 mg/dL. Other parameters were within normal limits. The patient received intravenous

ceftriaxone at a dose of 600 mg twice a day, vitamin K at 5 mg once a day, aspirin at 50 mg once a day, and symptomatic drugs.

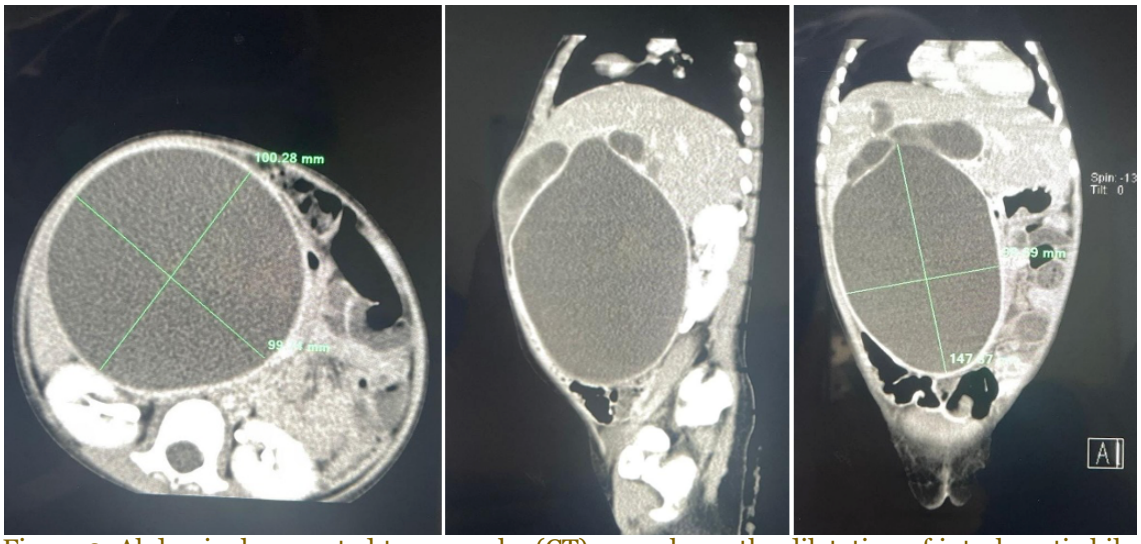

Figure 2. Abdominal computed tomography (CT) scan shows the dilatation of intrahepatic bile duct, common hepatic duct, cystic duct, and common bile duct, which pressed the pancreas and intestines.

On the 9th day of the admission, the laboratory results revealed a hemoglobin level of 12.6 g/dL, leukocyte count of 17,400/mm3, platelet count of 994,000/mm3, SGOT level of 108 U/L, SGPT level of 83 U/L, total bilirubin level of 6.60 mg/dL, direct bilirubin level of 4.80 mg/dL, and indirect bilirubin level of 1.80 mg/dL. As the leukocytosis improved, the patient was prepared for the surgery. The patient underwent Roux-en-Y hepaticojejunostomy, revealing a retroperitoneal cyst measuring 20x10 cm with drained fluid amounting to 200 ml. An end-to-side anastomosis from the Roux and Y limb was constructed to ensure biliary system continuity.

After surgery, the patient experienced episodes of green-colored vomitus amounting to 500 ml, accompanied by intermittent fever. The abdominal pain decreased and the jaundice showed signs of improvement (**Figure 3**). The laboratory results indicated improved levels of SGOT, SGPT, total bilirubin, direct bilirubin, and indirect bilirubin. The patient was treated with intravenous meropenem at a dose of 450 mg three times a day, along with maintenance of adequate fluid balance. After being afebrile for a period of three days, the patient was discharged. Follow-up of the patient indicated that no early complications (wound infections, acute pancreatitis, and the formation of pancreatic or biliary fistula) occurred.

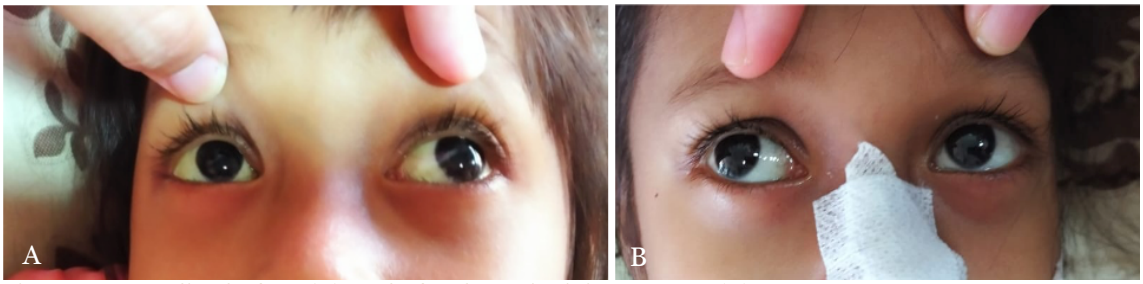

Figure 3. Jaundice before (A) and after hepaticojejunostomy (B).

Discussion

Choledochal cyst mainly occurs in 80% of children, with the triad of jaundice, stomach pain, and a palpable mass in the right upper quadrant being primarily observed in young patients. These manifestations might be accompanied by coagulopathy, pancreatitis, cholangitis, portal hypertension, and abnormal liver function. Notably, jaundice is especially prevalent in choledochal cyst type I and IV due to the abnormal pancreaticobiliary ductal union (APBDU), leading to pancreaticobiliary reflux [7].

The symptoms associated with choledochal cysts display distinct patterns in infants (≤1 year old) compared to the pediatric group (1–18 years old). Newborns are more prone to abdominal pain and clay-colored stools than their pediatric counterparts [8,9,10]. Pediatric patients, on the other hand, are more likely to present with an abdominal mass and abnormal pancreaticobiliary ductual union (APBDU) than adults [5]. Prenatal diagnosis allows early treatment of choledochal cysts, a practice followed in most developed countries. In contrast, choledochal cysts are often diagnosed after the appearance of symptoms in developing countries [7]. Cholestasis and jaundice are more common in infants while in older children and adults, abdominal pain and pancreatitis are more common [11,12]. A study assessing the cholangiography characteristics of 85 children with common bile duct dilatation revealed that most choledochal cysts were symptomatic during childhood, although some were diagnosed prenatally (15%) or in adulthood (20%) [13]. In the present case-report, the patient presented with complaints of painful abdominal enlargement that occurred two years prior, but was further exacerbated with jaundice in the month leading up to admission.

The classification of choledochal cysts is determined by imaging examinations. USG, the preferred initial imaging technique, is quick, inexpensive, noninvasive, and widely accessible. USG can reveal the cyst's position, size, and fusion with other organs, such as the liver, duodenum, and portal vein [10,11]. In our case, the intrahepatic bile duct dilatation was initially visualized with USG.

Dilatation of the intrahepatic bile duct necessitates further imaging to differentiate choledochal cyst types I and IVa. Moreover, the cyst and bile duct might resemble other intra-abdominal cysts, such as pancreatic pseudocyst, echinococcal cyst, and cystadenoma [12]. In this case, we confirmed the diagnosis of choledochal cyst type IVa after obtaining abdominal CT scan results, which indicated a cystic and fusiform dilatation of the intrahepatic bile duct, common hepatic duct, cystic duct, and common bile duct. The CT scan not only confirms the extent of ductal involvement or extrahepatic disease [10,11], but it also demonstrates the location of the choledochal cyst based on the mass and fluid density while determining the involvement of intrahepatic or extrahepatic bile ducts [5].

The standard approach for managing choledochal cyst type I and IV is extrahepatic bile duct excision which eases the choledochal cyst removal. The biliary-enteric continuity is achieved with anastomosis technique known as Roux-en-Y hepaticojejunostomy [10,11]. The patient in our case-study had no more jaundice and abdominal enlargement following the prompt diagnosis and Roux-en-Y hepaticojejunostomy.

Resection of pediatric choledochal cysts is generally well tolerated [13,14]. Early complications may include anastomotic leaks, post-operative bleeding, wound infections, acute pancreatitis, and the formation of pancreatic or biliary fistulas. Acute complications subsequent to the resection procedure, such as wound infection, have been reported in 0–17% of cases [15, 16]. Late complications include anastomotic stricture, hepatolithiasis, cirrhosis, cholangitis, and malignancy. Benign anastomotic stricture with recurrent cholangitis is less common in children (10–25%) compared to adults and might be associated with gallstone formation [15,16]. In the presented case, no early complications occurred; nevertheless, we have scheduled follow-up visits in order to detect any late complications.

Conclusion

A choledochal cyst, particularly of type IVa, is an infrequent biliary tract anomaly that can hinder prompt diagnosis. Imaging modalities such as USG and CT scan, are necessary to identify the choledochal cyst's type, location, and size. As the definitive therapeutic approach, Roux-en-Y hepaticojejunostomy results in an immediate good prognosis for the child patient.

Ethics approval

The patient family provided written informed consent to be published as a case report.

Competing interests

The authors declare that there is no conflict of interest.

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Underlying data

All data underlying the results are available as part of the article and no additional source data are required.

How to cite

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