INTRODUCTION

Choledochal cysts (C.C.s) or biliary cysts are congenital dilatations of extrahepatic and/or intrahepatic bile ducts. Approximately 80% of C.C.s are diagnosed in infants and young children in the first decade of life. In the Asian population, the incidence can be as high as the population.1-4 The exact etiology of choledochal cysts is still unknown. There are several classifications for choledochal cysts. The modified Todani classification with five types of CC was implemented in 1977 and is the most widely used.3,5 In most cases of C.C.s are diagnosed during childhood. C.C.s present in children as a right upper quadrant mass, abdominal pain, and jaundice, also known as the classic triad for C.C.s, found in only 20% of cases. 85% of children present with 2 of these classic clinical features, including an abdominal mass and jaundice. Infants under 12 months of age may present with jaundice, alcoholic stools, and vomiting.

Other features of choledochal cysts are cholangitis, pancreatitis, and biliary peritonitis due to cyst rupture. Biliary malignancy is seen in 10% to 30% C.C.s. Malignancy is rarely seen in pediatric C.C.s. 5-7

The treatment of choledochal Cyst commonly involves excision of the cyst, but the biliary enteric continuity can be achieved with different reconstruction techniques. Some surgeons prefer hepaticojejunostomy (H.J.) anastomosis with a Roux-en-Y limb, while others use hepaticodudodenostomy (H.D.) reconstruction. Both techniques have advantages and disadvantages. Hepaticojejunostomy (H.J.) techniques have fewer complications of bile leak, and the incidence of cholangitis postoperative is less common. The advantages of H.D. include faster operative time and faster recovery of bowel function.8-10

Based on those mentioned above, this case study aims to evaluate the choledochal cyst Todani Type IA case in early childhood from the clinical diagnosis and treatment of choledochal cysts.

CASE REPORT

A two-year-old girl was referred to our hospital with a chief complaint of abdominal pain in the upper right quadrant one month before admission. Pain is felt in the upper right quadrant. The patient had recurrent fever with the highest temperature of 39.7°C since one month ago. History of cloudy urine about one week before admission to the hospital. Pain when urinating is denied. There was no consanguineous in her parents.

There was no history of previous hepatobiliary disorders, routine medication or hospitalization since birth. There was no familial history of recurrent cholestasis or jaundice. There was no history of congenital anomalies in her family. There was no consanguineous in her parents. No history of illness or consuming any medicine during pregnancy was noted. There was no abnormality during

Choledochal Cyst (CC) Todani type I in two years old girl: a case report

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ABSTRACT

Backgrounds: Choledochal cyst (CC) is a rare congenital cystic dilation of the bile duct and can be associated with severe complications, including malignancy and inflammation of the surrounding anatomy. Approximately 80% of CC is diagnosed in infants and young children in the first decade of life. Complete surgical resection is recommended in patients with choledochal malformations with excellent outcomes.

Case Presentation: A two-year-old girl had intermittent abdominal pain in the upper right quadrant. This complaint was accompanied by fever, nausea and vomiting, decreased appetite and drinking. The physical examination revealed jaundice on the sclera and palpable liver, two fingers below the costal arch. Abdominal ultrasound revealed a cystic mass in the right lower lobe suggestive of the hepatic cyst with inflammation of the cystic head of the pancreas and minimal free fluid per lumen. An abdominal C.T. scan without contrast showed a choledochal cyst classified as Todani Type IV. A cyst in the bile duct was found during the surgical procedure.

Conclusion: Choledochal cyst is one of the differential diagnoses in children with symptoms of abdominal pain. Appropriate surgical measures should be undertaken to avoid complications.

Keywords: Children, Choledochal Cysts, Cholestasis.


Published by Intisari Sains Medis
pregnancy or delivery. The patient was the second child in the family. He was born full term spontaneously, with a birth weight of 3100 grams, healthy condition, and breastfed for years. The immunization record was completed according to the government's recommendation. His food recall was following the recommended daily allowance (R.D.A.). The patient never had a blood transfusion history.

The patient was fully alert when admitted to our hospital. The patient's vital signs were pulse 100 bpm, temperature 36.9°C, respiratory rate 24 bpm, oxygen saturation 99% with room air, and pain scale 4. Abdomen examination was souple, and no tenderness was observed. The liver was palpated 1 cm below the costal arch, sharp and flat. The spleen was not palpable. Other examinations were within normal limits. The laboratory investigation revealed a complete blood count with hemoglobin (Hb) 9.3 g/dL, hematocrit (Ht) 29.4%, leukocytes 22.38 mm3 and thrombocytes 357,000 mm3 (Table 1). Liver function test showed an increase of serum aspartate transaminase (AST) 165 U/L, alanine aminotransferase (ALT) 256 U/L, and gamma-glutamyl transferase (GGT) 254 U/L (Table 2). Renal function test has shown BUN 18.9 mg/dL and creatinine serum 0.5 mg/dL. Urinalysis revealed yellow color, pH 6, leucocyte esterase negative, nitrite negative, protein +1, ketone negative, urobilinogen normal, bilirubin negative, erythrocytes 0 per field of view, leucocytes 1 per field of view (Table 3). Abdominal ultrasonography revealed a suspect biliary cyst (Figure 1). Magnetic resonance cholangiopancreatoigraphy (M.R.C.P.) showed fusiform dilatation of the extrahepatic bile ducts with moderate dilatation of the right and left intrahepatic bile duct, suggesting a choledocal cyst, with Todani classification type Ia (Figure 2).

Based on the clinical and other examinations, the patient was diagnosed with Choledocal Cyst Type 1 Todany classification. The patient was given maintenance fluids, cefoperazone, amikacin, metronidazole, and paracetamol and consulted a Pediatric Surgery. The patient was planned to have laparotomy, choledochal cyst excision and hepaticojunostomy Roux-en-Y. During the surgical procedure, the surgeon found a cyst in the bile duct, and then they performed common bile duct cyst excision and hepaticojunostomy Roux-en-Y (Figure 3 and Figure 4).

After surgery, the patient was admitted to the Pediatric Intensive Care Unit (PICU) and given intravenous antibiotics: cefoperazone, amikacin, and metronidazole. The patient was fasting and given total parenteral nutrition for three days. Three days after surgery, tropic
sample confirmed the diagnosis of a choledochal cyst.

DISCUSSION

Choledochal cysts (C.C.s) or biliary cyst is congenital dilatation of extrahepatic and/or intrahepatic bile ducts. CC involves widening the biliary tree, which may affect the extrahepatic and/or intrahepatic segments. Approximately 80% of CC is diagnosed in infants and young children in the first decade of life. Evident regional variation exists for choledochal cysts, with two-thirds of cases reported in Asia occurring in Japan. Choledoccal cysts are four times more common in women. C.C.s type I and IV have a female-to-male ratio of 4:1 or 3:1.

The patient was a two-year-old girl with complaints of abdominal pain, fever, and choledochal cysts without a history of congenital anomalies in her family. The precise etiology of the extrahepatic cyst is still unclear. Type I cysts are associated with an abnormal arrangement of the pancreaticobiliary ducts (A.P.B.D.), also known as "common channel". A long common channel can cause various pathologic conditions, such as pancreatitis, stenosis of the papilla of Vater, and choledochal cyst. A typical channel may enhance the reflux of pancreatic juice into the bile duct, exposing the common bile duct wall to pancreatic enzymes and increasing pressure in the choledochal duct, resulting in cyst formation. The complete type of choledochal cysts are as follows: type IA: cystic dilatation of the extrahepatic duct; type I.B.: focal segmental dilatation of the extrahepatic duct; type I.C.: fusiform dilatation of the entire extrahepatic bile duct and common bile duct; type II: simple diverticulum of the common bile duct; type III: cyst/choledochoele distal intramural dilation of the common bile duct; type I.V.A.: combined intrahepatic and extrahaepatic bile duct dilatation; type I.V.B.: multiple extrahepatic bile duct dilatations; type V: multiple intrahepatic bile duct dilatation. In this case, no history of illness nor consuming any medicine during pregnancy was noted. There was no abnormality during pregnancy or delivery.

The classic triad of choledochal cysts is abdominal pain, jaundice, and a surgical procedure, and the patient was discharged in good condition. The patient came for a follow-up in good condition. Pathologic examination from the biopsy feeding from the feeding tube was given. The patient was admitted to the regular ward and was fully fed on the eighth day of surgery. No complication was noted post-surgical procedure, and the patient was discharged in good condition. The patient came for a follow-up in good condition.
choledochal cysts with a high sensitivity of 90% to 100%. M.R.C.P is noninvasive, does not expose the patient to ionizing radiation, and has no associated complications such as bleeding, perforation, cholangitis or acute pancreatitis as seen on ERCP. There are no specific laboratory studies for choledochal cysts. However, the presence of high amylose levels and several studies have reported increased levels of phospholipase A2 and trypsinogen in bile CCs. Performing the bile enteric anastomosis in the lower portion of the common hepatic duct is safer and has a lower risk of complications. In this case, abdominal ultrasonography revealed a suspect biliary cyst. Magnetic resonance cholangiopancreatography (M.R.C.P) showed fusiform dilatation of the extrahepatic bile ducts with moderate dilatation of the right and left intrahepatic bile duct, suggesting a choledocal cyst, according to the Todani classification type I.A.

The management approach of choledochal cysts depends on the cyst type and the extent of hepatobiliary pathology. All cysts should be resected, and bile flow should be restored. Type I and type IV CCs are managed with complete excision of the choledochus and bile flow restoration, preferably Roux-en-Y hepaticojejunostomy (H.J.) or hepaticodudenumostomy (H.D.). The risk of type II and type III cysts to transform into malignancies is lower. The hypothesis is that the cause of malignancy after choledochal cyst resection is performed, the development of malignancy after choledochal cyst surgery can occur. The risk of developing malignancy among patients with choledochal cyst malformation was almost 11%, and the risk following cystic drainage surgery was four times higher after complete cyst excision. Therefore, complete surgical resection is recommended in patients with choledochal malformations.

We conclude that we should consider choledocal cysts as one of the differential diagnoses in children with symptoms of abdominal pain. Awareness of this may reduce complaints and avoid any delay in definitive surgical treatment. Once a choledocal cyst is diagnosed, appropriate surgical measures should be undertaken to prevent complications. The limitation of this case report is that it does not include biopsy results.

CONCLUSION

Cholelcal cyst is one of the differential diagnoses in children with symptoms of abdominal pain. Appropriate surgical measures should be undertaken to avoid complications.
CONFLICT OF INTEREST
None.

ETHICAL CONSIDERATIONS
This case report has received informed consent from her parents.

FUNDING
This research uses private funds from researchers.

AUTHOR CONTRIBUTIONS
All authors provide a balanced contribution.

REFERENCES