A giant choledochal cyst in 8 years old female

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ABSTRACT

Choledochal cyst incidence in the Asian population is 1:1000 live births and more common in females. The classic triad that can be found in younger children is jaundice, abdominal pain, and a palpable mass in the abdomen. Various techniques have been performed, but cyst excision and Roux-en-Y hepaticojejunostomy still preferred. Inappropriate management can cause several complications. We reported a rare case of giant choledochal cyst in 8 years old female presents with abdominal pain and a palpable mass in the abdomen from below arcus costae to right inguinal region. Laboratory findings showed an elevation of serum bilirubin, alkaline phosphatase, and gamma-glutamyl transferase level. Computed tomography scans showed a type IV choledochal cyst, with a size 117.1x83.5 mm. First step surgery, external drainage was performed. The definitive surgery was incomplete cyst excision and Roux-en-Y hepaticojejunostomy. The patient was discharged in good condition. The long-term prognostic is dubia ad bonam but biliary tract malignancy may still occur.

Keywords: giant choledochal cyst, Roux-en-Y hepaticojejunostomy


INTRODUCTION

Choledochal cysts are rare congenital, but not familial anomalies of the intrahepatic or extrahepatic biliary tract.1 The incidence in the Asian population is 1 in 1000 live births. The reason for this Asian preponderance is still unclear.2 There is also an unexplained female: male dominance, commonly reported as 1.5:1 in pediatric population.3

Choledochal cysts are usually classified according to Todani in five types: type I consists of dilatation of the extrahepatic bile ducts. Type II describes a diverticulum in the common bile duct (CBD). Type III is a focal dilatation of the distal CBD in the papillary region into which the pancreatic ducts drain. Type IVa shows multiple dilatations in the extra and intrahepatic bile ducts, whereas type IVb represents multiple dilatations in only the extrahepatic bile duct. Type V is synonymous with Caroli disease and consists of cystic dilatation of the intrahepatic biliary system.4,5

Imaging techniques confirm the diagnosis of choledochal cyst. Abdominal ultrasonography (USG) is the initial diagnostic modality of choice, allowing for precise measurements of intra or extrahepatic duct dilatation and identification of stones and sludge. Abdominal computed tomography (CT) is highly accurate and also helps in planning surgical approaches. Magnetic resonance cholangiopancreatography (MRCP) is regarded as the gold standard for the diagnosis of choledochal cyst, with sensitivity 90-100%.6,7

Total cyst excision and biliary reconstruction is the treatment of choice. Biliary reconstruction may
be achieved by one of several techniques based on the surgeon's personal preference. Roux-en-Y hepaticojejunostomy (RYHJ) is commonly used.  

We present a rare case of giant choledochal cyst in a child who had undergone incomplete cyst excision and biliary reconstruction using the RYHJ technique.

**CASE ILLUSTRATION**

IPS, an 8-year-old female was referred with suspected hepatoma, differential diagnose of cirrhotic hepatic. The chief complaint was abdominal pain since three days before admission. The pain was started at epigastrium and then spread to the right upper quadrant of the abdomen; it was colicky and felt more painful. The pain was not getting better or worse after taking a meal. History of trauma was denied. The patient also complains about the abdomen was getting distended when she was admitted at B public hospital.

Fever was felt since two days before admission but never checked by parents. They only took antipyretics. Vomiting complained since two days before hospitalization, contains food, and always happened after taking a meal. There was no nausea. Furthermore, the vomiting contains blood, three times daily, without blood clots.

The patient had a history of abdominal pain and jaundice when she was three years old. At the time, the patient was admitted to the B public hospital for one week with a diagnose of liver swelling. The patient was getting better after getting blood transfusion and medicine, but the parents forgot the kind of medicine.

On physical examination, we found no jaundice sclera, the abdomen looks distended, tenderness, liver swelling 5 cm below arcus costae, and 3 cm below xhypoid processus. Erythema palmaris and spider nevi were not found. During treatment at our hospital, the patient complained about the abdomen getting more distended than before, and on physical examination, a cystic mass was palpable below the arcus costae until the right inguinal region, and the liver evaluation was difficult.

Laboratory findings showed leukocytosis with neutrophil dominant (leucocyte 18.75 K/µL, neutrophil 70.47%, lymphocyte 18.22%), elevated serum transaminase (SGOT: 94.8 u/L; SGPT 164.5 u/L), elevated serum bilirubin level (bilirubin total 3.18 mg/dL, bilirubin direct 1.57 mg/dL), elevated serum alkali phosphatase (194 u/L), gamma-glutamyl transferase (GGT) 117 u/L. Abdominal USG revealed heterogeneous solid lesion on right lobe suspected liver abscess and suspected biliary duct involvement with a sign of biliary tract obstruction. Abdominal CT revealed choledochal cyst type 4 with size 117.1 x 83.5 mm (Figure 1).

Initial management were antibiotics, ursodeoxycholic acid (UDCA), vitamins A, D, E, K, omeprazole, and analgesics. Because of the pain worsening, fentanyl was added. The pediatric surgeon performed external drainage as initial management. Drain production was 400–600 ml per 24 hours and dark green in color. The drain was maintained for seven days. During the first surgery, we found a massive cyst, and there was a part of the cyst was attached to the abdominal aorta (Figure 2).

The second surgery was performed, there was incomplete cyst excision to prevent vascular trauma and biliary reconstruction by RYHJ. The patient was discharged in good condition.

**DISCUSSION**

Choledochal cyst is a rare congenital anomaly with more than 60% present during the first year of life and about 20% present in adulthood. Choledochal cyst is defined as an abnormal, disproportionate, cystic dilatations of the biliary duct. Choledochal cyst is 3-4 times more common in females.

Choledochal cyst in childhood are frequently categorized into an infantile group (in the 1-year-old patient) and a classical pediatric group (in more
than the 1-year-old patient), the two groups differ markedly in their clinical presentation and the pathological anatomy. The classic triad can be found in younger children is jaundice, abdominal pain, and a palpable mass in the abdomen but symptoms depend on the age at presentation. Obstructive jaundice is the main presenting symptoms in the infantile group which is similar to jaundice in biliary atresia, but abdominal pain is the most typical symptoms in younger children. Our case 8 years old female, referred to our hospital with chief complaint abdominal pain and the other complaints were distended abdomen. History was abdominal pain and jaundice at three years old. She was diagnosed with liver swelling and fully recover with blood transfusion and some medicine. In observation during hospitalization, the abdomen becomes more distended and painful.

Choledochal cyst classified into five types according to Todani, based on the location of biliary duct dilatation or kind of cyst. Type I is by far the most common, accounting for 90% to 95% of cases, and constitutes the cystic/saccular or fusiform dilation of the common bile duct. Type II is a diverticulum of the CBD with no dilatation of the common bile, extrahepatic, or intrahepatic ducts. Type III also referred to as a choledochocele usually has a normal CBD and main pancreatic duct with cystic dilatation of the distal CBD that is either intraduodenal or intrapancreatic in location. The ducts may either enter the choledochocele separately or in the union at the wall of the duodenum, but are usually stenotic at their openings due to chronic inflammation. Type IV is composed of multiple cysts located intrahepatically, extrahepatically, or in both locations. Type IV can be subdivided into two types: IVa involving various intra and extrahepatic, IVb involving only multiple extrahepatic biliary dilatations. Type V cysts in conjunction with hepatic fibrosis are commonly referred to as Caroli disease. The size of the choledochal cyst usually is two until 9 cm. If the choledochal cyst size was 117.1 x 83.5 mm.

The principle of treatment of the choledochal cyst is a cyst resection, improves and ensures the integrity of bile and pancreatic duct attention may also experience an anomaly. To prevent the occurrence of malignancies, total resection of the choledochal cyst is considered the best measure. If there are adhesions between the cyst with a network behind it so as difficult and traumatic vascular released, part of the posterior wall of cysts can be abandoned, but the mucosa removed by means peeled. Drainage is necessary for a giant choledochal cyst in children because of excessive fluid exudation from broad dissection area, inflammatory oozing, ascites caused by liver dysfunction. In the giant choledochal cyst case, the surgeon has to open the cyst wall and drain bile, which may be sometimes as much as 3L to decompress the cyst. It gives better access to the surrounding structure and allows the identification of the luminal appearance grossly to determine the proximal transaction line.
resection followed by hepaticoenterostomy anastomosis high layout. The aim of definitive surgery for choledochal cyst includes excision of the cyst wall and the gall bladder, furthermore can reduce risk of malignancy, interruption of pancreatic-biliary reflux, reconstruction of the biliary-enteric channel for optimal drainage of bile. In our case, first step surgery, external drainage already was performed to decrease the intra-abdominal pressure and decompress the cyst. In this step, the pediatric surgeon found part of the choledochal cyst attaches to the abdominal aorta. Therefore, the definitive surgery was incomplete cyst excision and RYHJ. Incomplete cyst excision is done to prevent vascular trauma that can cause severe bleeding during the operation.

The best management of the giant choledochal cyst is total excision, but it challenging to be done because of the big size of the cyst. Partial removal can induce reflux of pancreatic enzymes into the biliary tract, thereby increasing the incidence of carcinoma of the bile duct. Postoperative complications can be visible immediately and lately. The immediate complexity is fluid and electrolyte derangement caused by multifactorial such as vomiting, bile ascites, electrolyte sequestration into the cysts, and reduced intake due to inadequate feeding during the illness. Furthermore, drainages from the surgical site postoperatively may have accentuated the electrolyte and fluid deficits. The electrolyte levels in our case post-drainage were sodium serum 138 mmol/L, potassium serum 4.2 mmol/L, calcium serum 8.33 mg/dL, and chloride 100.70 mmol/L — all of that level within the normal range. We need to monitor the electrolyte level to observe the late complication.

Postoperative morbidity and mortality are low incidences in children. The early complication such as bile leaks and wound infection. The late complications are biochemical liver dysfunction, persistent dilatation of intrahepatic bile duct, recurrent abdominal pain, recurrence of the CBD adenocarcinoma, and repeated cholangitis. Giant choledochal cyst causing more often mortality compared to a choledochal cyst in a smaller size. Our case, there was no early complication, but we have not already known the late complication yet, therefore our case need long-term monitoring.

**SUMMARY**

Giant choledochal cyst, a rare congenital anomaly in 8 years old female was reported. The diagnosis was established based on clinical manifestation and laboratory findings. The case was treated with medicine and surgery and has a good prognosis.

**REFERENCES**


