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CASE REPORT

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Choledochal Cysts on 2 Years-Old Girl : A Case Report

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Keyword:
Choledochal cysts children, cholesistojejunostomy, jejunojejunostomy, cholecystectomy

Abstract

A choledochal cyst is a swelling / widening or dilatation of the bile ducts. Choledochal cysts are diagnosed in children and adults, although most (80%) develop in children younger than 10 years. Choledochal cysts are also 4 times more common in females than males, The exact cause of CCs is unknown, but described as a reflux of pancreatic enzymes into the biliary tree through an anomalous pancreaticobiliary duct union (APBDU), thereby leading to biliary dilation. Although rare in the general population, APBDU has been reported in more than 80% of children with CCs.The classic symptoms in children are intermittent abdominal pain, jaundice, and the presence of a right upper quadrant abdominal mass. We present a case of 2 year-old girl with choledochal cysts and pancreatitis as a complication. She underwent biliodigestive bypass cholesis-tojejunostomyjejunojejunostomy and cholecystectomy, got total parenteral nutrition for 2 days and treated with intravenous meropenem. There was improvement in clinical manifestations after surgery.

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INTRODUCTION

A choledochal cyst is a swelling/widening or dilatation of the bile ducts. This condition could affects the part of the bile ducts at the outside (the common bile duct and the hepatic ducts) and the inside of the liver (the intrahepatic ducts). First reported by Vater and Ezle in 1723, choledochal cysts (CCs) are rare cystic dilations of the biliary tract. Choledochal cysts are diagnosed in children and adults, although most (80%) develop in children younger than 10 years. Choledochal cysts are also 4 times more common in females than males,higher incidence in Asian populations ranging from 1 in 100 000 Western individuals to 1 in 13 000 people within

the Japanese population. The exact cause of CCs is unknown; however, in 1969, Babbitt first described the reflux of pancreatic enzymes into the biliary tree through an anomalous pancreaticobiliary duct union (APBDU), thereby leading to biliary dilation. Although rare in the general population, APBDU has been reported in more than 80% of children with CCs.

The classic symptoms in children are intermittent abdominal pain, jaundice, and the presence of a right upper quadrant abdominal mass. According to Todani's classification system, choledochal cysts include five main types. The treatment of the CCs will vary accordingly to the Todani's classification. For the commonest type of Choledochal cyst (Type I and IV) the treatment is surgery, although, the reconstruction technique of the bile duct has no gold standard. The surgical approach has evolved from drainage procedures to the excision of the cyst and reconstruction of the bile duct as the treatment of choice. There are two reconstruction techniques, hepaticojejunal anastomosis in Roux-en-Y and hepaticoduodenal anastomosis, with no apparent benefit of any of them. The aim of this case presentation is to point out the clinical manifestation, diagnosis and management of choledochal cysts in children.

CASE REPORT

A girl, 2 year and 5 months old suffered with Intermittent abdominal pain since 1 months ago. The pain has gotten worse since the past 15 days. Both eyes looked yellow since 2 weeks ago, increases throughout the body. There was history of putty white defecation 2 weeks ago. Vomit since one week ago, 1-2 times a day, amount about 5 tablespoons until a quarter of glass each time. Decreased of appetite during illness. Patient was referred from district hospital with diagnosis choledochal cyts type I based on ultrasound, and treated with ursodeoxycolat acid 3x250 mg po, curcuma 3x1 tablet.

Patient was the only child, orphan, had complete basic immunization. Growth and development was normal. She lived with her mother, grandmother and grandfather in a permanent house with good hygiene and sanitation. She had good quality and quantity of nutrition.

General appearance was moderately ill, alert, blood pressure was 90/60 mmHg, heart rate was 98 beats/minute, respiratory rate was 24 times/minute and body temperature 37^{0}C.Body weight was 11 kgs (body weight for age -2<SD<0, z score), body height 87 cms (Length for age -2<SD<0), nutritional status was wellnourished (weight for height -1<SD<0). There was icteric and anemic but no edema and cyanotic. The skin was pale and warm, jaundice all over the body. There was no enlargement of lymph node. The head was round, symmetrical and normochephal (48 cm, -2<SD<2 Nellhauss standard). The conjunctiva was anemic, sclera was icteric and the pupils were normal. The chest and heart within normal limit. Abdominal examination not distended, supple palpated, liver palpated and spleen was not palpable, normal bowel sounds.

Hemoglobin was 8,7 gr/dl, white blood cell17.200/mm^3, and platelet 771.000/mm^3. Differential count 0/3/2/69/24/2, erythrocyte 3,4x10^6/mm^3, reticulocyte 2,3% and haematocryte 27 vol %. Increased of liver function level. Amylase pancreatic was 1 523 U/L (13-53) and lipase was 994 U/L (<37). Bilirubin was +2 in the urinalysis examination, and feces examination the color was pale yellow, other results within normal limit.

From the ultrasound there was dilatation of the intrahepatic bile duct, saccular shape, suitable with impression of choledochal cyts type I. From MRI-MRCP found the gallbladder appeared slightly enlarged. There was no mass in the liver. Intra and extrahepatic bile ducts appeared to be widened. The size of intrahepatic biliary duct wss enlarged with a homogeneous signal intensity.The size of the extrahepatic bile duct was enlarged with a homogeneous signal intensity. Conclusion was choledochal duct cyst type IV A (Todani classification).

Patient was treated with liver diet 1100 kcal, ursodeoxycholat acid 3x80 mg, curcuma syr 3x1 cth, vitamin A 1x 10 000 IU, vitamin D 1x 800 IU, vitamin E 1x400 IU, vitamin K 1x5 mg (alternating day), meropenem 3x 450 mg intravena and consult to Surgery Department. Patient has undergone

biliodigestive bypass surgery cholesistojejunostomyj ejunojejunostomy and cholecystectomy, treated in the PICU room and got total parenteral nutrition for 2 days.

From the pathology anatomy of the gallbladder tissue found piece of gallbladder, greenish, chewy, solid, with greenish mucosa smooth , thick wall (macroscopic) and piece of gallbladder tissue like cyst with thick wall consisted of mucosa layer coated with epitelcolumna cell, grow forming crypts. Below it, muscularis layer and loose connective tissue, consist of capillaries and lymphocytes at mucosa (microscopic). Conclusion was choledochal cyst.

Since patient had no complaints and in stable condition, she was planned to discharge after 38 days of hospitalization.

DISCUSSION

Choledochal cysts are abnormal dilatation intra and extrahepatic of the biliary tree. First reported by Vater and Ezler in 1723.[3, 19] They may occur as a single or multiple cysts within the biliary tree and are more common in Asian populations with the incidence of 1 in 13,000 versus 1 in 100,000 in Western populations. Females are at higher risk for the disease with a nearly 4:1 female preponderance compared with males. Most of the cysts (85%) are reported to be diagnosed in the first decade or under 15 years of age. Approximately 20% of cysts are diagnosed in older patients. Research by Sumathi et al in 2016 found that mean age of presentation was 5.2 years old. Sujit Kumar et al found that commonly affected cohort is the age group of 6-10 years old with female preponderance.[8] Our patient was 2 year and 5 months old and was a female patient.

There are some classifications for choledochal cysts but classification by Todani is the most widely used. Todani and colleages modified classification previously found by Alonso-Lei et al by added the fifth type biliary cysts or Caroli disease. This patient was referred with diagnosis choledochal cyst type I based on ultrasound examination. In our institution after MRI-MRCP was performed, team concluded that it was choledochal cyst type IVA. Type I CCs are the most common encountered CCs (80-90%). Type I cysts can be further subdivided into Type IA, IB and IC cysts. Type IA CC has the gall- bladder arising directly from the CC with a dilated extrahepatic biliary tree and a non-dilated intrahepatic tree. Type IB CC contains no evidence of APBDU and a focal segment of the common bile duct is dilated. Finally, type IC CC is represented by a fusiform dilatation of the common hepatic duct and common bile duct in the presence of APBDU.

Type IV CC is multiple cysts which can involve both the intrahepatic and extrahepatic biliary tree. Type IV CCs can be further subdivided into Type IVa and IVb cysts depend on intrahepatic involvement. Type Iva CC refers to extrahepatic biliary dilatation with at least one intrahepatic cystic dilatation. Type IVb refers to multiple extrahepatic biliary cysts without intrahepatic involvement. Type IV CC is the second most common CC representing 15–20% of all reported CC. Type I CC along with type IV cysts, have the highest risk of malignancy. This is not surprising given that both types of these CCs have extrahepatic involvement and are typically associated with APBDU.

Clinical presentation varies and most often consists of nonspecific abdominal pain. The classic triad of jaundice, abdominal pain and right upper quadrant mass is rare and seen mainly in the pediatric population (only 10-20% of patient). Jaundice, cholangitis, pancreatitis, portal hypertension, liver function abnormalities and coagulopathy are also seen. Jaundice mainly occurs in type I and IV CCs where APBDU allows the reflux of biliary and pancreatic juices leading to protein plugs and stone formation. Biliary amylase levels may be elevated and correlate with clinical severity. Type V CCs typically presents with cholangitis and stone formation. In 1–2% of cases, CCs may present with rupture and biliary peritonitis prompting emergency biliary drainage. This presentation is typically seen in neonates and infants. Incidental identification is rare in the pediatric population, but seen in nearly one-third of adult CCs patients. Presentation in infancy (<1 year old) compared with the classical pediatric group (1–18 years old) is different. Specifically, infants are

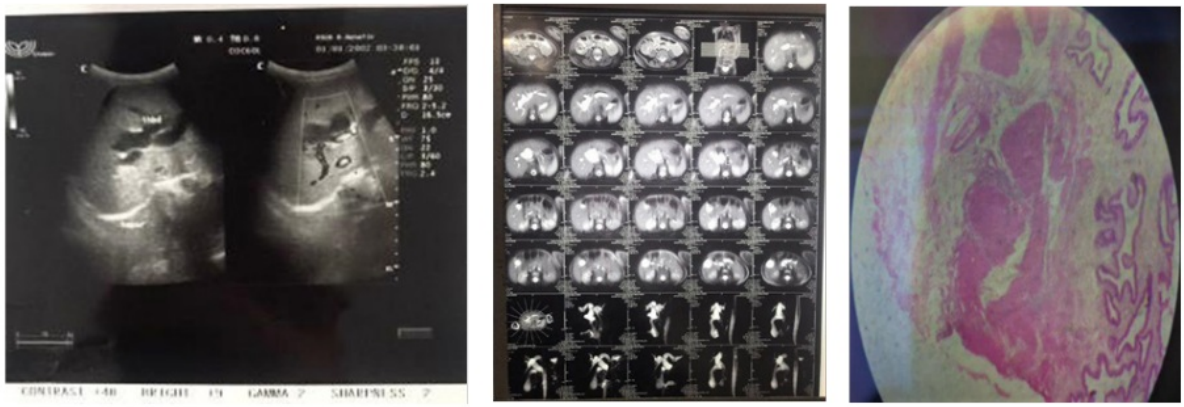

Figure 1. Abdominal ultrasound, MRI- MRCP and Pathology anatomy

more likely to present with jaundice, clay colored stools whereas the classic pediatric group is more likely to present with abdominal pain. Less commonly seen presentation includes duodenal obstruction and perforation. Compared with adults, the pediatric CCs patient is more likely to present with an abdominal mass and an APBDU. Forny et al done a review of 30 cases of choledochal cysts observed that the most common clinical manifestation was jaundice that is in 70% of cases (21 patients), followed by abdominal pain presented in 46.6% of patients (14 cases). Acute cholangitis was present in one child (3.3%). Four children (13.3%) were asymptomatic at the first surgical evaluation: two had prenatal diagnosis by obstetric ultrasonography and two were evaluated due to hepatomegaly. One patient had a palpable abdominal mass (3.3%).[11] Our patient was the classical pediatric group (1-18 years old) had the symptoms of jaundice, abdominal pain but also had the symptoms of infancy which is clay-colored stool. There was no abdominal mass found in our patient.

No laboratory studies are specific for the diagnosis of choledochal cyst. An elevated white blood cell count with increased number of neutrophils and immature neutrophil forms may be observed in the presence of pancreatitis. Liver function tests may be useful in narrowing the differential diagnosis. Hepatocellular enzyme and alkaline phosphatase levels may be elevated. None of these tests are specific for the diagnosis of a choledochal cyst.

We diagnosed pancreatitis based on clinical findings of abdominal pain and an elevated lipase and/or amylase. Pancreatitis is a common complication of choledochal duct cyst. This condition is as a result of reflux of pancreatic enzymes into the pancreatic duct which may lead to pancreatitis. Muthucumaru *et al*, found that patient with type IV A cysts that represented the largest proportion of cyst types, mostly presented with pancreatitis (53%).[28] We gave antibiotic meropenem 3x450 mg iv to treat the pancreatitis.

Results of serum chemistry may be abnormal if the patient is vomiting. One might expect to see a hypochloremic, hypokalemic metabolic alkalosis in this clinical picture.[12] Our patient was performed bypass biliodigestive. The treatment of choice for choledochal cysts is complete excision with construction of a biliary-enteric anastomosis to restore continuity with the gastrointestinal tract. The surgical management for each choledochal cyst type is described as follows:[1,4,8] Type I: Treatment of choice is complete excision of the involved portion of the extrahepatic bile duct; a Roux-en-Y hepaticojejunostomy is performed to restore biliary-enteric continuity.[7] Type III (choledochocele): Therapeutic choice depends on the size of the cyst; choledochoceles measured 3 cm or less can be treated effectively with endoscopic sphincterotomy, whereas lesions larger than 3 cm (which typically produce some degree of duodenal obstruction) are excised surgically via a transduodenal approach

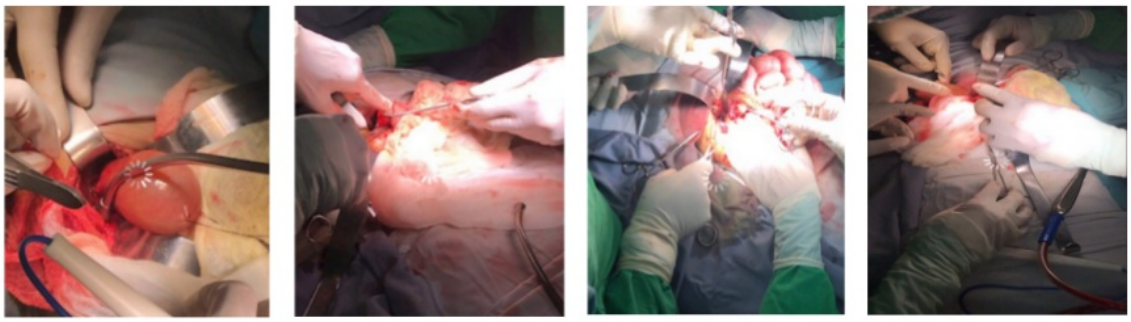

Figure 2. biliodigestive bypass surgery cholesistojejunostomy jejunojejunostomy and cholecystectomy

if the pancreatic duct enters the choledochocele, reimplantation into the duodenum may be required following excision of the cyst. Type IV: Complete excision of the dilated extrahepatic duct, followed by a Roux-en-Y hepaticojejunostomy to restore continuity; intrahepatic ductal disease does not require dedicated therapy unless hepatolithiasis, intrahepatic ductal strictures, and hepatic abscesses are present (in such instances, resection of the affected hepatic segment or lobe is performed). Type V (Caroli disease): Hepatic lobectomy for disease limited to one hepatic lobe (left lobe usually affected); however, one should carefully examine the hepatic functional reserve before committing to such therapy; patients with bilobar disease manifesting signs of liver failure, biliary cirrhosis, or portal hypertension may require liver transplantation. Lilly technique: When the cyst adheres densely to the portal vein secondary to long-standing inflammatory reaction, it may not be possible to perform a complete, full-thickness excision of the cyst; the Lilly technique allows the serosal surface of the duct to be left adhering to the portal vein, while the mucosa of the cyst wall is obliterated by curettage or cautery—theoretically, this removes the risk of malignant transformation in that segment of the duct.

After surgery the patient gradually recovered. The jaundice decreased and the abdominal pain diminished after several days. Resection of pediatric CC is generally well tolerated. Intestine movements are usually slow after surgery. Most children are not allowed to have a meal after surgery until there is a return of intestinal function as evidenced by passing gas or having stool. Often, a tube is placed through the nose, with the tip in the stomach to suction stomach contents while intestines are not working yet. This avoid the child to vomit and retch, which is uncomfortable especially with new incision.[26]

In the postoperative period, early complications can include anastomotic leak, postoperative bleeding, wound infection, acute pancreatitis, and pancreatic or biliary fistula. However, most series are without early mortality and report rates of acute complications including wound infections from 0 to 17%, without significant difference between infants and children. Late complications include anastomotic stricture, cholangitis, hepatolithiasis, cirrhosis, and malignancy. Benign anastomotic stricture with recurrent cholangitis is less common than in adults but is still seen in as many as 10–25% of patients and can be associated with both intrahepatic and bile duct stone formation. Rigorous long-term follow-up after pediatric CC resection is limited, but the risk of biliary carcinoma, most often cholangiocarcinoma, clearly remains elevated even after CC excision compared to the general population. The incidence of gallbladder or bile duct cancer increases with age. In patients with choledochal cysts under the age of 10 years, the risk of developing biliary duct cancer is 0.7%, and the risk increases to 14.3% for patients over 20 years of age in Western countries. The incidence of gallbladder or bile duct cancer is 0.3% in children, whereas it is 15.6% in adults. Gallbladder or bile duct cancer occurs in 26% of patients under 40 years old and in 45.5% among those over 70 years old in the Japanese literature. The incidence of gallbladder cancer (67.8%) is more frequent than that of bile duct cancer (32.1%) in patients with choledochal cysts, according to the register of the Japanese Study

Group of Pancreaticobiliary Maljunction (JSPBM). [26]

Restricting enteral fat intake is not indicated. In choosing enteral formulae, standard products may be adequate. Since she has had cholesistectomy, our patient may be benefit from formulae rich in medium-chain triglycerides or formulae supplemented with medium-chain triglycerides oil. This could prevent essential fatty acid deficiency.[29]

CONCLUSION

A case of a 2 year-old girl with choledochal cysts with pancreatitis as a complication has been reported. The diagnosis of choledochal cyst and reported. The diagnosis of choledochal cysts and pancreatitis based on patient's hystory, physical examination, laboratory findings and imaging modalities.

Intermitten abdominal pain dan icteric. The laboratory findings revealed an increase of liver function level, amylase and lipase serum. Imaging modalities found choledochal cysts type I from ultrasound but type IVa from MRI-MRCP. Treatment of choledochal cysts was surgery which biliodigestive bypass surgery cholesis to jejunos to my jejuno jejunos to my and cholecystectomy for this patient. Treatment of pancreatitis was meropenem as drug of choice of pancreatitis. The result of this treatment was satisfiying with no complication of surgery. Nutritional diet for this patient was medium chain tryglycerides to prevent deficiency of fatty acid.

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