CASE REPORT : RUPTURED CHOLEDOCHAL CYST IN INFANT AND CHILDREN ; 3 CASES

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ABSTRACT

Choledochal cysts are uncommon and appear as cystic or fusiform dilatation of the biliary tract including both intra- and extra-hepatic sites. They are usually diagnosed in infancy and childhood. Ruptured choledochal cyst is a rare complication of choledochal cyst. The etiology of ruptured choledochal cyst remain obscure but it may be associated with blunt trauma, cholelithiasis, an additional anatomical abnormality such as ductal stenosis distal to the cyst, or pancreaticobiliary malunion, and inspissated bile. However the cause of rupture is unknown in many cases and is considered to be spontaneous. The perforation site may be single or multiple and mostly in the posterior wall of the cyst. In this report, there were 3 female patients admitted in our hospital with ruptured choledochal cyst type 1. The first and second patients were 3 years old and complained of fever, abdominal pain, and nonbilious vomiting. The third patient, 19 days old, was presented by obstructive jaundice and clinical sepsis. The operative findings of exploratory laparotomy in all 3 patients were saccular dilatation of CBD with intraabdominal bilelike fluid, leading us to make the postoperative diagnosis of ruptured choledochal cyst. The possible causes of ruptured choledochal cyst in the three patients of this report could be cholelithiasis, blunt trauma with a long common channel of the pancreaticobiliary system and inspissated bile respectively. All 3 patients had choledochal cyst type1 and were treated by cyst excision in the first operation and Roux-en-Y hepaticojejunostomy in the second operation with good result and no complication.

CASE 1

A 3-year-old girl was admitted with complaints of RUQ abdominal pain, nonbilious vomiting, and fever for 20 days. After admission there were worsened abdominal pain with a palpable RUQ abdominal mass. A right upper quadrant soft tissue density mass was seen in the plain abdomen, an exploratory laparotomy was underwent. The operative findings were saccular dilatation of choledochal cyst about 5 cm in size and dilated gallbladder with no stone, bile fluid collection about 10 ml at supracolic gutter but no perforation site of bile leakage was detected. So a postoperative diagnosis of concealed rupture of choledochal cyst type 1 with bile leakage to supracolic gutter was made and then T-tube choledochostomy was performed. Postoperative results were good and recovered 2 weeks after the operation. T-tube cholangiography showed saccular dilatation of the common hepatic duct and proximal common bile duct; measuring about 2.7x4.5 cm consistent with choledochal cyst.

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She was then referred to Siriraj Hospital. Laboratory studies on admission showed hemoglobin level of 12.1 g/dL, WBC of 7,000/mL, serum alkaline phosphatase level of 329 U/L, serum glutamic-oxaloacetic transaminase (SGOT) level of 62 U/L, serum glutamic-pyruvic transaminase (SGPT) level of 119 U/L with normal bilirubin level.

The second operation with total excision of choledochal cyst and Roux-en-Y hepaticojejunostomy were performed. Histopathologic findings were choledochal cyst type1, 3.5 cm in length, 1.5 cm in circumference with recent and old hemorrhage and nonspecific chronic inflammation at the cyst wall whereas the gallbladder contained pigmented stones with congestion and focal hemorrhage. Postoperative results were good and no complication after follow-up for 1 year and 2 months.

CASE 2

A 3-year-old girl was presented with RUQ abdominal pain, nonbilious vomiting and diarrhea for 4 days which firstly she was diagnosed and treated as acute gastroenteritis. Two days later, she had fever with progressive vomiting and abdominal pain so she was admitted in Siriraj Hospital. There was a history of preceeding blunt abdominal trauma of falling from bicycle one week ago.

Physical examination revealed a body temperature of 38.8 °C, generalized abdominal tenderness, rebound tenderness, guarding with a maximal tender point in RUQ and decreased bowel sound. Laboratory studies showed a hemoglobin level of 12.3 g/dL, WBC of 2,630/mL, neutrophil 73%.

She had undergone exploratory laparotomy for peritonitis with suspected of ruptured appendicitis or ruptured liver abscess. The operative findings were bile-stained purulent peritoneal fluid about 300 ml, dilated gallbladder, contusion and dilatation of CBD about 2 cm in diameter with no perforation site seen, unusually firm pancreas, small perforation at the mid-portion of appendix with minimal fluid in lesser sac. Postoperative diagnosis were traumatic contusion and minute perforation of CBD, pancreatitis and ruptured appendicitis. Then cholecystostomy with T-tube drainage, transperitoneal drainage and appendectomy were performed. Histopathologic findings of appendix were compatible with acute appendicitis. T-tube cholangiography showed choledochal cyst type1 about 3x5 cm in size with distal CBD narrowing and a long common channel of the pancreaticobiliary junction. Followed up laboratory studies showed serum amylase level of 401 U/L, GGT 75 U/L, SGOT 88 U/L, SGPT 62 U/L, serum alkaline phosphatase 142 U/L with normal bilirubin level. After operation, she was given antibiotic and postoperative care. Then she recovered well and could be discharged 2 weeks after operation. Later the second operation was done with excision of the choledochal cyst (just beneath the confluence of Rt. IHD and Lt. IHD to 1 cm from the pancreaticobiliary junction) and Roux-en-Y hepaticojejunostomy was done successfully without complication.

Histopathologic findings were choledochal cyst about 2x5 cm in size and a short distal duct o.5 cm in length and 1 cm in diameter with chronic cholecystitis.

The result after 4-month follow-up was good.

CASE 3

A 19-day-old female infant was admitted because of jaundice, acholic stool, abdominal distension and clinical sepsis.

Laboratory studies showed a hemoglobin level of 13.1 g/dL, WBC of 16,550/mL, neutro-

phil 56%, lymphocyte20%, monocyte10%. The liver function test showed a total bilirubin level of 8.8 mg/dL, direct bilirubin level of 4.8 mg/dL, GGT 403 U/L with normal SGOT and SGPT levels. Radiologic examinations including plain * IHD = Intrahepatic Duct abdomen and abdominal ultrasound were performed. Plain abdomen showed ascites with bowel ileus. Abdominal ultrasound revealed ascites and a RUQ cystic mass communicating with the biliary system but no normal gallbladder could be identified. So the differential diagnosis was choledochal cyst or dilated gallbladder from sepsis. Peritoneal tapping was performed showing bilious ascites. Therefore exploratory laparotomy with peritoneal toilet and cholecystostomy were performed. The operative findings were intraperitoneal bile about 500 ml with fusiform dilatation of CBD about 5 cm in size consistent with ruptured choledochal cyst type 1. Later the second operation with excision of choledochal cyst and Roux-en-Y hepatico-jejunostomy were done with operative findings of bile plug in the choledochal cyst and perforation at the posterior wall of the choledochal cyst. She recovered well without complication after operation.

Histopathologic findings were compatible with choledochal cyst about 1.3x1.3x1 cm in size without stone formation.

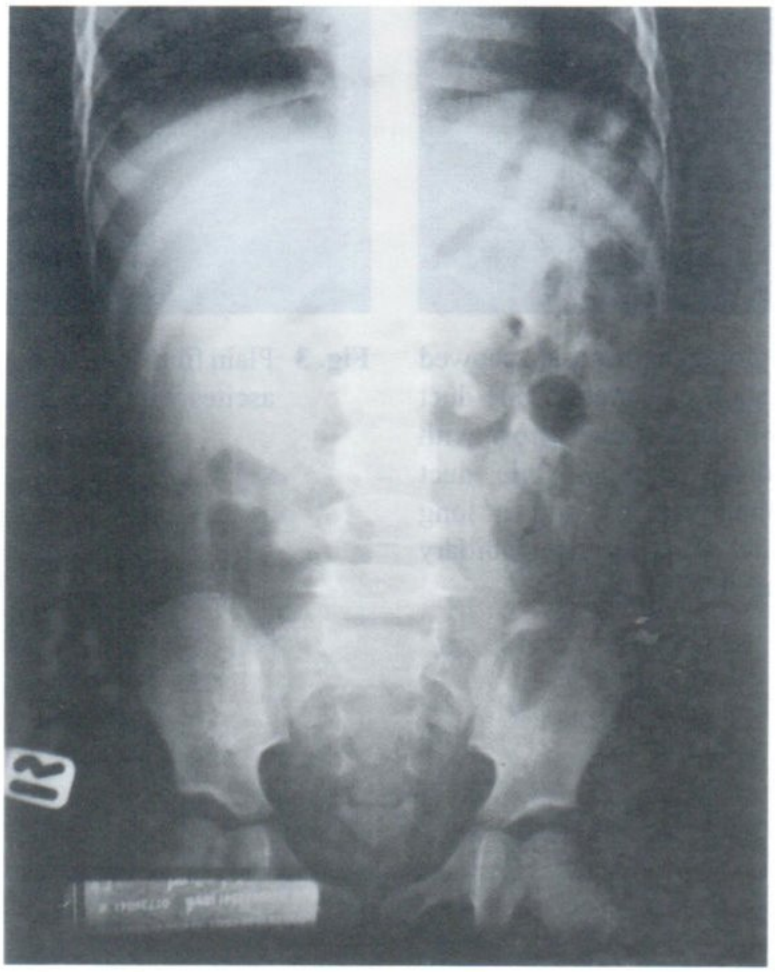

Fig. 1 Plain film of the abdomen in case 2 showed bowel ileus.

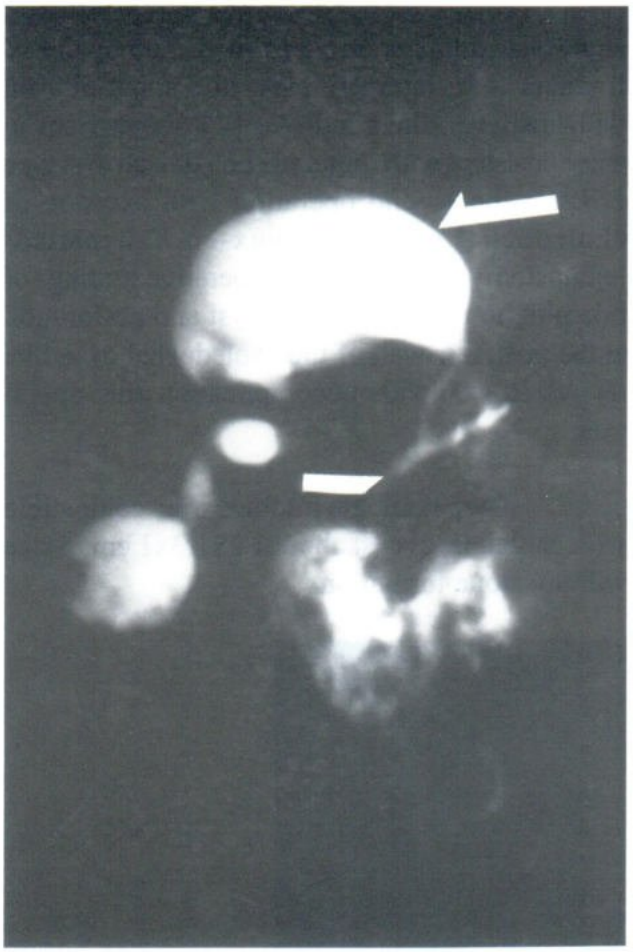

Fig. 2 T-tube cholangiography in case 2 showed fusiform dilatation of common bile duct about 3x5 cm in size (Long arrow with head) with distal common bile duct narrowing (Short Arrow) and a long common channel of the pancreaticobiliary junction. (Long Arrow)

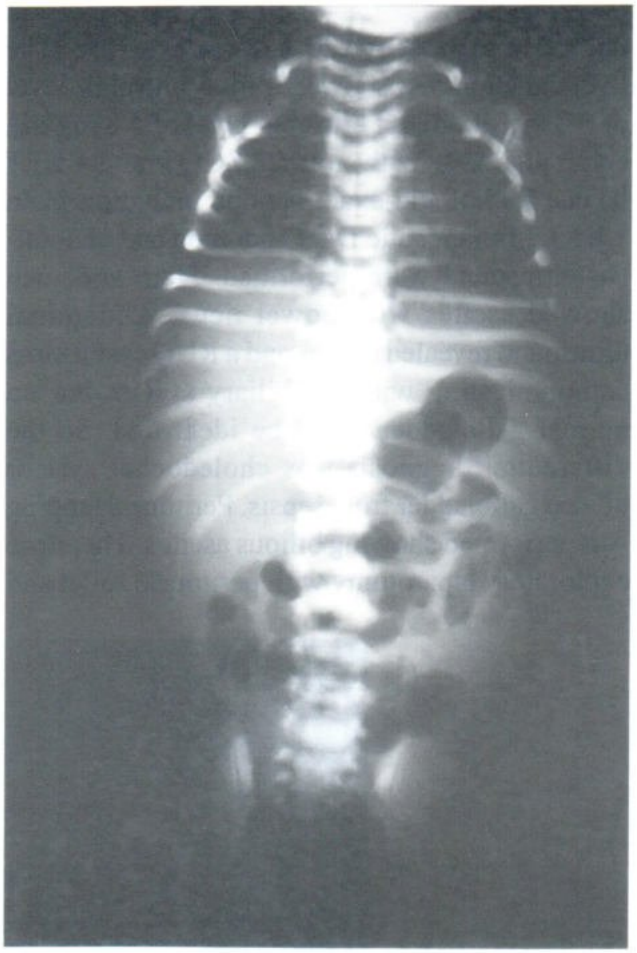

Fig. 3 Plain film of the abdomen in case 3 showed ascites with bowel ileus

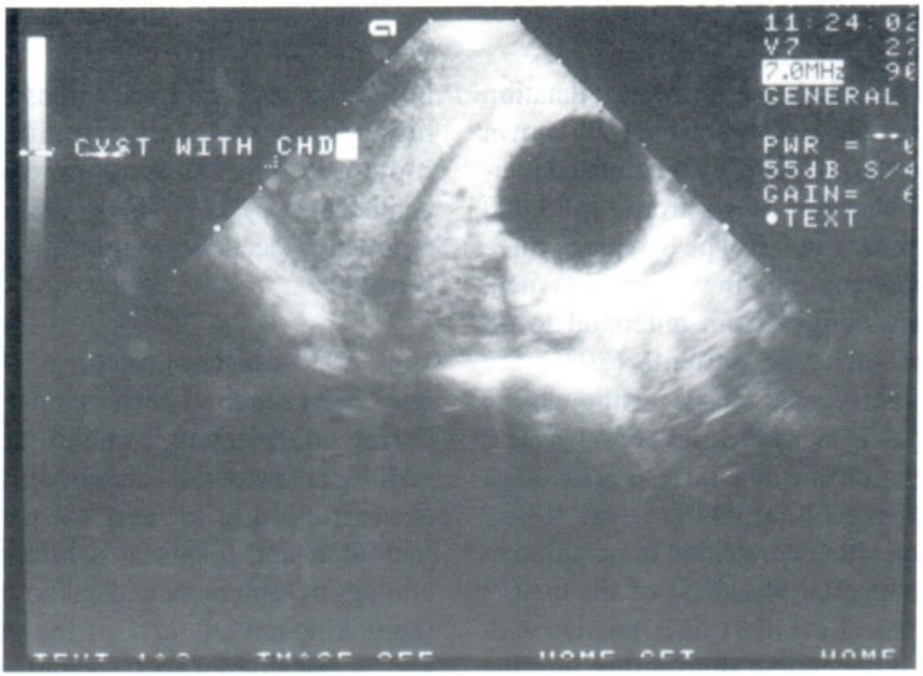

Fig. 4

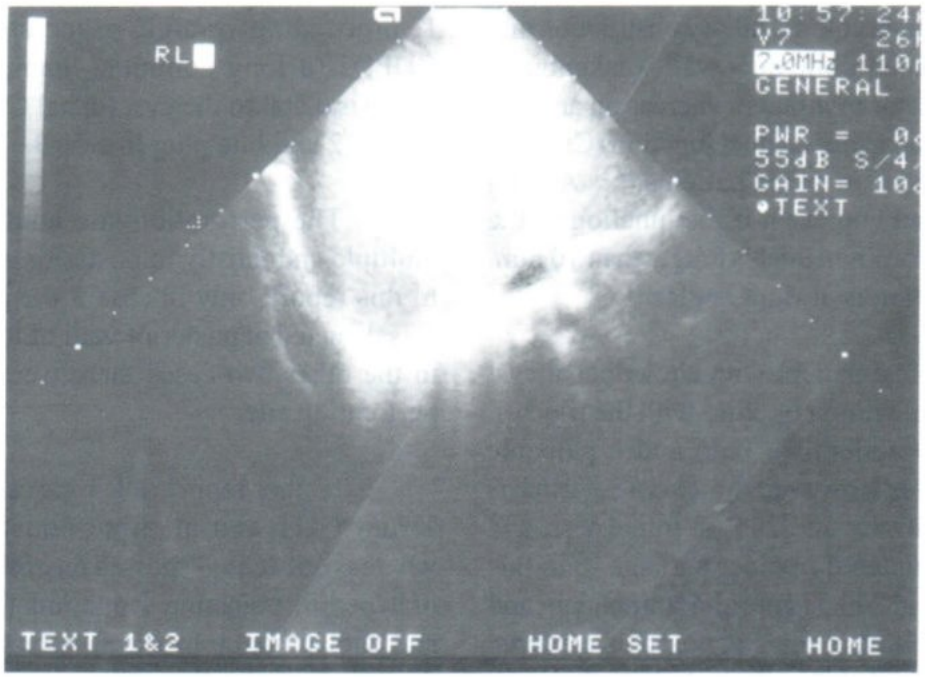

Fig. 5

Fig. 4 & 5 Abdominal ultrasound in case 3 showed ascites and a RUQ cystic mass communicating with biliary system but no visualization of a normal gallbladder

DISCUSSION

The choledochal cyst, a cystic dilatation of the biliary tree, is an uncommon but well-known cause of obstructive jaundice. The estimated incidence of choledochal cyst ranges from 1 in 13,000 to 1 in 2 million patients.[15,16] The disease is three to four times more common in women than men. Age at diagnosis ranges from antenatal to 68 years.[14] The majority of patients (80%) are diagnosed before 10 years of age. Although the etiology is unknown, many theories are suggested. Unequal proliferation of epithelial cells at the stage when the primitive bile ducts are still solid,[6] distal obstruction either congenital or acquired in additional to congenital weakness of the duct,[6,7] an abnormal pancreaticobiliary duct junction with resultant chronic pancreatic fluid reflux into the biliary tree,[8] and the ischemia of the bile duct[9] are among the proposed theories. Some authors have suggested that embryologic biliary obstruction and subsequent common bile duct wall weakening is an important factor in infants, whereas an anomalous choledochopancreatic duct junction (CDPDJ) is more important in older patients.[18-21] A long common channel is present if the junction of the pancreatic and common duct is longer than 10 mm. Often this junction is at right angles.[22]

The typical patient with choledochal cyst has been a female infant or child with the triad of, jaundice, RUQ abdominal pain and a palpable abdominal mass. However the classical triad is encountered only 15% to 25% at initial visit and is usually encountered in older patients.[1-6] In this report, case 1 and 2 had acute abdominal pain and peritonitis which were misdiagnosed as ruptured appendicitis. So the ruptured choledochal cyst should be considered as an unusual cause of acute abdomen in children similar to the previous study.[13] The patients may present with the complications of choledochal cysts such as ascending cholangitis, recurrent pancreatitis, progressive biliary cirrhosis and portal hypertension, stones in the cyst and malignant transformation in the biliary tract.[3-6] Bile peritonitis secondary to rupture is one of the rarest complications of choledochal cysts.[1,2] This complication has been reported to occur at rates of 1.8%- 2.8% in large series.[1,8-12]

The causes of rupture of choledochal cyst have been reported such as after blunt trauma[1], during confinement,[10] cholelithiasis,[22] with an additional anatomical abnormality such as ductal stenosis distal to the cyst,[7] or pancreaticobiliary malunion and inspissated bile.[22] However the etiology of rupture remain unknown in many cases (approximately 20-46% of the published cases) and is considered to be spontaneous.[13,22] This report showed the possible causes of ruptured choledochal cyst including cholelithiasis in case 1, preceeding trauma, associated anomalous CDPDJ (a long common channel) and ductal stenosis distal to the cyst (distal CBD narrowing) in case 2 and bile plug in case 3.

The perforation site could be single or multiple and mostly in posterior wall of the cyst. In this report, only in case 3 the perforation site was detected at posterior wall of the cyst whereas in the other two cases there were no detectable perforation site.

In this report, all 3 cases had increased serum GGT and alkaline phosphatase levels whereas only case3 showed an obstructive pattern of hyperbilirubinemia. Abdominal ultrasound, CT and cholangiography are all effective in defining the presence of biliary dilatation and in the diagnosis of choledochal cyst.[4-6,8] It has been generally accepted that abdominal ultrasound is the most useful diagnostic tool for detection of choledochal cyst. Cholangiography is essential in differentiating the type of choledochal cyst and in planning the extent of operative resection. Preoperative

percutaneous cholangiography has been the preferred mode of cholangiography to endoscopic retrograde cholangiography because it has the ability to define the proximal extent of biliary dilatation and this information can be used in the preoperative plan for resection. Endoscopic retrograde cholangiography is best to visualize the pancreaticobiliary junction but the superior intrahepatic extent of the cysts may be not defined if the cysts are redundant and sequester large amount of contrast material.[14]

An abdominal ultrasound was performed in only case 3 showing RUQ cystic mass communicating with biliary tract but no visualization of a normal gallbladder. The differential diagnosis could be either a choledochal cyst or a distended gallbladder due to sepsis. Plain abdomen showed ascites and bowel ileus, possibly from perforation of GI tract or a ruptured choledochal cyst. However bile-stained peritoneal fluid after peritoneal tapping was helpful and suggestive of biliary tract pathology. Although spontaneous perforation of choledochal cyst is rare, it can sometimes be the initial manifestation of choledochal cyst and should be considered in the presence of bilelike peritoneal fluid. In the presence of bile peritonitis, it is important to differentiate spontaneous perforation of the bile duct with a wall-off bile collection, from a ruptured choledochal cyst. Spontaneous perforation of the extrahepatic bile duct occurs almost exclusively in infants less than 20 weeks of age, and most often occurs at junction of the cystic and common hepatic ducts.[2,12]

Differentiation between these two entities is definitely required because a perforated bile duct could be cured and spontaneously close after simple surgical drainage whereas a ruptured choledochal cyst would require cyst excision and Roux-en-Y choledochoenterostomy.[2] On the other hand, intestinal anastomosis to the site of perforation or sac under the misdiagnosis of a ruptured choledochal cyst is generally lethal.[12] Preoperative diagnosis of bile leakage from spontaneous rupture of the extrahepatic bile duct could be differentiated from ruptured choledochal cyst in which a definite preoperative diagnosis could be made by hepatobiliary scan.[11] If bile duct pathology has not been considered preoperatively, the presence of any fluid suggestive of bile should urge the surgeon to evaluate the biliary tree for the possibilities of spontaneous perforation of bile duct or ruptured choledochal cyst.

The definite diagnosis of ruptured choledochal cyst may be difficult even at laparotomy. The main reasons for this difficulty are the lack of history suggesting a choledochal cyst, overlooking the cyst because of the collapse following the perforation, the rupture of a relatively small cyst, and the usual location of the perforation on the posterior wall of the cyst.[13] Perioperative cholangiography has been suggested to be helpful for the diagnosis of biliary pathology[4,8,9,12] and may be useful especially in the absence of apparent ruptured choledochal cyst or perforation of bile ducts.

Currently, excision of the cyst and Roux-en-Y hepaticojejunostomy are considered to be appropriated for most patients with a choledochal cyst type 1.

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