

Spontaneous rupture of choledochal cyst presenting in childhood

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Spontaneous rupture of choledochal cysts is one of the rare complications, and can sometimes be the initial manifestation. It should be considered in the presence of bile-like fluid. A 10-year-old girl had acute onset of abdominal pain, vomiting, and elevated bilirubin, alkaline phosphatase, glutamic oxaloacetic transaminase and glutamic-pyruvic transaminase levels. There was no trauma in her history. In ultrasonography and computed tomography, dilated common bile duct, cystic mass of 10x6 cm, and free intraperitoneal fluid in abdominal cavity were demonstrated. Radiological methods, especially intraoperative cholangiography, should be performed for evaluation. We report a case of spontaneous rupture of the choledochal cyst with clinical and radiological findings.

Key words: spontaneous rupture, childhood, choledochal cyst.

Choledochal cyst has long been known and considered an uncommon congenital abnormality of the biliary tract¹. The ages of reported cases range from neonates to 80 years old; however, 60% are diagnosed before the age of 10². The classic triad of jaundice, abdominal pain, and palpable mass is encountered among 15% to 25% of cases at initial visit^{3,4}. Additionally, the patients may present with the complication of choledochal cyst. Rupture is one of the rarest complications of choledochal cysts¹. This complication has been reported to occur at rates of 1.8%, 2.1%, and 2.8% in series^{5,6}. Preoperative diagnosis is important and radiological imaging techniques can help in diagnosis of the complications of choledochal cysts.

We report a case of spontaneous rupture of choledochal cyst on ultrasound, computed tomography, and cholangiography.

Case Report

A 10-year-old girl was admitted with complaints of abdominal pain, vomiting, and jaundice of two days' duration. There was no previous abdominal pain, fever, jaundice, or trauma in her medical history. Physical examination

revealed abdominal distension, tenderness, defense and abdominal mass in the right upper quadrant. Laboratory studies on admission showed a hemoglobin level of 11.8 g/dl, WBC of 21,600/mm³, direct bilirubin level of 5.04 mg/dl, indirect bilirubin level of 4.39 mg/dl, alkaline phosphatase level of 503 IU/L, serum glutamic-oxaloacetic transaminase level of 379 U/L, and serum glutamic-pyruvic transaminase of 309 U/L. She had no hepatitis antigens.

Both abdominal ultrasonography (USG) and computed tomography (CT) demonstrated a dilated 2.5 cm structure in the region of the common bile duct and a cystic mass (10x6 cm) communicating with this structure. This structure and cystic mass were separated from a thick-walled, partially collapsed gallbladder. There was a moderate amount of free intraperitoneal fluid in the abdominal cavity (Figs. 1, 2). Preoperative diagnosis was type I and type II choledochal cyst with cholecystitis. The child underwent laparotomy. The ductus choledochus was dilated fusiformly (type I choledochal cyst) and perforated on the posterior wall of the cyst. Approximately 500 ml of bile surrounded by a reactive pseudocapsule was found at this perforation region. Billious fluid was drained and intraoperative

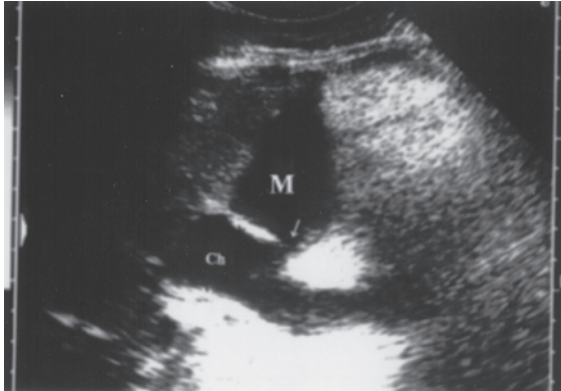
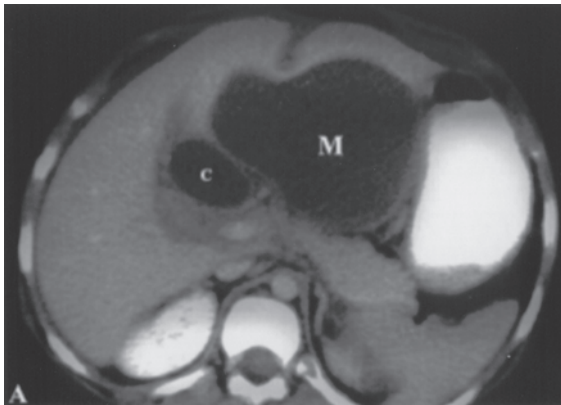
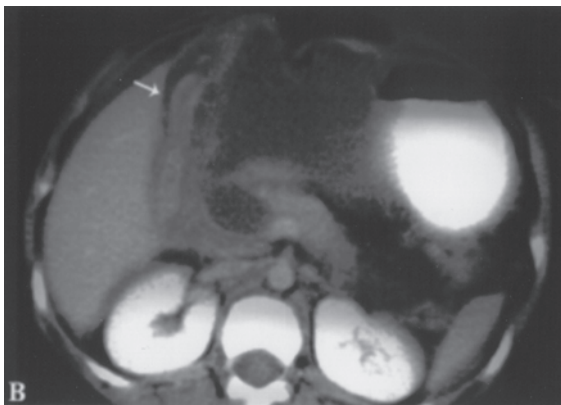


Fig. 1: Ultrasonography shows dilated common bile duct (Ch) (type I choledochal cyst) and cystic mass (M) with communicating (arrow) common bile duct due to perforation.



(a)



(b)

Fig. 2 a, b: CT scans show dilated common bile duct (C), and smooth-walled, large, without septae cystic mass (M). Note that gallbladder is collapsed and there is a moderate amount of free intraperitoneal fluid (arrow) in abdominal cavity.

cholangiogram was performed, which confirmed the presence of a type I fusiform choledochal cyst (Fig. 3). After continuous external drainage of the biliary tree for six weeks, the patient underwent a cholecystectomy, removal of the choledochal cyst, and Roux-en-Y hepaticoporoenterostomy. Histopathological examination of the cyst wall showed no malignant change in the biliary epithelium but thickened cyst wall with inflammation. She is well one year after the operation.

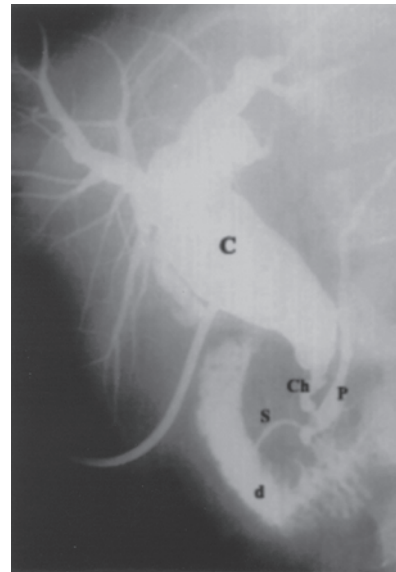


Fig. 3. Intraoperative cholangiogram demonstrates type I choledochal cyst (C) and mildly dilated hepatic ducts. Pancreatic duct (P) and distally choledochal duct (Ch) are of normal caliber and they drain to the third portion of the duodenum (d) with connection to one another. In addition, duct of Santorini (S) drainage to the second portion of the duodenum is shown.

Discussion

Bile duct cysts are generally considered congenital since they occur in fetuses and neonates, although the etiology is unknown. Among the proposed theories are: unequal proliferation of epithelial cells at a stage when the primitive bile ducts are still solid, distal obstruction either congenital or acquired in addition to congenital weakness of the duct, an abnormal pancreatic-biliary duct junction with resultant chronic pancreatic fluid reflux into the biliary tree, and ischemia of the bile duct^{7,8}.

Yamashita and colleagues⁹ proposed the five types of congenital choledochal cysts, which have gained widespread acceptance. Type I =

spherical or fusiform dilatation of extrahepatic bile duct; type II = single diverticulum of the extrahepatic duct; type III = cystic dilatation of the distal common bile duct which protrudes into the duodenum; type IV = combined intra- and extrahepatic bile duct cyst or cysts; and type V = isolated or diffuse cystic dilatation of intrahepatic bile ducts⁹.

The patients with choledochal cyst may present the complications of these cysts, including ascending cholangitis, recurrent pancreatitis, progressive biliary cirrhosis, portal hypertension, stones in the cyst, and malignant transformation in the biliary tract. Bile peritonitis secondary to rupture is one of rarest complications of choledochal cysts^{1,3,4,7}.

Several mechanisms have been postulated to explain the spontaneous rupture of choledochal cysts. Although blunt abdominal trauma, pregnancy and labor, and an additional anatomical abnormality have been reported, the etiology remains obscure in many cases⁸. The history of our case did not show a cause of rupture such as trauma.

The diagnosis of perforation should be made without paracentesis, which carries the potential risk of contaminating sterile bile, especially in the peritoneal cavity, which can cause the acute and rapid deterioration of the patient. In addition, the pre-operative diagnosis of this condition is of importance because fatal treatment disasters such as the anastomosis of the 'capsule' of a pseudo-cyst to a loop of bowel are to be avoided¹⁰.

Ultrasonography is the best initial method of investigation for the detection of choledochal cyst, the presence of ascites and of the pseudo-cyst⁴. Three-dimensional (3D) sonography promises to be a valuable adjunct to conventional two-dimensional imaging for the evaluation of choledochal cysts in pediatric patients¹¹. CT is considered to be a more accurate diagnostic modality in delineating the intrahepatic biliary tree⁷. Either endoscopic retrograde cholangiopancreatography (ERCP) or percutaneous transhepatic cholangiography (PTC) are definitive tests, demonstrating anatomical details of the biliary tree preoperatively^{2,12}. It has been reported that definitive preoperative diagnosis of bile leakage from extrahepatic bile ducts could be made using hepatobiliary scintigraphy¹. Intraoperative

cholangiography is the most important in order to confirm the diagnosis². We used abdominal USG, CT and intraoperative cholangiography in our case. We erroneously thought that there were two cysts communicating with each other on USG and CT. It is important to differentiate a spontaneous perforation of the bile duct with a walled-off collection of bile from a ruptured choledochal cyst¹³. Differentiation of these two entities is definitely required because simple surgical drainage often leads to spontaneous closure and cure of a perforated bile duct, whereas a ruptured choledochal cyst would require excision and choledochoenterostomy or portoenterostomy¹⁴. We detected only type I choledochal cyst and bile collection surrounded by a reactive pseudo-capsule on intraoperative cholangiogram.

Although rare in childhood, spontaneous rupture should be considered in patients with choledochal cyst. Preoperative diagnosis is important, as the presence of the reactive pseudo-capsule that surrounds the perforation has been incorrectly interpreted. USG and CT may be helpful in diagnosis of rupture of choledochal cysts. Intraoperative cholangiography is most important in order to confirm the diagnosis.

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