

Koledok kisti; Olgu sunumu.

CHOLEDOCHAL CYST; A CASE REPORT.

Shibajyoti Ghosh, Pabitra Goswami, Arkaprovo Roy, Prasenjit Mukherjee

R G Kar Medical College, Department of Surgery, Kolkata / Indian

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ABSTRACT

Cystic dilation of the biliary ducts, also known as a choledochal cyst, is an uncommon but serious condition that requires surgical treatment. Although choledochal cysts frequently present in infancy and childhood, the disease is more commonly diagnosed in adults. They occur three to eight times more commonly in women. Here a case report of obstructive jaundice in a 12 year old girl due to choledochal cyst is presented.

Key words : Obstructive jaundice; Choledochal cyst.

ÖZET

Safra kanallarının kistik genişlemesi koledok kisti olarak tanımlanır. Hastalık nadir görülmekle beraber cerrahi tedavi gerektiren ciddi bir durumdur. Hastalık her ne kadar infant ve çocukluk çağında olmakla beraber tanı sıklıkla adölesan çağda konulmaktadır. Kızlarda 3-8 katı daha fazla görülür. Burada, 12 yaşındaki bir kız çocuğunda tıkanma sarılığı ile beraber ortaya çıkan bir koledok kisti vakası sunulmuştur.

Anahtar kelimeler: Tıkanıcı sarılık; Koledok kisti.

INTRODUCTION

The most commonly accepted theory for the development of choledochal cyst is an anomalous pancreatic duct of biliary duct junction (which involves isolated or combined dilation of the extrahepatic or intrahepatic biliary tree) (1). A high common bile duct-pancreatic duct junction creates a long common channel. This results in reflux of pancreatic fluid into the distal common hepatic duct and results in mucosal injury, chronic inflammation, and weakening of the bile duct wall. This proposed mechanism is supported by elevated levels of amylase in choledochal cysts.

In 1977, Todani modified the Alonso-Lej classification and combined the extrahepatic and intrahepatic types into a classification that is currently used by most surgeons. Type I cysts are the most common and make up 50% of choledochal cysts (1).

Case report

A 12 year old girl presented with pain mainly in the upper abdomen for last 1 year, which became more intense and colicky in nature for last 1 month. Patient had also developed mild jaundice with a gradually increasing swelling in the mid and upper abdomen for last 2 months. Patient is complaining of occasional low grade intermittent fever with chill and rigor for last 1 month. She had lost weight in last 3 months. There is no history of haematemesis or melaena. Stool colour is normal.

On examination patient was icteric. There was a tense cystic lump measuring approximately 20x15 cm was found in the upper and mid abdomen.

Blood report showed that normochromic normocytic anaemia (Hb: 8.8 g/dL) with polymorphonuclear leukocytosis (White blood count: 11,800, N: 86%). Platelet count: 110,000 / μ L. Liver function tests

revealed conjugated hyperbilirubinemia (Total bilirubin: 4.1 mg/dL with conjugated fraction: 2.8 mg/dL) with elevated alkaline phosphatase level. Total protein was 6.7 g/dL with albumin 3.8 g/dL. Amylase and lipase levels also were increased. Patient was negative for HBsAg.

USG showed that mild dilatation of intrahepatic biliary radicals with no calculi in the common bile duct (CBD), but CBD was dilated. Features were suggestive of choledochal cyst. MRCP revealed Type III choledochal cyst (Figure 1).



Figure 1: MRCP of the patient showing huge choledochal cyst

Patient was put up for operation after transfusing 2 units of blood. On exploration a huge cyst of size 15x12 cm involving almost entire length of the CBD was found adhering with stomach, duodenum and pancreas (Figure 2 and 3). After that cholecystectomy and choledochojejunostomy was done. Patient was discharged after 2 weeks of operation.

DISCUSSION

Choledochal cysts are congenital conditions involving cystic dilatation of bile ducts. They are uncommon in western countries but not as rare in East Asian nations (2).

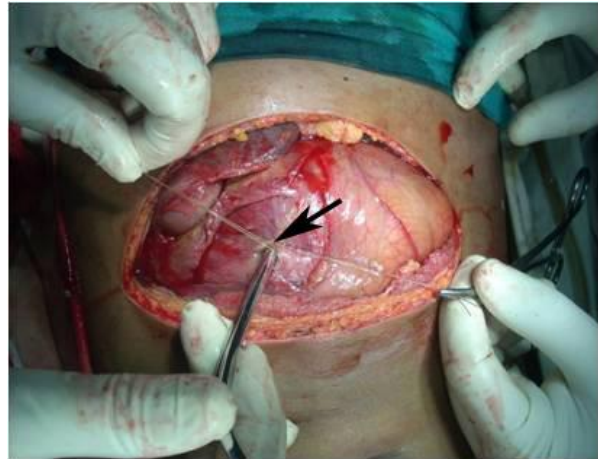


Figure 2: The cyst after opening the abdomen (arrow)

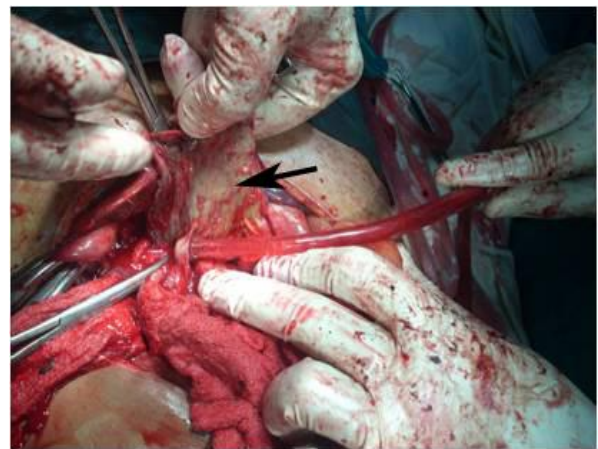


Figure 3: Cyst is excised: It was technically difficult to take out the cyst completely. Cyst was aspirated and was dissected out from the surrounding structures and excised (arrow).

In 1977, Todani modified the Alonso-Lej classification and combined the extrahepatic and intrahepatic types into a classification that is currently used by most surgeons. Type I cysts are the most common and make up 50% of choledochal cysts (Table 1).

Some patients do not present with the disease until adulthood. In many adult patients, subclinical bile duct inflammation and biliary stasis have been ongoing for years. Adults with choledochal cysts can present with hepatic abscesses, cirrhosis, recurrent pancreatitis, cholelithiasis, and portal hypertension.

The clinical history and presentation of a patient with a choledochal cyst varies with the patient's age. Overt, dramatic signs and symptoms are more common in infancy, whereas manifestations are more subtle and protean in adulthood.

Table 1: Todani Modification of Alonso-Lej Classification of Choledochal Cysts	
Type I	Dilation of the extrahepatic biliary tree. Type I dilations are further classified according to the shape of the affected segment into type Ia: cystic dilation; type Ib: focal segmental dilation; type Ic, fusiform dilation.
Type II	Diverticular dilation of the extrahepatic biliary tree.
Type III	Cystic dilation of the intraduodenal portion of the common bile duct (choledochoceles).
Type IVa	Dilation of the extrahepatic and intrahepatic biliary tree.
Type IVb	Dilation of multiple sections of the extrahepatic bile ducts.
Type V	Dilation confined to the intrahepatic bile ducts (Caroli's disease).

Infants frequently come to clinical attention with jaundice and the passage of acholic stools. If this presentation occurs in early infancy, a workup to exclude biliary atresia may be initiated. Infants with choledochal cysts can have a palpable mass in the right upper abdominal quadrant; this may be accompanied by hepatomegaly.

Children in whom the condition is diagnosed after infancy present with a different clinical constellation, which includes intermittent bouts of biliary obstructive symptoms or recurrent episodes of acute pancreatitis. Children in whom biliary obstruction is present may also have jaundice and a palpable mass in the right upper quadrant. The correct diagnosis is occasionally more difficult in children with pancreatitis. Often, the only clinical symptoms are intermittent attacks of colicky abdominal pain. Eventually, an analysis of biochemical laboratory values reveals elevations in amylase and lipase levels. This leads to the proper diagnostic imaging workup (3).

According to Miyano and Yamataka, the preferred initial radiologic examination in the diagnostic workup of a choledochal cyst is an abdominal ultrasonography (US) scan (4). US scanning is non-invasive and involves no radiation exposure, and its findings are sensitive and specific for the diagnosis. Clinically, these features make sense. Patients with choledochal cysts most often have symptoms referable to the hepatobiliary system, and most US operators are familiar with the anatomy of this area (5).

Once a preliminary diagnosis is made using US scanning, other supportive studies may be ordered, including abdominal computed tomography (CT) scans, magnetic resonance imaging (MRI) studies, or magnetic resonance cholangiopancreatographic (MRCP) examinations. These studies demonstrate the cyst with more precise anatomic detail. In addition, important anatomic relationships to surrounding structures are better defined than with other modalities.

In 1959, preoperative diagnosis of choledochal cyst was possible in only 30% of cases (6) but now it is possible in > 90% cases with the help of ultrasound and ERCP (7).

Total excision of the cyst with adequate bile drainage is the standard treatment for choledochal cyst. However, in the setting of extensive inflammation, a cyst mucosectomy and cystoenterostomy can be performed (8).

Morbidity and mortality rates after laparoscopic management are comparable with published results of the open procedure. Laparoscopic surgery for choledochal cysts is feasible, safe, and even advantageous (8).

In conclusion, choledochal cyst is usually diagnosed in childhood. Early treatment can prevent further complication. Complete excision of cyst with Roux-en-Y hepaticojejunostomy is the treatment of choice, and the late result is good. Long-term follow-up is necessary.

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