Case Report

Choledochal cyst

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ABSTRACT

Choledochal cyst, which is characterized by dilatation of the biliary ducts, is common in Asian countries, mainly Japan, but relatively rare worldwide. This report describes 2 Saudi female children with choledochal cysts, with emphasis on long term follow-up.

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Choledochal cyst is a rare malformation, characterized by dilatation of the biliary tract. Vater in 1723 reported the first case of choledochal cyst,[1] and although common in Japan, it is considered a relatively rare disease worldwide.[2] This report describes 2 Saudi female children with choledochal cyst outlining aspects of presentation, diagnosis, management and long-term follow-up.

Case Report. ***Patient One.*** A 14-month-old female was admitted to our hospital due to intermittent jaundice and itching of 3 weeks duration. She was a product of full term normal vaginal delivery. There was no history of other problems except for glucose-6-phosphate dehydrogenase deficiency. Examination revealed a well, healthy looking but jaundiced child. There was also a right-sided palpable upper abdominal mass. Investigations showed total bilirubin of 5.32 mg/dl (normal range <1mg/dl), direct bilirubin of 2.3 mg/dl (normal range <0.2 mg/dl), alkaline phosphatase (ALP) of 650 IU (normal range 26 - 88), alanine aminotransferase (ALT) of 109 IU (normal range 10 - 60), aspartate transaminase (AST) of 182 IU (normal range 10 - 42), amylase of 42 IU (normal range 0-88 IU), and hemoglobin of 11.0 gl/dl (normal range 12-15gl/dl). Abdominal ultrasound showed a large, more than 6.5 cm in diameter, rounded cyst occupying the area of the porta hepatis and overlapping the region of the inferior vena cava. Abdominal computerized tomography (CT) scan (**Figure 1**) showed a large, rounded cyst arising at the porta hepatis area and extending down, an appearance highly suggestive of a choledochal cyst. Percutaneous transhepatic cholangiogram (**Figure 2**) revealed normal right and left hepatic ducts with a large choledochal cyst. There was no evidence of an anomalous junction of the pancreatic and common bile ducts. In the delayed films, the gallbladder filled with contrast and it was of normal size, but the cystic duct was elongated, tortuous and dilated. She was operated upon, and intraoperatively, was found to have a large choledochal cyst (Type 1) involving the lower part of the common hepatic duct up to the supraduodenal part of the common bile duct. There were adhesions around the choledochal cyst, which were carefully released using both sharp and blunt dissection until the whole cyst was isolated. She underwent cholecystectomy, and total excision of the choledochal cyst with Roux-Y-hepaticojejunostomy. Postoperatively, she did well and was discharged home on the 14th postoperative day. Histological examination of the resected specimen revealed a cystic swelling composed of a fibrotic wall with chronic inflammatory cell infiltrate and a destroyed epithelial lining. The gallbladder showed chronic inflammation. There was no evidence of any malignant transformation. On follow-up, she is now 12 years postoperative and well. There was no evidence of

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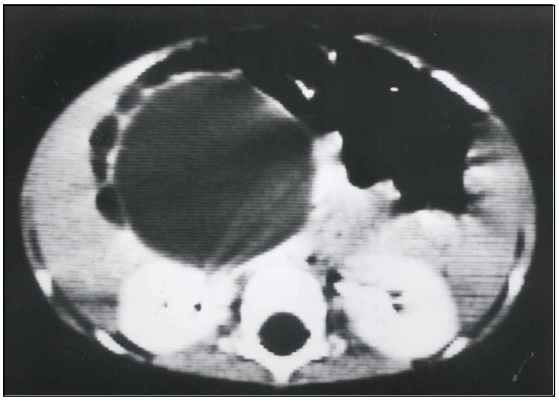

Figure 1

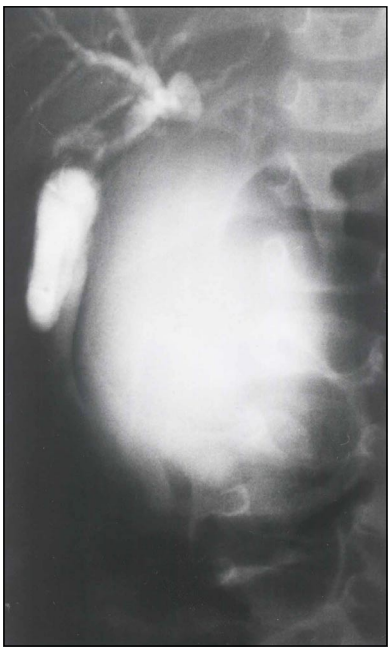

Figure 2

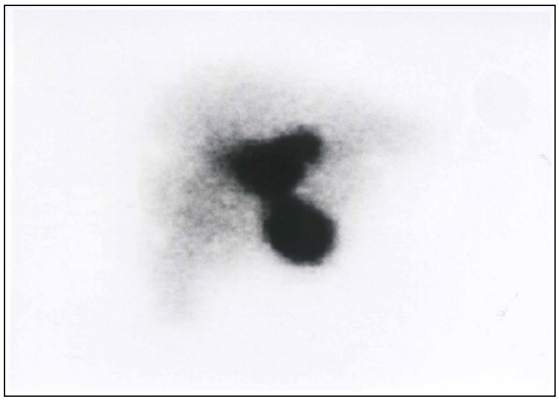

Figure 3

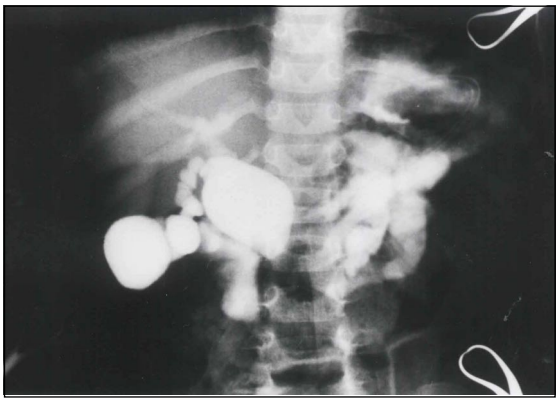

Figure 4

Figure 1 - Abdominal CT scan showing a large choledochal cyst.

Figure 2 - Percutaneous transhepatic cholangiogram showing normal right and left hepatic ducts and a very large choledochal cyst.

Figure 3 - Hepato-iminodiacetic scan showing choledochal cyst.

Figure 4 - Intraoperative cholangiogram showing normal right and left hepatic ducts and choledochal cyst.

cholangitis, and her liver function tests were normal. Her last abdominal ultrasound showed no abnormalities and her liver and intrahepatic biliary radicals were normal.

Patient 2. A 3-year and 9-month-old female was admitted to the hospital with recurrent abdominal pain of 10 months duration. She was seen in the emergency room on several occasions and each time she was treated symptomatically. The pain was associated with vomiting. Clinically, she was not jaundiced, and on abdominal examination no palpable masses were detected but there was mild upper abdominal tenderness. Her total bilirubin was 0.7 mg/dl, ALP 326 IU, ALT 242 IU, and her amylase was normal. Abdominal ultrasound revealed a cystic mass measuring 2.9 x 2.3 cm in size with echogenic contents and in the line of the common bile duct. The biliary ducts above the cystic mass were normal, and there was no intrahepatic biliary duct dilation. There was no evidence of an anomalous junction between the pancreatic and common bile ducts. She had a hepato-iminodiacetic (HIDA) scan (**Figure 3**) which was consistent with a choledochal cyst. She was operated on and intraoperative cholangiogram (**Figure 4**) showed a fusiform choledochal cyst (Type 1) with normal proximal biliary ducts. She was operated on and intraoperatively was found to have a fusiform choledochal cyst extending from the lower part of the common hepatic duct to the supraduodenal part of the common bile duct. There were adhesions between the choledochal cyst and surrounding structures, which were released using both sharp and blunt dissection. She underwent cholecystectomy and total excision of choledochal cyst with Roux-Y-hepaticojejunostomy.

Postoperatively, she did well and was discharged home on the 14th postoperative day. Histology showed a cystic swelling composed of a fibrotic wall infiltrated with chronic inflammatory cell infiltrate. The lining epithelium was destroyed in most of the wall. There was no evidence of any malignant transformation. The gallbladder showed chronic inflammation. She is now 8 years postoperative and well. Her last liver function tests were normal and abdominal ultrasound revealed no intrahepatic biliary dilatation.

Discussion. Choledochal cyst is a relatively rare disease characterized by dilatation of the biliary ducts, and although Vater is credited as the first to describe it in 1723,[1] it was the comprehensive review published in 1959 by Alfonso-Leje et al that shed light on choledochal cyst including its clinical manifestations and classification.[3] Choledochal cyst is a relatively rare disease worldwide, and the exact incidence is unknown ranging from 1:13,000 - 1:2,000,000,[4,5] but there is a definite geographical variation in its incidence being more common in Asian races mainly from Japan where there seems to be an unusually high incidence.[2,6,7] In Saudi Arabia, the incidence of choledochal cyst is not known, but like in other parts of the world, it is considered to be rare. There is also a definite female preponderance with a male:female ratio of 1:4, and the majority present in the first decade of life. In a large collective series of 1433 patients with choledochal cysts in the Japanese literature, their ages at the time of presentation ranged from 6 days - 78 years, however, 45% of them were infants.[2] The most common clinical presentation of choledochal cyst is recurrent attacks of abdominal pain, and the classic triad of abdominal pain, jaundice and abdominal mass are present in only 13% of cases.[2] Jaundice in these patients can be persistent obstructive seen in infants mainly or mild intermittent seen in older children and adults. The rarity of the condition as well as variations in its clinical presentation are contributing factors for the delay in diagnosis. Both our patients were female. The first presented with intermittent jaundice, itching and was found to have a large abdominal mass, while the second presented with recurrent abdominal pain. The diagnosis in our first patient was not difficult to reach, while in the second it was delayed for almost one year because of non-specificity of symptoms. In both patients, the diagnosis was made on abdominal ultrasound. To obviate this delay, physicians caring for these patients should be aware of this, and choledochal cyst should be included in the differential diagnosis of infants and children presenting with recurrent abdominal pain with or without jaundice and abdominal mass. Abdominal ultrasound a being simple, none invasive investigation should form part of the investigative evaluation of children presenting with such symptoms.

The exact etiology of choledochal cyst is not known. Yotuyanagi postulated an inequality of epithelial proliferation at the stage of physiological epithelial occlusion of the primitive choledochus.[8] Many workers have demonstrated the occurrence of an anomalous junction of the pancreatic and common bile ducts in association with choledochal cyst.[9,10] This however was not demonstrated in our patients. In 1969, Babbitt postulated that reflux of pancreatic juice into the common bile duct as a result of this anomalous junction will weaken the common bile duct wall giving rise to its dilatation.[11] Others postulated an associated distal congenital stenosis as the cause of dilatation.[12] Choledochal cysts have been classified into 5 different types.[2,13] Type I, which is the most common (75-85%) is characterized by cystic dilatation of the common bile duct. Both our patients had type 1 choledochal cyst. Type 2 (2-3%) is a diverticulum of the common bile duct while type 3 (1-5%) which is also called choledochocele is a cystic dilation of the intraduodenal part of the common bile duct. In type 4 (9-19%), there is a choledochal cyst associated with intrahepatic biliary cysts. In type 5, also called Caroli's disease (1-2%), there are multiple intrahepatic biliary cysts.[14]

The treatment of choledochal cyst is surgical. This is to obviate the associated complications (16.5%) and the risks of increased morbidity and mortality. External drainage as well as internal drainage in the form of choledocho-cysto-duodenostomy or choledocho-cysto-jejunostomy are no longer advocated because of the associated high incidence of cholangitis and anastomotic stricture as well as the occurrence of malignant change in the residual cyst. We like others, advocate total cyst excision and Roux-Y-hepaticojejunostomy.[2,6,7] In patients with pericystic inflammation and to obviate injury to surrounding structures, the cyst can be dissected from within excising all the lining mucosa and leaving the posterior wall. The importance of long-term follow-up in these patients needs to be emphasized. This is because of known associated long term complications including cholelithiasis, cystolithiasis, pancreatolithiasis, stenosis at the site of anastomosis with ascending cholangitis, and malignant transformation. These complications are however, much less or non-existing following total excision of the cyst and Roux-Y-hepaticojejunostomy. There is also an increased incidence of carcinoma arising in choledochal cyst, which is greater than that in the general population and this is commonly seen in adults.[15] According to the report by Todani et al,[16] the ages of patients with carcinoma arising in a choledochal cyst ranged from 15-73 years (mean 37 year) but the youngest patient reported so far with adenocarcinoma was 12 years old.[17]

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