



Common Presentation of Choledochal Cyst in Adult Female – A Case Report

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Abstract:

The Choledochal cyst is a rare disease of unknown etiology. Incidence of diagnosis of choledochal cyst is much higher in children (80%) than in adult (20%). It is prevalent among Asian & female populations & the incidence is 1:100000-150000. We are reporting one such case, a 57-year-old female presenting with recurrent pain with right hypochondriac region and investigations revealed type -1 choledochal cyst. This case was managed successfully by complete excision of cyst & Roux-en-Y hepatico-jejunostomy.

Introduction:

Choledochal cysts are congenital anomalies characterized by cystic dilatation of the biliary tree, extra hepatic or intra hepatic or both. In 1956 Alonso-LEJ et al. were the first to classify choledochal cyst into 3 types based on the site of the biliary duct dilatation¹. It was not until 1977 when Todani et al. modified the later classification by adding 2 types of choledochal cyst². This 5 category classification is the most commonly used by the clinicians today. Diagnosis is made based on disproportionate dilatation of biliary duct excluding possibility of a tumor, stone or inflammation as a cause of this dilatation.

Incidence of diagnosis of choledochal cyst is much higher in children (80%) than adult³. The clinical presentation is not specific, even the classic triad with abdominal pain, mass & jaundice. In adults, intermittent abdominal pain is the most common symptoms. Recurrent features of cholangitis may also occur.

The main diagnostic tool for detection of a choledochal cyst especially in children is Ultrasound scan. In adult, computed tomography can confirm the diagnosis. However, ERCP and MRCP are the most valuable diagnostic methods & can accurately show cystic segment of the biliary tree⁴.

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Surgery is the treatment of choice for choledochal cyst. Complete excision of all cystic tissue is recommended because of the risk of recurrent cholangitis & high risk of malignant transformation.⁵ Total excision of cyst together with the gall bladder and part of bile duct followed by biliary reconstruction is the standard procedure. Our case report of a 57-year old patient on whom we performed this surgery with a Roux-en-Y hepatico-jejunostomy.

Case Report:

Our patient was a 57-year old woman that presented at our hospital with abdominal pain. The patient presented with recurrent history of abdominal pain lasting for 3 months in right upper quadrant of the abdomen. Clinical examination revealed neither jaundice nor palpable abdominal mass.

The clinical laboratory data were normal. USG revealed Gallbladder normal in size, wall thickness within normal limit. There was no evidence of any

gall stone or sludge. Intra hepatic biliary tree was not dilated. Mild fusiform dilatation was seen in the mid CBD measuring about (26×14 mm) up to distal & proximal CBD. The findings were most compatible with choledochal cyst. MRCP revealed fusiform dilatation (measuring about 19mm in diameter) involving entire length of CHD & CBD with smooth tapering & concentric narrowing at terminal CBD involving ampullary region. While the liver presented to be normal.

Intra-operative exploration revealed a type I choledochal cyst. Resection of the cyst with surgical Roux-en-Y hepatico-jejunostomy was performed. Resected specimen was sent for histopathological examination. The patient had a steady post-operative course. Histopathology revealed compatible with choledochal cyst without any malignancy.

Discussion:

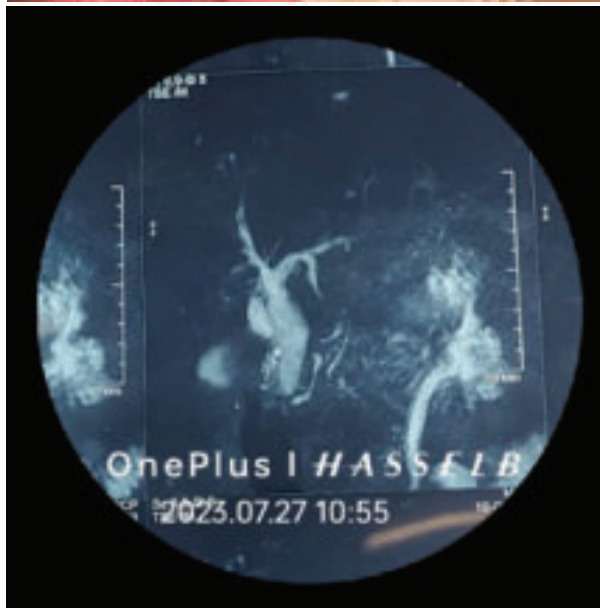


Fig.-1 : Fusiform dilatation of CBD

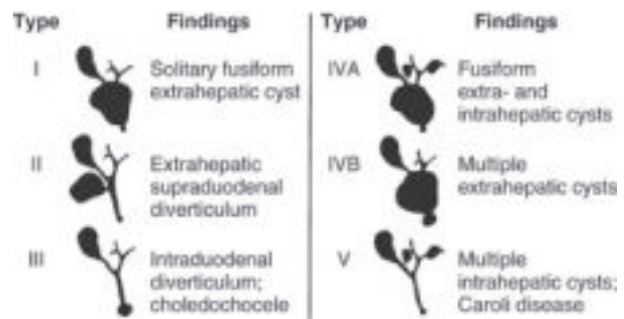


Fig.-2: Todani Classification of bile duct cysts

The reported incidence of choledochal cyst is around 1:100000-150000 live birth where two third of the reported cases in Asia occur in Japan⁷. Female to male ratio is 4:1. The region for the Asian & female predominance remain unknown. The Todani classification of bile duct cysts divides cysts of bile duct into five groups.

The etiology of choledochal cyst still remains unclear. The most popular theory is Babbit's which states that the cyst develops from an anomalous pancreatobiliary junction (APBJ). APBJ join 1 to 2 cm proximal to the sphincter of oddi. As a result, the long channel formed is not covered by sphincter and allows backflow and mixing of pancreatic and biliary secretion, which leads to the activation of pancreatic enzymes. This backflows causes dilatation, inflammation and epithelial change,

dysplasia and malignancy of the biliary tree.⁸ Oligogangliosis at the terminal portion of the bile duct causing functional obstruction and proximal dilatation has been implicated recently in the etiology.

Children present with the classic triads of abdominal mass, abdominal pain and jaundice. Adult however present mainly pain⁹ and complications of long standing cysts such as recurrent cholangitis, pancreatitis and malignancy. A palpable mass is a rare as a presenting feature in adult.

This condition can be identified by using specific imaging, such as ultrasound, CT scan, MRCP and ERCP. In most cases, Ultrasound is the first investigation to identify this condition. In case of suspicious diagnosis of biliary cyst at ultrasound, a CT scan or MRCP is required. MRCP is considered as Gold standard investigation for diagnostic evaluation as it is non-invasive technique which doesn't use radiating ionizing agents.

There is increased risk of malignancy in choledochal cyst and occurrence of cancer in most marked in adult. Type 1 has highest predominance of cancer followed by Type 4 cyst. Type 3 cysts are the least common types of harboring cancer. Most of the tumors are found adenocarcinoma, although squamous and anaplastic cell carcinomas are occasionally discovered. The most common site of occurrence is the posterior cyst wall. Prognosis is poor and most patient die within 2 years of diagnosis.

The management approach of choledochal cysts depends on the cyst types and the extent of hepatobiliary pathology. As a rule, all cysts should be resected and bile flow should be restored.

Type 1 and Type 4 choledochal cysts are managed with complete excision and restoration of bile flow by Roux-en-Y hepatico-jejunostomy. We did cholecystectomy and complete excision of choledochal cyst with Roux-en-Y hepatico-jejunostomy in our patient.

The risk of type-2 and type-3 cysts to malignancy is lower. The management of type-2 consists of simple excision of the cyst. Type-3 choledochal cyst can be treated by transduodenal cyst excision with or without sphincterotomy that was the treatment of choice. Type-5, caroli's disease, management

depends on the extent of disease. If it is localized or unilobar, then segmental hepatic resection is the best option. If it is diffused or bilobar, then liver transplantation is the option.

Conclusion:

Choledochal cyst should be considered in the differential diagnosis in all patients with history of biliary colic pain, intrabiliary calculus, mechanical jaundice and dilatation of bile duct. Complete cyst excision should be carried out early. Long term follow up is required in these patients as they are prone to cholangitis, anastomotic stricture and malignancy in the residual biliary tree.

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