

Choledochal Cyst in an Adult

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Abstract

Choledochal cyst is a cystic dilatation of the biliary passages. It is a disease which was more commonly seen in the pediatric age group; however, more and more such cases are seen in the adult population. The high complication rate including the risk of developing malignancy in adults makes it important to be aware of its presentation and the various surgical options for treatment. A case of choledochal cyst in an adult is presented along with a review of literature

INTRODUCTION

Choledochal cyst is an aneurysmal dilatation of the bile duct. It is a rare condition usually seen in the pediatric age group. There are less than 3500 cases reported till now, the majority of which are from Japanese literature.[1] Adult patients presenting with choledochal cyst are quite uncommon. In contrast to the pediatric experience, adults have an increased incidence of associated pathology of the hepatobiliary system, giving rise to a multitude of complications.[2,3] Hence, only prompt diagnosis and optimum treatment can prevent lethal complications such as cholangiocarcinoma. A case of a choledochal cyst in an adult female is presented along with review of literature

CASE REPORT

A 36-year-old female patient presented to the outpatient department with a history of right sided upper abdominal pain. The pain was dull aching in nature with no specific aggravating or relieving factors. There was no history of jaundice or fever associated with the pain.

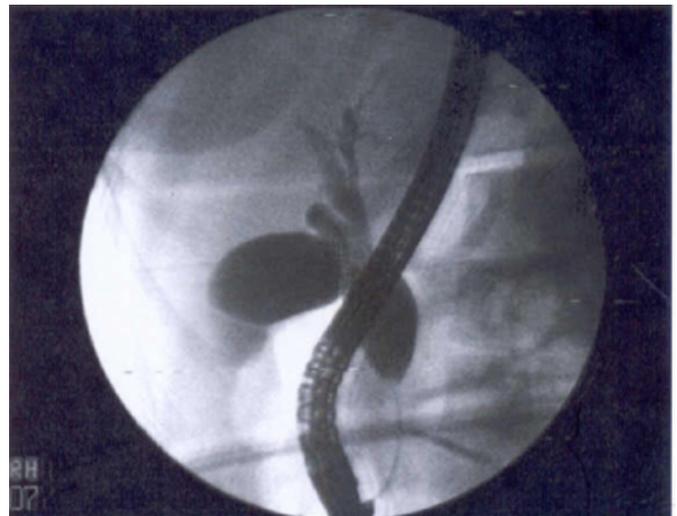
Physical examination revealed tenderness on deep palpation in the right hypochondrium. No mass was palpable. There was no free fluid in the abdomen. Hematological evaluation including liver function tests did not reveal any abnormality.

Abdominal ultrasound revealed dilatation of the common bile duct. However, no stones were detected. Liver and pancreas were normal.

ERCP revealed a dilated common bile duct (Figure 1). The cystic duct was arising from the dilated part of the common bile duct (CBD). The lower end of the cyst extended up to the duodenum (type I cyst).

Figure 1

Figure 1. ERCP showing cystic dilatation of the common bile duct..



The patient underwent exploratory laparotomy. The duodenum was kocherized. The cyst was dissected from the portal vein. The cyst was resected along with gallbladder, cystic duct and a part of the common hepatic duct, ensuring normal caliber of the cut end of the common hepatic duct (Figure 2). The distal part of the CBD at the duodenal end was sutured and closed.

Figure 2

Figure 2. Specimen removed showing the cyst, cystic duct, gall bladder and part of the normal hepatic duct.



No abnormal pancreaticobiliary communication was found. A Roux-en-Y hepaticojejunostomy was performed. The patient tolerated the procedure well and was started on oral fluids on day 4. She was discharged from hospital on day 7. Histopathological examination of the specimen did not reveal any malignancy. The patient has been following up for the last year with no complications.

DISCUSSION

Choledochal cysts presenting in adulthood are uncommon.[4] Choledochal cysts are classified on the basis of the part of the biliary system involved as described by Todani’s modification of Alonso-Lej’s classification (Table 1).[5.6]

Table 1: Classification of Choledochal Cysts

Type I: Dilatation of the extrahepatic biliary tree

Ia: Cystic

Ib: Focal

Ic: Fusiform

Type II: Saccular diverticulum of the extrahepatic bile duct.

Type III: Bile duct dilatation within the duodenum (Choledochoceles)

Type IV: Dilatation of the intrahepatic and extrahepatic biliary tree.

Type V: Intrahepatic cysts (Caroli’s Disease)

The condition is due to a weakness of the whole CBD wall or of a part of it. The majority of patients with choledochal cysts have an abnormal pancreaticobiliary junction (APBJ) i.e., the pancreatic duct enters the CBD 1cm or more proximal to where the CBD reaches the ampulla of Vater. This causes pancreatic juices to become activated causing inflammation and weakening of the bile duct wall. Pathologically, the cyst wall is composed of fibrous tissue with interspersed elastic and sparse smooth muscle fibers, the internal lining consists of columnar epithelium but it is often destroyed by inflammation and the presence of the distending fluid.[8] The condition is more common in women.[3] The majority of these patients presents with abdominal pain or discomfort. The classic triad of abdominal pain, abdominal mass and jaundice is not classically seen in all, but only 6% of patients with choledochal cysts.[7] The complications are intrahepatic abscesses, portal hypertension and malignancy. These complications are most often seen in patients suffering from Caroli’s disease.

USG is the best initial investigation. It reveals the extent of the lesion as well as the presence of stones. It helps in differentiating a choledochal cyst from an extrahepatic cystadenoma by the absence of internal septations. Pancreatic complications such as pancreatitis may be picked up.

ERCP is the most specific investigation for diagnosing choledochal cysts. It provides an excellent road map for planning treatment. ERCP shows the extent of the disease as well as an abnormal pancreaticobiliary junction if present. Filling defects due to stones or malignancy, other pancreatic anomalies such as pancreas divisum and curled main pancreatic duct can also be detected.

CT scan provides information regarding the relationship of the cyst to the portal vein. This helps in planning the surgical approach.

Surgical treatment of choledochal cysts is based on specific principles, viz. restoration of normal bile flow, abolition of pancreatic reflux and reduction of the risk of malignancy by removing the most common sites of malignant transformation, namely the cyst wall or the gall bladder.[9]

Type I & II cysts are treated by complete excision of the cyst along with cystic duct and gall bladder. The proximal margin of resection should be at the junction of the cyst and the normal appearing common hepatic duct. Distally, the cyst is

divided just above the pancreatic duct entry. The periampullary duct is oversewn. Reconstruction is accomplished by Roux-en-Y hepaticojejunostomy. If adhesions are dense especially in relation to the portal vein, complete resection is precluded. The anterior cyst wall is incised transversely. An arbitrary submucosal plane is created and the entire inner mucosal lining is removed. Full thickness of the anterior and lateral cyst wall is removed while a submucosal resection is done posteriorly. The outermost cyst wall adherent to the vascular structures, namely the portal vein and the hepatic artery, is left behind. Reconstruction is done by hepaticojejunostomy.

Type III cysts are partially excised and opened into the duodenum by means of either trans-duodenal sphincteroplasty or endoscopic sphincterotomy.[10]

Type IV cystic lesions require complex segmental resection of the liver if associated with lesions like stones, strictures or abscesses along with hepaticojejunostomy in order to restore biliary continuity. For advanced type IV and type V lesions, liver transplantation is required. Continuous follow-up is necessary especially in those patients who present for surgery in adulthood. The incidence of malignant change in these patients may reach 10% [10] and 75% of these tumors are adenocarcinomas. Hence, the malignant potential of this condition mandates complete and meticulous excision of the

cyst.

The prognosis for most surgically treated patients with type I & II choledochal cyst is excellent

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