CASE REPORT

Choledochocele: A Case Report and Review of Literatures

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ABSTRACT:

Choledochocele is a congenital cystic dilatation of the terminal portion of the common bile duct, which classified as type III choledochal cyst.

We present a patient with large choledochocele which is 6cm in diameter treated by transduodenal excision of the cyst with sphincteroplasty; this case report is one of the largest reported cases of choledochocele in the literature.

INTRODUCTION:

A choledochocele is a cystic dilation of the distal, intramural portion of the common bile duct, typically protruding into the descending duodenum. It is classified as a type III choledochal cyst according to the system originally presented by Alonso-Lej et al. [(1)].

Todani and colleagues expanded this system in 1977 to include the occurrence of the intrahepatic and the multiple cysts, and this modified classification is now most commonly used by clinicians. [(2)]

Choledochoceles are seen in about 1.5% of congenital choledochal cysts. [(3)]

Different from type I choledochal cyst, choledochocele anomaly was not generally regarded as being associated with abnormal arrangement of the pancreatobiliary ductal system or biliary malignancy, but recently, there have been some reports of patients with choledochocele associated with biliary malignancy, [(4,5)] as well such patients with high levels of amylase in bile. The possibility of choledochocele as a high-risk state for biliary malignancy is gaining interest. [(6-8)]

CASE REPORT:

Twenty year old mother of 2 children was complaining from recurrent upper abdominal pain during the last 18months, but the last few weeks the pain became more severe, frequent and radiated to the back, associated with bilious vomiting and sometimes with diarrhea. She lost five kilograms of her weight during this period.

She was slightly under weight, pale but not jaundiced, abdomen was soft but there was slight tenderness on epigastric area. Blood test revealed anemia (Hb=9.5gm/dl) others were normal including liver enzymes. Abdominal ultrasound revealed clear cyst related to the wall of the second part of duodenum 5cm in diameter, CBD 6mm, pancreatic duct 3mm diameter. Esophagogastroduodenoscopy was normal.

Abdominal C.T scan revealed 6cm diameter cyst at pancreatic head no septation or solid part. Endoscopic ultrasound showed multilayered cystic lesion in submucosa of second part of duodenum, pancreatic parenchyma as well as pancreatic duct was normal.

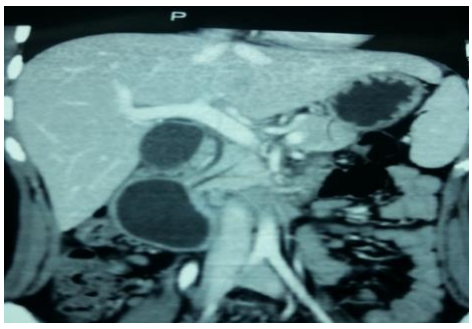

Figure 1: CT-scan of the abdomen revealed cystic lesion related to duodenum and head of pancreas.

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Operation done by midline incision, kocherization of the descending duodenum and 5cm longitudinal lateral duodenotomy revealed 5x6cm cm cystic mass covered with normal duodenal mucosa along the medial duodenal wall communicating with ampulla of Vater 1cm superior to the papilla. After safe guarding of CBD and pancreatic duct complete excision of the cyst with overlying duodenal mucosa done, the defect in ampulla was extended to the papilla to became one opening upon which sphincteroplasty done. The duodenal wall was closed by continuous extramucosal suturing using 3/0 Vicryl.

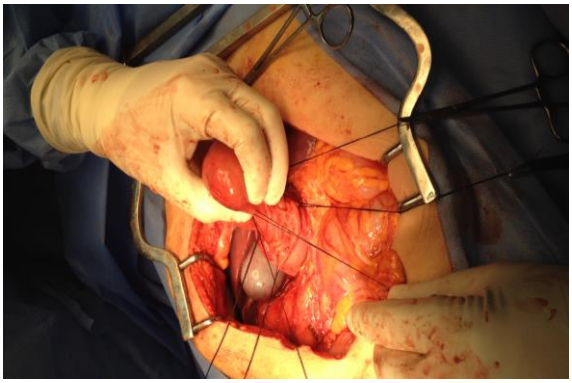

Figure 2: Intraoperative picture showing the choledochoceles protrude from medial wall of duodenum.

The postoperative period was smooth.

Histopathology study showed that the cyst lined by cuboidal epithelium similar to that of biliary ducts.

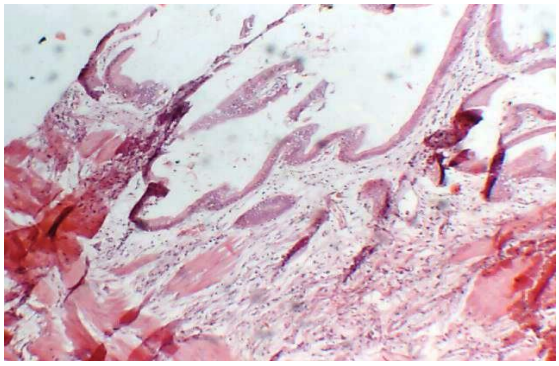

Figure 3: Showed the histopathology of choledochocele lined by cuboidal epithelium similar to that of biliary ducts.

DISCUSSION:

The definition of choledochocele is not well established, although there have been several proposals. [9,10] In general, enlargement or protrusion of the papilla of Vater and cystic dilatation of the terminal bile duct are considered to be mandatory. However, there are some conditions mimicking choledochocele, such patients with duplication cysts, those with impacted gallstones at the papilla of Vater, the development of cystic change at the terminal bile duct in patients who have undergone endoscopic sphincterotomy for bile duct stones, [9] and some patients with abnormal arrangement of the pancreatobiliary ductal system, necessitates great care in making the diagnosis.[11]

Histologic evaluation of the choledochocele wall was reported, in the pediatric cases; the cyst was lined with duodenal mucosa. In adult cases reviewed, both duodenal and biliary mucosa have been reported with equal frequency.[12]

Theoretically, type III cysts may lead to findings of the intestinal system (such as obstruction, bleeding or perforation) as they protrude to the duodenal lumen. However, as stated above,

choledochocele constitutes only a small fraction of total cases and there is little information in the literature regarding different symptoms and findings, as most of these cysts may be asymptomatic. Our patient had neither biliaryobstruction findings such as jaundice and hyperbilirubinemia nor any sign or findings in favor of infection.[13]

The choice of therapy depends upon the size the cyst. Choledochoceles measuring 3 cm or less can be treated effectively with endoscopic sphincterotomy. Lesions larger than 3 cm typically produce some degree of duodenal obstruction. These lesions are excised surgically through a transduodenal approach. If the pancreatic duct enters the choledochocele, it may have to be reimplanted into the duodenum following excision of the cyst. [14]

Because of their rarity and lower overall rate of malignant transformation, reports of choledochocele excision are uncommon. However, excision may be necessary in symptomatic patients, but many patients may benefit from endoscopic sphincterotomy. Surgical resection is typically approached via a transverse duodenotomy in the second or third portion of the duodenum. Prior to duodenotomy, cholecystectomy is performed and then the ampulla can be localized by passing a biliary Fogarty catheter into the duodenum via the transected cystic duct. The anatomy can also be better defined prior to duodenotomy by performing a Kocher maneuver and intraoperative ultrasound. Duodenotomy allows both the biliary and pancreatic ducts to be identified individually. The pancreatic duct can be intubated using a small Silastic tube so that the intraduodenal biliary cyst can be excised. Both the bile duct mucosa and pancreatic duct mucosa are then sutured individually to the duodenal mucosa using interrupted absorbable sutures. A piece of 5F or 8F plastic tubing can be placed into the pancreatic duct and secured with a single absorbable suture as a temporary stent to prevent postoperative acute pancreatitis. Finally the duodenostomy is closed in a transverse fashion. Type I and IV cysts have the highest risk of cancer, while cancer is rare in type II and III cysts. In type III cysts, cancer risk may be limited to those choledochoceles lined by biliary and not duodenal epithelium [15], as in our case.

CONCLUSION:

The choledochocele can be as large as 6cm in diameter, we thought that surgical excision rather than endoscopic sphincterotomy would be the best treatment to abolish the risk of malignancy completely.

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