Choledochal cyst: Histological structure, classification and clinical significance

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ABSTRACT: For this paper we present a very rare case of a diverticulum arising from the posterior wall of the common bile duct, 1 cm distal to the junction of the cystic duct. That diverticulum appeared a wide basis of communication with the common bile duct lumen and directed onto the right side without causing compression phenomena. Remarkable notice was the presence of ectopic tissue of gallbladder within the choledochal cyst wall fact which happens to be a very rare condition. We analyse and discuss the histology, pathogenesis, classification, implications and in general the possible clinical significance of that congenital abnormality.

Key Words: Choledochal cyst, Structure, Clinical significance.

INTRODUCTION
The choledochal cyst is a relatively rare congenital abnormality of the extrahepatic biliary tract, with an incidence of 1:13,000 - 1: 2,000,000 births¹. The first report of the presentation of such an anomaly was made by Vater and Ezel in 1723². It is noteworthy to be reported that the 70% of the approximately 3,000 cases reported until nowadays, are derived from Japan, while the 80% of those cases concern females³.

We incidentally have found a saccular cyst of the inferior wall of the supraduodenal portion of the common bile duct without association of another congenital abnormality. We refer to that anomaly, macroscopically and we discuss the pathogenesis, classification and clinical significance of that anomaly.

Case report
During routine preparation of the cadavers into the dissection room of our Department of Anatomy and after excluding the organs of the upper abdomen, we prepared the region of the extrahepatic biliary ducts. During this procedure, we found out a hemispherical diverticulum arising from the posterior wall of the supraduodenal portion of the common bile duct, 1 cm approximately distal to cystic duct junction. That cyst was directed onto the right side and appeared to have a wide communication with the lumen of common bile of the duct and had a diameter of approximately 1,2 cm (Figure 1). From its location did not seem to cause any compression to surrounding structures, such as the portal vein.

Discussion
For the explanation of the saccular distention of the common bile duct many theories have been stated. The most predominated one is the «common-channel theory» stated by Babbit in 1969. Babbit claimed that the abnormal junction of the terminal portion of common bile duct with the pancreatic duct especially when this occurs proximal to Oddi’s sphincter results in pancreatic fluid regurgitation to the common bile duct. The resulting activation of pancreatic enzymes causes common bile duct epithelium destruction. The following chronic inflammatory reaction of submucous layer causes thinning and distention of the common bile duct wall. However, that theory cannot explain the total number of cases, since in 30% of pa
tients with cystic dilatation of the extrahepatic biliary ducts does not appear any abnormal opening of common bile duct into the duodenum.

Another theory supports that pancreatic enzymes activation is derived from ectopic pancreatic tissue of the common bile duct. That tissue has been described in 4-98% of individuals with extrahepatic biliary tree cysts. The diagnosis of the extrahepatic biliary tree dilatation through sonography even in fetal life cannot be explained with the previous mentioned mechanisms, since the pancreatic enzymes are activated after the second month of life. Absence of ganglionic cells in the common bile duct wall, especially distal to gallbladder may produce choledochal cysts in the similar manner with Hirschsprung’s disease in the large intestine.

The most current classification of choledochal cysts is that of Todani formulated in 1977. According to that classification there are five types of common bile duct cysts: 1) Type Ia: single spindle-shaped cystic dilatation of the common bile duct (5%), type Ib: segmental dilatation (1%), type Ic: diffuse dilatation. 2) Type II: common bile duct diverticulum proximal to the duodenum (3%). 3) Type III: Cystic dilatation of the intramural segment of the common bile duct (5%). 4) Type IVa: intrahepatic and extrahepatic multiple cysts (10%), type IVb: only extrahepatic multiple cysts (<1%). 5) Type V: single or multiple intrahepatic cysts (Caroli’s disease) (<1%) with unilobar or diffuse location (Figure 3).

The histological features of those cysts include: a) Presence of thick layer of dense connective tissue with few bundles of smooth muscular fibres. b) Existence of non-cellular mucous membrane, with the rare co-existence of few cylindrical epithelium insulæ. c) Presence of chronic inflammation, especially in elderly individuals.

Choledochal cysts have been described in combination with duodenal atresia, annular pancreas, ectopic papilla, pancreas divisum, congenital agenesis of the portal vein, intestinal malrotation, polycystic kidney disease, congenital biliary atresia, accessory...
hepatic ducts, supranumerary hepatic artery, atresia of colon, hypoplasia of upper limbs, agenesis of the gallbladder, multiseptated gallbladder, diaphragm of the common bile duct, familial adenomatous polyposis and primary biliary cirrhosis.

Choledochal cysts predispose to the development of gallstones either within the gallbladder, or in other sites of the biliary tract. Choledochal cysts, especially of type I and IV are related to tumor formation in liver and biliary tract. The most common type is choledochus carcinoma. Only 57% of carcinomas are derived from the cyst’s epithelium. Malignant tumors associated with choledochal cysts are very rare in childhood, while non-malignant neoplasms have been reported and specifically associated to common bile duct adenomas. These patients, also, appear to have increased incidence of pancreatitis (2-70%). This may be due to abnormal pancreatic and common bile duct junction or gallstones formation within the choledochal cyst. Finally, spontaneous perforation either to the hepatic artery or, the common bile duct, especially in children are very rare implications of the choledochal cysts.

Figure 3. Classification of choledochal cysts according to Todani.
REFERENCES


