Introduction

A choledochal cyst is defined as a cystic dilation of the common bile duct. In 1723, VATER was the first to show a fusiform dilation of the common bile duct and DOUGLAS published on the possible congenital origin of this cyst in 1852 (1-2). The incidence of a choledochal cyst is about 1:100,000-150,000 with a men to women ratio of 1:3 (3-4). The aetiology is unknown. It is frequently seen in Asiatic countries, especially Japan. The authors would like to present a case of this pathology, rather rare in Western countries.

Case report

A previously healthy 20-year old woman presented with nausea, progressive jaundice and epigastric pain since 2 months. She also had tea colour urine and clay colour stools.

At clinical examination, she had right upper quadrant and epigastric pain.

Liver function tests showed total bilirubin of 12.56 mg/dl (normal 0.20 - 1.10 mg/dl) with conjugated bilirubin of 9.48 mg/dl (normal 0.00 - 0.30 mg/dl), alkaline phosphatase of 357 U/L (normal 35 - 104 U/L), gammaglutamyltranspeptidase of 218 U/L (normal 5-39 U/L), alanine aminotransferase of 315 U/L (normal 10 - 31 U/L) and aspartate aminotransferase of 149 U/L (normal 10 - 32 U/L).

Abdominal ultrasound revealed a dilation of the common bile duct with a diameter of 3 cm. On CT-scan the common bile duct was dilated fusiformly, with normal diameter at the head of the pancreas (Fig. 1). An anomalous position of the gall-bladder was evidenced. MRI with T2-weighted images and magnetic resonance cholangiopancreatography (MRCP) showed a massive dilation of the common bile duct, as seen on CT-scan, with a maximal diameter of 3.3 cm (Fig. 2). A choledochal cyst was therefore diagnosed. Because of its possible malignant degeneration resection was indicated. We performed an upper median laparotomy with cholecystectomy and resection of the cyst en-bloc (Fig. 3). A Roux-en-Y hepaticojejunostomy reconstruction was made. Anatomopathology showed a choledochal cyst with a single-layered cylindrical epithelium without...
evidence of dysplasia or carcinoma. There were no postoperative complications and the patient was discharged after 1 week. One year after the operation, she is asymptomatic.

Discussion

The first classification on choledochal cysts dates from 1959 and is from Alonzo-Levy et al (5). Nowadays, different types of choledochal cysts are classified according to the Todani’s classification (6). Type I is a fusiform dilation of the common bile duct, as seen in our case. It is the most frequent cyst with 80 to 90% prevalence. Most of the literature relating to diagnosis and treatment of choledochal cysts refers to type I lesions. It classically presents as a triad of jaundice, mass and right upper quadrant abdominal pain; however this triad is rarely seen in adults (7). Jaundice and abdominal pain, together with lightening of stools and darkening of the urine appeared in the reported case. Other possible symptoms are nausea, vomiting, abdominal distension, hepatosplenomegaly, diarrhoea and fever (7).

In 80% of the cases the diagnosis is made before the age of 10 (4). Diagnosis can be made by ultrasonography, CT-scan, endoscopic retrograde cholangiopancreatography (ERCP) and MRCP (4, 8). Ultrasonography remains the most useful initial method of evaluating biliary tract abnormalities. CT-scan can be used to confirm the diagnosis (4). A form of cholangiography is mandatory to define the precise anatomy and can either be ERCP or MRCP (3). In the case presented, 3 important investigations were done. Ultrasonography was performed as initial screening method, CT-scan to establish the diagnosis and MRCP to show the precise anatomy of the biliary tract. Because abnormal laboratory values often suggest obstructive biliary disease, initial imaging should focus on the presence of an obstructing lesion on the biliary tract and pancreaticobiliary junction. Therefore a choledochal cyst has to be differentiated from pancreatic pseudocysts, echinococcal cysts and cystic biliary neoplasms such as biliary cystadenoma and biliary cystadenocarcinoma (9).

The most common complications of a choledochal cyst are: cholangitis, pancreatitis, biliary cirrhosis, liver abscess, cholelithiasis, portal hypertension, cyst rupture, portal aneurysm and malignant degeneration (7). The risk of complications increases with age. The most important complication is malignant degeneration, with an incidence of 2.5 to 26% (4). Surgical management varies according to the type of cyst. Type I cyst requires surgical resection with Roux-en-Y hepaticojejunostomy (3-4, 7-8). In the past, the treatment of choledochal cysts consisted of internal drainage by cystenterostomy. Because of the increased risk of carcinoma, high rate of recurrent cholangitis and pancreatitis, drainage was abandoned in favour of cyst excision (4). Long-term follow-up of patients is necessary after any operation, because biliary tract carcinoma may be a late postoperative complication of choledochal cysts (10).

Although choledochal cysts are rare in Western countries, the authors would like to stress the need for a good diagnostic work-up and careful surgical treatment, as shown in this case.
References


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