Choledochal Cyst – Presentation and Treatment in an Adult

Ferat Sallahu, Antigona Hasani, Dalip Limani, Skender Shabani, Fadil Beka, Skender Murati, Hysni Jashari
Clinic of Surgery, University Clinical Centre of Kosovo, Prishtina, Kosovo

Corresponding author: Ferat Sallahu, MD, Lagja e Spitalit pp, 10000 Prishtina, Kosovo. E-mail: feratsa@yahoo.com

1. INTRODUCTION

Choledochal cyst is a congenital cystic dilation of a part of bile duct that occurs most commonly in the main portion of common bile duct (1). Diagnosis of choledochal cyst is made based on a disproportionate dilation of extrahepatic biliary duct, without ruling out the possibility of a tumor, stone, or inflammation as a cause of this dilation.

Incidence of diagnosis of choledochal cyst is much higher in children (80%) than in adults (20%) (2). Symptom trias are: abdominal pain, jaundice and abdominal mass represent clinical guideline signs of diagnosis.

Furthermore, hepato-biliary diseases in adults can conceal the primary condition. In addition to this, ultrasound, CT, MRI, cholangiopancreatography (ERCP), transhepatic percutaneous cholangiography (PTC) guide us for a detailed examination in order to verify the diagnosis.

2. CASE REPORT

Our patient was a 57 year old women that presented at our clinic with abdominal pain, vomiting, fever and jaundice. The patient presented with a history of moderate abdominal pain lasting for 3 months in the right higher quadrant of the abdomen and jaundice. The patient had previous history of abdominal pain without jaundice, and was treated by a pulmonologist with the suspicion of pulmonary infiltration. In our department, during the physical examination, the patient complained of pain in the right hypocondrium radiating to her back.

No abdominal mass was palpated. The bilirubine level was 3.8 mg/dl. Ultrasound showed thickening of the gall bladder, masses of stones in different diameters and a cystic dilation of common bile duct (diameter of CBD was 4.5 cm). MRCP revealed a fusiform dilation of CBD and oedema of the wall of the gall blader, common bile duct dilation, while the liver presented to be normal. Intraoperative exploration revealed a type Ic cyst; cholecystectomy and resection of the cyst with surgical Roux-en-Y jejuno-hepatic anastomosis. The patient had a steady postoperative course.

3. DISCUSSION

Incidence of the choledochal cyst in western countries is 1 in 100,000-150,000 individuals. Rate of incidence is higher in Asia and more common in women (1m: 6f). Alonso described classification of choledochal cysts as in 3 types, which were later modified by Todani et al. presented in Table 1 (1977).

This is the most commonly used classification. A type of cyst is a fusiform dilation of common bile duct. An isolated diverticulum that protrudes through the wall is considered to be a type II of choledochal cyst. Type III is also known as choledocholesteatoma.

| Type I | Type II | Type III | Type IVa | Type IVb | Type V |
|-------|--------|---------|---------|---------|-------|
| Dilatation of extrahepatic biliary ducts. Type I of dilation is further classified according to the segment in type Ia: cystic dilation; type Ib: focal dilation of the segment; type Ic fusiform dilation. | Divertercular dilation of extrahepatic biliary ducts. | Cystic dilation of intraduodenal portion of common bile duct (choledochocele). | Extrahepatic and intrahepatic dilation of bile ducts. | Dilatation of many perotions of extrahepatic biliary ducts. | Limited dilatation of intrahepatic biliary ducts (Caroli disease). |

Table 1. Classification of choledochal cysts – modified by Todani and Alonso-Lej
choledochal cysts is unknown. An anomalous pancreatico-biliary junction resulting in a joint extremely long common bile duct has been suggested to result in antenatal pancreatico-hepatic reflux and leads to infection and dilation of the biliary tree. Choledochal cysts occur most commonly in women. Adults with initial manifestation of choledochal cysts have non-specific symptoms in the right upper abdominal region, jaundice, pancreatitis or cholangitis. A palpable mass is rare and usually associated with children.

4. CONCLUSION

The two basic treatments of choledochal cysts are enterostomy of the cyst and hepatico-jejunostomy incision. Enterotomy of the cyst is technically easy to perform, but it is related to complications like anastomosis stricture, residual calculus and malignant alteration of the wall of cyst. Hepatico-duodenostomy is technically demanding, but is linked with associated morbidity. Cholecystectomy is performed on regular basis every time when the cyst needs to be removed completely. In difficult situations, when the cyst wall is attached to the wall of hepatic artery and Portal vein, biliary technique is used for removing the cyst. With this technique, we enter the cyst through anterior opening and we perform incision on the mucosa of the cyst in the external portion of the cyst attached to blood vessels. Procedure of removing the cyst reduces the risk of biliary tree inflammation and reduces the chances of development of cholangiocarcinoma and it’s dissemination which is reported to be from 9% to 28%. Due to the high risk of a well documented cancer, cholecystectomy and resection of cysts with Roux-en-Y hepatico-jejunostomy is the best solution for the patient. Choledochal cysts in adults should be considered separate entities from those in children.

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