Choledochus cyst: a case report

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Abstract. The etiologies of choledochus cysts are not exactly known. Its classic triads are abdominal pain, mass in abdominal area and icter. A 2-year-old girl was brought in with temperature, vomiting and abdominal pain complaints. In her physical examination, there was no sensitivity, defense and rebound in the abdomen. There was also no icter in her body. In the laboratory examination aspartate transaminase and alanine aminotransferase were slightly high, gama-glutamyl transferase and alkaline phosphatase also significantly high. Her abdomen ultrasonography showed a dimensionally cystic dilatation with dimensions of 2x1,5 cm and cholecytitis was seen in choledochus proximals. In abdomen computerized tomography a dimensionally 13 mm cystic structure was observed in choledochus distal terminus adjacency associated with choledochus. Our case was orientated to pediatric surgery in terms of follow-up and therapy.

Key words: Abdominal pain, Ultrasonography, child, computerized tomography

1. Introduction

Choledochal cyst is a rare malformation, characterized by dilatation of the biliary tract. The etiologies of choledochus cysts are not exactly known. It is mostly seen in children and young adults. It is more frequent in women than in girls than boys. Its classic triad is abdominal pain, mass in abdominal area and icter. Its most common complications are stone formation, stasis, cyst rupture, abscess, pancreatitis, biliary cirrhosis, portal hypertension and malignancy (1-5). We presented a 2-year-old girl with type 2 of choledochal cyst.

2. Case report

A 2-year-old girl patient was brought in with temperature, vomiting and abdominal pain complaints lasting for 10 days. There were no pathological characteristics in her history and family tree. Her general situation was good. She was conscious and awake during her physical examination. Her body weight was measured as 12,5 kg (25-50 percentile) and her height was 83 cm (10-25 percentile). Her blood pressure was

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measured to be 90/60 mmHg. Oropharynx was hyperemic. Tonsils were hypertrophic. There was no sensitivity, defense and rebound in abdomen. Other system examinations were normal. In laboratory evaluation aspartate aminotransferase was 51 U/L (0-38 U/L), alanine aminotransferase 56 U/L (0-41 U/L), gamma-glutamyl transferase 349.5 U/L (5-66 U/L), alkaline phosphatase 1661 U/L. Complete blood count, direct and indirect bilirubin values were normal.

Renal function tests, prothrombin time and active partial thromboplastin time were normal. Sedimentation was 18 mm/hours. Total protein was 7.39 g/dL, albumin 4.19 g/dL, anti-HBs was positive in evaluation in terms of viral hepatitis and other viral markers were negative. In abdomen ultrasonography (US) choledochus was 8.5 mm in width and cystic dilatation with 2x1.5 cm dimension in proximal and contrast formation was observed in gall bladder membrane and fluid values, cholecystitis were observed in pericholecystitis area.

In abdomen computerized tomography (CT) choledochus size was 5.5 mm in proximal and slightly explicit. A cystic structure 13 mm in size was observed in choledochus distal terminus adjacency associated with choledochus (Figure 1). Our case was orientated to pediatric surgery in terms of section before possible complications and cholecytitis therapy was not developed.

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Fig. 1. In abdomen computerized tomography a 13 mm cystic structure was observed in choledochus distal terminus adjacency associated with choledochus.

3. Discussion

The etiologies of choledochus cysts are not exactly known. The etiology of choledochal cysts remains speculative. The most widely accepted hypothesis is the presence of an anomalous pancreatico-biliary ductal confluence proximal to the regulatory control of the sphincter mechanism within the duodenal wall. The condition predisposes to reflux of pancreatic juice into the bile duct, leading to activation of pancreatic enzymes and deconjugation of bile acids. The combination of activated pancreatic enzymes and deconjugated bile acids could induce chronic inflammatory changes in the bile duct and weaken portions of the biliary tree, predisposing the latter to abnormal dilatation. The observation that biliary cysts are more common in the extrahepatic biliary tree supports this hypothesis (6,7). Oligoganglionosis at the terminal portion of the bile duct causing functional obstruction and proximal dilatation has been implicated recently in the etiology (8). 60% of patients with choledochus cyst is under the age of ten (9). The frequency rate of choledochus cyst is 4:1 in girls/boys and the incidence of it is between 50.000 and 200.000:1 in births (10). Our case was a girl at the age of two compatible with literature. Mass in abdomen and icter were not seen in our patient except from abdominal pain which is a part of the classic triad. This situation can be explained by the early age of our patient. High alkaline phosphatase and gama-glutamyl transferase enzyme levels show that our patient was affected. Probably, growing body mass will be concluded with growing cyst as age proceeds and icter will come up as a result of pressure to intrahepatic and/or extrahepatic bile ducts and

mass in the abdomen can be diagnosed upon growing of cyst.

Todani classifications are used most commonly in the evaluation of choledochus cysts (11). First group is found in 80-90% of cases. Cystic dilatation of 1A choledoc is 1B focal segmental dilatation and 1C fusiform dilatation. Second type is a real choledoc diverticulum. Third type is focal dilatation of intraduodenal part of choledoc and it is named as "choledochal". Type 4 is divided into two sub-groups. 4A involves multiple intra and extrahepatic cysts and 4B involves extrahepatic cysts less than 4A. Type 5 is "Caroli disease". Single or multiple intrahepatic cysts are seen (11). According to this classification our case complies with the type 2 cyst. In a series with 26 cases which was announced from Turkey type 1 cyst has been found at a rate of 96% and type 2 cyst has been found at a rate of 4% (12).

US. endoscopic CT. retrograde cholangiopancreatography (ERCP), percutaneous transhepatic cholangiography, magnetic resonance imaging and magnetic resonance cholangiography are carried out in choledochus adequate cysts diagnosis. US provides information about the intrahepatic and extrahepatic biliary tree and is an extremely useful initial investigation. The observation of a cystic formation complying with choledochus trace in US and hepatic hilus apart from gall bladder brings the probability of a choledochus cyst to mind. However, the relation through the interways of cyst and gall should be designated and cyst should be separated from non-biliary cysts and fluid collections for an exact diagnosis. While, they are observed as masses in fluid density in abdomen CT, identifying the biliary origin of the cyst is mostly not possible (1.3.13.14).multidetector Α computed tomography with reformatted imaging is another important technique which has the ability to demonstrate the anatomic details of the biliary tree and the pancreaticobile duct union (15).

The choledochus cyst in our case, which was diagnosed in abdomen US, was affirmed in abdomen CT. Keeping choledochus cysts in mind during the retractor diagnosis of cystic lesions in hepatic portal hilus is important for early diagnosis. US, CT and magnetic resonance cholangiography provide an advantage in diagnoses of choledochus cyst cases because of their non-invasive and practical usage especially in pediatric age group for which it is commonly observed (16). An ERCP is generally regarded as the gold standard for the diagnosis of choledochal cysts and associated anomalies. However, the

ERCP has inherent morbidity because of its invasiveness. Alternatively, the magnetic resonance cholangiopancreatography is a useful, noninvasive tool which shows good overall accuracy in the detection and classification of choledochal cysts. Furthermore, the magnetic resonance cholangiopancreatography can simultaneously and effortlessly delineate both the biliary and pancreatic duct structure, whereas the ERCP may fail to delineate these structures in patients with choledochal cysts due to the possibility of the tight structure in the distal portion of the cyst. Therefore, the magnetic resonance cholangiopancreatography may be a superior diagnostic tool over the ERCP for patients with choledochal cysts (17).

The incidence of biliary tract cancer in patients with choledochal cysts increases with age (18). The ideal treatment of choledochal cyst is total excision. Choledochus cysts bear high potential necessitate surgical intervention and in consequence of complications that can go to malign degeneration. Total cyst excision and biliary enteric anastomose are performed in surgical theraphy (19,20). Laparoscopic practices have been performed successfully for choledochus cyst therapy in recent years. Also, good results have been obtained in both pediatric and adult population and they have been issued as respective groups (21). The most common complications of choledochus cysts are known as biliary cirrhosis and cholangitis associated with level and duration of obstruction following inadequate therapy and prior to surgery (22). Our case has been directed to pediatric surgery in terms of cholecystitis therapy and operation before probable complications develop.

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