Echinococcus Cyst Mimicking Choledochal Cyst in Childhood

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ABSTRACT
Choledochal cyst is a rare congenital dilatation of the common bile duct, often associated with a congenital or acquired dilatation of intra-hepatic ducts. The classic symptoms of abdominal mass, pain and jaundice is rarely seen during childhood. Some children may not show symptoms for years. In most patients the diagnosis can be confirmed by using ultrasound pictures or by injecting a radioactive substance and performing a tomography scan which gives an “image” of the abnormal duct. Echinococcus granulosus is often seen in the pediatric surgical practice in endemic areas, and present with various clinical and surgical pictures such as obstructive jaundice. We had under our care a 2-year old girl who choledochal cyst with the posterior wall of choledochus, which resulted extrahepatic ductal obstruction and choledochus displaced anterolaterally. Any suspected radiologic lesion observed in infant, or child patient in an endemic area should be evaluated for hydatic cyst. ©2008, Fırat University, Medical Faculty.  

Key words: Hydatic cyst, choledochal cyst

Laboratory findings were; hemoglobin 11.3 g/dL, white blood cell count 7850/mm³, total bilirubin 8.9 mg/dL (normal range: 0.1-1.2), “conjugate bilirubin 7.4 mg/dL (normal range: 0-0.3), alkaline phosphatase 1589 U/L (normal range: 250-1000), aspartate aminotransferase (AST) 186 U/L (normal range: 0-37), alanine aminotransferase (ALT) 214 U/L (normal range: 0-42).

Figure 1. Abdominal ultrasonography showed intrahepatic bilier ductal dilatation and cystic dilatation of choledochus.
Abdominal ultrasonography (US) showed intrahepatic biliary ductal dilatation and cystic dilatation of choledochus (Figure 1). “Computed tomography (CT) showed 6x6x5 cm cystic mass at choledochal sites” (Figure 2). Iminodiumicrtic acid (IDA) biliary system siitigraphy showed normal excretion function of hepatocit and stasis of extrahepatic biliary ductal system (Figure 3). US, CT and, siitigraphy findings were interpreted as type 1 choledochal cyst.

Postoperative second day, all laboratory data came the normal level. The patient was discharged at postoperative 8.day with albendazole therapy.

DISCUSSION

Hydatid disease caused by “Echinococcus granulosus” is encountered frequently in Mediterranean countries. More than 33% of all reported cases are from Japan, where Miyano and Yamataka have reported a prevalence of as high as 1 case per 1000 population (6). Female predominance (female/male ratio:4/1) is shown in many reports of choledochal cyst, our case report proved this. Hydatid disease can occur in all visceras and soft tissues. Involvement of organs except the liver, lungs and, the nervous system are uncommon, but such ectopic locations of the disease as the pancreas, kidney, pelvis, thyroid, heart and vertebral column have been described (5,6).

Hydatids cause symptoms because of their size, the involvement of important organs, the release of hepatic sand into the biliary channels, and their rupture. The association of jaundice and hydatid disease is unusual but has been reported to occur up to 15% of cases with hepatic hydatid cysts (3). Our patient was admitted with a two month history of jaundice, pruritis and abdominal mass to the pediatric surgery clinic. In the pediatric population, there are some cases presenting with obstructive jaundice resulting from extrahepatic bile duct obstruction caused by intrabiliary rupture of hepatic cysts and a few cases with isolated intrinsic compression of bile ducts by a hydatid cyst located around the biliopancreatic junction. The reported rate of this malignancy in patients with choledochal cysts is 9-28% (3-5). Delay in treatment can cause this lethal complications and increased morbidity in patients.

The pathogenesis of choledochal cysts is most likely multifactorial (7,8). Some aspects of the disease are consistent with a congenital etiology, others with a congenital predisposition to acquiring the disease under the right conditions. The vast majority of patients with choledochal cysts have an anomalous junction of the common bile duct with the pancreatic duct (anomalous pancreatobiliary junction). This results in inflammation and weakening of the bile duct wall. Severe damage may result in complete denuding of the common bile duct mucosa. From a congenital standpoint, defects in epithelialization and recanalization of the developing bile ducts during organogenesis and congenital weakness of the duct wall also have been implicated. The result is formation of a choledochal cyst (7).

The following discussion of the pertinent anatomy of choledochal cysts is based on the Todani classification, published in 1977 (8): Type I choledochal cysts - These are the most common, representing 80-90% of the lesions. Type I cysts are dilatations of the entire common hepatic and common bile ducts or of segments of each. They can be saccular or fusiform in configuration. Type II choledochal cysts - These are relatively isolated protrusions or diverticula that project from the common bile duct wall. Type III choledochal cysts - Also called choledochoceles, these are found in the intraduodenal portion of the common bile duct. Type IV cysts - These are characterized by multiple dilatations of the intrahepatic and extrahepatic biliary tree. Most frequently, a large, solitary cyst of the extrahepatic duct is accompanied by multiple cysts of the intrahepatic ducts. Type V choledochal cysts - These are defined multiple or soliter intrahepatic biliary cysts. This type of clustering of cysts is also known as Caroli’s disease. We found of our case as type I choledochal cyst.
Most patients with choledochal cysts have undergone abdominal US imaging prior to CT scanning (2,9). US findings suggest the diagnosis in most patients and may be conclusive in many. According to Lipsett and colleagues, CT scanning confirms an unclear diagnosis and provides information concerning the relationships of the cyst to surrounding structures (2,9,10). The authors recommended the use of MR cholangiography as the confirmatory imaging study in children with choledochal cysts because it does not require breath holding, is noninvasive, does not require the administration of contrast material, and is not associated with ionizing radiation. Hepatobiliary sintroigraphy has reasonably good accuracy in the diagnosis of choledochal cysts Before USG to diagnose choledochal cyst, percutaneous transhepatic cholangiography were definitive tests with 80-90% diagnostic accuracy (11,12). We support the use of abdominal USG, CT and biliary sintroigraphy findings (IDA) as choledochal cyst.

In our patient, giant cyst caused compression of choledochus and resulted extrahepatic ductal obstruction. Radiologic and clinical presentation of the cyst was mimicking the choledochal cyst. The cyst was showed no specific US or CT findings in our case and imaging techniques failure established the diagnosis. The diagnosis of an Echinococcus cyst is usually based on suspicion resulting from an unexpected finding. Therefore any discrete radiologic lesion observed in any infant or child patient in an endemic area should be considered a hydatid cyst. The Casoni and indirect hemaglutination tests were found to be diagnostically unreliable (12).

The pediatric surgeon may decide only to remove the cyst lining, to protect the underlying portal structures such as our patient. Once the cyst or cysts are surgically removed, the biliary duct requires reconstruction (1,8).

As a conclusion in an endemic area, hydatid disease should be considered in the differential diagnosis of all cystic masses in all anatomical locations. Isolated choledochal cyst hydatid is extremely rare in children and it causes symptoms, because of its pressure on adjacent organs, and should be simulated choledochal cyst. If the wall of choledochus is not destroyed and if there is no cystobiliary communication, enucleation is the procedure of choice. We underline prompt diagnosis and treatment to prevent late complications of disease.

REFERENCES