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CLINICAL CASE REPORT

Type I choledocal cyst: imaging diagnosis and hepaticoduodenostomy as a therapeutic measure

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ABSTRACT

Background. Choledochal cyst is a congenital cystic dilatation of the intra- or extrahepatic biliary tree. It may affect only the extrahepatic bile duct (type I, II and III), intrahepatic (type V) or both (type IVa). Diagnosis is primarily made with imaging, abdominal ultrasound being the first modality used. However, in case of a diagnostic doubt using this means, a technectium-99 HIDA scan, which has a sensitivity of 100% for type I cysts (sacciform), may be performed. It may be useful for distinguishing between choledochal cyst and biliary atresia. Computed tomography scan with contrast can also be used.

Case report. We present the case of a 2-year-old female who was admitted to the Emergency Department because of abdominal pain. She underwent abdominal USG, which reported acute cholecystitis and a cystic image adjacent to the cystic duct fibrosis consistent with a choledochal cyst.

Conclusions. The elective treatment for type I choledochal cyst is resection of the cyst with Roux-en-Y hepaticojejunostomy or hepaticoduodenostomy. The latter is considered a useful and reliable alternative in our context, although initially it fell into disuse due to reports of ascending cholangitis. However, this has not been recently documented.

Key words: choledochal cyst, technectium-99 HIDA scan, computed tomography, hepaticoduodenostomy.

INTRODUCTION

Choledochal cyst is a congenital and rare cystic dilatation of any portion of the biliary tract. It occurs with greater frequency in the main part of the common bile duct.

Choledochal cysts were described for the first time by Vater in 1723. In 1959, Alonso-Lej et al. described three types of choledochal cyst. Subsequently, Todani et al. modified this classification. There are several theories to explain the etiology of the choledochal cyst. It is believed that its origin may be related to an abnormal choledochal-pancreatic ductal union that allows chronic reflux of pancreatic enzymes into the bile duct, resulting in weakening and widening of the pathway, as well as the subsequent formation of a cyst.[1] The widely accepted system of choledochal cyst classification, which was devised by Todani et al., is based on cholangiographic morphology, location and number of intra- and extrahepatic cysts of the biliary tract (Figure 1). Type I is the most frequent (80-90%) and represent cysts that are limited to the extrahepatic biliary pathways. They are subdivided into IA (sacciform dilatation that affects all or almost all of the common bile duct), IB (sacciform dilatation that affects only one segment of the common bile duct), and IC (diffuse dilatation of the entire extrahepatic bile duct).[2] Type II or cystic diverticula and type III or choledochocele are the most rare types (2%). Type IV is second in frequency (10-15%). It is characterized by multiple cystic dilatations of the intra- and extrahepatic bile duct (IVa) or only of the ex-

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trahepatic (IVb). Type V or Caroli disease is a diffuse cystic disorder of the intrahepatic bile duct and fibrosis.[3]

There is a well-documented predominance of females (3-4:1) in the international literature. In contrast, in Mexican publications the ratio has been reported as 2:1 (male:female). In Western countries, the incidence of choledochal cyst is one case per 100,000-150,000 live births, of which 75% are diagnosed during infancy and 20% during adulthood.[4]

Abdominal ultrasound (USG) is the initial and easiest test to carry out. It allows images to be taken of the intra- and extrahepatic bile duct with measurements of the diameter of the choledochus or common hepatic duct as well as of the choledochal cyst. It shows a cystic mass in the right upper quadrant separate from the gallbladder (with the exception of type III and type V cysts).[5] In case of a suspected choledochal cyst on ultrasound but without diagnostic certainty, a cholangiogram with ^{99m}Tc-HIDA may provide more specific information. It consists of uptake of the drug at the site of the cyst followed by filling and delay in emptying. There is 100% sensitivity for type I cysts and up to 67% for type IV cysts. ^{99m}Tc-HIDA scan has less sensitivity for intrahepatic cysts. However, it can be of great use to distinguish between a choledochal cyst and biliary atresia. In biliary atresia, one can see lack of emptying into the duodenum, whereas in choledochal cyst, contrast will enter into the duodenum. In cases of cyst rupture, passage of the contrast media to the peritoneal cavity can be observed.[6] Computed tomography (CT) is useful to demonstrate continuity of the cyst with the biliary tract, its relationship with the adjacent structures, the presence and the stage of associated malignant tumors. It allows for better visualization of the intrahepatic bile ducts, the distal bile duct and the head of the pancreas. In patients with type IVa cysts and Caroli disease, it is useful for describing the intrahepatic dilatations and extension of the disease (diffuse hepatic or localized segment involvement).[6] However, it tends to underestimate the extent of dilation and does not provide information on the biliopancreatic junction.[7] Magnetic resonance cholangiopancreatography study is now considered as the gold standard for the diagnosis of choledochal cyst, with a diagnostic sensitivity of 90-100%. However, one must take into account that it is not an invasive procedure and allows for better visualization of the biliary tract.[2,5,6] With regard to the therapeutic measure, generally a cholecystectomy and resection of the cyst with bilioenteric reconstruction has been used. The most utilized is the hepatojejunal Roux-en-Y anastomosis. However, in our environment, hepatoduodenal anastomosis is done as a safe alternative with good results.[4,8]

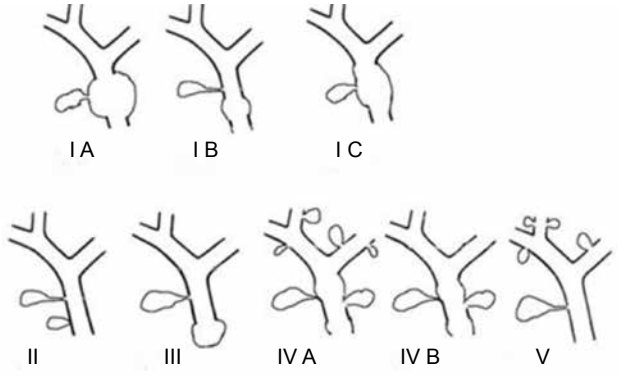

Figure 1. Todani classification for choledochal cysts.

The prognosis for type I, II, III and IVb choledochal cyst is good when patients are operated on. Follow-up is of utmost importance. A monthly examination is necessary during the first 3 months postoperative and then each 3 months for 2 years to detect the presence of cholangitis.[9]

CLINICAL CASE

We present the case of a 2-year-old female with no family hereditary or personal history of disease and without important information regarding the actual disease. According to the personal pathological history there is a report of mild traumatic brain injury 6 months prior to her admission that evolved without complications and only required observation in the Pediatric Emergency Department. She had been hospitalized on two occasions: the first occurred 2 years prior for infectious gastroenteritis; the second hospitalization was 1 year prior due to abdominal pain. On that occasion she was diagnosed with type A viral hepatitis with positive serology for IgG anti-hepatitis antibodies and was treated with ribavirin.

She was admitted to the emergency department due to abdominal pain with history of repeated episodes of abdominal pain in the right upper quadrant. She arrived at the clinic where an abdominal USG was done. Acute cholecystitis and a cystic image adjacent to the cystic duct was reported, compatible with choledochal cyst. During the previous year she presented various episodes of gastroenteritis. She was treated with amoxicillin and trimethoprim/sulfamethoxazole. The symptomatology decreased; however, abdominal

pain continued. According to parental decision, an abdominal USG was done 10 months after where multiple bladder microlithiasis was reported as well as significant dilatation of the intra- and extrahepatic bile ducts. For this reason she was hospitalized to continue with diagnostic studies and to regulate the processes to follow.

At the time of the patient's admission her vital signs were within normal limits. There was no alteration of the exterior habitus. The abdomen was soft, depressible with tenderness to deep palpation in the right upper quadrant. There were no organomegalies or masses identified. Peristalsis was established without data of peritoneal irritation.

Baseline analysis showed increase in bilirubin and transaminases: total bilirubin (TB) 3.07 mg/dl, direct bilirubin (DB) 1.78 mg/dl, indirect bilirubin (IB), 1.29 mg/dl, TGP/ALT 349.7 U/L, TGO/AST 140.4 U/L.

A new abdominal USG was done where no bile duct pathology was noted. Therefore, due to the discordance with the previous results, ^{99m}Tc-HIDA cholangiogram was performed (Figure 2). This study reported the liver to be normal in size, shape, and location. A heterogeneous pattern of collection and distribution of the radiopharmaceutical in the hepatic parenchyma was observed. Elimination of the material towards the intestine was delayed

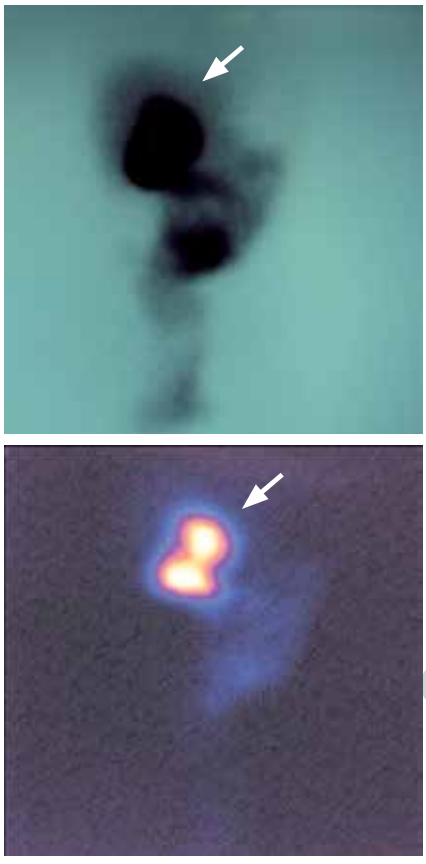

Figure 2. ^{99m}Tc-HIDA cholangiogram.

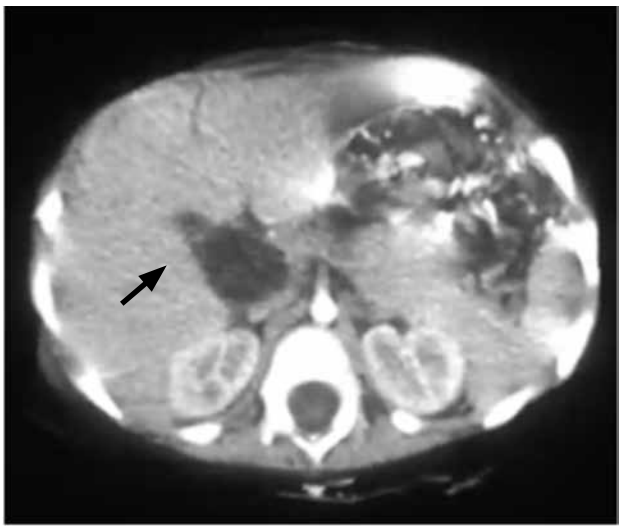

Figure 3. Contrast computed tomography (CT) of abdomen. Arrow: cyst.

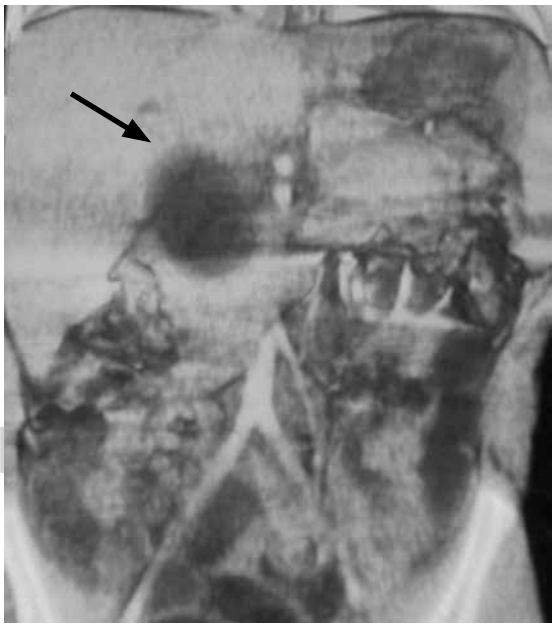

Figure 4. Contrast CT of abdomen. Arrow: cyst.

and partial, showing elimination in the post-food stimulus, suggesting an obstructive pattern.

CT of the abdomen with contrast (Figure 3) demonstrated a tumor at the level of the choledochus, which is not enhanced with contrast media and suggests a choledochal cyst without dilatation of the intrahepatic bile duct (Figure 4).

Surgical treatment was carried out with cyst resection, cholecystectomy and biliodigestive bypass with hepatoduodenal anastomosis (Figure 5).[4] Surgical findings were a type I common hepatic choledochal cyst (Figure 6) with anastomotic mouth of 13 mm, ~2 cm in length from the portahepatis (Figure 7). The pathology report was negative for malignancy. Postoperative management included fasting for 5 days, nasogastric tube, proton pump inhibitor, ketorolac, cefotaxime and metronidazole. At 6 months of evolution the patient did not present a clinical picture of cholangitis.

DISCUSSION

We can attest to the fact that USG is a very important auxiliary method for diagnosis of type I choledochal cyst. However, one of its limitations is that it depends, in large part, on the expertise of the radiologist. For this reason, and due to diagnostic doubt, two imaging studies were also evaluated: contrast CT of the abdomen and ^{99m}Tc-HIDA cholangiography because they are more sensitive and specific than USG and are not as operator-dependent for arriving at an accurate diagnosis.

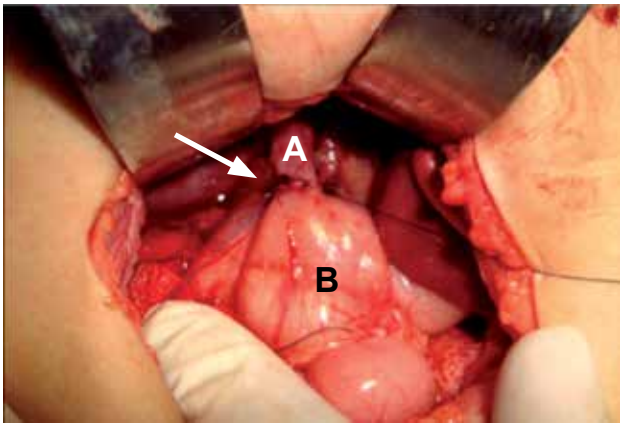

Figure 5. Hepaticoduodenal anastomosis. **(A)** Common bile duct. **(B)** Duodenum. Arrow: anastomosis.

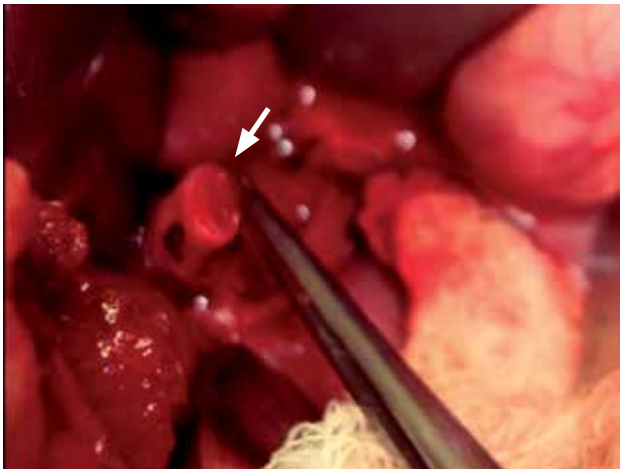

Figure 7. Arrow: mouth of anastomosis of common bile duct.

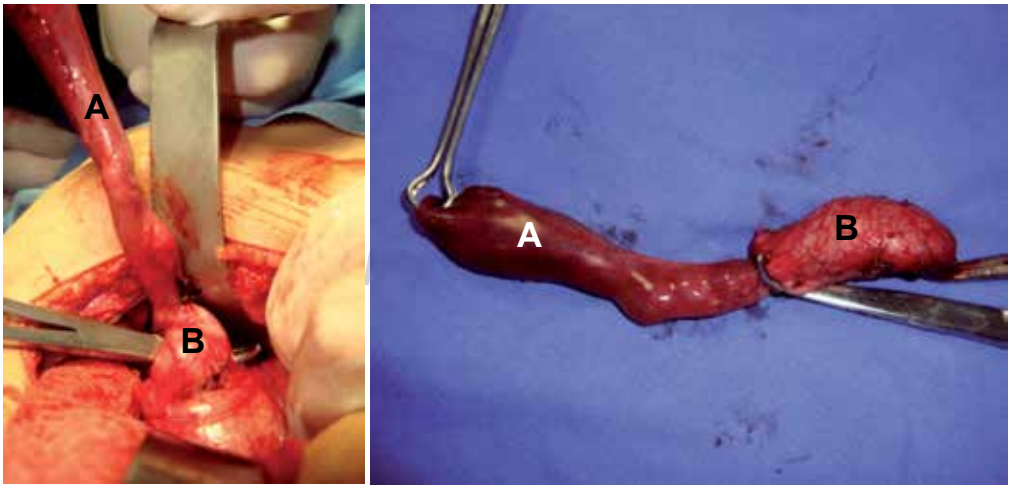

Figure 6.

(A) Gallbladder. **(B)** Choledochal cyst.

When comparing the cholangiogram with CT, it was found that the former helps to determine if there is adequate hepatic and biliary function, it has a sensitivity of 100%, is less costly and is a fast and simple procedure not requiring any special preparation. Risk of presenting secondary adverse effects from the study is very low. CT is useful for arriving at the diagnosis because it evaluates the intra- and extrahepatic ducts, adjacent structures and the presence and stage of associated malignancies. However, disadvantages are adverse effects to the contrast media and exposure to radiation.

With regard to treatment, we believe that hepatoduodenal anastomosis is a surgical technique that today can be considered for the management of patients with type I, II, III and IVb choledochal cysts. It has advantages such as decreasing surgical time, allowing for a more rapid recovery of the intestinal function, and causing fewer complications that require reintervention.[8] In addition, the number of anastomoses performed are less, it is an easily reproducible technique and, according to the results of Gallardo et al., the presence of cholangitis is practically zero as shown in the monitoring of their cases and, so far, in this patient.[4]

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