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Case report

CHOLEDOCHAL CYSTS: THE DIAGNOSTIC RELIABILITY OF ULTRASOUND

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ABSTRACT

Choledochal cysts are rare cystic dilatations of the bile ducts, which are commonly found in children. We report two young patients affected by this anomaly. The clinical symptoms of the patients were minimal: mild pain localized in the upper-right quadrant and a serum increase in transaminases. Both of the patients were affected by type I choledochal cysts (B and A, respectively, according to the Todani classification). The diagnosis was made using abdominal ultrasound and then confirmed by MRI. We wish to emphasize the relevant contribution of ultrasound as an easy, reliable, rapid, low-cost modality for the diagnosis of choledochal cysts.

1. Introduction

Choledochal cysts (CCs) are extremely rare congenital disorders which occur largely during childhood, and are more common in Asian populations with an incidence of 1 in 13,000 versus 1 in 100,000 in Western populations [1]. Females are at higher risk for the disease with a nearly 4:1 female preponderance compared with males [2]. Choledochal cysts are congenital abnormalities consisting of dilatation of the intra and/or extrahepatic biliary ductal system. The etiopathogenesis of this anomaly is widely debated. There are many theories that try to explain its pathophysiology. The most widely accepted hypothesis is Babbitt's theory, which states that cystic dilation of the bile ducts is related to an anomalous arrangement of the pancreaticobiliary ductal junction [3]. This anomaly promotes the reflux of pancreatic fluid into the biliary tree, thereby supporting infections and the destruction of the bile duct wall accompanied by the formation of cysts [4]. Most of the reported cases have a normal intra- and extrahepatic biliary ductal system, which indicates distinct pathology. The reason for isolated involvement of the cystic duct excluding the common bile duct (CBD) is unclear.

It is possible that the junction of the cystic duct with CBD is the weakest part, due to low vascularity, causing an ecstatic change that continues as a vicious cycle, resulting in further dilatation [5]. Other etiopathogenetic hypotheses have been proposed, such as defective neurons and ganglion innervations, sphincter of Oddi dysfunction, and distal obstruction of the common bile duct (CBD) [6]. Here, we report two young patients affected by CCs and we emphasize the use of ultrasound (US) as a reliable and low-cost modality to ensure a correct and rapid diagnosis.

2. Case presentations

Case 1

A 5-year-old girl from North Africa visited us presenting with vomiting and abdominal pain located in the upper-right quadrant. No information regarding family history, maternal gestation, or neonatal period was reported. Her physical examination yielded good results. Her head circumference was 51 cm, her height was 102 cm and her weight was 18 Kg, all within normal limits.

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No anomalies were found upon physical examination of her heart, eyes, ears, pulmonary apparatus, spleen and liver. Her abdomen was soft and slightly painful; no masses or hernias were reported. A neurological examination was also normal. Laboratory tests revealed no alterations in blood cell count, erythrocyte sedimentation rate, reactive protein, or ammonia and lactate. Her serum glutamic oxaloacetic transaminase value was 1300 U/L, and her serum glutamate pyruvate transaminase value was 747 U/L. The patient's total bilirubin and amylase were normal. A complete abdominal US revealed normal echogenecity and echotexture of the liver with no crosses. An intrahepatic biliary ductal dilatation with a small cystic mass (3×2 cm in diameter) originating from the CBD was found (Figure 1 a and b) . No other abdominal anomalies were found in the patient's gallbladder, pancreas or kidneys. We performed a cholangiopancreatography MRI (Figure 2) which confirmed the presence of a type 1 B CC. The girl was sent to surgical treatment for extrahepatic biliary tree removal, hepato-jejunal anastomosis to Roux anse, and dijuno-dijunals anastomosis. At follow-up, the girl showed good, rapid improvement with gradual normalization of her transaminases. At the 35-week follow-up, her abdomen ultrasonography yielded normal results.

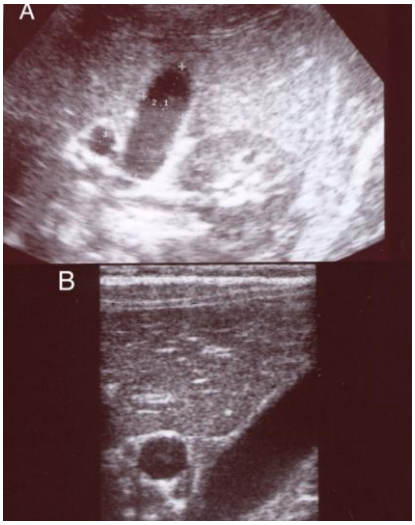

Figure 1. (A and B). US of case 1 showing intrahepatic biliary ductal dilatation with the small cystic mass.

Figure 2. Cholangio-MRI showing the presence of the small choledoctal cyst type 1 B

Case 2

We examined a 4-year-old girl who first came to our attention for addominal pain localized in the upper-right quadrant and episodes of somnolence . The family history was not relevant. The girl was born at 39 weeks of gestation by spontaneous delivery with a birth weight of 3200 g, a length of 50 cm and a head circumference of 35 cm. Her Apgar score was 9 and 10 at 1 and 5 minutes, respectively. The perinatal period was uneventful and the stages of psychomotor development were normally attained. Upon physical examination, she revealed to be in good condition. Palpation showed that her liver was mildly painful. The remainder of the physical examination was normal. The patient's weight and height were within normal limits. Laboratory tests revealed an increase in serum glutamic oxalatetic transaminase with a value of 340 U/L and serum glutamate pyruvate of 240 U/I. Hemogram, blood glucose and total bilirubin were normal, and an amylase test was normal. Upon US examination, we found a large, ovoidal cyst 47×24 cm in diameter with well-delimitated borders and an asonic content (Figure 3). According to Todani classification, the patient's CCs were type I A2. The CC diagnosis was confirmed by colangio-RM. The patient underwent surgical intervention for the removal of her cysts, and she experienced complete recovery. At a 41-week follow-up, no abdominal anomalies were found upon US sonography.

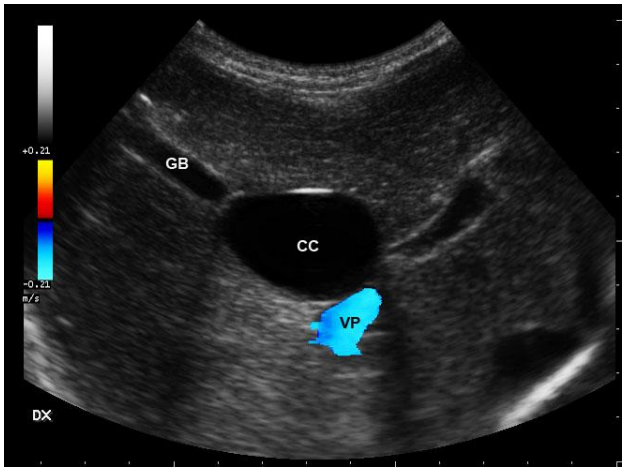

Figure 3. US of the case showing the large cyst (47x24 in diameter with asonic content and delimitated borders).

3. Discussion

Choledochal cysts (CC) are a rare congenital cystic dilation of the biliary tract, first described by Vater and Ezler in 1723 [7]. They present primarily in female infants and young children, and are more prevalent in East Asian populations. Common presentations include abdominal pain, jaundice, and right upper quadrant mass. In our patients, the clinical presentation was not impressive. Both of the girls had pain that was localized in the upper-right abdominal quadrant, but without signs of jaundice or other clinical signs; no masses were palpable in their abdomens and laboratory analyses only revealed an increase of transaminases. Choledochal cysts may appear as isolated anomalies, but they can sometimes be associated with other congenital malformations including double CBD, sclerosing cholangitis, congenital hepatic fibrosis, pancreatic cysts and annular pancreas [8].

In a nationwide study, congenital cardiac anomalies occurred in 31% of pediatric patients with CC and are most commonly manifested in infancy [9]. Biliary malignancies are rarely observed with CCs in childhood.

Different classifications of CCs have been proposed. The most widely accepted current classification is that of Todani et al. based on the site of cystic change [10]. According to this system, five subtypes of CCs may be distinguished. In type I CCs, the cysts communicate with the biliary tract. Type 1 cysts can be subdivided into type 1 A, 1 B and 1 C, depending on the relationship between the gallbladder and the cystic duct location. In type I A cysts, the gallbladder rises from the CC and the extrahepatic biliary tree appears dilated. In type I B cysts, the extrahepatic biliary tree is normal with a dilatation involving the most distal area of the CBD. Type I C cysts are characterized by a smooth fusiform dilatation of the CBD with pancreaticobiliary malocclusion. Type II cysts are less frequent, and their anomalies consist of extrahepatic duct diverticula. Type III cysts have a intraduodenal location in the pancreatic-biliary junction, and type IV cysts can be subclassified as IV A and B. In the first case, the dilatation extends from the CBD to the common hepatic duct into the intrahepatic biliary tree; in type IV B cysts, there are multiple dilatations of the extrahepatic biliary tree. In type V cysts, intrahepatic saccular or fusiform dilatation are present. Among all types of CCs, the most frequent are type I, which constitute 80–90% of all CCs. In our patients, the first patient had a type I B cyst, and the second patient had a type I A cyst. The diagnosis was made with the use of abdominal US. This modality has been extremely useful for diagnoses because it is low cost and reliable. Additionally, this modality doesn't produce any side effects and can be performed easily. Ultrasound has been shown to be highly specific in identifying gallbladder anomalies. It has a sensitivity of 71–97% for detecting CCs [11]. Furthermore, according to a large study by Formy et al. [12], abdominal US demonstrated a sensitivity of 56.6% in 30 cases of CCs reported by the authors with diagnostic definition in 17 children. Confirming the presence of CCs after US with magnetic resonance cholangiopancreatography (MRCP) may be useful, as it is both non-invasive and raditation free. For many years, surgical treatment for CCs consisted of cystenterostomies. However, this approach was criticized for the malignancy complications linked to cyst wall residue. For type 1 cysts, surgical treatment consists of complete extrahepatic bile duct cyst excision down to the level of communication with the pancreatic duct, cholecystectomy, and restoration of bilioenteric continuity. More recently, patients treated with laparoscopic re-section of the cyst with hepatico-dijunostomy have exhibited good results [13].

Although malignancy is rare, CC resection does not reduce it to baseline levels, so long-term surveillance is recommended, given the increased likelihood of developing post-excision biliary malignancy.

4. Conclusion

Choledochal cysts are rare cystic dilatations of the bile ducts, which are commonly found in children. The diagnosis typically occurs using multi-modality imaging including ultrasound, CT, and MRI, including MRCP. Clearly, the US evaluation cannot replace performing additional, more sophisticated diagnostic radiologic investigations, but US remains a rapid and reliable modality in identifying abdominal masses.

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References

1. Nittis L, Soares KC, Arnaoutakis DJ, Kamel I, Rastegar N, Anders R, Maithel S, Pawlik TM. Choledochal cysts: presentation, clinical differentiation, and management. J Am Coll Surg 2014;219(6):1167–1180
2. Lipsett PA, Pitt HA. Surgical treatment of choledochal cysts. J Hepatobiliary Pancreat Surg 2003; 10(5):352–359.
3. Babbitt DP, Starshak RJ, Clemett AR. Choledochal cyst: a concept of etiology. Am J Roentgenol Radium Ther Nucl Med 1973;119(1):57–62.
4. Altıntoprak F, Uzunoğlu MY, Dikicier E, Zengin İ. Choledocal Cyt-Classificaton, Physiopathology, and Clinical Course 2016;3:588–592.
5. Singham J, Yoshida EM, Scudamore CH. Choledochal cysts: part 1 of 3: classification and pathogenesis. Can J Surg 2009;52(5):434–40.
6. Hill R, Parsons C, Farrant P, Sellars M, Davenport M. Intrahepatic duct dilatation in type 4 choledochal malformation: pressure-related, postoperative resolution. J Pediatr Surg 2011;46:299-303.
7. Vater A, Ezler C. Dissertatio de Scirrhis viserum occasione sections viri tymponite defunte. Wittenburgae Pamphlets 1723;4:22.
8. Xie XY, Strauch E, Sun CC. Choledochal cysts and multilocular cysts of the pancreas.Hum Pathol 2003;34:99-101.
9. Murphy AJ, Axt JR, Lovvorn HN. 3rd Associations between pediatric choledochal cysts, biliary atresia, and congenital cardiac anomalies. J Surg Res 2012;177:59–63.
10. Todani T, Watanabe Y, Narusue M, Tabuchi K, Okajima K. Congenital bile duct cysts: Classification, operative procedures, and review of thirty-seven cases including cancer arising from choledochal cyst. Am J Surg 1977;134:263-9.
11. Subramony R, Kittisarapong N, Barata I, Nelson M. Choledochal Cyst Mimicking Gallbladder with Stones in a Six-Year-Old with Right-sided Abdominal Pain. West J Emerg Med 2015;16:568-71.
12. Forny DN, Ferrante SM, Silveira VG, Siviero I, Chagas VL, Méio IB. Choledochal cyst in childhood: review of 30 cases. Rev Col Bras Cir 2014;41:331-5.
13. She WH, Chung HY, Lan LC, et al. Management of choledochal cyst: 30 years of experience and results in a single center. J Pediatr Surg 2009;44:2307–2311.