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Choledochal cysts: the diagnostic reliability of ultrasound

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Summary

We report two young patients affected by choledochal cysts. The clinical symptoms of the patients were minimal: mild pain localized in the upper-right quadrant and a serum increase in transaminases. The patients were affected by choledochal cysts type 1 B and type 2, respectively, according to the classification of Todani. The diagnosis was made using abdominal ultrasound (US) and then confirmed by MRI (Magnetic Resonance Imaging). We wish to emphasize the relevant contribution of ultrasound as an easy, reliable, rapid, low-cost modality for the diagnosis of choledochal cysts. Clearly the ultrasound evaluation can't get out of performing more sophisticated radiologic procedures.

Key Words

Ultrasound; Choledochal Cyst; Magnetic Resonance Imaging.

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Introduction

Choledochal cysts (CCs) are uncommon clinical manifestations that occur largely during childhood. In affected individuals, a diagnosis of CCs is made prior to an age of 10 in 80% of cases (Yamaguchi, 1980; Kim et al., 1995; Wiseman et al., 2005). The anomaly is more frequently reported in females with a male: female report ratio of 1:4. The incidence of CCs ranges from 1 in 100,000 individuals to 1 in 150,000 individuals, but the prevalence of CCs has been reported to be higher in Japan (Yamaguchi, 1980; Sato et al., 2001).

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. It has been hypothesized that the anomaly arises during the phase of pancreatic development in which the ventral and dorsal buds rotate, fuse and insert into the biliary tree. A new etiopathogenetic hypothesis on the origin of CCs has been advanced on the basis of the presence of an anomalous congenital pancreaticobiliary duct union (APBDH) outside the duodenum in 30–70% of patients with CCs (Babbitt, 1969; Babbitt et al., 1973; Komi et al., 1977; Iwai et al., 1992; Cha et al., 2008). The large common channel (APBDH) is believed to be secondary to the arrest in migration of the choledochopancreatic junction into the duodenal wall. 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.

As reported by Soares et al. (2014), other etiopathogenetic hypotheses have been advanced such as a weak bile duct wall, defective neurons and ganglions innervations, sphincter of Oddi dysfunction, and distal obstruction of the common bile duct (CBD) (Alonso-Lej et al., 1959; Hill et al., 2011). Since the first report by Alonso-Lej et al. (1959) and then by Fornj et al. (1977), different classifications of CCs

have been proposed; the most widely accepted current classification is that of Todani et al. (1977).

Here, we report two young patients affected by CCs type I B and type 2, respectively, according to theTodani classification. We emphasize the use of ultrasound (US) as a reliable and low-cost modality to ensure a correct and rapid diagnosis.

Case Reports

Case 1

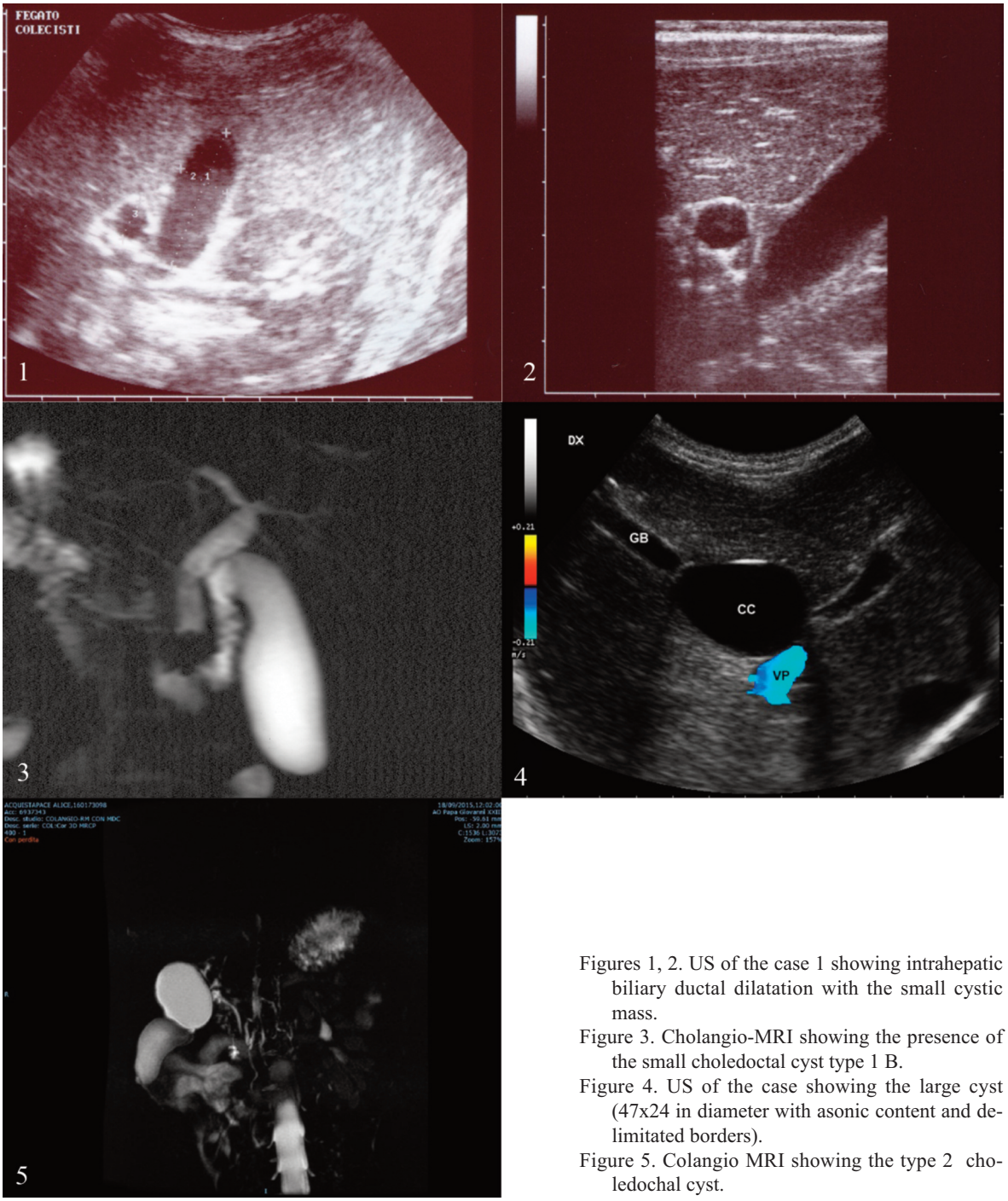
A 5-year-old girl from North Africa visited us based on vomiting and abdominal pain located in the upper-right quadrant. No information regarding family history, maternal gestation, or neonatal period was reported. Her physical examination yielded good results. Her head circumference 51 cm, her height was 102 cm and her weight was 18 Kg, all within normal limits. 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 bilirubine 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 (3×2cm in diameter) originating from the CBD was found (Fig. 1, 2) . No other abdominal anomalies were found in the patient's gallbladder, pancreas or kidneys. We performed a cholangiopancreatography MRI (Fig. 3) that confirmed the presence of a type 1 B CC. The girl was sent to surgical treatment for extrahepatic biliary tree removal, hepato-dijiunal 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 3-years follow-up, her abdomen ultrasonography yielded normal results.

Case 2

We examined a 4-year-old girl who first came to our attention at the Emergency Section of the Vittorio Emanuele Hospital 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 gr, 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 was 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 inserum glutamic oxalacetic transaminase with a value of 340 U/L and serum glutamate pyruvate of 240 U/l. Hemogram, blood glucose and total bilirubine 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 (Fig 4). According to Todani classification (Todani et al., 1977), the patient's CCs was suspected to be type 2. The CC diagnosis was confirmed by colangio-RM. The patient received surgical intervention for the removal of her cysts, and she experienced complete recovery. At a 4-year follow-up, no abdominal anomalies were found upon US sonography.

Discussion

Both girls had pain that was localized in the upper-right abdominal quadrant without signs of jaundice or other clinical signs; no masses were palpable in their abdomens. The CCs reported in the patients were type 1B and type 2 according to the classification of Todani et al. (1977). According to this classification, 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 IA cysts, the gallbladder arises 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 IC 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 intrahepa-



Figures 1, 2. US of the case 1 showing intrahepatic biliary ductal dilatation with the small cystic mass.
Figure 3. Cholangio-MRI showing the presence of the small choledoctal cyst type 1 B.
Figure 4. US of the case showing the large cyst (47x24 in diameter with asonic content and delimitated borders).
Figure 5. Colangio MRI showing the type 2 choledochal cyst.

tic 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 (Soares et al., 2014).

Among all types of CCs, the most frequent are type I, which constitute 80–90% of all CCs. In our patients, we classified the CCs according the subtype of Todani et al. (1977) the first patient had a type IB cyst, and the second patient had a type 2 cyst, which is of rare observation.

The clinical presentation of CCs includes abdominal pain, jaundice and an upper-right quadrant mass. Choledochal cysts may appear as isolated anomalies, but they can sometimes be associated with others congenital malformations including double CBD, sclerosing cholangitis, congenital hepatic fibrosis, pancreatic cysts and annular pancreas (Crittenden & McKinley, 1985; Xie e al., 2003). Biliary malignancies are rarely observed with CCs in childhood. In our patients, the clinical presentation was not impressive. Abdominal pain increased suddenly, and laboratory analyses only revealed an increase of transaminases. The diagnosis was made with the use of abdominal US. This modality has been extremely useful for diagnosis because it is low cost and reliable. Moreover, this modality is free of 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 (Visser et al., 2004; Forny et al., 2014; Subramony et al., 2015). Confirming the presence of CCs may be useful after US to perform magnetic resonance cholangiopancreatography that is both not invasive and free of irradiation. According to a large study by Forny et al. (2014), abdominal US demonstrated a sensitivity of 56.6% among 30 cases of CCs reported by these Authors with diagnostic definition in 17 children.

For many years, surgical treatment for CCs consisted of cyst enterostomies. However, the malignancy complications linked to cyst wall residual resulted in criticism. 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 resection of the cyst with hepaticodijunostomy have exhibited good results (Soares et al., 2014).

Clearly the US evaluation can't get out of performing others more sophisticated diagnostic radiologic investigations, but US as widely demonstrated remains a rapid and reliable modality in identifying abdominal masses.

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