Infant Cystic Duct Cyst - Case Report and Literature Review

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Abstract

The cystic malformation of the cystic duct is a rare disease and was first reported by Bode and Aust in 1983. There were several different nomenclatures since then, including cyst of the cystic duct. Rarely diagnosed in adult life, this case occurred in infants and evolved with perforation of the common hepatic duct. This malformation manifests as abdominal pain, jaundice, cholangitis, choledocholithiasis, and pancreatitis. Retrograde cholangiopancreatography is the gold standard for diagnosis; however, it is not uncommon to be an intra-operative finding. The treatment is surgical with excellent results. In this case, we highlight the importance of imaging exams in the evaluation, describing an infant with a cystic malformation of the cystic duct with perforation of the common bile duct.

Keywords: Children; Congenital Biliary Dilatation; Cystic Duct; Bile Duct Diseases; Case Report

Abbreviations
CT: Computerized Tomography; MR: Magnetic Resonance; US: Ultrasound

Introduction

Brode and Aust described the first case report of a cystic duct cyst alone in 1983, in a seven years old child [1]. In 1987, Champetier, et al. reported two cases [2] and only in 1991, Serrena, et al. described one case and suggested that anatomic variation should be included in Todani's classification as type VI [3]. Loke, et al. proposed that this lesion would be a type II variant [4]. The choledochal cysts are congenital abnormalities with intra and/or extrahepatic bile ducts dilatations, with a prevalence between 1:100,000 to 1:150,000 live births in the western countries, with a female predisposition in a proportion of 3 to 4:1 and usually diagnosed in the first years of life [5]. The cystic duct cyst alone is extremely rare, and there are few cases reported in the literature [6]. Some authors propose to include on Todani's classification as type VI and others as a variant of type II [7]. The most accepted theory is that it is a result of the combination of a biliopancreatic junction anomaly, with an acute angulation, and a large implantation orifice of the cystic duct in the hepatic duct. Other factors also reported as possible causes of the dilatation of the cystic duct are a congenital stenosis or an aganglionar segment of the bile duct with a proximal dilatation [8].

Materials and Methods

We retrospectively collected data of one patient from the clinical records between 2018 and 2019 at Conjunto Hospitalar do Mandaqui, Sao Paulo, Brazil. Ethics committee approval by Secretaria de Saude do Estado de Sao Paulo (Plataforma Brasil no 30619220.0.0000.5551). We obtained the informed consent from both patient’s parents and followed the SCARE guideline in this report [9].

Case Report and Discussion

A female infant, eight months old, white, started with vomiting for seven days, mainly after eating. After three days, she started with liquid diarrhea. She had no previous episode. She was in non-exclusive breastfeeding. She was born at term without pregnancy complications, no similar family history. In physical examination, she was in regular general condition, anicteric, afebrile, active, and reactive, weighting 8 Kg heart rate of 100 beats per minute, 20 breaths per minute, 98% oxygen saturation. Abdomen without collateral circulation, without scars, without bulging, normotympanic, normal bowel sounds, with diffuse pain but no peritoneal reactivity, no palpable masses, but positive fluid wave test. At admission the laboratory exams were: hemoglobin - 9,6 g/dL, amylase 59 U/L, direct (conjugated) bilirubin - 1,7 mg/dL and indirect bilirubin - 0,53 mg/dL, alkaline phosphatase - 589 U/L e γ-gt - 1220 U/L; abdomen US showing dilation of intrahepatic bile ducts, with an anechoic cystic image with well-defined limits over the portal vein and below the gallbladder, measuring 1,1 cm x 1,2 cm and moderate quantity of ascites. During the investigation, the patient evolved with a better general condition, without new episodes of vomiting, diarrhea, or abdominal pain, but started with progressive jaundice, with an increase of conjugated bilirubin. CT with intravenous contrast of the abdomen showed a considerable amount of free liquid in the abdominal cavity and a distended gallbladder with no well-defined limits at the infundibular site, highlighting a cystic formation between the gallbladder and the common bile duct, reaching near 3,3 cm (Figure 1).

![Figure 1: A: CT scan with cystic duct cyst (*) and cystic duct (***); B: gallbladder (***); C: common duct perforation (arrow sign).](image)

We perform a diagnostic video laparoscopy and found a considerable quantity of free bile in the cavity, fibrin near the choledochus and an extensive inflammatory process over the bile ducts (Figure 2), opted for conversion to open laparotomy. We found a cystic duct cyst and perform an intra-operatory cholangiography (Figure 3).

During the dissection of the gallbladder, it was observed a dilated cystic formation anteriorly to the choledochus, confirmed by another cholangiography, that showed that was a segment of the cystic duct after this dilation, with usual caliber, connected to choledochus. It was also evident contrast overflow to the abdominal cavity from the proximal common hepatic duct (Figure 4 and 5).

**Figure 2:** Video laparoscopy, with a large amount of fibrin on the bile duct.

**Figure 3:** Intraoperative cholangiography showing the gallbladder (*), the cystic duct cyst (**) and the bile duct.
Figure 4: Deep cystic gallbladder dissection (*) with long cystic duct (**) and the appearance of cystic formation inside the cavity (***)

Figure 5: Cholangiography after gallbladder removal with an image of the main bile duct (*), the cystic duct cyst (**) and leakage of contrast into the cavity through the perforation of the common hepatic duct (***)

A cholecystectomy, including the cystic duct cyst, was performed (Figure 6) with the closure of common hepatic duct perforation. We drain the abdominal cavity with a tubule-laminar drain, and a Kehr drain in the choledochal duct. The post-operative was uneventful, with hospital discharge on the 10th postoperative day with the Kehr drain closed. At follow-up, we withdrew the Kehr drain at the 24th postoperative day, with the patient asymptomatic.

The classic triad of biliary cysts is jaundice, pain, and a palpable mass in the right hypochondrium, present in 6 - 38% of cases. In children, it is possible to find biliary ascites due to spontaneous or traumatic perforation of these cysts, and at long term follow up, we can find about 2,5 to 39,4% of biliary carcinoma [10]. At this time, in medical literature, there are few cases reports in infants that presented with cystic duct cyst. In our search, we achieved the following papers (Table 1). In this case report, we noted that, differently than other cases described in infants, there was ascites at first presentation, justified by the intraoperative finding of a common hepatic duct perforation in the medial position, 0.5 cm from the insertion of the cystic duct, which again surprises us, because in the literature the most common perforation described is in the cyst wall [11].

In most cases, the cause of perforation of the common hepatic duct remains unknown. Theoretically, the etiology of this perforation includes a bile duct with intrinsic flaccidity (aganglionosis), ischemia on perforation site due to congenital vascular anomaly [12], anomalous biliopancreatic junction [13] and distal biliary obstruction due to biliary or biliopancreatic malformation [14]. The most usual site, of the biliary tree perforations, is in the junction of the cystic duct with the common hepatic duct. It makes us believe that due to the position of the cystic duct, in this case, it compressed the common hepatic duct, leading to increased pressure inside it and associated with ischemia by extrinsic compression, led to a perforation.

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<table>
<thead>
<tr>
<th>Author</th>
<th>Age</th>
<th>Clinical condition</th>
<th>Image</th>
<th>Subtype and Associations</th>
<th>Treatment</th>
</tr>
</thead>
<tbody>
<tr>
<td>Maheshwari et al.</td>
<td>6 days</td>
<td>Jaundice, vomiting, bloating</td>
<td>Ultrasound and magnetic resonance cholangiography</td>
<td>Fusiform expansion; associated gastric perforation</td>
<td>Not specified</td>
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<td></td>
<td>4 months</td>
<td>Not specified</td>
<td>Ultrasound and magnetic resonance cholangiography</td>
<td>Fusiform expansion</td>
<td>Not specified</td>
</tr>
<tr>
<td>Youn., et al.</td>
<td>4 months</td>
<td>Abdominal pain, vomiting</td>
<td>Ultrasound and magnetic resonance cholangiography</td>
<td>Fusiform expansion and anomalous junction of the pancreatic duct</td>
<td>Video laparoscopic cholecystectomy converted</td>
</tr>
<tr>
<td>Kotzias C., et al.</td>
<td>8 months</td>
<td>Abdominal pain, vomiting, jaundice, and ascites</td>
<td>Ultrasound and Computerized Tomography</td>
<td>Fusiform expansion; Perforation of common hepatic duct</td>
<td>Video laparoscopic cholecystectomy converted and raffia of perforation of the common hepatic duct with biliary drainage</td>
</tr>
</tbody>
</table>

Table 1: Literature review: Infant cases of cystic duct cyst.

Imaging exams assist in the diagnosis and surgical planning, and it is crucial to know the location and size of the cystic duct. At the beginning of the investigation, the abdominal US can suggest the presence of a cystic mass between the cystic duct and the gallbladder, also is capable of showing cholelithiasis and dilation of the intra and extrahepatic bile ducts [15]. The MR cholangiography is indicated for better evaluation of the dimensions of the bile ducts and differential diagnoses [15]. The difficulty of doing this exam in infants is the time to accomplish and the necessity of sedation. The retrograde cholangiopancreatography is considered the gold standard for diagnosis [15] but is not used routinely due to its invasive approach, and in the pediatric population, another difficulty is the appropriate size of the necessary material for the exam.

The cystic duct, in the adult, has a maximum diameter of 5 mm and its dilations can be saccular or fusiform, the latter being the more common variation [8]. In infants, we cannot define the standard size of the cystic duct. The treatment for this lesion is surgical excision, which not only will relieve the symptoms, but also decrease the future malignization risk [11]. Cholecystectomy with cystic duct cyst resection is the recommended procedure, but if the lesion is too close to the common hepatic duct, Ray S., et al. suggest, perform a biliodigestive reconstruction, for complete resection [6]. After surgery, follow-up is required because there is a risk of malignant transformation of 0.5 to 6% due to a residual lesion or subclinical neoplasm [14].

Conclusion

The cystic duct cyst is a rare anatomic abnormality, with difficult diagnoses, although symptomatic and relevant due to the risk of future malignancy. We believe it represents the same features of choledochal cysts and advocates a simple resection as the treatment of choice as an open or laparoscopically assisted cholecystectomy. If this malformation compromises the junction of the cystic duct with the common hepatic duct, we believe the best option would be a Roux-en-Y hepaticojejunostomy. The diagnoses can be clinically suspected, even in infants, and defined by images, despite the inherent difficulties of the patient size. We should not delay the treatment after the diagnosis and must do a close follow-up.

Conflict of Interest

The authors state that there is no conflict of interest related to this case report.