Choledochal cyst- unusual presentation in the adult phase: Case report

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ABSTRACT

INTRODUCTION: Bile duct cysts are congenital malformations characterized by bile duct dilatation with intra and/or extrahepatic localization. About 80% of cases are diagnosed in childhood, so their presentation in adults is rare and repeatedly associated with complications.

PRESENTATION OF CASE: A 38-year-old female patient complained of severe abdominal pain for one month, with no associated cholestatic signs. She underwent abdominal ultrasonography that showed cholelithiasis with extrahepatic bile duct dilatation and underwent cholangiography (MRI), which revealed fusiform dilatation of the proximal portion of the hepatobiliary ductal system, compatible with choledochal cyst.

DISCUSSION: In adult patients, clinical manifestations are usually nonspecific, and the diagnosis is established by medical imaging. Magnetic resonance cholangiopancreatography (MRCP) is the gold standard diagnostic today. Treatment with better prognosis consists of total cyst resection with hepaticojejunostomy and intestinal transit reconstruction with Roux-en-Y.

CONCLUSION: Bile duct cysts are a rare condition in adults. Currently, increasing access to more accurate medical imaging enables early diagnosis, contributing to the use of therapeutic methods in appropriate time, avoiding development of complications.

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1. Introduction

Bile duct cysts are congenital malformations characterized by biliary duct dilatation with an intra and/or extrahepatic localization. There are incidences of 1:100,000 to 150,000 live births in the West and 1:1,000 in Asia, being most prevailing in female at a ration from 3 to 4:1 [1–8]. Around 80% of cases are diagnosed in childhood, then, the presentation in adults is rare and it is repeatedly associated with complications such as cholangitis, stone formation, cyst rupture, secondary biliary cirrhosis, obstructive jaundice and malignancy (cholangiocarcinoma) [1,3,9,10]. The typical clinical presentation features abdominal pain, jaundice and palpable mass in the right upper quadrant. However, in adults the presentation can be difficult with nonspecific abdominal pain, under considera-

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recent weight loss. Physical examination was without semiological changes. She underwent abdominal US that revealed cholelithiasis with extrahepatic bile duct dilatation and thus requested MRI and referred to the general surgery outpatient clinic. She denied any other complaints, such as comorbidities, medication use, allergies or previous surgery.

Imaging (MRI) revealed normal-walled gallbladder with multiple thin septations within it, notably in the body and fundus, as well as fusiform dilatation of the proximal portion of the hepatobiliary duct, measuring up to 2.5 cm, compatible with cyst of Todani type I choledochal cyst (Fig. 1).

After two months, the patient underwent cholecystectomy and complete surgical resection of the extrahepatic bile duct cyst segment to about 1 cm below the bifurcation of the hepatic ducts, followed by Roux-en-Y end-lateral hepatic jejunostomy (Fig. 2). The postoperative evolution was satisfactory, being discharged on the eighth day after surgery, being followed so far without evidence of complications.

3. Discussion

Reports of bile duct cysts have been gone up in recent years, with more than half of the cases occurring in Japan. A report from Finland estimates that the incidence of bile duct cysts has been increased from 1:128,000 to 1:38,000 for the past 40 years; and although most cases were reported in children in the past, some studies have registered similar data in adults and children [11]. Despite various theories created, the exact etiology remains uncertain. The most accepted one is the proposal by Babitts in 1969 [17], in which states that choledochal cysts are caused by an anomalous junction of the pancreatic and choledochal duct outside the duodenal wall, proximal to Oddi’s sphincter during the embryonic period; with a formation of a common duct about 15 mm and that generates reflux from pancreatic bile duct secretions [1,3–9,11–13,17,18]. The activation of pancreatic enzymes in the biliary drainage system generates an increased intra-ductal pressure and inflammation, leading to a dilatation with cyst formation [7,8,18]. Despite being common, representing 57–96% of cases, it does not represent the etiologic factor from all cases of biliary cysts [5].

Regarding classification, Alonso-Lej and colleagues elaborated the first system in 1959, which encompassed 4 types of biliary cysts (type I to IV) [19]. In 1977, Todani and colleagues made modifications and added a fifth category characterized by the presence of intrahepatic cysts (Type V or Caroli Disease) [20]. In 2003 the presence of anomalous pancreaticobiliary junction was added to a classification modified by Todani, in which is currently the most utilized one [11]. It was summarized in Table 1 and it divides the patients in 5 groups according to place, extension and shape of cystic alterations [1,3–9,11–13,19].

Concerning the clinical presentation, there is no consensus in the literature about the most common one. Some authors affirm that jaundice is the most observed symptom [2], whereas others claim to be an abdominal pain [12]. Both symptoms added up to the presence of palpable abdominal mass in the right upper quadrant make up the classic triad seen in a minority of patients (about
less than 20%), with reports of higher expression in the pediatric age group [3,5,6,8–10]. In adult patients, the initial manifestations are unspecific, ranging from right upper quadrant abdominal pain, fever, nausea, vomiting to jaundice [1,11,13]. In children, about 85% have at least 2 classic triad symptoms [1].

The diagnosis of bile duct cysts is defined by the identification of bile duct dilation after exclusion from other etiologies (neoplastic, calculus or inflammatory) [3]. This condition can be identified by using specific imaging exams, such as ultrasound (US), computed tomography (CT), magnetic resonance cholangiopancreatography (MRCP), endoscopic retrograde cholangiopancreatography (ERCP) and endoscopic ultrasound [1–6,8,9,11–13]. In most cases ultrasound is the first examination performed to investigate the condition, being effective to evaluate the presence of dilatations and gallstones in the biliary duct [3,10]. In case of suspicion for the diagnosis of biliary cyst at a US, a CT or MRCP is requested; the last one, however, would be considered gold standard these days and the main choice for diagnostic evaluation, as it is a non-invasive technique which does not use radiation ionizing agent [2,5,6,8,9,11].

The standard treatment from bile duct cysts is surgical with specific approaches for each type according to its classification. Those classified as type I, II or IV are generally undergone resection surgery due to the risk of neoplastic development. The types I and V are completely resected with cholecystectomy completion and biliary reconstruction by Roux-en-Y hepaticojunostomy, being required a partial heptectomy on IVA type [3,11]. The technique previously mentioned is considered a gold standard for traffic reconstruction in pathologies which affect the biliary ducts, with studies demonstrating deepness rates between 2% to 6%. Colocoloduodenostomy or hepaticocolodenostomy configure other possible approaches, however, studies have shown that the occurrence of biliary gastritis was a recurring consequence to this technique when used for benign conditions treatment [21]. Types II are treated with cystic lesion resection and in specific cases, it may be required extension resection associated with biliary [4,9,11] transit reconstruction. In type IIIA cysts, it is recommended endoscopic sphincterectomy whilst types IIIB can be resected surgically or endoscopically [1,4,9,11]. Those classified as type V require a multidisciplinary approach (endoscopy, interventional radiology and surgery). In some cases, the only possible treatment is supporting, in order to prevent development from cholangitis and sepsis. Nevertheless, in other cases it is possible to use surgical approaches such as: segmental liver resection, lobar resection or liver transplantation [1,3,4,9,11].

4. Conclusions

Bile duct cysts are rare pathology in adults, happening to be more common in the pediatric age group, especially in Asian population. Currently, improvements in health care and the increase in accessing more accurate medical imaging enabling early diagnosis, contributing to use of therapeutic methods in appropriate time, avoiding development complication. The classic triad signs and symptoms are rarely present, as such vital the awareness of the disease to include it in diagnostic hypothesis and make use of optimal screening tests with a favourable cost-benefit, hence ultrasound to be the most used in the initial approach. The main treatment is consisted in surgical resection from cyst(s) when feasible, due to the risk of future malignancy.

Ethical approval

As the manuscript is not a research study, we only have the patient consent for writing and others forms of publication. Also, the ethical approval for this case report has been exempted by our institution.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Author contribution

Zuriel Seixas and Ketlen Sousa made contributions to conception and design. collected the patient details and wrote the paper. Frank Macedo made contributions to patient management. Victor Albuquerque and Rubem Silva Junior critically reviewed the article. All authors read and approved the final manuscript.

Registration of research studies

The manuscript is a case report, not considered a formal research involving participants.

Guarantor

Dr. Victor Albuquerque.

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Provenance and peer review

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Declaration of Competing Interest

The authors declare that there are no conflicts of interest.

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