Choledochal cysts in adults: A case report and review of literature

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ABSTRACT
Choledochal cyst is an uncommon disease that affects especially the pediatric population, although the diagnosis in adulthood has become frequent in the last few years. The treatment depends on the type of the cyst, but total excision of the cyst must be performed when it is possible. We report a 16-year-old female patient who presented with long-term unspecific symptoms alike colic biliary pain. Although abdominal ultrasound revealed no cholelithiasis, it suggested focal dilatation in the choledochal duct, which was confirmed by Endoscopic Retrograde Cholangiopancreatography (ERCP) as choledochal cyst Type IB of Todani. The patient was submitted to a laparoscopic cholecystectomy, excision of the cyst, and Roux-en-Y hepaticojejunostomy reconstruction. There were no postoperative complications. The diagnosis of choledochal cyst in adulthood has some particularities compared to its diagnosis in childhood, such as a different presentation and a greater association with other hepatobiliary diseases. Total excision of the cyst is the general principle for the treatment, which reduces the risk for cancer development. The way of reconstruction is discussed, comparing hepaticoduodenostomy and hepaticojejunostomy, and both the procedures are acceptable. Laparoscopic surgery is feasible and the preferable approach. Excision of the cyst is the main step of the treatment. Laparoscopic approach is feasible, but must be reserved for experienced surgeons in complex biliary procedures and in advanced laparoscopic surgery.

Keywords: Choledochal cyst, hepaticoduodenostomy versus hepaticojejunostomy, laparoscopic surgery.

INTRODUCTION
Choledochal cyst is a rare biliary disease that affects mostly the pediatric population, but has been diagnosed in a later age more frequently in the last few years. In adults, there are some particularities related to the biliary tract, such as previous manipulations, that deserve special care in the clinical management. Despite the low incidence rate, choledochal cysts can be associated with many hepatobiliary diseases, including cholangiocarcinoma.

The treatment depends on the type of the cyst classified according to the Todani’s classification, and has the excision of the cyst as a general principle. There are many options for the biliary tract reconstruction, and some studies have been conducted to compare them. The laparoscopic surgery is a feasible approach (such as the robotic surgery), with the advantage of being a minimally invasive procedure.

CASE REPORT
A 16-year-old female patient, who was a student without any associated disease, presented with epigastric pain...
since she was 6 years old and with post-alimentary vomits after fatty food ingestion in the last year. There was no history of jaundice.

Investigation with abdominal ultrasound and Endoscopic Retrograde Cholangiopancreatography (ERCP) revealed a severe cystic dilation of 5 cm in diameter in the choledochal duct, classified as Type IB of Todani’s [Figure 1]. On admission, the serum bilirubin level was 0.2 mg/dl, alkaline phosphatase was 239 U/l, and the activities of gamma glutamyl transpeptidase, aspartate transaminase and alanine transaminasewere 40 U/l, 56 U/l, and 68 U/l, respectively.

Surgical treatment was indicated and a laparoscopic cholecystectomy plus excision of the biliary cyst with Roux-en-Y hepaticojejunostomy reconstruction was performed [Figures 2 and 3]. The surgical specimen is shown in Figure 4 and the histopathologic analysis did not reveal any signs of malignancy. The postoperative curse was uneventful.

**DISCUSSION**

Choledochal cyst is a disease almost exclusively affecting the pediatric population, but recent studies are showing increased diagnosis in adults in the West. The improvement of imaging techniques, particularly Magnetic Resonance Imaging (MRI), in pancreatic and biliary ducts’ studies, the evolution of laparoscopic surgery and interventional techniques are some of the reasons for this change in presentation, with more cases of incidental diagnosis.

In the adulthood, it is more common in association with other hepatobiliary diseases, often leading to previous manipulation on the biliary tract, which may enhance the difficulties in the clinical management and surgical technique.

![Figure 1. ERCP showing a 5-cm-diameter cystic dilation in choledochal duct – Type IB of Todani’s classification (arrow).](Image)

![Figure 2. Laparoscopic view: (A) choledochal cyst; (B) cystic duct with clips.](Image)

![Figure 3. Laparoscopic surgery: hepaticojejunostomy.](Image)

![Figure 4. Surgical specimen: (A) gallbladder; (B) choledochal cyst.](Image)
As reported in this case, Type I of Todani’s classification is the most common presentation in 50%–80% of cases.\(^1\)\(^-\)\(^3\)\(^-\)\(^5\) The classic triad of symptoms consisting of abdominal pain, jaundice, and a palpable abdominal mass occurs in less than 20% of the patients.\(^6\) Mukhopadhyay et al., reported abdominal pain as the most common presentation in a pediatric population.\(^4\) Dhupar et al., in a retrospective review of choledochal cysts in 14 adult patients, reported the clinical presentation as biliary sepsis (3), pancreatitis (2), abdominal pain (3), and painless jaundice (1). Three patients had the cyst identified during laparoscopic cholecystectomy, and two had an incidental finding after CT scan.\(^2\) It is possible to conclude that adults may have a different presentation from children and there are no specific symptoms for the choledochal cyst diagnosis. Thus, the suspicion of this pathology requires a valorization of these common symptoms and a special attention given to the imaging date and incidental findings.

Association with other hepatobiliary pathologies occurs in 80% of the cases and includes benign diseases such as lithiasis in the cyst, intrahepatic lithiasis, acute cholecystitis and pancreatitis, and also malignant neoplasms, with cholangiocarcinoma being the most common. Patients with choledochal cysts have 20 times greater risk for the development of cholangiocarcinoma than the general population. Other malignant neoplasms have been reported, such as neuroendocrine tumors, but are much less common.\(^7\)

Technically, cyst excision is the main step to diminish the risk for cancer development, and must be performed when it is possible.\(^8\) Unfortunately, a cholangiocarcinoma may arise even after the cyst excision, as described by Nishiyama et al.\(^9\)

The surgical strategy depends on the type of the cyst. For the Type I cyst, complete excision of the cyst is usually feasible and Roux-en-Y hepaticojejunostomy is the preferable reconstruction technique. This approach was associated with low morbidity and mortality rates and few long-term complications.\(^8\) Reconstruction with a hepaticoduodenostomy has also been studied and found to have good outcomes. Mukhopadhyay et al., reported a review of 79 cases with hepaticoduodenostomy reconstruction and concluded that it is a quick procedure, with preservation of normal anatomy and physiology, and avoids multiple intestinal anastomoses. They suggested that this should be the preferred approach, as there were minimum complications.\(^4\)

Santore et al., compared hepaticoduodenostomy versus hepaticojejunostomy reconstructions in their series, stating that hepaticoduodenostomy required less operative time, allowed faster recovery of bowel function, and produced fewer complications requiring reoperation.\(^10\) With this data, it is reasonable to conclude that both the reconstruction techniques can be accepted. Further studies should be conducted to better understand the advantages and disadvantages of such techniques.

The laparoscopic approach for the choledochal cyst excision is the technique of choice instead of open laparotomy.\(^11\) It is a safe procedure and has the advantages of a minimally invasive surgery, such as lower pain, less wound complications, lower hospital stay, and faster surgical recovery. Robotic approach has also been described and seems to be as feasible as laparoscopy, although cost issues could be raised in this case.\(^12\)

It has to be emphasized that despite the advantages and feasibility of the laparoscopic approach for the treatment of choledochal cysts, it must be reserved for experienced surgeons in complex biliary procedures and advanced laparoscopic surgery, in order to avoid lesions and complications on the biliary tract with a negative impact in the patient outcome.

**CONCLUSION**

We conclude that there are some peculiarities in the clinical presentation of this disease in adults compared to the pediatric population. The aim of treatment is directed at the cyst excision, with reconstruction of biliary tract through hepaticoduodenostomy or hepaticojejunostomy. The laparoscopic approach is the preferable technique and must be performed only by experienced surgeons.

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Yoshida, et al.: Choledochal cysts in adults