v. 3, n. 10, 2023

International Journal of Health Science

# CHOLEDOCUS CYST TYPE I: CASE REPORT

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**Abstract**: Case report of a 16-year-old female patient with a rare diagnosis of type I choledochal cyst of the Todani classification, presenting with abdominal pain since childhood without episodes of cholestasis throughout the course of the disease. The treatment of choice was cholecystectomy and exeresis of the choledochal cyst, with a Roux-en-Y hepatojejunostomy type of biliary-digestive anastomosis.
**Keywords**: Choledocus cyst, ultrasonography, computerized axial tomography, surgery.

## INTRODUCTION

Initially described in 1852, the choledochal cyst is a congenital anomaly that affects both the extrahepatic and intrahepatic bile ducts, being classified into five types. Its incidence reaches 1:1000 in Asian countries, and 1:100,000 to 1:150,000 in Western countries, with women being affected two to four times more often than men. Most cases are discovered in childhood, with 60% before the age of 10 and 25% after the age of 20. (2)

The diagnosis is usually performed before 10 years of age and has been increasing due to the facilities and improvement of non-invasive imaging techniques.

The classic choledochal cyst triad (abdominal pain, jaundice, and palpable mass) is more common in children than in adults. However, most patients will have only one or two elements of the triad. (3)

Complications of biliary cysts are usually associated with an increased risk of cancer, especially cholangiocarcinoma, cholangitis, rupture, among others. There is a 20- to 30-fold increased risk of biliary cysts evolving into cholangiocarcinoma compared to the general population. The different types of dilations must undergo different surgical approaches, based on the principle of total resection of the dilation and restoration of biliary drainage through biliodigestive anastomosis. (1,2)

We will present a case report of a type I biliary cyst according to the Alonso Lej and Todani classification, a rare disease with low incidence and prevalence. We will discuss the clinical presentation, the diagnostic formulation and, with emphasis, the complex surgical approach to a cyst with distortion of the gastrointestinal anatomy and biliary tract.

The objective is to report a case of complex surgical approach to a biliary cyst and describe the possibility of good prognosis and quality of life for patients with choledochal cyst.

## CASE REPORT

J.N, 16 years old, female, born and raised in Itabaianinha (SE), presented for evaluation at the Digestive System Surgery outpatient clinic, complaining of intermittent and nonspecific epigastric pain since childhood, which worsened after eating. There was no association of episodes of cholestasis during the entire evolution of the pathology. The patient reported difficulty in gaining weight and that in the last four years she had had a non-significant weight loss. On physical examination, she was anicteric, with pain on palpation in the epigastrium and right hypochondrium, with the presence of a large palpable mass in the right hypochondrium, painful and massive on percussion. Laboratory tests showed no changes.

Ultrasonography of the entire abdomen revealed marked dilation of the intrahepatic bile ducts, up to the confluence of the hepatic ducts, where a large thick-walled cyst with debris inside could be seen adjacent to the bile duct. The ultrasonography findings were confirmed by magnetic resonance imaging of the upper abdomen, in which a circumscribed cystic formation was observed, with thin and regular walls, with cystic dilatation of the bile duct and common hepatic duct (in the transition between these two segments), measuring 8.8 cm. x 4.7 x 3.7cm, with no gaps

inside. Pancreas unchanged - choledochal cyst type 1a. (Figure 1).

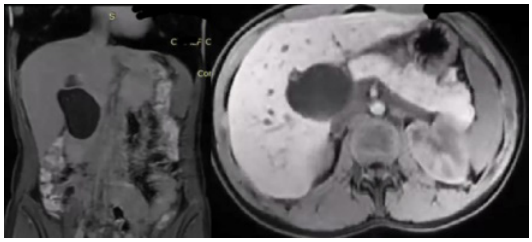

Figure 1

Based on these findings and after evaluating the patient, it was decided to perform surgical intervention to excise the cyst. The surgical procedure began with a median incision. Inventory of the cavity showed a large choledochal cyst extending from the insertion of the head of the pancreas to the confluence of the right and left livers with dilatation of the cystic duct (Figure 2). The choledochus was isolated under direct view of the portal vein and hepatic artery, followed by cyst dissection up to the intrapancreatic portion. A section and burial of the distal portion of the common bile duct was then performed. The proximal portion of the cyst was then dissected down to the hepatic duct below the confluence of the right and left hepatic ducts. To reestablish bile flow, a transmesocolic Roux-en-Y bileodigestive anastomosis was performed (Figure 3). The result of the anatomopathological analysis of the surgical specimen confirmed the diagnosis as choledochal cyst without atypia in the sample.

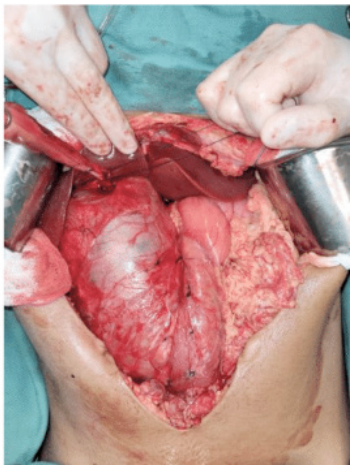

Figure 2

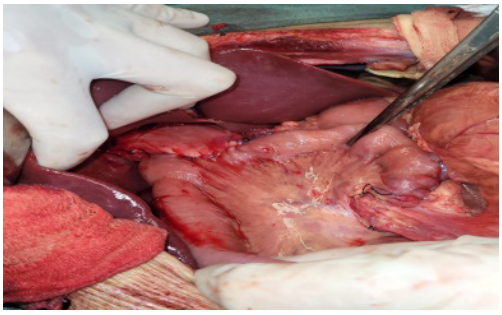

Figure 3

The patient evolved well in the postoperative period, being discharged on the 5th postoperative day, with adequate oral feeding, without pain complaints and with a suction drain with serous-looking secretion at low output and surgical wound with no secretion output, to outpatient follow-up, and the drain was removed on the 9th postoperative day.

She returned to the outpatient clinic 67 days after excision of the cyst with Roux-en-Y bileodigestive anastomosis, remaining asymptomatic, without postoperative complications, with a weight gain of 5 kg, being monitored every six months in the specialty service.

## DISCUSSION

In 1959, Alonzo-Lej presented two cases of choledochal cyst and analyzed another 94 cases of patients described in the literature at

the time, proposing the first classification for this anomaly, which in his work included only extrahepatic dilations. With the development and improvement of diagnostic methods, intrahepatic dilations were added to the initial classification. (3, 6)

The highest prevalence of these cysts is observed in eastern countries, especially in Japan, which has an incidence of 1:1000. (1)

Biliary cysts are classified according to location, extent and shape. As previously mentioned, in 1959, Alonso-Lej proposed a classification that included only extrahepatic biliary cysts, which was later modified by Todani et al (1977) (Figure 4), being subdivided into 5 types:

Type I - solitary extrahepatic fusiform cyst;

Type II- extrahepatic supraduodenal diverticulum;

Type III- intraduodenal diverticulum (choledococele);

Type IV A- intra and extrahepatic fusiform cysts;

Type IV B- multiple extrahepatic cysts;

Type V - multiple intrahepatic cysts (Caroli's disease).

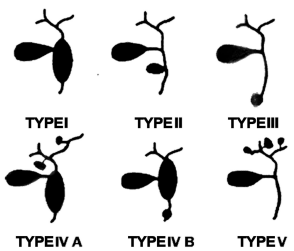


Figure 4

Type I corresponds to 50% of choledochal cysts, type IV corresponds to 35% of cases, while types II, III and V are less common and are diagnosed in less than 10% of patients with cystic choledochal formations. The symptomatological triad – jaundice, abdominal pain and palpable mass in the upper right quadrant of the abdomen – is present in a minority of patients (0%-17%). Abdominal pain and fluctuating jaundice are the most frequent diagnostic findings, but not mandatory during the investigation, while palpation of an abdominal tumor occurs in less than 20% of cases.

The main diagnostic tool, especially in children, is ultrasound. In adults, computed tomography can confirm the diagnosis, however ERCP (endoscopic retrograde cholangiopancreatography) and magnetic resonance imaging are the most accurate methods.

Among the most common complications of choledochal cyst, in addition to evolution to cholangiocarcinoma, pancreatitis and cholangitis stand out, which are frequently described. There is also a chance of evolution to secondary biliary cirrhosis, which may affect approximately 50% of cases, while malignancy, that is, the development of a cholangiocarinoma, affects between 10 and 30% of cases. (5.6)

Exeresis of the cyst and reconstruction of the biliary tree through a Roux-en-Y hepatojejunostomy type of biliary-digestive anastomosis is the most indicated surgical procedure. Among the different lines of treatment for choledochal cysts, there are external drainage, internal drainage, which includes cystoduodenostomy or cystojejunostomy, and surgical removal of the cyst. (5)

External drainage is considered a good treatment for emergency patients, being a simple procedure with a short operating time, but with a high rate of reoperation (86%). Internal drainage is related to a high rate of reapproachment (50%) and a high incidence of carcinoma (30%), not being the treatment of choice electively, being only indicated in cases with contraindications or urgency due to previously observed complications. definitive surgery for this pathology cannot

be considered. (1,2,6)

The treatment with the best prognosis, with a low rate of reoperation (6%) and incidence of carcinoma (6%), is cyst excision, followed by Roux-en-Y hepatojejunostomy, with liver lobectomies or liver transplantation. normally indicated for types IVa and V (Caroli's Disease) of the Todani classification respectively. (6)

## CONCLUSION

The choledochal cyst may be asymptomatic or with an abdominal mass, jaundice and abdominal pain. The exam of choice to start the investigation is still the abdominal ultrasound. Once suggested, diagnostic complementation is generally necessary due to the low sensitivity of ultrasound and the need to carry out a differential diagnosis. In this respect, magnetic resonance cholangiography allows better definition of the lesion and its classification as proposed by Todani.

Once the diagnosis is made, treatment will always be surgical due to the risk of malignancy, with cholecystectomy and resection of the lesion, regardless of the type of disease. The variation is related to the need for biliodigestive diversion during surgery. Type I disease, as in the reported case, can only be treated with excision of the lesion and cholecystectomy, excluding dilatation or other changes in the biliary tract.

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