Reoperation for the treatment of choledochal cyst in adults

Reoperação para tratamento de cisto de colédoco no adulto

Manoel Lisboa1, José Salizi2, Fernando Queiroz2, Isadora Maia Nobre1, Isabela Lemos de Almeida Castro1, Maria Alice Soares Silveira Cardoso2, Luana Magalhaes Bernardo2, Neander Neves Alves Ezidório3, Vivian Resende4

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ABSTRACT

Choledochal cyst refers to congenital dilatation of the main biliary route associated or not with the segmental intrahepatic ducts. We report the case of a female patient, 40 years old, with the diagnosis of choledochal cyst, who at 17 years of age had undergone surgical treatment. She evolved with cholangitis of repetition and obstructive jaundice. The imaging exams showed dilation of the biliary intra-hepatic routes, mainly on the left, hepatolithiasis, and a large cystic lesion in the liver pedicle. The laparotomy evidenced that the previous treatment had been a cholecystectomy and cyst draining through Roux cystojejunostomy in Y, which was blocked. The treatment consisted of resection of the choledochal cyst, hepatectomy of the 2 and 3 segments, and Roux hepaticojejunostomy in Y. The patient is in the fourth year of outpatient follow-up with no complications. On the basis of its association with cancer, resection and not just draining is the recommended treatment of choledochal cyst.

Key words: Choledochal Cyst; Choledochal Cyst/surgery; Reoperation; Cholangiocarcinoma.

INTRODUCTION

The choledochal cyst (CC) is a congenital dilatation of the biliary tree, which can be associated to the malformation of the biliopancreatic junction, which is considered a risk factor for malignant biliary tract disease. Cholangiocarcinoma has been described in patients under 20 years of age with a choledochal cyst. There are reports of carcino-
ma developed in the intra-hepatic ducts and remaining stump after resection of a choledochal cyst.

The general CC diagnosis is reached in the pediatric patient and, currently, due to the accuracy of propaedutic imaging methods, it is possible to be conducted intra-uterus. The diagnosis can also be made in adulthood when symptoms are late.

Due to its association with biliary cancer for most type I cases, according to the classification of Todani, total resection of the extrahepatic biliary duct and Roux hepaticojejunostomy in Y is the preferred treatment. An early diagnosis and surgical treatment provide a good prognosis.

The objective of this report is to describe the case of a patient who had choledochal cyst derivation by means of cistojejunal anastomosis as the initial treatment.

CASE REPORT

The patient’s consent for this case report was obtained. A female 40-year-old patient, melanodermic, healthy, complaining of recurrent abdominal pain. She reported having had episodes of cholangitis and had been submitted to conventional cholecystectomy and choledochal cyst treatment, at the same time, 22 years ago. Abdomen magnetic resonance imaging showed large cystic lesion near the hepatic hilum, dilatation of the biliary intra-hepatic route, predominantly on the left, with multiple filling defects suggestive of calculi (Figure 1).

The patient underwent laparotomy during which, an extra-hepatic biliary route cyst, cistojejunal Roux Y anastomosis, and internal hernia with dilatation of the proximal RouxY loop were observed (Figure 2). The pre-operative cholangiography demonstrated dilatation of the biliary intra-hepatic routes to the left, with multiple filling defects. Lysis of adhesions, resection of the biliary cyst and dilated Y de Roux loop, hepatectomy of segments 2 and 3 (Figure 3), and Roux hepaticojejunostomy in Y were conducted. The patient evolved with no complications and was discharged on the fifth postoperative day. She is in the fourth year of outpatient follow-up and showed no intercurrences during this period.

Figure 1 - Abdomen MRI showing dilatation of intrahepatic bile ducts, hepatolithiasis, cystic lesion near the hepatic hilum, dilatation of intrahepatic bile ducts, predominantly on the left, with multiple filling defects suggestive of calculi.

Figure 2 - Cistojejunal Roux anastomosis in Y. The tweezer shows the site of obstruction and dilatation of the loop proximal to the Y in Roux.

Figure 3 - Dried liver segments II and III showing dilatation and lithiasis in intrahepatic bile ducts.
DISCUSSION

The incidence of choledochal cyst is very variable. The incidence estimate in Western populations is 1/200,000 and 1/13,000 live births, while in the East, Japan presents the highest number of reported cases. The prevalence is in females with a ratio of 4:1 (approximately 80% of cases). The disease is typical of young children, with less than 20% of the diagnoses in adults.10,12

The pathogenesis of choledochal cyst is probably multifactorial.14-17 It can be related to the malformation of the biliopancreatic junction.14,15 In autopsy studies in normal individuals, it was found that the union of the choledochal duct and main pancreatic duct results in the formation of the hepatopancreatic ampulla inside the duodenal wall, subjected to the control of a sphincter mechanism, in 80% of cases.14 In the normal child, the maximum length of the common channel is up to 4 mm, and 10 mm in adults. The sphincter action prevents pancreatic juice reflux into the biliary route. Similarly, this sphincter mechanism prevents the passage of bile into the pancreatic duct.14

In the embryological point of view, the biliopancreatic junction malformation occurs due to the very proximal origin of the ventral pancreas diverticulum that, after subsequent rotation, determines a biliopancreatic channel of greater length.14 The anomalous biliopancreatic junction, unlike the normal situation, is not inside the sphincter of Oddi and thus, the contraction of the sphincter is not able to prevent the reflux of pancreatic enzymes into the biliary route, with the activation of pancreatic proenzymes, epithelial lesion, and wall weakening in the bile ducts. As a result, inflammation, hypertension, and ductal dilatation occur.25 Despite the high incidence of the anomalous biliopancreatic junction in patients with choledochal cysts, this change is not present in all cases.

The isolated dysfunction of the sphincter of Oddi and alteration in the autonomous innervation of the choledoctal duct terminal has been considered as causing biliary dilatation.16,17 There is also the hypothesis that the viral infection can be related to the etiology of choledochal duct cysts because retroviral RNA was detected in the tissues of patients. In this case, the viral infection would have caused aganglionosis of the distal choledochal by an immunological mechanism.17

The hereditary pattern or genetic predisposition to the formation of choledochal cysts is not yet well defined, although it has been demonstrated that several members of the same family may be affected.16,17 Defects in epithelialization and bile duct recanalization during the embryonic development and congenital weakness of its walls have also been implicated.16,17

In the classification of Todani11, type I choledochal cysts are divided into three subtypes (IA, IB, IC). The IA subdivision is characterized by coledochus dilation, affecting part or all of the extrahepatic biliary route with a normal intrahepatic biliary route. Type IB cysts are characterized by focal and segmental dilatation of the distal coledochus. Type IC cysts display coledochus fusiform dilatation with cylindrical dilatation of the common hepatic duct with the normal intrahepatic biliary route.

Type II cysts present with a saccular diverticulum, also suggesting duplication of gallbladder.11 Type III cysts are also called choledochoceles.11 In this type, the coledochus ends up forming a small intramural cyst, which creates a protrusion into the duodenal light covered by mucous membrane.

Type IV cysts are subdivided into IVA and IVB. These are more frequent in adults and associated with intrahepatic lithiasis, complicated with bile duct dilatation and hepatic abscesses.11 Subtype IVA cysts correspond to multiple cysts, affecting extra- and intra-hepatic bile routes. These latter are more frequent in the left liver lobe and was the type presented by the patient in this report. Subtype IVB cysts are also multiple. However, they only affect extra-hepatic bile routes. Type V cysts correspond to the Caroli disease.18

On the basis of its association with cancer,14-17 most authors have proposed surgical resection of type I and II choledochal cysts followed by traffic reconstruction of Roux in Y.10-13,19 This conduct provides the best results, with low morbidity and mortality.12,13 In patients with type III cyst, the recommended treatment is endoscopic papillosectomy.19 Procedures range from hepatectomies to segmental liver transplantation in cases where there is impairment of the intra-hepatic biliary tree.20 When not operated, patients can evolve to complete biliary obstruction, secondary biliary cirrhosis, hypotension, and septic cholangitis due to severe infection.19

In conclusion, it should be noted that, in the past, as in the case of this patient's report, choledochal cysts were only drained into an intestinal loop instead of being dried. This conduct should be avoided because these cysts can evolve to serious complications; the main one being the development of cholangiocarcinoma.
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REFERENCES


