CASE REPORT

Choledochal cyst: a case report

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ABSTRACT

A choledochal cyst is a cystic dilatation that affects both the intrahepatic and extrahepatic biliary tree. The most commonly used classification to describe this pathology is that of Todani et al. (1997), which includes five categories. It is usually asymptomatic, but may present with symptoms such as abdominal pain, jaundice, and fever. The gold standard for diagnosis is magnetic resonance cholangiopancreatography (MRCP). Patients with choledochal cysts have an increased risk of carcinogenesis. The neoplasms most commonly associated with choledochal cysts are cholangiocarcinoma and gallbladder cancer. Type I and V cysts are the most likely to undergo malignant transformation, while it is rarer in types II and III. Treatment consists of the surgical excision of the cyst with Roux-en-Y biliodigestive reconstruction. The laparoscopic approach is currently the most commonly used. Biliary drainage reconstruction is primarily performed using the Roux-en-Y procedure with hepaticojejunostomy (HJ) or hepaticoduodenostomy (HD). The literature on the choice of anastomosis and its benefits for the patient is limited. Several factors, such as age, cyst type, histologic findings, and location, influence the prognosis of each patient. We present the case of a 29-year-old female patient who sought medical consultation due to colicky abdominal pain lasting one month, associated with persistent nausea and located in the right upper quadrant of the abdomen. Magnetic resonance imaging (MRI) revealed a Todani type Ia choledochal cyst. Therefore, she underwent a complete surgical excision, with a favorable postoperative course.

Keywords: Case Reports; Choledochal Cyst; General Surgery (Source: MeSH NLM).

INTRODUCTION

A choledochal cyst is an uncommon condition caused by cystic dilatation of the biliary tree, which may involve one or more segments of the bile ducts, both extrahepatic and intrahepatic [1]. It mainly affects the Asian population and females [2]. Todani et al. (1997) classified this pathology into five categories (Figure 1); this classification is the most widely used today. Type I choledochal cysts account for 60 % to 80 % of cases and are characterized by a single cystic dilatation of the common bile duct [1].

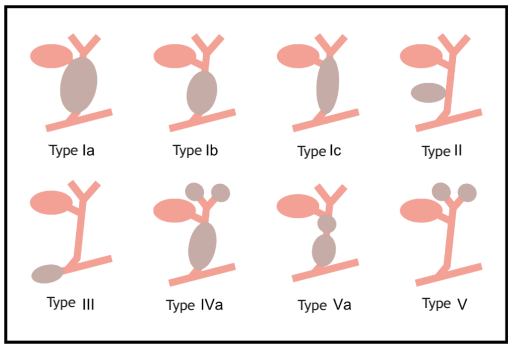

Figure 1. Todani classification

Two main pathophysiological theories have been proposed: the first is based on an abnormal pancreatobiliary junction, and the second on congenital distal biliary stenosis. Both propose that there is reflux and mixing of pancreatic and biliary secretions, leading to activation of pancreatic enzymes and trophic changes in the bile ducts [3].

Choledochal cysts are usually asymptomatic until adulthood, but they can also manifest with symptoms such as abdominal pain, jaundice, and fever [4]. The diagnostic approach usually begins with ultrasound, but is confirmed with magnetic resonance cholangiopancreatography (MRCP) [5]. The therapeutic measure used is cholecystectomy with complete surgical resection of the cyst along with Roux-en-Y biliodigestive reconstruction [6]. We herein present the case of a patient with a choledochal cyst.

CLINICAL CASE

We report the case of a 29-year-old female patient with no medical or surgical history. She was evaluated in an outpatient clinic after one month of symptoms characterized by colicky abdominal pain in the right upper abdomen, associated with persistent nausea. During the

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physical examination, mild tenderness was detected on deep palpation in the right hypochondrium, with no palpable masses or other findings suggestive of pathology.

Initial laboratory tests revealed hemoglobin of 12.5 g/dL, white blood cell count within normal limits, and C-reactive protein within the normal range. Liver and renal function were preserved, with normal values of transaminases, total and fractionated bilirubin, alkaline phosphatase, gamma-glutamyl transferase (GGT), albumin, urea, and creatinine

An initial CT scan was also performed, revealing a hypodense image projected at the level of the extrapancreatic common bile duct, reported as possible ectasia of the extrapancreatic common bile duct, with a lower probability of being a cystic component. No focal lesions were identified using this method (Figure 2).

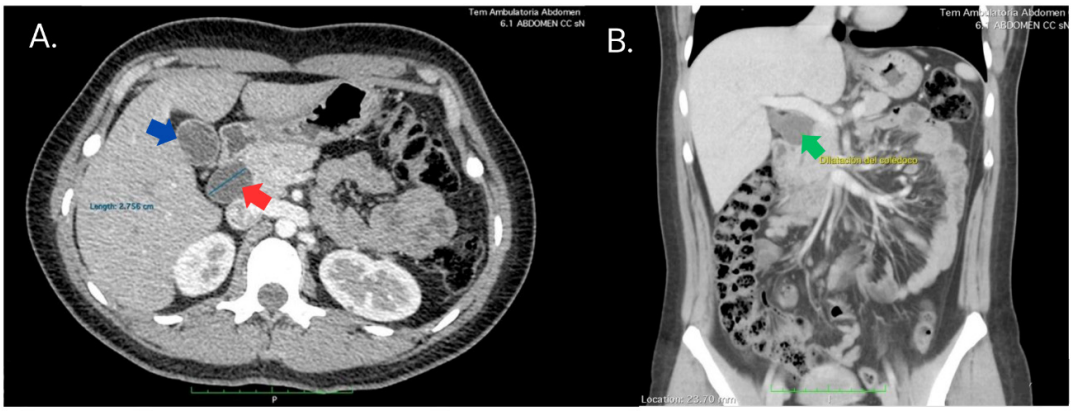

Figure 2. A. Axial image from a MDCT scan showing dilatation of the extrahepatic biliary tract measuring approximately 2.75 cm in diameter (red arrow), the same size as the gallbladder (blue arrow). B. Coronal image of an abdominal MDCT scan showing dilatation of the extrahepatic bile tract (green arrow).

MDCT = multidetector computed tomography

Further studies included an upper abdominal MRI with MRCP, which revealed seven polyps in the fundus of the gallbladder measuring up to 4 mm in transverse diameter. In addition, a cystic dilatation of the extrapancreatic common bile duct was observed, measuring 22 mm in transverse diameter and 32 mm in longitudinal diameter, with no evidence of wall thickening, intraluminal nodules, enhancement, or pathological restriction, consistent with a choledochal cyst (Todani type Ia). Likewise, a cystic dilatation of the cystic duct was identified, measuring 8 mm in transverse diameter and 13 mm in length. The intrapancreatic common bile duct measured 3.5 mm in caliber with a homogeneous signal, with no intraluminal lesions, choledocholithiasis, or extrinsic lesions (Figure 3).

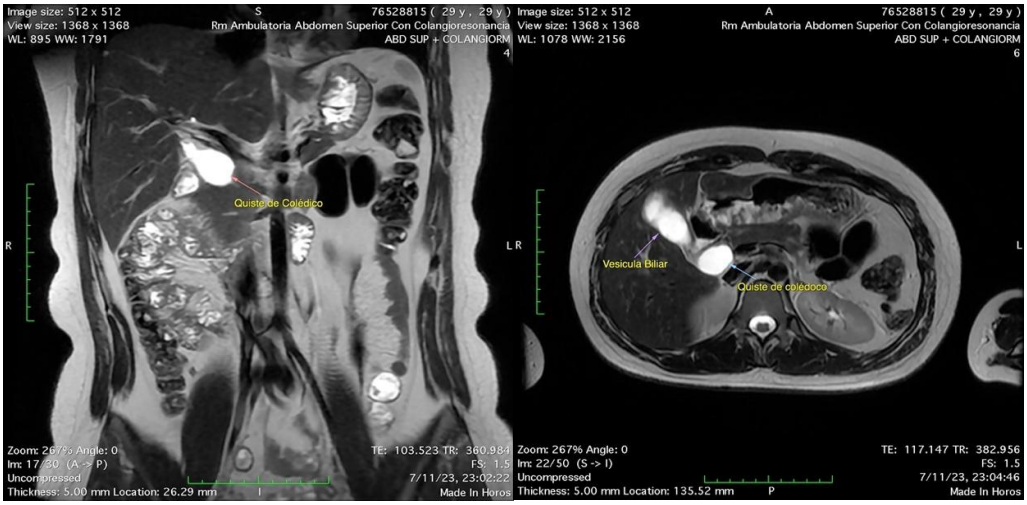

Figure 3. Fusiform dilatation of the extrahepatic biliary tract, corresponding to a type Ia dilatation according to Todani classification.

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In view of the diagnosis of a choledochal cyst and gallbladder polyps, surgical intervention was indicated. The patient underwent laparoscopic cholecystectomy with choledochal resection and Roux-en-Y biliary-digestive reconstruction. During the surgical procedure, a gallbladder of 8 x 5 cm was found, and the cystic duct was approximately 5–6 cm length and 3 cm in diameter. In addition, at the level of the common bile duct, a fusiform dilatation was evident, extending from the gallbladder confluence to the retroduodenal duct (Figure 4).

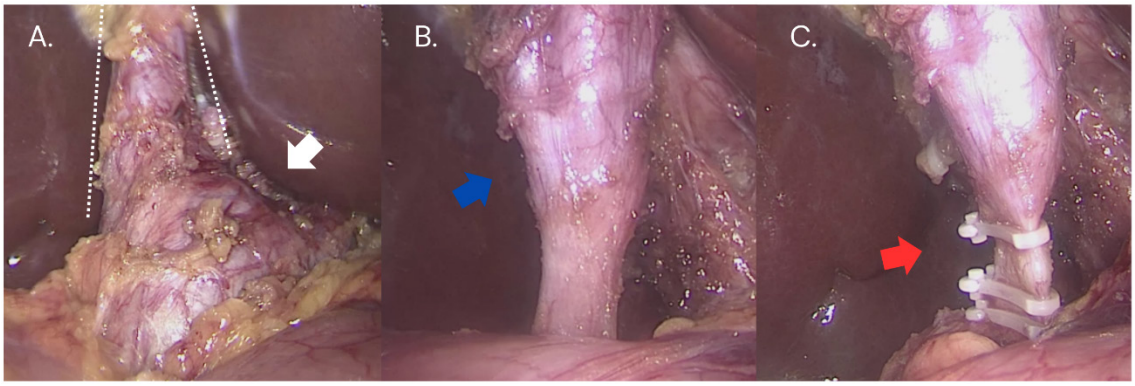

Figure 4. A. Intraoperative image showing the cystic duct (dotted lines) draining into the choledochal cyst (white arrow), type Ia according to Todani classification. B. Intraoperative image showing the end of the cystic dilatation (blue arrow) in the distal portion of the biliary tract. C. Intraoperative image showing the distal section of the choledochal cyst (red arrow).

A laparoscopic cholecystectomy with resection of the affected biliary tract was performed. During the surgical procedure, the common bile duct was identified, and resection was carried out at its distal intrapancreatic portion with clipping of the remaining biliary tract, while the proximal section was transected 1.5 cm from the bifurcation of the proximal biliary tract. Subsequently, an extracorporeal Roux-en-Y biliary-digestive reconstruction was performed. The cystic lesion of the common bile duct was completely resected (Figure 5) and sent for pathological examination, which reported a choledochal cyst, chronic cholecystitis, cholesterolosis, a gallbladder polyp, and a cystic lymph node with reactive changes, with no evidence of malignant cells.

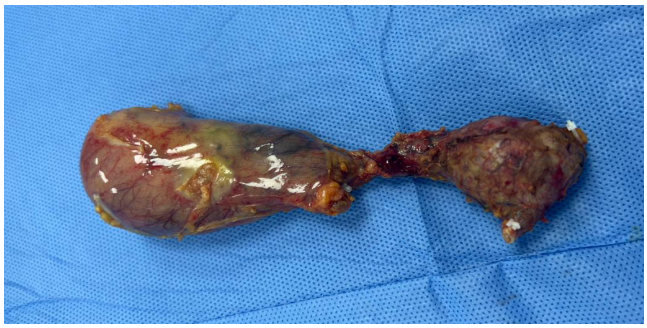

Figure 5. Surgical specimen showing the gallbladder and biliary tract dilatation, with clips at the proximal and distal resection edges.

The patient had a favorable postoperative course and was discharged five days after surgery. During the postoperative follow-up, she showed good progress and tolerated the diet without nausea or vomiting. At the outpatient follow-up visit, she reported no complications. She is currently asymptomatic.

DISCUSSION

Choledochal cysts are congenital malformations characterized by abnormal dilatation of the extrahepatic and intrahepatic ducts. They are usually diagnosed in childhood but may remain asymptomatic until adulthood. The most common symptoms, in order of frequency, are abdominal pain, jaundice, and fever [4]. The gold standard for diagnosis is MRCP. Since 1977, the Todani classification has been the most widely used worldwide, and divides choledochal cysts into five types, with type I being the most common [5].

Although the pathophysiology is not fully understood, Babbitt's theory is the most widely accepted. It states that an abnormal pancreatobiliary junction causes reflux of pancreatic secretions, which weakens the bile duct wall and predisposes to cyst formation [7]. Therefore, early radical surgical excision of the bile ducts is recommended to prevent reflux of pancreatic enzymes and restore normal biliary flow. This approach helps prevent complications such as recurrent cholangitis, pancreatitis, and liver cirrhosis, and minimizes the risk of hyperplasia/metaplasia that may promote carcinogenesis [8].

Patients with choledochal cysts have an increased risk of developing cancer. Ten Hove et al. found that, in most cases, the risk of malignancy transformation was approximately 11 % [9]. Sastry et al. reported that the neoplasms most frequently associated with choledochal cysts were cholangiocarcinoma (70.4 %) and gallbladder cancer (23.5 %). After the age of 60, this risk increased proportionally with age, reaching up to 38 % [10]. Type I and type V cysts show the highest propensity for malignant transformation, whereas this is rare in types II and III [9].

The standard treatment is based on complete resection of the cyst with reconstruction using a bilioenteric anastomosis. This procedure is performed laparoscopically, which is currently the most widely used technique. However, it is recommended that the Roux-en-Y reconstruction be performed extracorporeally through an extended incision at the umbilical trocar site to ensure shorter operative time and greater patient safety [6].

According to Todani classification, type I requires complete excision of the extrahepatic biliary tree, as well as cholecystectomy and Roux-en-Y hepaticojejunostomy (HJ). Type II only requires excision of the cyst due to the low risk of malignant transformation. In type III, endoscopic sphincterotomy is preferred, especially in cysts smaller than 2 cm. Types Ic, IVa, and V, due to intrahepatic involvement, usually require complete excision and liver resection, and may ultimately require liver transplantation [5].

The laparoscopic surgical approach has recently been shown to be a feasible, safe, and effective procedure. Yuan Liu et al. found that blood loss was relatively lower, facilitating rapid recovery of intestinal peristalsis, early resumption of diet, and a shorter hospital stay. However, no significant results were observed in the postoperative complication rates [9]. Therefore, it is important to individualize each patient and perform a proper preoperative assessment to select the best approach.

Biliary drainage reconstruction is mainly performed using the Roux-en-Y technique with HJ or hepaticoduodenostomy (HD). Narayanan et al. found that HD was associated with higher rates of postoperative reflux and gastritis compared to HJ, but resulted in a shorter hospital stay. In conclusion, both techniques have similar results in terms of other complications and surgical benefits [11]. In this case, HJ was chosen to avoid complications such as recurrent cholangitis and biliary reflux.

Complete excision of the choledochal cyst has shown excellent outcomes, with event-free survival and overall 5-year survival rates exceeding 90 % [9]. However, there remains a risk of malignant neoplasia developing in the residual bile duct, due to recurrent cholangitis or preexisting precancerous transformation prior to surgery [9]. Watanabe et al. found that the most frequent sites of carcinoma after excision were at the anastomotic site (35 %), intrahepatically (26 %), and in the intrapancreatic ducts (26 %) [12]. Similarly, He et al. reported a higher incidence of malignancy in the common bile duct [13]. Therefore, postoperative follow-up is recommended with annual liver function tests, Ca 19-9 measurement, and semiannual ultrasound evaluation, preferably performed in specialized hepatobiliary centers [14, 15].

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