SCIREA Journal of Clinical Medicine
ISSN: 2706-8870
http://www.scirea.org/journal/CM
December 11, 2022
Volume 7, Issue 6, December 2022
https://doi.org/10.54647/cm32918

TYPE B CHOLEDOCHOCELE: A RARE PRESENTATION WITH A COMMON TREATMENT

Review of Literature and a case report

Erick Servin Torres [1,*], Silvana Castelán-Sánchez[1], Eulalio Jiménez-González[1]

[1]Servicio de Cirugía General, Hospital de Especialidades "Dr. Antonio Fraga Mouret" Centro Médico Nacional La Raza, IMSS. Mexico

[*]**Corresponding Author:** drerickservin@gmail.com *(*Erick Servin Torres*)*

ABSTRACT

Introduction: Biliary dilation is a rare disease involving intrahepatic and extrahepatic biliary tract abnormalities. Choledochocele or type III choledochal cyst (CC) is a lesion present in less than 2% of all reported cases and is defined as a cystic dilation of the distal common bile duct protruding into the duodenal lumen.

Objective: To report on the endoscopic treatment of type III common bile duct cyst, as well as a review of the literature on the subject.

Case presentation: A 61-year-old female patient complained of severe abdominal pain for one month, with no associated cholestatic signs. She underwent abdominal ultrasonography that showed cholelithiasis. First diagnostic suspicion was cholecystitis, so she underwent emergency open cholecystectomy. The patient presented a torpid post-surgical evolution with jaundice, where control ultrasound showed extrahepatic bile duct dilation. MRI

cholangiography revealed a fusiform dilation of the proximal portion of the hepatocholedochal, compatible with a choledochal cyst.

Conclusion: Our case presented with relatively rare manifestations and involved one of the undiscussed categories of the Todani classification system; thus, highly relevant. Endoscopic sphincterotomy is a feasible and effective treatment for choledochocele including large and protruding ones.

Keywords: biliary dilatation, choledochocele, CC, MRI, sphincterotomy.

BACKGROUND

Choledochal cysts are a heterogeneous group of anomalous and uncommon dilatations of the extrahepatic and/or intrahepatic biliary system first described by Abraham Vater in 1973. While some theories regarding the etiopathogenesis of this entity have been proposed no definitive consensus has been reached. Several complications of choledochal cysts, some serious in nature, have been described, including malignant transformation. Therefore, accurate diagnosis and early treatment are paramount.

Initial classification by Alonso-Lej et al. in 1959 described 3 types of choledochal cysts, type I–III. Later, in 1977, Todani et al. modified the classification according to the location of biliary duct dilatation adding types IV and V. Currently this classification is the most used by surgeons.

In this paper we will focus on type III. Choledochoceles. A cystic dilatation of the intra-ampullary portion of the common bile duct (CBD). They have been reported as having distinctive demographic and anatomic features and a lower risk of malignant transformation than other types of choledochal cysts.

Alonso-Lej classification for bile duct cysts as modified by Todani:		
Type I	Cystic or fusiform dilatation of the bile duct without affecting intrahepatic bile duct.	Type IA: cystic dilatation of the common bile duct, as well as part of the hepatic common duct and some portions from the right and left hepatic duct

	.	Type IB: segmental/focal dilatation of the extrahepatic common bile duct
		Type IC: fusiform dilatation of the extrahepatic biliary tree.
		Type ID: cystic dilatation of the common and cystic duct.
Type II	Real diverticulum of the extrahepatic bile duct.	
Type III	Choledococeles	Type IIIA: both common bile duct and pancreatic duct enter the cyst which connects to the duodenum through another orifice.
		Type IIIB: an intra-ampullary common bile duct diverticulum
Type IV	Presence of multiple intrahepatic and extrahepatic cysts, or only the extrahepatic ones.	Type IVA: intra and extrahepatic dilatations.
		Type IVB: multiple dilatations in the extrahepatic bile ducts only.
Type V	One or more cystic dilatations of the intrahepatic biliary tract (without involvement of extrahepatic biliary duct).	

The prefix *choledocho-* refers to the common bile duct (CBD), and the suffix *-cele* refers to a swelling or cavity. The term *choledochocele* was coined by Wheeler[1] in 1940 to describe a cystically dilated intraduodenal portion of the CBD.

% Of frequency and malignancy according to Todani classification		
	Frequency	**Malignancy**
Type I	50 - 85%	68%
Type II	2%	5%

Type III	1 - 5%	2.5%
Type IV	15 - 35%	21%
Type V	20%	7 - 15%

Type A choledochoceles are cystic dilatations of a segment of the intra-ampullary bile duct and are located proximal to the ampullary orifice. Type B choledochoceles are diverticula of the intra-ampullary common channel and are located distal to the ampullary orifice; they can be distinguished from duodenal duplication cysts both anatomically and histologically. Both types of choledochocele may present with pancreatitis, biliary obstruction, or nonspecific gastrointestinal symptoms.

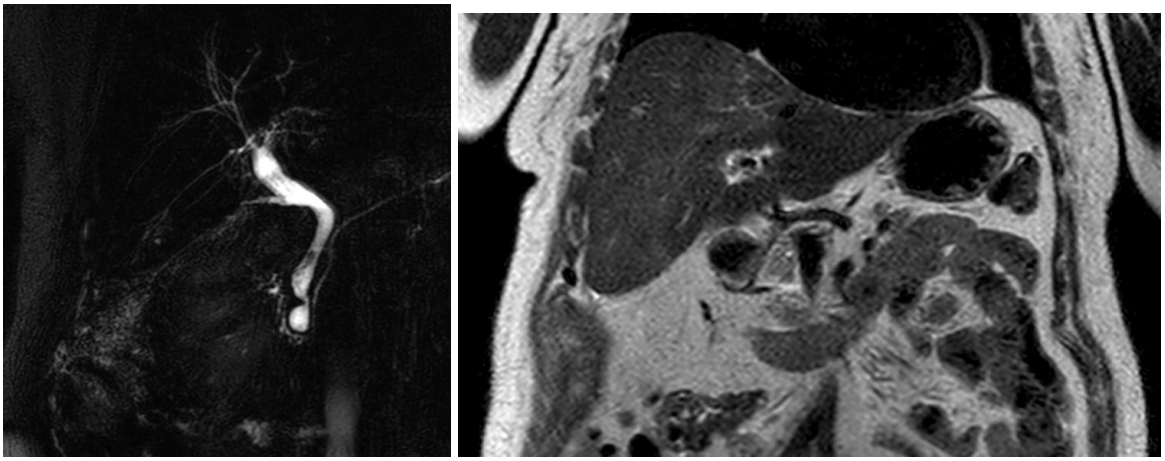
Given its anatomical presentation, choledochocele can be treated with sphincterotomy or endoscopic papillectomy with few complications.

This manuscript focuses on Type III choledochal cysts or choledochoceles, describing a case and its treatment in a specialty hospital in Mexico City.

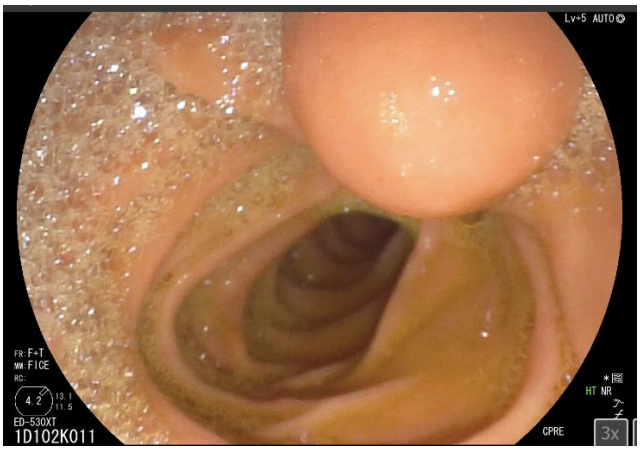
CASE PRESENTATION

A 61-year-old woman presented with a history of intermittent colicky right hypochondrial pain not relieved by medication for the past 2 months. Initially, she was treated with proton pump inhibitors, but her pain was not relieved, and later she developed a fever. She was admitted to a general hospital and diagnosed with cholecystitis. An open cholecystectomy was performed, with findings described as pyocholecystitis with adhesions. She was managed with parenteral antibiotics and discharged. She developed jaundice and was referred to a high specialty hospital with suspicion of a biliary lesion.

She was admitted to our hospital with severe abdominal pain. After multidisciplinary team discussion, an abdominal ultrasonogram (USG) was performed. It showed a clear thick wall cyst measuring 4.6 × 2.6 cm between the second part of the duodenum and the head of the pancreas. A magnetic resonance cholangiopancreatography showed an isolated cystic-like dilatation of 43 x 20 mm in the intraduodenal portion of the CBD.



ERCP revealed an 80 x70 mm subepithelial swelling proximal to the major papilla protruding into the duodenum. Based on these findings, the lesion was reported as type III choledochal cyst. The patient was deemed a good candidate for endoscopic treatment (▶ Video 1).



Biliary sphincterotomy was performed with a 30W Endocut achieving cannulation of the biliary tract. No post-procedural complications were observed. At the 3-month follow-up the patient has remained asymptomatic with normal bilirubin levels.

In our opinion, this case confirms that endoscopic papillectomy is a good option for the treatment of patients with choledochocele.

DISCUSSION

Choledochal cysts (CCs) are uncommon congenital anomalies of the biliary tract with an incidence of 1 in 100,000– 150,000 live births in western populations but reported to be as high as 1 in 13,500 live births in the United States and 1 in 15,000 in Australia. The incidence is higher amongst Asian populations and are usually diagnosed during childhood. About 25% are detected later in adult life. Choledochal cyst have an unexplained female:male

preponderance, commonly reported between 4:1 and 3:1. In Mexico there is little information about the incidence and prevalence of this entity.

The true incidence of choledochoceles is likely dependent on the definition of the lesion, the diagnostic modalities available, and the population studied. Choledochoceles frequently present at an older age when compared with other choledochal cysts, with an average age at presentation of 51 years compared with 29 years for other cystic lesions of the biliary ducts. Pediatric presentations are unusual.

In contrast to other choledochal cysts (CCs), the risk of malignancy appears to be low for choledochoceles. These considerations have led some to conclude that choledochoceles should not be classified as choledochal cysts.

CLASSIFICATION

Cyst anatomy, histology, and radiology have all been used to classify periampullary cystic lesions, resulting in a variety of systems and terminologies.

In Todani's classification type III represents a cystic dilation of the intramural portion of distal CBD which bulges into the duodenum. Ziegler et al. in their analysis, comparing choledochoceles with Todani's types I, II, IV, and V, with respect to age, sex, complications, and management concluded that classification of CCs should not include choledochoceles. Sarris and Tsang subdivided choledochoceles into Type A and Type B lesions based on their anatomic appearance.

Type A lesions are those in which the intramural bile duct opens into a cystically dilated segment, which communicates to the duodenal lumen via a separate orifice.

Type B choledochoceles are characterized by a bile duct that opens normally into the duodenal lumen, with the choledochocele arising as a diverticulum of the intra-ampullary common channel.

Additional choledochocele variants (Types C–E) have been described by Kagiyama et al with each variant demonstrating slightly different pancreaticobiliary anatomy.

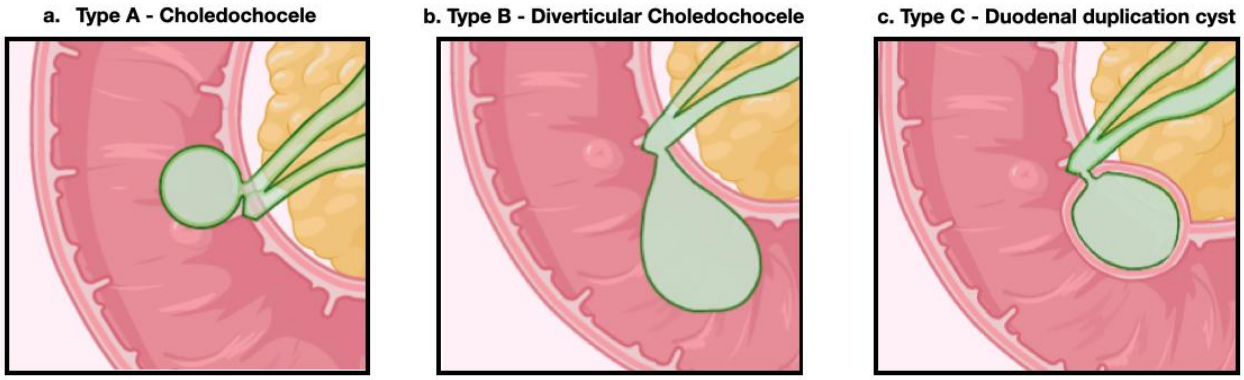


Fig. 1 Shows type A choledochocele, **b.** Shows type B choledochocele and **c.** shows Duodenal duplication cyst. See that B and C communicating to the ampulla. C covered with muscle coat that differentiate this from the type B choledochocele.

Etiopathogenesis

The etiology of CCs is the subject of an ongoing debate with supporters for both theories: a congenital or an acquired origin.

It is likely that most choledochal cysts arise in utero or in early infancy; however, the origin of choledochoceles is still debatable.

Schwegler and Boyden postulated that the primitive ampulla arises from 2 ostia during embryonic development, with the inferior ostium subsequently regressing and the superior ostium becoming the ampulla of Vater. Based on this paradigm, a choledochocele could develop from a rudimentary bile duct that, after failed or transposed regression, grow larger in size later in life.

Additional theories hypothesize that these lesions represent a congenital cyst or diverticulum of the intramural segment of the CBD.

Choledochoceles might also be acquired, possibly because of papillary inflammation leading to obstructive ballooning of the intramural CBD or a secondary effect of sphincter of Oddi dysfunction or stenosis, with the increased sphincter pressure causing development of a cystic dilatation.

The most accepted theory is Babbitt's theory, where Choledochoceles are supposed to be caused by an anomalous pancreaticobiliary duct junction (APBDJ) in which the pancreatic duct joins the bile duct 1–2 cm proximal to the sphincter of Oddi. The pancreatobiliary reflux is then followed by wall weakening and a diverticulum forms, with possible mucosal

dysplasia and malignization. This theory is supported by finding of high amylase levels in Choledochoceles bile. But this theory is questioned by some authors because APBDJ is observed in only 50–80% cases of Choledochoceles, and Choledochoceles detected antenatally do not have pancreatic juice reflux and neonatal acini do not secrete sufficient pancreatic enzymes. Obstruction of distal CBD is another theory, which is supported by studies on animal models. Sphincter of Oddi dysfunction has been reported in some studies as a predisposing factor to Choledochocele.

Malignancy

The increased risk of malignancy in CCs is well known and is proposed to occur because of chronic inflammation, cell regeneration, and DNA breaks, leading to dysplasia. Pancreatic reflux is also suspected to cause K-ras mutation, cellular atypia, P53 over expression, and carcinogenesis.

The reported incidence varies from 2.5% to 17.5% in patients with CCs. The incidence of malignancy increases with age, which means early diagnosis and treatment has a favorable outcome.

Choledochoceles and duodenal duplication cysts are mostly benign, but malignancy within these lesions has been described. The incidence of carcinoma in patients with symptomatic choledochoceles has been estimated at 2.5%, and only a handful of cases with concurrent ampullary carcinoma or cholangiocarcinoma have been reported.

Pancreatic cancer also rarely occurs in association with choledochoceles

Diagnosis

Abdominal ultrasound scan (US) is the first step toward confirmation of diagnosis. Sensitivity of ultrasound scan is about 71– 97%.

Magnetic resonance cholangiopancreatography is regarded as the “gold standard” for the diagnosis of CCs. Sensitivity has been reported to be as high as 90–100%.

Endoscopic retrograde cholangiopancreatography (ERCP) is reported to be the most sensitive diagnostic modality for CCs. ERCP is the choice of imaging modality in choledochoceles because therapeutic sphincterotomy can be performed at the time of diagnosis.

Type III CCs may need multiple imaging modalities before making a diagnosis.

Treatment

Type III CCs were historically treated by transduodenal excision and sphincteroplasty. But recently endoscopic sphincterotomy has become accepted to be sufficient treatment.

Conclusion

Clinical suspicion of CCs should be followed by early diagnosis and management in view of life-threatening complications and high risk of malignancy. A late diagnosis will worsen the prognosis.

Choledochocele unlike other CCs presents at an older age, with the average age at presentation being 51 years.

The standard treatment from bile duct cysts is surgical with specific approaches for each type according to its classification. In type IIIA cysts, it is recommended endoscopic sphincterectomy whilst types IIIB can be resected surgically or endoscopically. Endoscopic drainage of choledococele is a good option for conservative treatment.

Abbreviations

CBD: Common bile duct; CT: Computed tomography; USG: Ultrasonogram

Ethics approval and consent to participate

Obtained. Patient is in regular follow-up to the author.

The authors

Erick Servin Torres, Silvana Castelán Sánchez, Eulalio Jiménez Gonzalez. General Surgery Department, Instituto Mexicano del Seguro Social, Mexico City.

Authors' contributions. All authors read and approved the final manuscript.

References

[1] Todani T, Watanabe Y, Narusue M et al. Congenital bile duct cysts: classification, operative procedures, and review of thirtyseven cases including cancer arising from choledochal cyst. Am J Surg 1977; 134: 263–269

[2] F. Alonzo-Lej, W.B. Revor Jr, D.J. Pessagno, Congenital choledochal cyst, with a report of 2, and an analysis of 94 cases, Surg. Gynecol. Obstet. Int. Abstr. Surg. 108 (1) (1959) 1–30.

[3] Law R, Topazian M. Diagnosis and treatment of choledochoceles. Clin Gastroenterol Hepatol 2014; 12: 196–203

[4] Ohtsuka T, Inoue K, Ohuchida J et al. Carcinoma arising in choledochocele. Endoscopy 2001; 33: 614–619

[5] Kagiyama S, Okazaki K, Yamamoto Y. Anatomic variants of choledochocele and manometric measurements of pressure in the cele and the orifice zone. Am J Gastroenterol 1987; 82:641–649.

[6] Law R, Topazian M. Diagnosis and treatment of choledochoceles. Clin Gastroenterol Hepatol. 2014;12(2):196–203

[7] Wheeler W. An unusual case of obstruction to the common bile duct (choledochocele?). Br J Surg 1940;27:446–448.