Type VI choledochal cyst diagnosed on ERCP with direct cholangioscopy

Lawrence Ku, MD,[1] Marco A. Paez, MD,[1] Michael P. O'Leary, MD,[2] Danielle M. Hari, MD, FACS,[2] Sofiya Reicher, MD, FACG[1]

Choledochal cysts are rare congenital dilations of the biliary system. They are more common in East Asia than in the West. Choledochal cysts have been traditionally classified into 5 types.[1] A 6th type of choledochal cyst has been described as either a cyst arising off the cystic duct or a dilation of the cystic duct (Fig. 1). Data on type VI choledochal cysts are limited, with fewer than 25 cases reported.

Malignant transformation of these cysts has been described; from a review of the literature, malignancy was found in 2 of 15 adult patients and 0 of 9 pediatric patients.[2-13] The overall rate of malignancy ranges from 5% to 7.5%, with the highest rates reported in types I and IV, and the lowest rates in types II and III.[14]

Type VI choledochal cysts are typically managed with cholecystectomy. The definitive diagnosis of type VI choledochal cysts is usually made intraoperatively; the differentiation between type VI and type II cysts can be challenging on preoperative imaging. Here we present the case of a type VI choledochal cyst diagnosed by ERCP with direct cholangioscopy, allowing for preoperative planning for laparoscopic cholecystectomy, instead of cyst resection with bile duct reconstruction by means of Roux-en-Y hepaticojejunostomy appropriate for a type II choledochal cyst.

A 62-year-old Korean woman was evaluated for abdominal pain. Her liver function test results were as follows: alkaline phosphatase 84 U/L, aspartate aminotransferase 20 U/L, alanine aminotransferase 12 U/L, and total bilirubin 0.6 mg/dL.

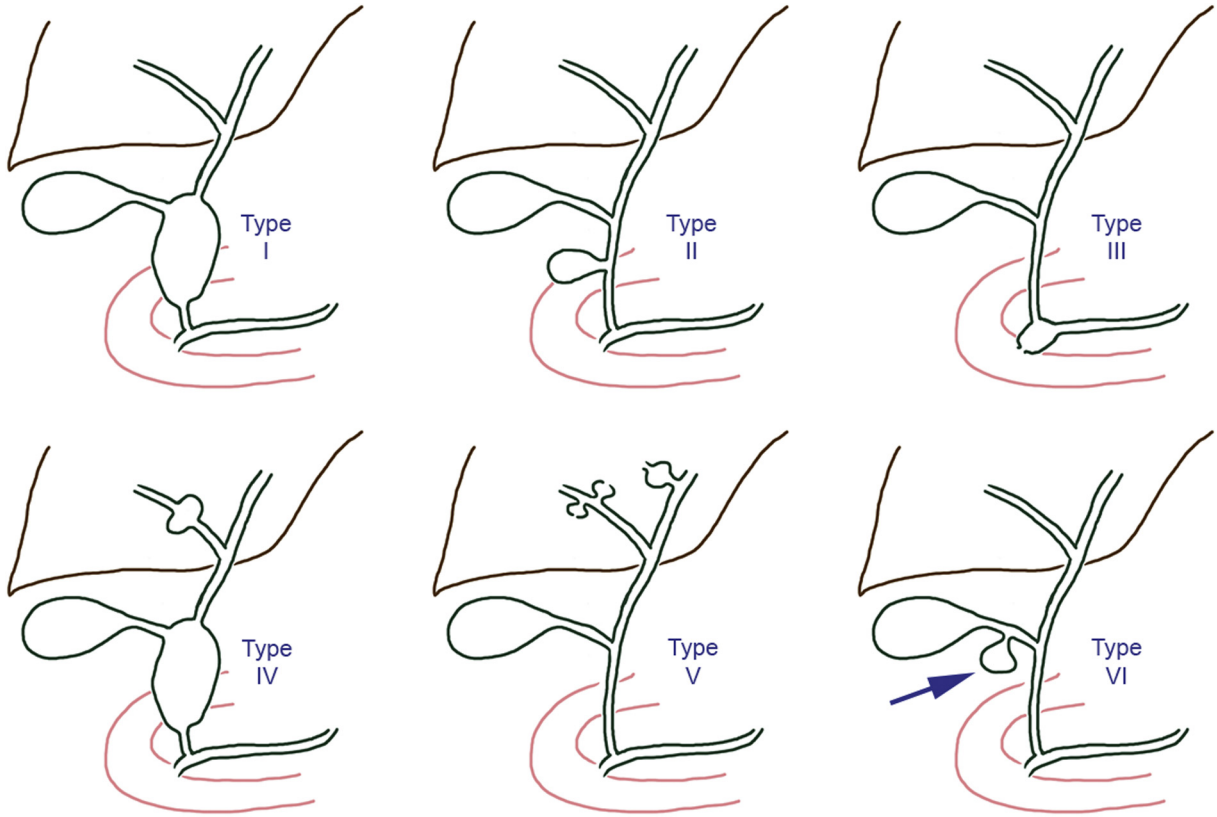

Figure 1. Choledochal cysts; type VI (*arrow*).

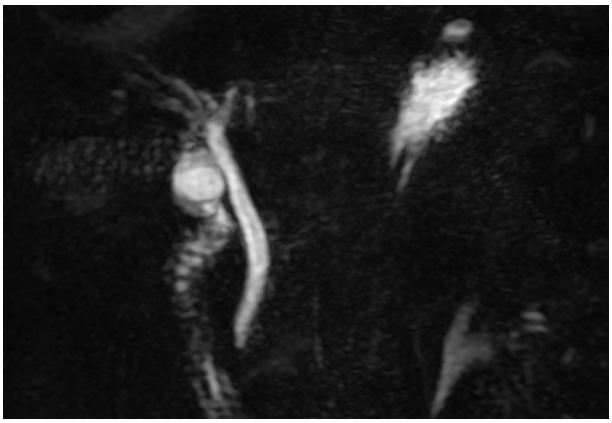

Figure 2. MRCP view of a choledochal cyst, likely type II.

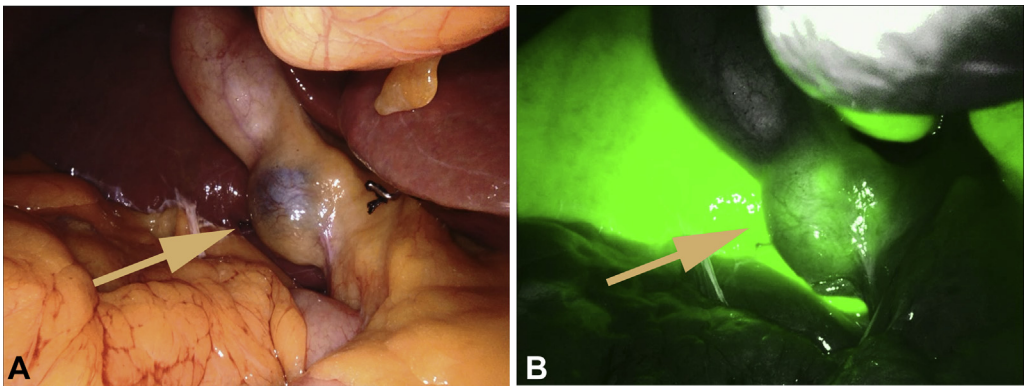

Figure 3. A, Type VI choledochal cyst (*arrow*) and gallbladder, intraoperative view. **B,** Type VI choledochal cyst (*arrow*) and gallbladder, intraoperative FireFly view.

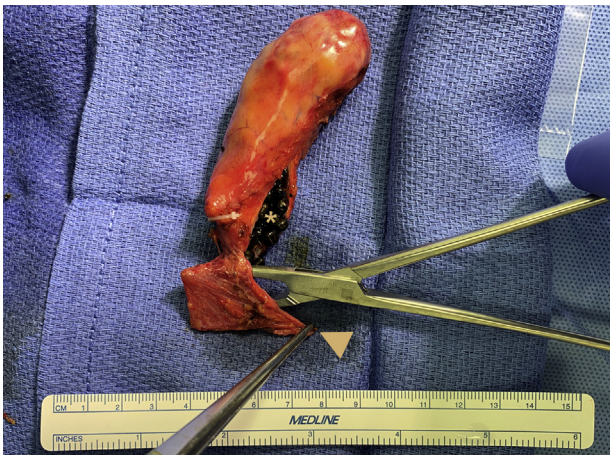

Figure 4. Resected en bloc specimen; Debakey forceps on the cystic duct (*arrow*), tonsil forceps inside the choledochal cyst, and gallbladder stones (*).

MRCP imaging revealed a 2- × 2-cm cystic structure adjacent to the cystic duct, common bile duct, and duodenum, suggestive of a type II choledochal cyst; however, a diagnosis could not be definitively made by radiology (Fig. 2).

Attendees of a multidisciplinary meeting, which included gastroenterology, surgical oncology, and radiology services, made the decision to proceed with EUS and ERCP to further characterize the cyst, inasmuch as differentiation of a type II from a type VI choledochal cyst would significantly change surgical management. The patient underwent EUS and ERCP with direct cholangioscopy (SpyGlass DS II; Boston Scientific, Inc, Marlborough, Mass, USA) (Video 1, available online at www.VideoGIE.org).

EUS demonstrated a 2-cm cystic structure adjacent to the common bile duct; no obvious mass was seen. ERCP with sphincterotomy and cholangioscopy identified an opening 4 cm distal to the bifurcation, which led to the cystic duct, and a cyst arising from the cystic duct; the ductal epithelium appeared normal, decreasing concerns for malignancy. A cholangiogram confirmed the cystic duct as the origin of the cyst and demonstrated contrast material filling the cyst followed by filling of the gallbladder immediately afterward.

The patient tolerated EUS/ERCP without adverse events. Subsequently, the patient underwent outpatient robotic multiport cholecystectomy with an intraoperative cholangiogram with the use of indocyanine green and the Da Vinci FireFly fluorescence system without adverse events

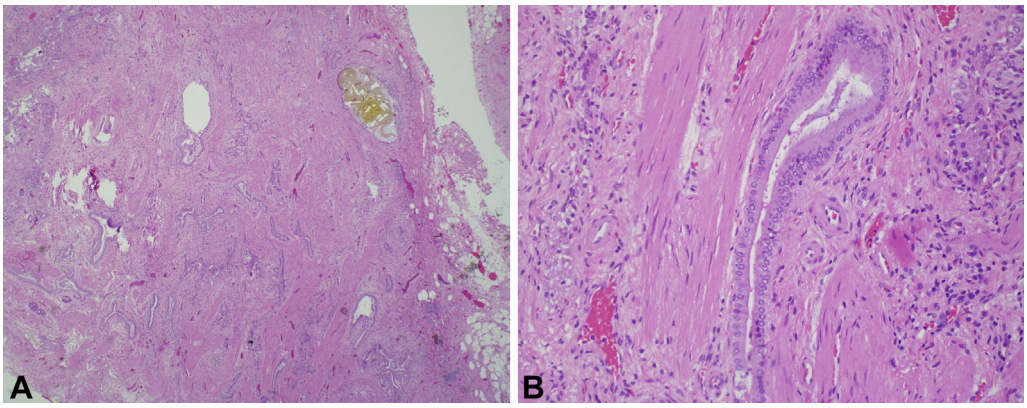

Figure 5. A, Type VI choledochal cyst without dysplasia (H&E, orig. mag. ×20). **B,** Type VI choledochal cyst without dysplasia (H&E, orig. mag. ×200).

(Fig. 3). The gallbladder and choledochal cyst were resected en bloc (Fig. 4). The patient was seen 2 weeks postoperatively and was asymptomatic. The final pathologic report confirmed the diagnosis of type VI choledochal cyst without dysplasia (Fig. 5).

DISCLOSURE

All authors disclosed no financial relationships relevant to this publication.

ACKNOWLEDGEMENTS

The authors thank Dr Michael J. Crabtree and Dr Samuel W. French of the Department of Pathology and Dr Alisa S. Han of the Department of Radiology for assisting with the interpretation of images for this case.

REFERENCES

1. Todani T, Watanabe Y, Narusue M, et al. Congenital bile duct cysts: classification, operative procedures, and review of thirty-seven cases including cancer arising from choledochal cyst. Am J Surg 1977;134:263-9.
2. Bode WE, Aust JB. Isolated cystic dilatation of the cystic duct. Am J Surg 1983;145:828-9.
3. Champetier P, Partensky C, Ponchon T. Cystic malformations of the cystic duct. Surg Radiol Anat 1987;9:287-91.
4. Kise Y, Uetsuji S, Takada H, et al. Dilatation of the cystic duct with its congenital low entry into the common hepatic duct. Am J Gastroenterol 1990;85:769-70.
5. Serena Serradel AF, Santamaría Linares E, Herrera Goepfert R. Cystic dilatation of the cystic duct: a new type of biliary cyst. Surgery 1991;109:320-2.
6. Bresciani C, Gama-Rodrigues J, Santos VR. Video-laparoscopic treatment of a sizeable cyst of the cystic duct: a case report. Surg Laparosc Endosc 1998;8:376-9.
7. Loke TK, Lam SH, Chan CS. Choledochal cyst: an unusual type of cystic dilatation of the cystic duct. AJR Am J Roentgenol 1999;173:619-20.
8. Conway WC, Telian SH, Wasif N, et al. Type VI biliary cyst: report of a case. Surg Today 2009;39:77-9.
9. Chan ES, Auyang ED, Hungness ES. Laparoscopic management of a cystic duct cyst. JSLS 2009;13:436-40.
10. Yoon JH. Magnetic resonance cholangiopancreatography diagnosis of choledochal cyst involving the cystic duct: report of three cases. Br J Radiol 2011;84:e18-22.
11. De U, Das S, Sarkar S. Type VI choledochal cyst revisited. Singapore Med J 2011;52:e91-3.
12. Maheshwari P. Cystic malformation of cystic duct: 10 cases and review of literature. World J Radiol 2012;4:413-7.
13. Shah OJ, Shera A, Shah P, et al. Cystic dilatation of the cystic duct: a type 6 biliary cyst. Indian J Surg 2013;75:500-2.
14. Ten Hove A, de Meijer VE, Hulscher JBF, et al. Meta-analysis of risk of developing malignancy in congenital choledochal malformation. Br J Surg 2018;105:482-90.

Division of Gastroenterology (1), Department of Surgery, Division of Surgical Oncology (2), Harbor-UCLA Medical Center, Torrance, California, USA.

https://doi.org/10.1016/j.vgie.2019.07.005