



Contents lists available at ScienceDirect

## International Journal of Surgery Case Reports

journal homepage: [www.casereports.com](http://www.casereports.com)

## An impressive choledochal cyst and its surgical resection



Nhu Thao Nguyen Galván\*, Kayla Kumm, Dor Yoeli, Ellen Witte, Michael Kueht, Ronald Timothy Cotton, Abbas Rana, Christine A. O'Mahony, John A. Goss

Division of Abdominal Transplantation and Hepatobiliary Surgery, Baylor College of Medicine, 6620 Main Street, Suite 1425, Houston, TX 77030, USA

## ARTICLE INFO

## Article history:

Received 26 October 2016

Received in revised form 19 February 2017

Accepted 19 February 2017

Available online 21 February 2017

## Keywords:

Hepaticojejunostomy

Choledochal cyst

Hepatobiliary surgery

Choledocholithiasis

Endoscopic retrograde

cholangiopancreatography or ERCP

Case report

## ABSTRACT

**INTRODUCTION:** Choledochal cysts are rare congenital dilations of the biliary tree that can present with non-specific symptoms such as abdominal pain, jaundice, cholelithiasis and pancreatitis. Although most commonly identified in children, they can be found in the adult population. However, because of the non-specific symptoms, this diagnosis may be difficult to make in the adult. A physician therefore must keep this diagnosis within their differential, as it may arise in an unexpected patient population who may present with a convoluted work up.

**CASE PRESENTATION:** In this report, we present the case of a 50-year-old African American woman with recurrent cholelithiasis, cholangitis and eventually obstructive jaundice despite undergoing a laparoscopic cholecystectomy six years prior. Her only work up at that point was a right upper quadrant ultrasound revealing gallbladder sludge, which led to her cholecystectomy. It was the persistence of her symptoms—abdominal pain, cholangitis and obstructive jaundice—previously attributed to chronic cholecystitis and choledocholithiasis that warranted further work up. After multiple physician visits, she was referred to our academic center after an ERCP was performed and she was found to have a dilation of her common bile duct consistent with a choledochal cyst. Furthermore, the ERCP identified multiple bile duct stones within the cyst. This was not identified on her original ultrasound or prior ERCPs. The patient underwent a complete cyst excision with Roux-en-Y hepaticojejunostomy and did well post-operatively.

**DISCUSSION:** This report illustrates how choledochal cysts can be an elusive diagnosis, but may present with repeated infections, recurrent biliary stones, and biliary obstruction despite a cholecystectomy. Had she an MRCP prior to her cholecystectomy, she would likely have avoided multiple surgeries, and years of persistent symptoms. Choledochal cysts are associated with an increased risk of biliary malignancy and therefore cyst excision is the standard of care.

**CONCLUSION:** Although rare, physicians need to keep this diagnosis in mind, and be aware of the clinical and imaging findings consistent with a choledochal cyst in order to facilitate appropriate work up, referral and treatment.

© 2017 The Author(s). Published by Elsevier Ltd on behalf of IJS Publishing Group Ltd. This is an open access article under the CC BY-NC-ND license (<http://creativecommons.org/licenses/by-nc-nd/4.0/>).

## 1. Introduction

Choledochal cysts are congenital dilations of the biliary tree and represent a rare diagnosis in the western hemisphere, afflicting females greater than males [1,2]. Classically they are diagnosed in children, though it can be diagnosed in adults [4]. Presentation in adults often includes abdominal pain, nausea, vomiting, cholelithiasis, cholangitis, pancreatitis or malignancy [3,4]. These are vague symptoms that can be misleading in the adult patient. A choledochal cysts carries up to a 10% risk of malignant transformation to cholangiocarcinoma [3] and even with resection, a patient's risk

for biliary malignancy does not return to the population baseline [4].

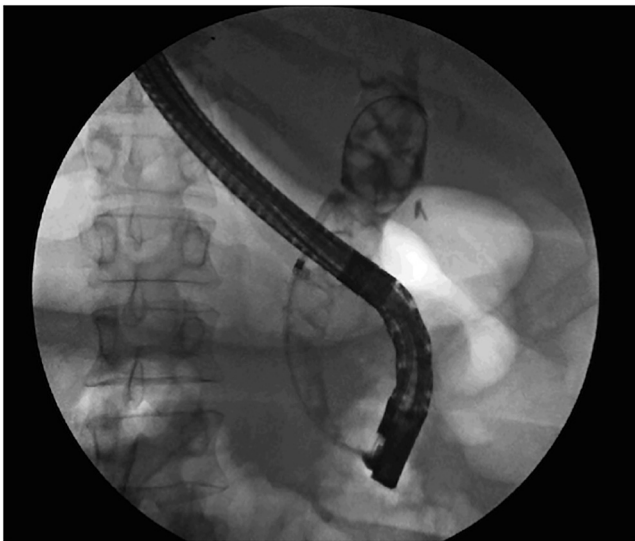
We present a rare case of a choledochal cyst that presented as recurrent cholangitis and choledocholithiasis years after her laparoscopic cholecystectomy. Delayed imaging revealed the choledochal cyst had fusiform dilation for the entirety of the extrahepatic bile duct, type Ia based on the Todani classification schema [5]. The patient was eventually referred to our institution for surgical management. This work has been reported in line with the SCARE criteria, a consensus on surgical case report guidelines [6].

## 2. Presentation of case

The patient is a 50-year-old obese African-American woman with a BMI of 43 and a past medical history significant for asthma, gastroesophageal reflux, hypertension and pancreatitis who underwent cholecystectomy 6 years prior to referral to our institution.

\* Corresponding author.

E-mail addresses: [thao.galvan@bcm.edu](mailto:thao.galvan@bcm.edu), [nhuthao.galvan@gmail.com](mailto:nhuthao.galvan@gmail.com) (N.T.N. Galván).



**Fig. 1.** ERCP demonstrating choledochal cyst with intraluminal biliary stones.

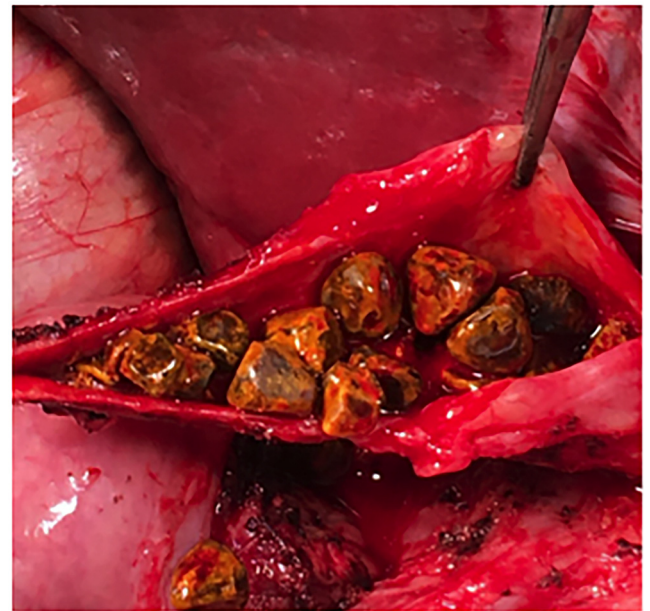


**Fig. 2.** ERCP demonstrating intrahepatic biliary dilation proximal to the choledochal cyst.

Following her initial operation, she continued to have multiple episodes of choledocholithiasis leading to cholangitis, manifested as colicky abdominal pain accompanied by nausea and vomiting. Her only work up at that point was a right upper quadrant ultrasound revealing gallbladder sludge, which led to her cholecystectomy. It was the persistence of her symptoms—abdominal pain, cholangitis and obstructive jaundice—previously attributed to chronic cholecystitis and choledocholithiasis that warranted further work up. These episodes required multiple ERCPs with intermittent biliary stent placements and stone removals for her recurrent symptoms. At the time of referral, she presented to an outside hospital with obstructive jaundice and scleral icterus, revealing hyperbilirubinemia and elevated alkaline phosphatase. Another ERCP at that time revealed something it had not previously— multiple dilated central intrahepatic ducts with a grossly dilated segment of the common hepatic duct containing large biliary stones, concerning for a type Ia choledochal cyst, with proximal biliary obstruction (Figs. 1 and 2). This corresponded to an abdominal CT performed during the same admission that revealed similar findings. Due to these diagnostic findings, she was referred to our institution for surgical management.

Upon review of her history and imaging, a complete surgical resection was indicated for her recurrent episodes of cholangitis and to reduce the risk of malignancy. To optimize her surgical outcome, we recommended weight loss and smoking cessation. She was also referred to pulmonology for risk stratification and optimization of her asthma.

She underwent an exploratory laparotomy with resection of the cyst and Roux-en-Y hepaticojejunostomy by an experienced hepatobiliary surgeon. Intra-operatively, the impressive choledochal cyst was opened and found filled with large biliary stones (Fig. 3); furthermore, a right posterior duct was identified which was included as a second anastomosis to the jejunal limb of the Roux-en-Y hepaticojejunostomy (Fig. 4). Considering the impressive size of this cyst, it is remarkable that it was not identified during her laparoscopic cholecystectomy. Histological examination was done at the time of the operation via frozen section microscopy and revealed no evidence of adenocarcinoma at either margin. This was confirmed on final pathology, which revealed congested biliary mucosa with chronic inflammation and focal squamous metaplasia. The patient did well post operatively and was discharged on POD 7.



**Fig. 3.** Intra-operative photograph of choledochal cyst containing large biliary stones.

At the time of this report the patient was 6 months post-op and continued to do well with resolution of her abdominal pain, nausea, vomiting and episodes of cholangitis.

### 3. Discussion

Choledochal cysts are rare in the US, afflicting 1 out of 150,000 patient, usually in their first year of life, and this report illustrates how they can lead to repeated infections, recurrent biliary stones, and biliary obstruction. More notably, this diagnosis was difficult to make, and in fact, was missed initially, despite an ultrasound, a laparoscopy and multiple ERCPs. Most

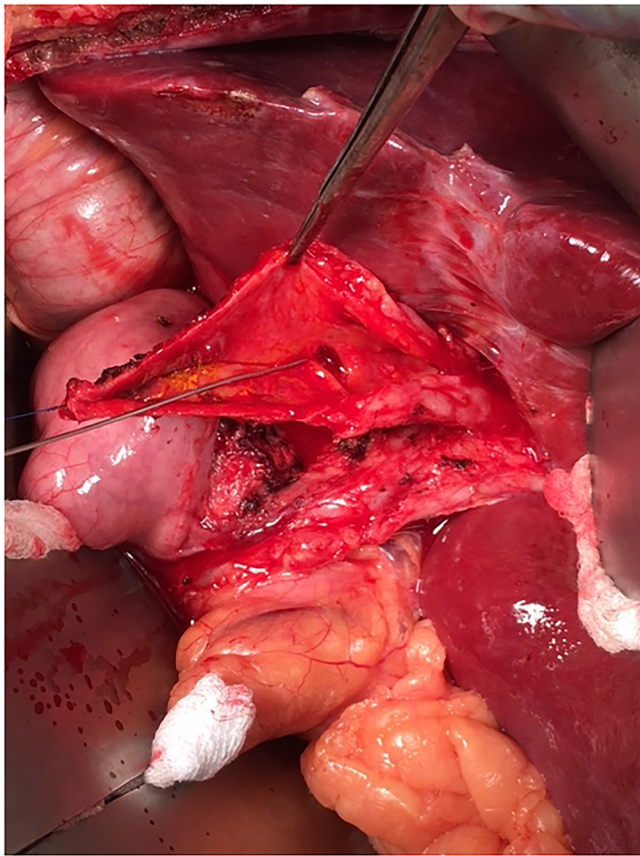


Fig. 4. Intra-operative photograph of a right posterior duct.

choledochal cysts are diagnosed before the age of 10, and are hypothesized to develop at least partially from anomalous formation of the pancreaticobiliary ductal junction, which could result in pancreobiliary reflux and mixing [1]. This leads to cholangitis and bile duct wall destruction, in turn resulting in biliary stricture from scarring and then proximal dilation [5]. Surgical intervention is the standard treatment modality for this disease process, and a risk reduction measure for the development of cholangiocarcinoma [7]. Had this patient an MRCP prior to her cholecystectomy, she would likely have avoided multiple surgeries, and years of persistent symptoms, as well as increased cancer risk.

In this case, the patient had undergone cholecystectomy without recognition of a choledochal cyst during preoperative planning or at the time of the operation. She then continued to have episodes of choledocholithiasis with repeated ERCP interventions prior to recognition of a choledochal cyst. Once recognized, appropriate surgical intervention was performed to remove the cyst completely. Doing so has led to a resolution of the patient's symptoms and reduction in her risk for cholangiocarcinoma. Although the symptoms of choledochal cysts can be vague, practitioners must have some degree of clinical suspicion in order to obtain the appropriate diagnostic work up and to facilitate appropriate referral and treatment of choledochal cysts.

#### Conflicts of interest

The authors have no financial or personal relationships to disclose.

#### Sources of funding

No sources of funding were necessary for this study.

#### Ethical approval

IRB approval was obtained through the Baylor College of Medicine.

Protocol: H-38333.

Furthermore, patient consent was obtained, allowing report of their medical disease, work up and treatment, so long as identifying information was omitted.

#### Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

#### Authors contribution

N. Thao Galvan – Study design, collection, analysis, writing.  
 Kayla Kumm – Study collection, analysis, writing.  
 Dor Yoeli BS – writing.  
 Ellen Witte BS – writing.  
 Michael Kueht MD – writing.  
 Ronald Timothy Cotton MD – Study design.  
 Abbas Rana MD – Study design, collection, analysis, writing.  
 Christine A. O'Mahony MD – Study design, collection.  
 John Goss – Study design, collection, analysis, writing.

#### Guarantor

N. Thao Galvan MD MPH.

#### References

- [1] M.A. Moslim, H. Takahashi, F.G. Seifarth, R.M. Walsh, G. Morris-Stiff, Choledochal cyst disease in a western center: a 30-year experience, *J. Gastrointest. Surg.* 20 (August (8)) (2016) 1453–1463.
- [2] K. Soreide, J.A. Soreide, Bile duct cyst as precursor to biliary tract cancer, *Ann. Surg. Oncol.* 14 (3) (2007) 1200–1211.
- [3] S.M. Ronnekleiv-Kelly, K.C. Soares, A. Ejaz, T.M. Pawlik, Management of choledochal cysts, *Curr. Opin. Gastroenterol.* (2016) (Ahead of print).
- [4] K.C. Soares, Y. Kim, G. Spolverato, et al., Presentation and clinical outcomes of choledochal cysts in children and adults: a multi-institutional analysis, *JAMA Surg.* 150 (2015) 577–584.
- [5] T. Todani, Y. Watanabe, M. Narusue, K. Tabuchi, K. Okajima, Congenital bile duct cysts: classification, operative procedures, and review of thirty-seven cases including cancer arising from choledochal cyst, *Am. J. Surg.* 134 (August (2)) (1977) 263–269.
- [6] R.A. Agha, A.J. Fowler, A. Saetta, I. Barai, S. Rajmohan, D.P. Orgill, the SCARE Group, The SCARE statement: consensus-based surgical case report guidelines, *Int. J. Surg.* 34 (2016) 180–186.
- [7] D. Mantas, P. Stamopoulos, E. Kouskos, D. Dimitroulis, Giant biliary cyst, *J. Gastrointest. Surg.* 26 (April) (2016).

#### Open Access

This article is published Open Access at [sciedirect.com](http://sciedirect.com). It is distributed under the [IJSCR Supplemental terms and conditions](#), which permits unrestricted non commercial use, distribution, and reproduction in any medium, provided the original authors and source are credited.