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Case Report

A Unique and Rare Presentation of Obstructed Choledochal Cyst in an Adult: A Case Report

Ahmad E. Al-Mulla, MD[1*]; Salah Termos, MD[2]; Fawzia Ashkanani, MD[1]; Ehab S. Imam, MD, PhD[1]; Abdulla E. Sultan, MD[1]; Ali Altabeekh, MD[1]; Mohammad Y. Saleh, MD[1]; Ahmad Al-Jafar, MD[1]

[1]*Department of Surgery, Farwaniya Hospital, Ministry of Health Kuwait, Farwaniya, Kuwait*
[2]*Hepato-Biliary and Pancreas and General Surgery, Al- Amiri Hospital, Kuwait City, Kuwait*

[*]***Corresponding author***
Ahmad E. Al-Mulla, MD
Senior Specialist General and Bariatric Surgery, Department of Surgery, Farwaniya Hospital, Ministry of Health Kuwait, P. O. Box 13373, Farwaniya 81004, Kuwait; Tel. 0096524888000, 00965-99833454; E-mail: draalmulla2007@gmail.com

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ABSTRACT

Choledochal cysts in adults are rare congenital abnormalities. Approximately 80% are found in childhood. Thus, their presentation in adults is always associated with complications, such as stone formation, inflammation and malignancies. The pathophysiology of this disease is yet uncertain. There are different types of choledochal cysts. Diagnosis can be challenging clinically; however, imaging techniques, such as ultrasound, magnetic resonance cholangiopancreatography (MRCP) and computed tomography (CT), can be helpful. We found several procedures performed in the extant literature, such as choledochoduodenostomy and choledocho-jejunostomy submucosal excision of the cyst; however, the best surgical option is excision with hepaticojejunostomy. We discuss the unusual presentation of a 33-year-old female patient with an obstructed choledochal cyst, despite having undergone a drainage procedure in childhood.

Keywords
Choledochal cyst; Hepato-jejunal anastomosis; Biliary system anomaly; Choledochal cyst in adults.

INTRODUCTION

A choledochal cyst is the unusual dilatation of the bile duct with intra- or extra-hepatic dilatation.[1] The incidence of choledochal cysts is one in 100,000-150,000 live births in the West. They are more prevalent in females than males, occurring at a ratio of 3-4:1.[2] Eighty percent of cases are diagnosed in childhood; therefore, cases in adulthood are always associated with complications. Choledochal cysts are associated with many complications, such as stone formation, secondary biliary cirrhosis, cholangitis, cyst rupture, obstructive jaundice and malignancy (cholangiocarcinoma).[3] This case study presents a rare case of a female with obstructive jaundice due to a choledochal cyst that was drained in childhood.

CASE PRESENTATION

A 33-year-old female Sri Lankan patient presented to our surgical casualty, complaining of four-days of sharp abdominal pain, mainly in the epigastric region and radiating to the back, associated with anorexia, vomiting and nausea. The patient mentioned having darker urine and pale stool. She had a background history of undergoing a previous surgical procedure when she was four years old but could not recall the type of procedure. Upon examination (pulse 90 beats per minute, temperature 37 °C, blood pressure 90/70 mmHg), we determined that she was jaundiced, with a laparotomy scar and mild tenderness at the epigastric region. Initial investigations revealed an increase in white blood cells (WBC) 26×109, haemoglobin (Hb) 9.7 and platelets (Plts) 298. The liver profile showed alkaline phosphatase (ALP) 168, alanine aminotransferase (ALT) 138, aspartate aminotransferase (AST) 430, gamma glutamyl transferase (GGT) 139, total bilirubin 60, direct bilirubin 30 and amylase 1170. The kidney profile showed creatinine (Cr) 50, potassium (K) 3.5 and sodium (Na) 135.

The chest and abdomen X-rays were unremarkable. The abdomen ultrasound showed interstitial pancreatitis with a normal gallbladder. The patient was resuscitated with intravenous fluids and antibiotics (piperacillin-tazobactam and metronidazole). Due to her uncertain history, the surgical team ordered a computed tomography (CT) scan for the abdomen and pelvis, which showed

a choledochal cyst (Type I) and acute cholecystitis (Figure 1). A magnetic retrograde cholangiopancreatography (MRCP) also was ordered, which showed distal common bile-duct (CBD) stones and dilated intra- and extra-hepatic radicles with a choledochal cyst (Type Ib) (Figure 2). The patient underwent endoscopic retrograde cholangiopancreatography (ERCP). The patient's previous drainage procedure for a congenital choledochal cyst has caused a blockage, resulting in her current symptoms. The case was discussed with the hepatobiliary team, who decided on a choledochal cyst and cholecystectomy with hepatic-jejunostomy and jejuno-jejunostomy.

INTRA-OPERATIVE

The hepatobiliary team elected to proceed with a laparotomy. Extensive adhesiolysis was performed due to severe adhesions from the patient's previous surgery. The team worked to identify the choledochal cyst and the old cholecystojejunostomy, which was obstructed by gallstones and sludge. The cyst was carefully dissected from the porta hepatis: the hepatic artery and portal vein. The surgical team performed *en bloc* excisions of the choledochal cyst, gallbladder and segmental jejunal with stent removal (Figure 3). A Roux-En-Y (three-ducts anastomosis) jejuno-jejunostomy and a Hutson Russel loop at the right upper quadrant were made. Two drains were placed at the anastomosis and pelvis.

POST-OPERATIVE

The patient's post-operative recovery on the ward was uneventful. She continued taking antibiotics while hospitalised and did not experience any bile leakage from her drains. After five-days, the patient was discharged and resumed a regular diet. The drains were removed, and her liver profile was normal. The first outpatient visit after 14-days was unremarkable. The patient tolerated a regular diet with no abdominal pain, fever or changes in bowel habits. The wound was clean, and the patient's histopathology result showed acute cholecystitis and a choledochal cyst.

DISCUSSION

Choledochal cysts are rare biliary system abnormalities; however, in recent years, incidences of choledochal cysts have increased from 1:128,000 to 1:38,000.[4] Nevertheless, they are primarily found in children rather than in adults. Various theories have described the aetiology of choledochal cysts, but none are specific. The most accepted theory is the presence of an anomalous junction between the pancreatic and the bile duct (APBDJ) outside the duodenal wall. The abnormal pancreatic bile-duct junction is located 1 cm proximal to where the CBD reaches the ampulla of Vater.[5,6] The anomalous pancreatic bile-duct junction allows the reflux of pancreatic enzymes, which enter the biliary system, leading to inflam-

Figure 1. *CT Scan of the Abdomen and Pelvis, Showing the Choledochal Cyst*

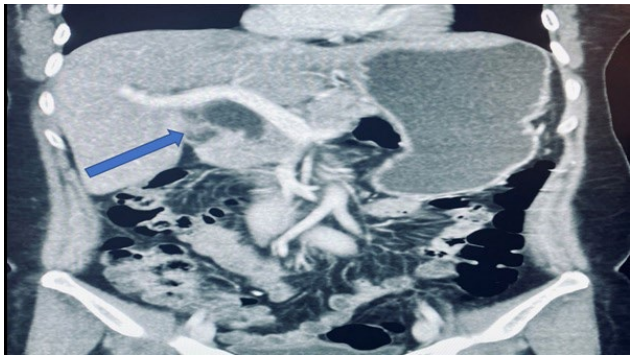


Figure 2. *Magnetic Resonance Cholangiopancreatography Highlighting a Type Ib Choledochal Cyst*

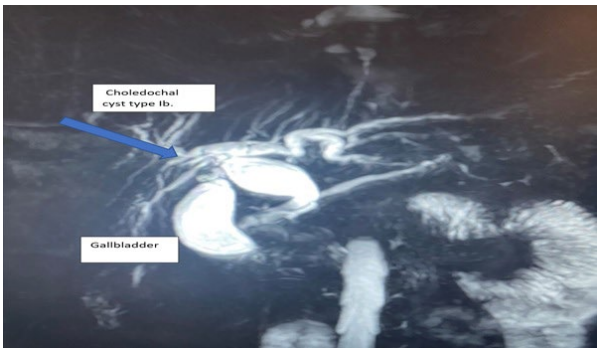
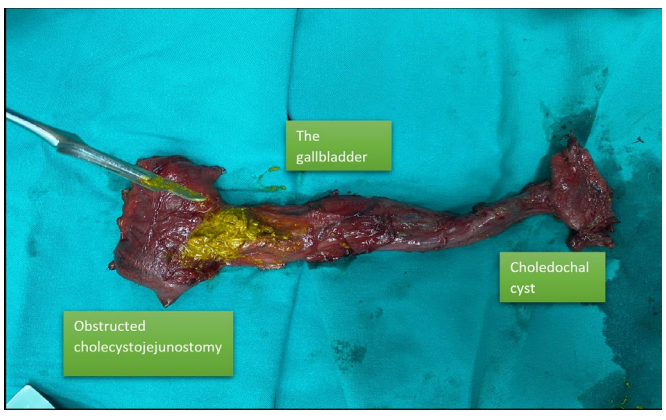


Figure 3. *En Bloc Resection of the Gallbladder, Choledochal Cyst and an Obstructed Cholecystojejunostomy*



mation, damage, dilatation, and cyst formation predisposition. The first systematic classification of choledochal cysts, presented in Alonso-Lej et al[7] described four types of cysts (I-VI). This classification system was later modified by Todani and colleagues, who added a fifth type due to intrahepatic dilatation referred to as caroli disease.[4,8] Between 50 to 80% of cases present with Type I, according to Todani's classification of choledochal cysts, and it is the most prevalent presentation.[9] It is associated with 80% of other hepatobiliary pathologies, such as intrahepatic lithiasis, pancreatitis, acute cholecystitis and malignant neoplasm. Patients with choledochal cysts are 20 times more likely to develop cholangiocarcinoma than the general population. Other neoplastic diseases have also been reported, such as neuroendocrine tumours.[4,10]

The most common presentation of a choledochal cyst is abdominal pain. The classic triad of abdominal pain, jaundice and a palpable abdominal mass occur in fewer than 20% of cases.[11] A retrospective study of 14 adults revealed different symptoms, including three biliary infections, three instances of pancreatitis, two abdominal pain and one painless jaundice. Three patients also had cysts identified during laparoscopic cholecystectomies, and two had incidental findings from CT scans.[12] Therefore, it is hard to diagnose a choledochal cyst symptomatically.

Bile-duct cysts are diagnosed *via* imaging techniques, such as ultrasound, CT, MRCP and ERCP. An abdominal ultrasound is an initial investigation to evaluate the presence of a dilated bile duct and gallbladder stones.[13] MRCP is the gold standard of choice for diagnosis, as it is a non-invasive technique that does not require any ionising contrast.[14]

Once the diagnosis is confirmed, the bile-duct cysts must be removed surgically to prevent future complications. Surgical resection depends on the type of choledochal cyst. Type I cysts need resection and reconstruction of the bile duct with a hepaticojejunostomy (also known as a Roux-en-Y procedure). Type II cysts can usually be excised, and the defect in the CBD can be sutured on a T-tube. Type III cysts can be partially excised and opened into the duodenum using either a trans-duodenal sphincteroplasty or endoscopic sphincterotomy.[15] Type VI and V cysts require a multiple disciplinary team approach (endoscopy, intervention radiology and surgery) and a possible partial hepatectomy.[16] In our case report, the patient underwent a drainage procedure as a child; thus, due to incomplete excision of the cyst, the patient developed an obstruction, leading to her current situation.

CONCLUSION

Choledochal cysts are extremely rare pathologies in adults because they mainly appear in childhood. They may present with ambiguous signs and symptoms; diagnosing such a disease is challenging and requires careful investigation. Treatments for this disease are diverse; however, resection and hepaticojejunostomy remain the best bilioenteric therapeutic options. Awareness of this congenital abnormality and familiarity with its potential complications can lead to better outcomes. We recommend referring those patients to hepatobiliary specialists to follow the appropriate therapeutic management to avoid future complications and have better prognoses.

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DISCLOSURE

None.

CONFLICTS OF INTEREST

The authors declare that they have no conflicts of interest.

REFERENCES

1. Mann CV, Russell RCG. Gall bladder and bile duct. In: Russell RCG, ed. *Bailey and Love Short Practice of Surgery*. 23rd ed. London, UK: Chapman and Hall; 2000: 973-994.

2. Baison GN, Bonds MM, Helton WS, Kozarek RA. Choledochal cysts: Similarities and differences between Asian and Western countries. *World J Gastroenterol.* 2019; 25(26): 3334-3343. doi: 10.3748/wjg.v25.i26.3334

3. Kumar M, Rajagopalan S. Choledochal cyst. *Med J Armed Forces India.* 2012; 68(3): 296-298. doi: 10.1016/j.mjafi.2012.04.011

4. de Albuquerque VVML, De Macedo FP, Costa KG, Nunes ZRS, da Silva Junior RA. Choledochal cyst - unusual presentation in the adult phase: Case report. *Int J Surg Case Rep.* 2020; 70: 33-36. doi: 10.1016/j.ijscr.2020.03.014

5. Stringer MD, Dhawan A, Davenport M, Mieli-Vergani G, Mowat AP, Howard ER. Choledochal cysts: Lessons from a 20-year experience. *Arch Dis Child.* 1995; 73(6): 528-531. doi: 10.1136/adc.73.6.528

6. Burkhalter E. Choledochal cyst. A case report. *Am Surg.* 1990; 56(6): 377-379.

7. Alonso-Lej F, Rever WB, Pessagno DJ. Congenital choledochal cyst, with a report of 2, and an analysis of 94, cases. *Int Abstr Surg.* 1959; 108(1): 1-30.

8. Todani T, Watanabe Y, Narusue M, Tabuchi K, Okajima K. Congenital bile duct cysts: Classification, operative procedures, and review of thirty-seven cases including cancer arising from choledochal cyst. *Am J Surg.* 1977; 134(2): 263-269. doi: 10.1016/0002-9610(77)90359-2

9. Singham J, Yoshida EM, Scudamore CH. Choledochal cysts: part 1 of 3: Classification and pathogenesis. *Can J Surg.* 2009; 52(5): 434-440.

10. Dahiya D, Raman K, Singh H. Neuroendocrine tumor in choledochal cyst – case report and review of literature. *Pol Przegl Chir.*

2011; 83: 627-629. doi: 10.2478/v10035-011-0099-0

11. Dhupar R, Gulack B, Geller DA, Marsh JW, Gamblin TC. The changing presentation of choledochal cyst disease: An incidental diagnosis. *HPB Surg*. 2009; 2009: 1037-1039. doi: 10.1155/2009/103739

12. Singham J, Yoshida EM, Scudamore CH. Choledochal cysts: Part 2 of 3: Diagnosis. *Can J Surg*. 2009; 52: 506-511.

13. Chijiiwa K, Koga A. Surgical management and long-term follow-up of patients with choledochal cysts. *Am J Surg*. 1993; 165(2): 238-242. doi: 10.1016/s0002-9610(05)80518-5

14. Govil S, Justus A, Korah I, Perakath A, Zachariah N, Sen S. Choledochal cysts: Evaluation with MR cholangiography. *Abdom Imaging*. 1998; 23(6): 616-619. doi: 10.1007/s002619900415

15. Liu CL, Fan ST, Lo CM, Lam CM, Poon RT, Wong J. Choledochal cysts in adults. *Arch Surg*. 2002; 137(4): 465-468. doi: 10.1001/archsurg.137.4.465

16. Metcalfe MS, Wemyss-Holden SA, Maddern GJ. Management dilemmas with choledochal cysts. *Arch Surg*. 2003; 138(3): 333-339. doi: 10.1001/archsurg.138.3.333