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Adult Giant Choledochal Cyst Conundrum: A Case Report

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1. Abstract

1.1. Introduction: Choledochal cyst is rare malformation of the biliary system, characterized by abnormal cystic dilation in the biliary duct. There is no clear explanation regarding the etiology of the disease. Choledochal cysts are more common in Asian population, mainly diagnosed in children, particularly in females. Due to the rare incidence of giant choledochal cyst, evidence regarding safe operative approach was still scarce. This study aims to present the management option of type 1 giant choledochal cyst.

1.2. Case presentation: We present a 26-year-old female patient with a chief complaint of intermittent right upper quadrant abdominal pain, which radiated to the back. The pain worsened while the patient was pregnant. The patient was then diagnosed with a giant choledochal cyst and concomitant left- and right- hepatic duct dilation with multiple gallstones.

1.3. Results: We utilize Lilly's technique, which involves cauterizing the mucosa while leaving the serosa connected to the adhering tissues. This method is helpful in giant long standing cyst, where the mucosa could not be entirely removed because it adhered to another structure, such as the pancreatic parenchyma, inferior vena cava, or hepatic hilar arteries.

Conclusion: A complicated gigantic Type 1 choledochal cyst case benefits from Lilly's method.

2. Introduction

Choledochal cyst are rare malformation in the biliary system, characterized by abnormal cystic dilation in the biliary duct. The incidence of choledochal cyst varies from 1 in 100,000 cases in Western countries to 1 in 13,000 in Asian countries. Epidemiologically, choledochal cysts are approximately four times more common in females [1]. There is no clear explanation regarding the etiology of choledochal cyst, but it is believed to be a congenital condition. Anomalous connection between the biliary duct and pancreas (pancreaticobiliary junction) that allows pancreatic enzymes reflux into the biliary system is the most widely accepted etiology theory for the disease. These conditions result in inflammation, metaplasia, and further transformation into malignancy of the endothelial lining [2,3].

In 1977, The Todani classification was established, which divided choledochal cyst into 5 types: type I, type II, type III, type IV, and type V. Among the five types of choledochal cyst, the majority of cases are type 1. The cyst with a diameter of more than 10 cm referred as giant choledochal cyst [4]. The treatment of choledochal cyst may involve supportive care, endoscopic procedures, resection, or liver transplantation [3]. Due to the rare incidence of giant choledochal cyst, evidence regarding safe operative approach was still scarce [5]. Therefore, we propose a case of giant choledochal cyst treated with total cystectomy with Roux-en-Y hepaticojejunostomy.

3. Case Presentation

A 26-year-old female came to our digestive surgery unit with a chief complaint of intermittent right upper quadrant abdominal pain. The pain radiated to the back, accompanied with nausea and loss of appetite. There was no jaundice, fever, and tenderness in physical examination. Laboratory investigations showed total bilirubin, direct bilirubin, and indirect bilirubin of 5.24 mg/dL, 4.43 mg/dL, 0.81 mg/dL respectively. Liver function test showed ALT 169 U/L and AST 173 U/L. Radiological examination was also performed on the patient. Magnetic Resonance Cholangio Pancreaticography (MRCP) with contrast examination revealed massive cystic common bile duct dilation measuring 11.1 cm x 21.8 cm x 17.2 cm in size (Figure 1). Dilation was also seen in the right and left intrahepatic duct. There were also multiple gallstones.

The patient was diagnosed with obstructive jaundice due to giant choledochal cyst. Total cystectomy with Roux-en-Y hepaticojejunostomy using laparotomy approach was performed. Patient was in a supine position with general anesthesia. Sharp adhesiolysis was initially performed due the presence of previous cholecystectomy adhesion. We further identified the choledochal cyst, with subsequent tunnelling of the cyst. Then, total excision was performed to remove the cyst. In our patient, there were gallstones in the left hepatic duct. Therefore, left lobe hepatectomy was done followed by stone extraction until none left. Reconstructive procedure using Roux-en-Y hepaticojejunostomy was utilized in the patient. Surgery was then closed after primary suture. Intraoperative operative was shown in (Figure 2). Postoperatively, laboratory investigations revealed decrease in total bilirubin (0,4 mg/dL), direct bilirubin (0,2 mg/dL), indirect bilirubin (0,2 mg/dL). There were also decrease in ALT (29 U/L) and AST (37 U/L). Patient reported being satisfied with her current condition.

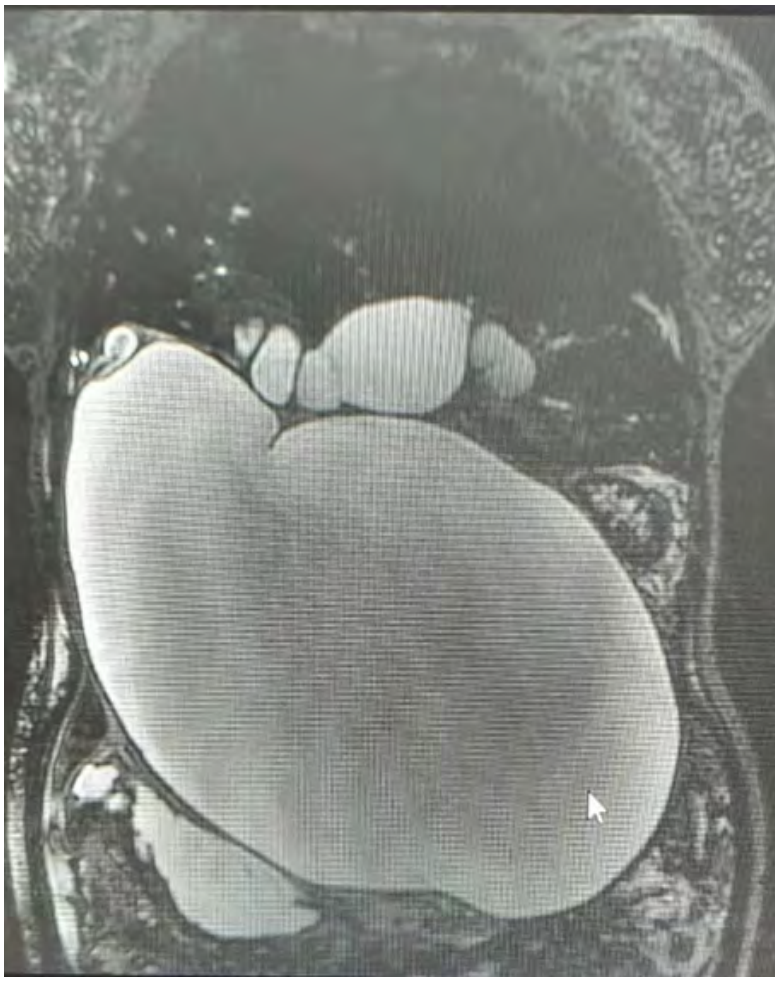

Figure 1: Preoperative MRCP revealed massive cystic common bile duct dilation

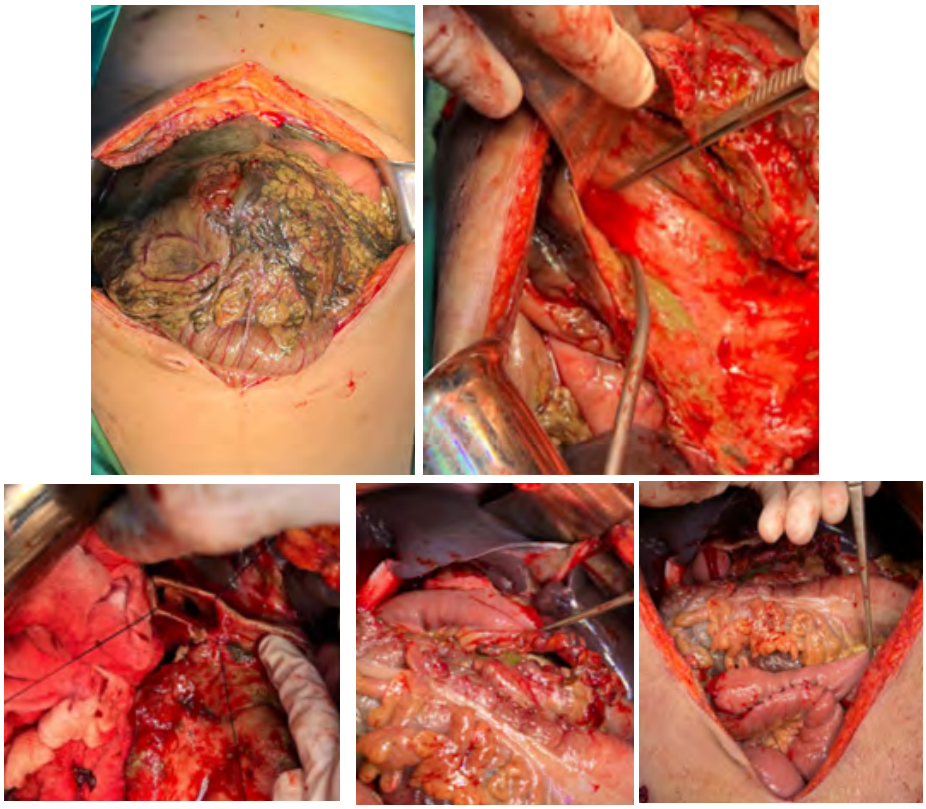

Figure 2: (a) Choledochal cyst; (b) Primary suture performed in common bile duct (c) Common hepatic duct lumen; (d) Roux-en-Y hepaticojejunostomy; (e) Jejuno-jejunostomy.

4. Discussion

Giant choledochal cyst is a rare entity of biliary system characterized by abnormal cystic dilation in the biliary duct. Although the discovery was vague, the patient may come with the usual triad of abdominal discomfort, jaundice, and palpable mass. According to a previous study, only 25% of adults had more than one of the choledochal cyst's defining characteristics. In a similar vein, our patient reported right upper quadrant stomach pain without any obvious signs of jaundice or a palpable tumor during a physical examination. Choledochal cyst should not be ruled out even if it lacks the majority of the traditional triads [6]. There were no particular laboratory tests performed to support the choledochal cyst diagnosis. Ultrasonography is one of the additional diagnostic tools (preferably in children). Regarding their ability to precisely visualize cystic regions in the biliary system in adults, ERCP and MRCP are more frequently used [7,8]. We first ran bilirubin and liver enzyme testing as part of our liver function analysis (ALT and AST). The presence of obstructive jaundice was indicated by our patient's high direct bilirubin level. We confirmed a diagnosis using MRCP as well.

Based on the anatomical position of the cyst, Todani classified choledochal cysts into Type I, Type II, Type III, Type IV, and Type V. The bulk of choledochal cyst cases (between 50 and 80 percent) fall within Type I. Common bile duct dilatation with cystic characteristics was a hallmark of type I. The common bile duct in our patient contained a massive choledochal cyst that was classified as Todani Type I [9]. Various clinical issues can arise in adult patients following choledochal cysts excision due to the variety of choledochal cysts features. To maintain a balance between the risk of surgery and the potential risk of late complications, surgical treatment for patients with complex choledochal cysts should be tailored to each patient's unique needs. The type of cyst and the degree of hepatobiliary system pathology will determine how to cure choledochal cysts [10]. Surgery's primary goal is to completely remove the cysts in an effort to prevent long-term complications like cholangitis, liver cirrhosis, pancreatitis, and malignant transformation [11]. Diao et al. [12] advise early surgical removal of choledochal cysts to decreased the risk of hepatic fibrosis and a quicker return to normal in liver function tests. Surgical treatment for adult patients with CCDs having complex features should be individualized to maintain a balance between risk of surgery and potential risk of late complications. Previous studies indicated that the best course of treatment for Type I choledochal cysts was total cyst removal followed by Roux-en-Y hepatojejunostomy to re-establish bile flow. Total cyst removal has been linked to superior clinical and laboratory results, including symptom alleviation and a reduced chance of malignant change. Total cyst excision and Roux-en-Y hepatojejunostomy led to positive clinical outcomes and early bilirubin and liver enzyme (AST and ALT) value normalization in our patient [6,13]. Lilly's approach could be employed when it is dangerous to completely remove the cyst mucosa due to adhesion to another structure, such as the pancreatic parenchyma, inferior vena cava, or hepatic hilar arteries. The serosa is left attached to the adherent structures while the mucosa is removed

with cauterization [2,14,15]. This surgery obviously avoids future malignancy of the residual mucosa as well as injury to the vena cava or pancreas [13].

5. Conclusion

Giant choledochal cysts are a rare congenital biliary system malformation that might pose a surgical conundrum. The use of MRCP is advised due to its ability to precisely diagnose huge choledochal cysts, which in turn results in an appropriate surgical strategy. In this particular situation, Lilly's technique helps to reduce damage to the main structures.

References

1. Soares KC, Arnaoutakis DJ, Kamel I, Rastegar N, Anders R, Maithel S, et al. Choledochal Cysts: Presentation, Clinical Differentiation, and Management. J Am Coll Surg. 2014; 219(6): 1167-80.
2. Anand U, Priyadarshi RN, Kumar B. A giant type IVA choledochal cyst. Ann Gastroenterol. 2012; 25(1): 73-5.
3. Gomes C, Tivnan P, McAneny D, Tseng JF, Tkacz J, Sachs TE, et al. Choledochal Cyst or Benign Biliary Dilation: Is Resection Always Necessary? J Gastrointest Surg. 2021; 25(9): 2353-7.
4. Harikrishnan S, Chandramohan SM, Chandramohan A. Giant choledochal cyst type 4A: A surgical challenge. Pan Afr Med J. 2020; 37(95): 1-5.
5. Kumar S, Singla S, Kumar S, Singh RK, Raj AK, Mandal M, et al. Tactics, techniques, and challenges in the management of giant choledochal cyst in adolescents and adults: a retrospective cohort study. Langenbeck's Arch Surg. 2021; 406(6): 1925-33.
6. Sikhondze MM, Dreque CC, Tayebwa E, Tumubugane G, Odongo CN, Ogwang E, et al. Giant choledochal cyst in an adult at a teaching hospital in south-western uganda: A case report. Int Med Case Rep J. 2021; 14: 789–95.
7. Hoilat GJ, John S. Choledochal cyst. In: StatPearls. Treasure Island (FL): StatPearls Publishing; 2022.
8. Siahaya FJ, Lalisang TJM, Jeo WS, Simanjuntak ABH, Philippi B. Uncommon Mixed Type I and II Choledochal Cyst: An Indonesian Experience. Case Rep Surg. 2013; 2013(4): 1-4.
9. Todani T, Watanabe Y, Toki A, Morotomi Y. Classification of congenital biliary cystic disease: Special reference to type Ic IV A cysts with primary ductal stricture. J Hepatobiliary Pancreat Surg. 2003; 10(5): 340-4.
10. Cho MJ, Hwang S, Lee YJ, Kim KH, Ahn CS, Moon DB, et al. Surgical experience of 204 cases of adult choledochal cyst disease over 14 years. World J Surg. 2011; 35(5): 1094-102.
11. Machado NO, Chopra PJ, Al-Zadjali A, Younas S. Choledochal Cyst in Adults: Etiopathogenesis, Presentation, Management, and Outcome-Case Series and Review. Gastroenterol Res Pract. 2015; 2015: 602591.
12. Diao M, Li L, Cheng W. Timing of surgery for prenatally diagnosed asymptomatic choledochal cysts: A prospective randomized study. J Pediatr Surg. 2012; 47(3): 506-12.
13. Mathur P, Gupta PK, Udawat P, Mittal P, Nunia V. Hepatobiliary malformations: proposed updation of classification system, clinicopathological profile and a report of largest pediatric giant choledochal cyst. Hpb [Internet]. 2022; 24(3): 422-32.
14. Lilly JR. The surgical treatment of choledochal cyst. Surg Gynecol Obstet. 1979; 149(1): 36-42.
15. Tannuri ACA, Hara LAA, Paganoti GF, Andrade WC, Tannuri U. Choledochal cysts in children: How to Diagnose and Operate on. Clinics (Sao Paulo). 2020; 23(75): e1539.