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CASE REPORT

Giant choledochal cyst: The largest reported!

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Abstract

Choledochal cyst (CC) is an atypical congenital abnormality of the biliary system. CC more than 10 cm is rare and only a few cases are reported. Herein, we reported a 25 × 18 cm CC and highlighted how definitive treatment after a timely diagnosis provides good prognosis irrespective of the size.

KEYWORDS

biliary tract, choledochal cyst, outcome, surgery

1 | INTRODUCTION

Choledochal cysts are atypical congenital abnormalities of the biliary system characterized by varying cystic dilatations of the extrahepatic biliary tree, intrahepatic biliary ducts, or both.[1] They were first reported by Ezler and Vader in the 18th century and later classified by Todani et al.[2] Giant choledochal cysts rarely exceed 10 cm in size.[3] The exact pathogenesis of choledochal cysts is unknown. The active enzymes cause inflammation and degradation of the biliary duct wall which leads to dilatation and cyst formation.[4,5]

2 | CASE REPORT

A 15-year-old boy presented with upper abdominal pain, non-bilious vomiting, and jaundice for the last 6 months associated with gradual distention of the abdomen. He also developed episodic fever with chills and rigor over the last month. Physical examination revealed icterus and a diffuse, large (approximately 20 × 15 cm), intraperitoneal abdominal mass.

Laboratory investigation revealed reduced hemoglobin (9.1 g/dL) and elevated bilirubin (total: 9.1 mg/dL, conjugated fraction: 6.1 mg/dL). Ultrasonographic evaluation of the abdomen showed a large fusiform cystic dilatation of the common bile duct. Central intrahepatic-biliary-radicle dilatation noted and primary confluence formed. Contrast-enhanced CT scan of the abdomen (Figure 1) revealed a 25.4 × 18.3 × 16 cm cystic lesion arising from the hilar confluence and extending till the 2nd part of the duodenum. There is also evidence of bilateral intrahepatic-biliary-radicle-dilatation. MRCP (Figure 2) revealed a T2 weighted hyperintense well-defined cystic dilatation of the common hepatic duct and common bile duct, suggestive of type 1 choledochal cyst (CC). Due to recurrent episodes of cholangitis and mass effect on the adjacent viscera, surgical intervention was warranted. He underwent excision of choledochal cyst with Roux-en-Y

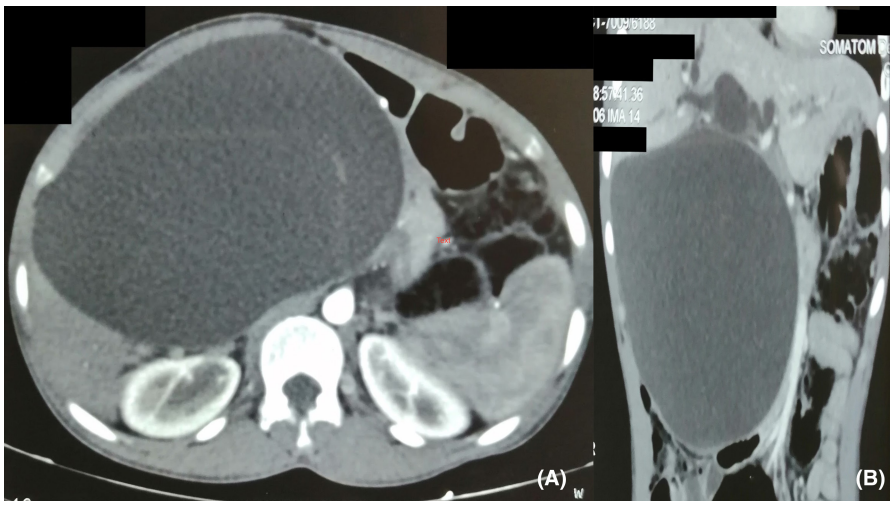

FIGURE 1 Contrast-enhanced computed tomography (A: transverse section, B: coronal section) showing a 25.4 × 18.3 × 16 cm cystic lesion arising from the hilar confluence and extending till the 2nd part of the duodenum.

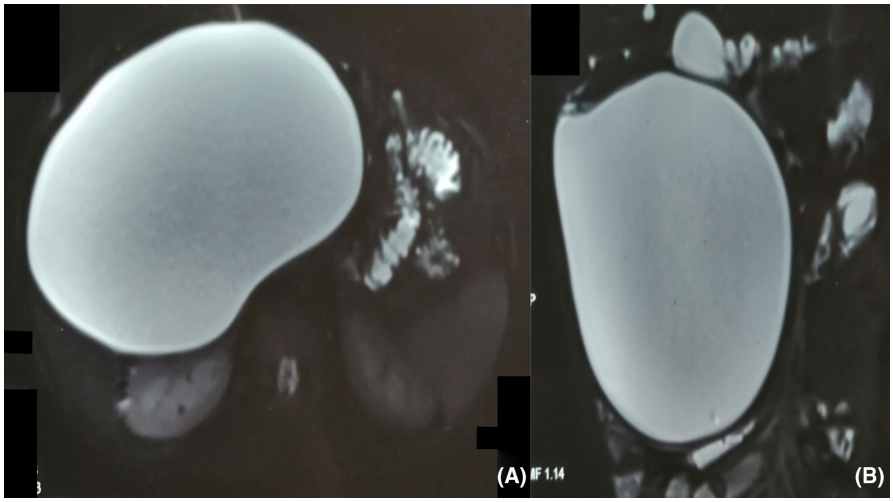

FIGURE 2 MRCP (A: transverse section, B: coronal section) showing T2 weighted hyperintense well-defined cystic dilatation of the common hepatic duct and common bile duct, suggestive of type 1 choledochal cyst.

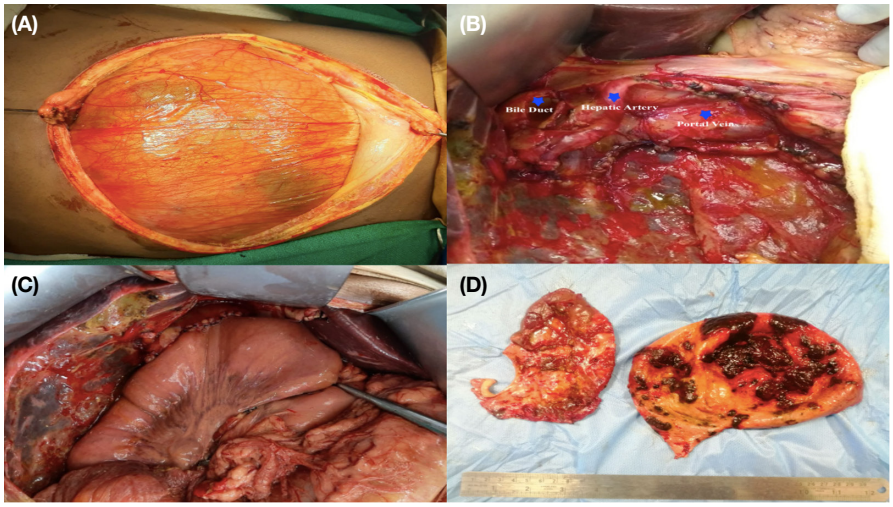

FIGURE 3 Intraoperative image showing (A) choledochal cyst, (B) portal structures after removal of cyst, (C) Bilio-enteric anastomosis, (D) resected choledochal cyst and gallbladder.

hepaticojejunostomy (Figure 3). 2600 mL of bile was removed from the lesion. Histopathological evaluation of the resected specimen confirmed the diagnosis.

He had an uneventful postoperative recovery with complete resolution of symptoms over 2 weeks. On follow-up at the 6th month, he had gained weight with no

recurrence of symptoms and had a normal liver function test.

3 | DISCUSSION

Choledochal cysts are rare atypical congenital malformations portrayed as cystic dilatations of the intrahepatic or extrahepatic biliary tree or both.[1] Todani et al.[2] categorized these into five main types based on anatomical main configurations. Type I is most common in the younger age group, whereas type IV occurs more in adults.[6] In our case, the patient had a type I choledochal cyst characterized by hyperintense well-defined cystic dilatation of the common hepatic duct and common bile duct.

The most widely approved etiological theory of choledochal cysts was given by Babbitt et al. They attributed it to the presence of an anomalous pancreaticobiliary junction outside the Ampulla of Vater, which leads to regurgitation of pancreatic secretions into common bile duct due to small diameter and high pressure of pancreatic duct.[4] Thus, the pancreatic and biliary juices mix and activate the pancreatic enzymes. The active pancreatic enzymes, along with an increase in pressure, lead to inflammation, degradation of the biliary duct wall, dilatation, dysplasia, and subsequent malignancy of the biliary tree.[7,8]

Choledochal cysts present a classical triad of abdominal pain, jaundice, and palpable abdominal mass, but it is present in only 20% of cases.[6,9–11] Other clinical findings include features of cholangitis, pancreatitis, coagulopathy, nausea, vomiting, fever, and portal hypertension.[12,13] The patient described in the report presented with upper abdominal pain, jaundice, and abdominal mass constituting the classical triad. He also had episodic fever and nonbilious vomiting.

The initial diagnosis of choledochal cysts is frequently made using abdominal ultrasound.[14] Other imaging techniques such as computed tomography(CT) scan, magnetic resonance imaging (MRI), endoscopic retrograde cholangiopancreatography (ERCP), and magnetic resonance cholangiopancreatography (MRCP) are key to preoperatively define biliary anatomy.[15] MRCP has high sensitivity (70%–100%) and specificity (90%–100%) and it easily differentiates between the pancreatic and biliary ducts helping in classification of the cyst according to Todani's classification. ERCP is highly sensitive but invasive and associated with many risks such as cholangitis, bleeding, perforation, and pancreatitis.[16] As a result, they are not used frequently.

Choledochal cysts are of various sizes and those greater than 10 cm are referred to as giant choledochal cysts.[17] Our patient had a 25.4 × 18.3 × 16 cm cystic lesion arising from the confluence and extending till the 2nd part of duodenum – thus fitting the criteria of a giant choledochal cyst. Moreover, to the best of our knowledge, this is the largest cyst ever reported.

The ultimate treatment for choledochal cysts is surgical management by cyst excision. Type I and IV require complete excision with cholecystectomy, and bile flow is restored via a bilio-enteric anastomosis. Simple excision and sphincterotomy are done in type II and type III cystic lesions, respectively. Partial hepatic resection may be done in type V lesions. Giant choledochal cysts ate comparatively challenging to be removed surgically as there are risks of post-operative complications like biliary leakage, pancreatic leakage, fistula, and pancreatitis.[18,19]

He had an uneventful postoperative recovery with complete resolution of symptoms over 2 weeks.

AUTHOR CONTRIBUTIONS

Jyotirmoy Biswas: Conceptualization; data curation; writing – original draft. **Siddhartha Nath:** Writing – original draft. **Kankana Karpha:** Writing – original draft.

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DATA AVAILABILITY STATEMENT

Data sharing is not applicable to this article as no new data were created or analyzed in this study.

CONSENT

Written informed consent was obtained from the patient to publish this report in accordance with the journal's patient consent policy.

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