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Giant Type IV Choledochal Cyst: Case Report and Review of Literature

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Abstract: ***Giant choledochal cysts are congenital cystic dilations of the biliary tree with a diameter ≥ 10cm. Most such cysts reported are of Todani type I class. A giant type IV choledochal cyst is uncommon, with only four studies having reported it. Magnetic resonance cholangiopancreatography is indispensable in diagnosing and anatomically classifying the cyst. Surgical intervention is the mainstay of treatment, however, the extent of resection remains controversial for type IV cysts. Here, we discuss the case of a 26year old lady presenting with a painful right sided abdominal lump.***

Keywords: choledochal cyst type IV, giant choledochal cyst, cystic duct cyst, cholecystectomy, biliary malformations

1. Case Report

A 26-year-old female with no comorbidity presented with complaints of an abdominal lump in the right side of abdomen (Figure 1), which wasgradually increasing in size for 1 year, and jaundicewith pale stools for one month. The patient had mild dragging type of pain in the right upper abdomen with no aggravating or relieving factors. There was no history of a sudden increase in the size of the abdominal lump or fever with chills and rigors.

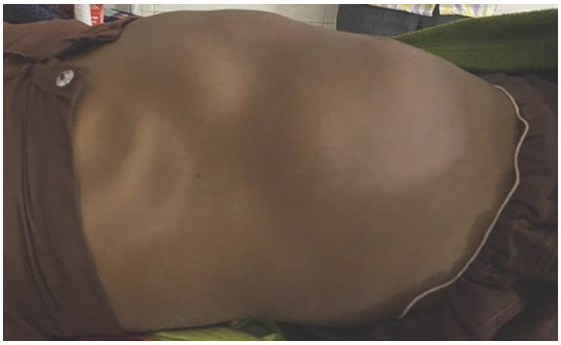

Figure 1: Abdominal distension with a 20 x 18 cm sized lump having a soft and cystic consistency with well-defined rounded borders, extending from the right hypochondrium and epigastrium, through the right lumbar, umbilicus, into the right iliac region.The lump was continuous with the liver dullness and had a dull note on percussion.

The patient was oriented, afebrile, and icteric. On abdominal examination, in addition to the lump shown (Figure 1), there was another smooth cystic swelling measuring around 4 cm horizontally and 4cm vertically in the epigastrium. The swelling had a tympanic note on percussion and gurgling sounds on auscultation. Liver function tests were suggestive of an obstructive aetiology (total bilirubin/direct bilirubin-7.1/3.2, AST/ALT/ALP –168/66/2236, total protein/serum albumin- 7.4/3.1). Coagulation profile was deranged (PT-14.5sec, PT- INR- 1.83, aPTT->60sec). Contrast-enhanced CT abdomen showed hepatosplenomegaly with a fusiform dilatation of the extrahepatic biliarytree along with dilatation of right and left hepatic duct, and intrahepatic biliary radicals suggestive of a type IVA choledochal cyst with pressure effects on the main pancreatic duct, bowel loops, and stomach. MRCP confirmed these findings (Figure 1).Conservative management with a hepatic diet, injection vitamin K, syrup lactulose, and antibiotics were provided as the patient denied consent for operative intervention.

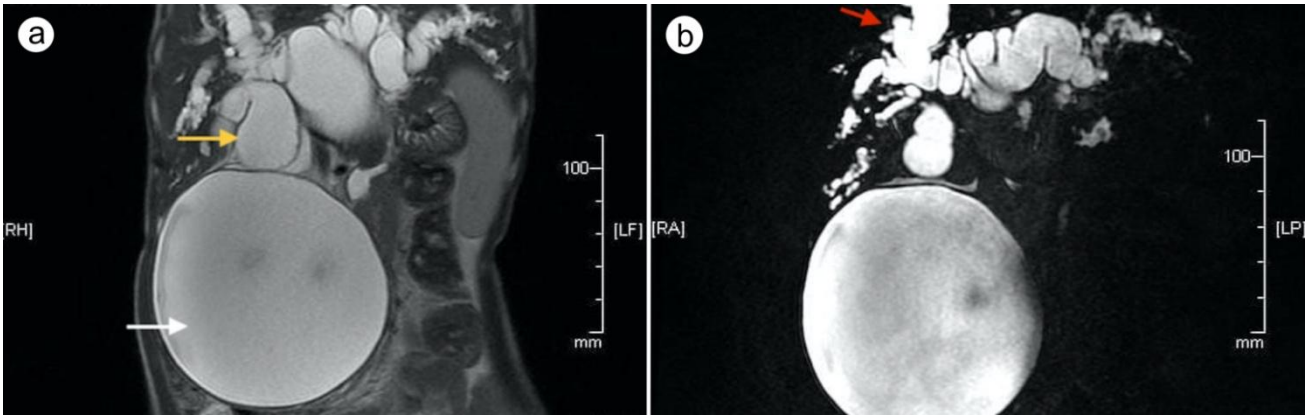

Figure 2: (a) Magnetic resonance cholangiopancreaticogram (MRCP) showing extrahepatic component of the type IVA cyst comprising of grossly dilated distal common bile duct(131mm, *white arrow*) and proximal common bile duct (78mm, *yellow arrow*). **(b)** MRCP showing intrahepatic component of the type IVA cyst comprising of dilated intrahepatic biliary radicles (*redarrow*s)

2. Discussion

Congenital cystic dilations of the biliary tract are called Choledochal cysts (CC), earliest reported in 1723.[1] Cysts with a diameter ≥ 10cm are referred to as Giant choledochal cysts (GCC). CC are more prevalent in adult East Asian populations and usually present with non-specific complaints like abdominal mass, right upper quadrant pain, jaundice, nausea or vomiting. They result from an anomalous formation of the pancreaticobiliary junction (ABPJ) near the Sphincter of Oddi complex, resulting in a common channel emptying the pancreatic and bile ducts.[2]

Diagnosis cannot be made from history and physical examination alone, making radiological investigations crucial. The initial investigation is an Ultrasound or CT scan in most settings, followed by MRCP to anatomically classify the cyst. ERCP is invasive but remains the the gold standard as it can pick up minor ductal anomalies. Depending on the anatomy, the widely accepted Todani classification categorizes CC into five subtypes, with a recently added sixth subtype.[3]Amongst these subtypes, type IVa is the most common subtypein adults and refers to the presence of both intrahepatic and extrahepatic components in the cyst.[3] Surgical management is the preferred treatment modality for all CC. In particular, the management of giant Type IV a cysts is more controversial. While only extrahepatic portion removal maybe associated with risk of developing biliary intraepithelial neoplasia, intrahepatic portion removal maybe associated with reduced residual liver function.[4] GCC excision is surgically challenging, and not without the risk of dreaded complications like postoperative pancreatitis and bile leak.

Most giant choledochal cysts reported in literature belong to Todani type I class. There are only four studies reporting a giant Todani type IV choledochal cyst to our knowledge (Table 1).[5–8]As seen in this case, the role of MRCP is indispensable in delineating the anatomy and guiding management of such cysts. The classic triad of right upper quadrant pain, abdominal mass and jaundice is found in 20% cases of CC but in as high as 60% cases of GCC.[1,7]Our case strengthens the consensus that adult patients with GCC tend to have severe symptoms and that the classical triad is more frequent with giant cysts due to an increased propensity to cause biliary obstruction. Furthermore, the risk for developing malignancy in unexcised cysts is as high as 45%, which increases with age.[9] With this case, we highlight the need for a timely, comprehensive follow-up schedule in patients denying surgery or managed conservatively for other medical or socio-cultural reasons.

Table 1: List of Type IV Giant Choledochal Cysts Reported in the Literature

Study/Report	Cyst Type	Size in Centimeters (cm)	Country
Sikhondze et al, 2021 [5]	IVA	20cm x 15cm	Uganda
Kumar et al, 2021[8]	I and IV	Median: 15cm, Range: 10-20cm	India
Harikrishnan et al, 2020 [6]	IVA	23cm x 15cm	India
Anand et al, 2013 [7]	I and IV	Mean: 14.2cm, Range: 12-20cm	India

3. Conclusion

With the availability of high resolution imaging in modern day practice, clinicians might increasingly encounter CC, which were once considered rare entities!A detailed understanding of the type IV CC anatomy, its findings on MRCP, and treatment options is thus essential.

References

[1] Soares KC, Arnaoutakis DJ, Kamel I, et al. Choledochal Cysts: Presentation, Clinical Differentiation, and Management. J AmColl Surg. 2014; 219(6):1167.
[2] Babbit DP. Congenital choledochal cyst: new etiological concept based on anomalous relationships of the common bile duct and pancreatic bulb. Ann Radiol. 1969; 12:231.

[3] Todani T, Watanabe Y, Toki A, et al. Classification of congenital biliary duct cystic disease: special reference to type Ic and IVA cysts with primary ductal stricture. J Hepatobiliary Pancreat Surg. 2003;10(5):340-344.
[4] Liu CL, Fan ST, lo CM, Lam CM, Poon RT, Wang J. Choledochal cysts in adults. Arch Surg. 2002;(137.4):465-468.
[5] Sikhondze MM, Dreque CC, Tayebwa E, Tumubugane G, Odongo CN, Ogwang E. Giant Choledochal Cyst in an Adult at a Teaching Hospital in South-Western Uganda: A Case Report. Int MedCase RepJ. 2021;14:789.
[6] Harikrishnan S, Chandramohan SM, Chandramohan A. Giant choledochal cyst type 4A: a surgical challenge. Pan AfrMedJ. 2020;37(95):1-5.
[7] Anand U, Priyadarshi RN, Kumar B, Khandelwal C. Diagnosis and management of giant choledochal cysts: complexities compared to smaller cysts. Ind J Gastroenterol. 2013;32(4):262-267.
[8] Kumar S, Singla S, Kumar S, Singh RK, Raj AK, Mandal M. Tactics, techniques, and challenges in the management of giant choledochal cyst in adolescents and adults: a retrospective cohort study. Langenbeck Arch Surg. 2021;406(6):1925-1933.
[9] Lipsett PA, Pitt HA, Colombani PM, Boitnott JK, Cameron JL. Choledochal cyst disease. A changing pattern of presentation. Ann Surg. 1994;220(5):644.

Author Profile

Dharmendra Shah is currently an Additional Professor and the Head of Unit, Department of General Surgery, Medical College Baroda & S.S.G. Hospital, Gujarat, India. He has served as the local principal investigator for international multicentric studies like COVID Surg 3, and has mentored numerous postgraduate and undergraduate students for more than a decade now. *Shivani Chaudhary*is an aspiring neurosurgeon and presently serves as a Senior Resident after completing her Masters in Surgery from Medical College Baroda & S.S.G. Hospital, Gujarat, India. Her works have been published in reputed journals like British Journal of Neurosurgery. *Nisarg Desai* is an aspiring thoracic surgeon and is currently a rotating house doctor after completing his medical degree from Medical College Baroda & S.S.G. Hospital, Gujarat, India.