

A New Variant of Choledochal Cyst Diagnosed on Magnetic Resonance Cholangiopancreatography

SHAILESH GUPTA¹, S. RAJESH², NITESH AGRAWAL³, SOMSHARAN S. BETGERI⁴, ANKUR ARORA⁵

ABSTRACT

Choledochal cysts (CDC) have been traditionally classified into five types and subtypes based on the pattern and location of involvement of intra and extra hepatic biliary tree. Herein, we describe a new variant of choledochal cyst which has not been previously described in the English-language medical literature.

Keywords: Biliary tree, Choledochal cyst, Cystic duct, MRCP

CASE REPORT

A 15-year-old male presented to the hepatology out-patient department with complaints of intermittent crampy upper abdominal pain for a month which got aggravated after meals. The pain was not associated with nausea, vomiting, and abdominal distension or altered bowel movements. There was no past history of fever or jaundice. Physical examination did not reveal any significant abnormality. Laboratory investigations including liver function tests were within normal limits. Ultrasound examination performed at an institution elsewhere had shown mildly thickened gall bladder walls with mild central bilobar intrahepatic biliary radical dilatation. In view of biliary dilatation, patient underwent magnetic resonance cholangiopancreatography (MRCP) which revealed that there was focal segmental fusiform dilatation of both right and left hepatic ducts just proximal to the primary biliary confluence. In addition, the cystic duct was also focally dilated [Table/Fig-1]. The common bile duct (CBD) was normal in caliber. There was no associated anomalous pancreatico-biliary junction. No evidence of any obstructive intraductal calculus or stricture was noted in any part of biliary tree. The patient was advised surgical management but was lost to follow-up.

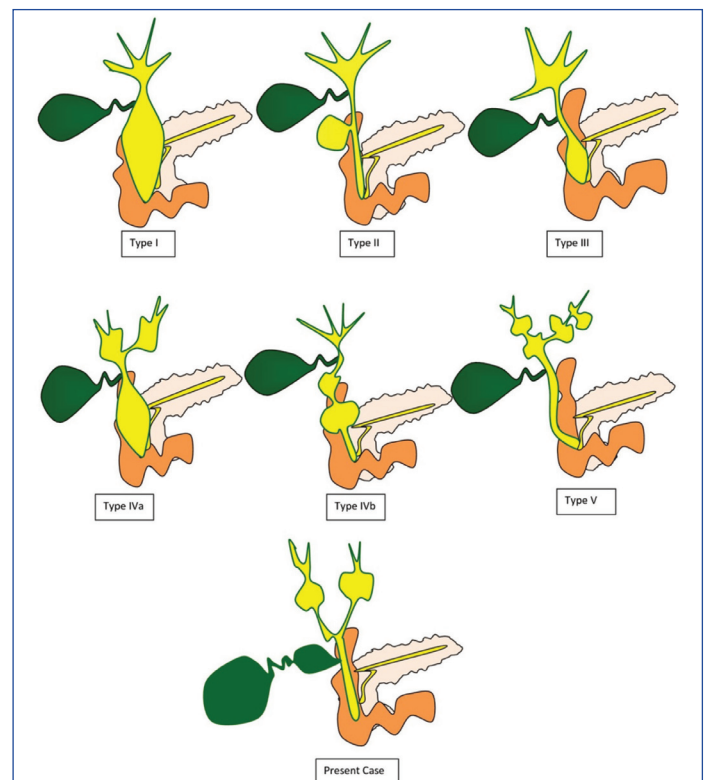
DISCUSSION

Choledochal cysts are an uncommon anomaly and are thought to be a result of abnormal long common channel between the distal CBD and pancreatic duct which can lead to reflux of pancreatic enzymes into the biliary tree causing weakness in ductal walls and

abnormal dilatation [1]. Another proposed theory is malfunctioning sphincter of oddi causing upstream reflux of pancreatic juices with resultant biliary dilatation [2]. Both extra and intrahepatic ducts can be involved. Based on the location of involvement, Todani et al., [3] have classified CDC into five types: Type 1 is the most common type and is further divided into three subtypes on the basis of morphology of dilated CBD – 1A(cystic dilatation), 1B(segmental dilatation) and type 1C corresponds to fusiform dilatation of common bile duct, type 2 represents diverticulum of CBD, while type 3 is focal cystic dilatation of terminal CBD known as choledochoceles; type 4 has been further subdivided into type 4a which is saccular dilatation of both intra and extra hepatic bile ducts and type 4b where there is saccular dilatation of only extra hepatic bile duct, type 5 is cystic dilatation of only intrahepatic bile ducts and popularly known as Caroli's disease. This classification does not take into account



[Table/Fig-1]: Thick-section (40-mm) MRCP images demonstrates the fusiform dilatation of Intrahepatic bile ducts (thin arrows) and cystic duct (thick arrows). Note normal caliber of common bile duct (arrow head). Curved arrow denotes gall bladder



[Table/Fig-2]: Schematic diagram depicting different types of choledochal cysts as proposed by Todani et al., [3] and the present case. Yellow colour- biliary tree, green- gall bladder and cystic duct, orange- duodenum, light pink- pancreas

the involvement of cystic duct. The tremendous advancements in imaging technology have resulted in incidental detection of a number of new variants most of which include involvement of cystic duct. However, all the previous literature is either on isolated cystic duct involvement or the involvement of cystic duct along with extra hepatic duct involvement [4-7]. The present case was unique as there was involvement of cystic duct along with bilobar central intra hepatic bile ducts [Table/Fig-2].

Preoperative diagnosis is very important as choledochal cysts with only extrahepatic duct involvement are managed by complete excision of cyst with hepaticoenterostomy and cholecystectomy, while if there is intrahepatic involvement then partial hepatectomy or liver transplantation may be needed according to the extent of disease [8,9]. As the use of laparoscopic management of CDC is increasing [10], preoperative diagnosis of such variants helps in deciding the surgical approach and to avoid complications of duct injury.

CONCLUSION

In conclusion, with the recent surge in the volume of literature describing new variants of CDC, especially including cystic ducts, we propose the inclusion of cystic duct variants in the classification and that a suffix "c" can be added after the parent subtype if cystic duct is also involved. Hence, our case can be considered as a variant

of Todani's type 4A with extrahepatic duct involvement in form of cystic duct dilatation. Hence it can be classified as type 4A'c' or 4c, while an isolated cystic duct can be considered as type 6.

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PARTICULARS OF CONTRIBUTORS:

1. Senior Resident, Department of Radiology, Institute of Liver and Biliary Sciences, D-1, Vasant Kunj, New Delhi, India.
2. Assistant Professor, Department of Radiology, Institute of Liver and Biliary Sciences, D-1, Vasant Kunj, New Delhi, India.
3. Senior Resident, Department of Radiology, Institute of Liver and Biliary Sciences, D-1, Vasant Kunj, New Delhi, India.
4. Senior Resident, Department of Radiology, Institute of Liver and Biliary Sciences, D-1, Vasant Kunj, New Delhi, India.
5. Associate Professor, Department of Radiology, Institute of Liver and Biliary Sciences, D-1, Vasant Kunj, New Delhi, India.

NAME, ADDRESS, E-MAIL ID OF THE CORRESPONDING AUTHOR:

Dr. S. Rajesh,
Assistant Professor, Department of Radiology, Institute of Liver and Biliary Sciences,
D-1, Vasant Kunj, New Delhi-11070, India.
E-mail: rajesh387@gmail.com

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