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Case Report

Radiology Section

MRCP-based Diagnosis of Todani Type IVa Choledochal Cyst with Associated Hepatic Duct Anatomical Variant in a Young Adult Female: A Case Report

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ABSTRACT

Choledochal cysts are rare congenital anomalies of the biliary system, with Todani type IVa involving both intrahepatic and extrahepatic bile ducts. Accurate classification and identification of associated biliary anatomical variants are essential for appropriate management and surgical planning. We report the case of a 20-year-old female who presented with postprandial abdominal pain and was referred for Magnetic Resonance Cholangiopancreatography (MRCP) following a suspicious ultrasound examination. MRCP demonstrated fusiform dilatation of the extrahepatic bile duct with multifocal dilatation of the intrahepatic biliary radicles, consistent with a Todani type IVa choledochal cyst. An associated hepatic duct anatomical variant (type II) was also identified. Laboratory parameters were within normal limits, and no surgical intervention had been undertaken at the time of presentation. This case highlights the pivotal role of MRCP in the non-invasive diagnosis, classification, and comprehensive biliary mapping of choledochal cysts. The co-existence of a choledochal cyst with a hepatic duct anatomical variant makes this case unique and has significant implications for surgical planning.

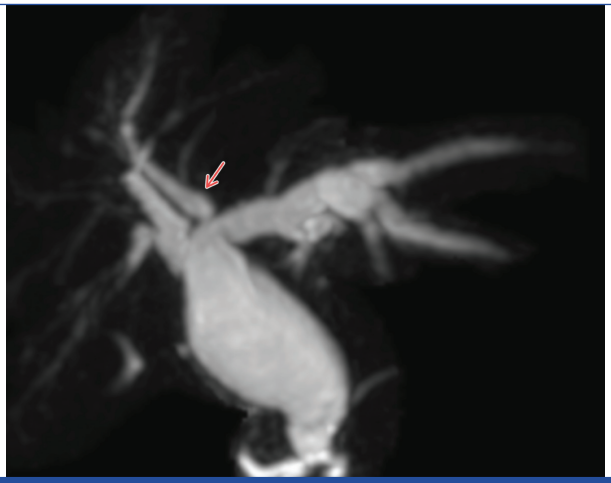
Keywords: Aberrant ductal drainage, Congenital biliary anomaly, Hepatobiliary anatomy, Non-invasive imaging, Preoperative biliary assessment

CASE REPORT

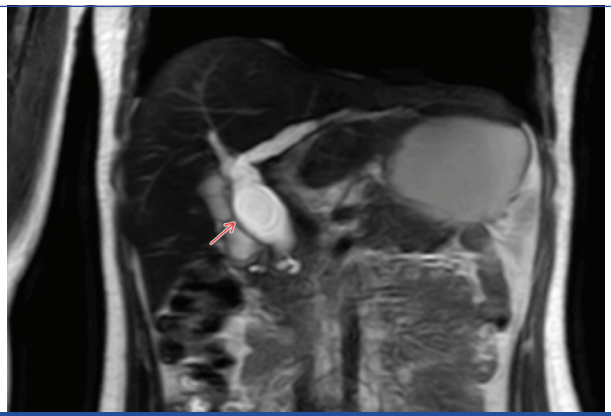
A 20-year-old female presented with intermittent abdominal pain aggravated after meals for the past six months. There was no history of jaundice, fever, vomiting, or weight loss. There was no prior surgical history, no history of similar illness in the family, and no known comorbid conditions. Physical examination was unremarkable. Laboratory investigations, including liver function tests, were within normal limits. The patient had not undergone any prior imaging or treatment apart from an initial ultrasound examination at an outside centre, which showed dilatation of the common bile duct with mild prominence of intrahepatic biliary radicles. No focal hepatic lesions were observed. Based on these findings, a provisional diagnosis of a choledochal cyst was suggested, and further evaluation with MRCP was recommended for detailed biliary mapping.

The MRCP demonstrated fusiform dilatation of the extrahepatic bile duct, involving the common hepatic duct and common bile duct, with smooth margins and distal tapering. Multifocal dilatation of the intrahepatic biliary radicles was also noted, consistent with a Todani type IVa choledochal cyst. No evidence of an Anomalous Pancreaticobiliary Junction (APBJ) was identified. The pancreatic duct and common bile duct joined normally within the duodenal wall, without formation of an abnormally long common channel. An associated hepatic duct anatomical variant corresponding to a type II variation was identified, characterised by aberrant drainage of the right posterior sectoral duct into the common hepatic duct [Table/Fig-1,2]. This hepatic duct variation is classified according to the Huang classification of intrahepatic biliary ductal anatomy. The gallbladder appeared normal. No intraductal calculi, mural thickening, restricted diffusion, or imaging features suggestive of malignancy were observed.

Following MRCP confirmation of a Todani type IVa choledochal cyst with associated hepatic duct anatomical variant (type II), the patient was referred to the hepatobiliary surgery department for definitive management. Given the involvement of both intrahepatic



[Table/Fig-1]: MRCP maximum intensity projection image demonstrating choledochal cyst type – IVa with an associated hepatic duct anatomical variant (type II), with aberrant drainage of the right posterior sectoral duct (arrow).

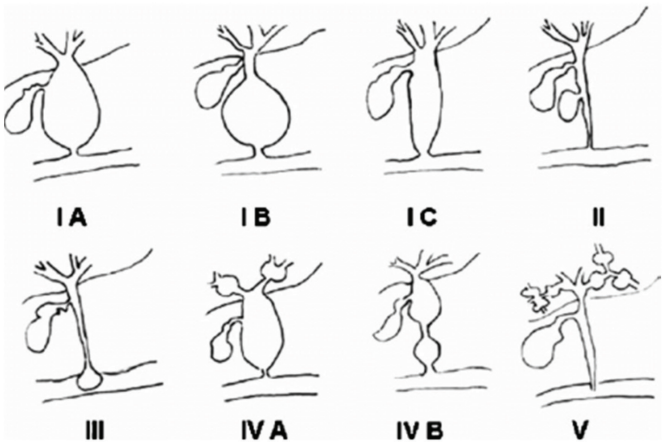


[Table/Fig-2]: MRCP thick slab image demonstrating fusiform dilatation of the extrahepatic bile duct (arrow) with associated multifocal dilatation of the intrahepatic biliary radicles, consistent with a Todani type IVa choledochal cyst.

and extrahepatic bile ducts (type IVa), surgical intervention was planned, consisting of complete excision of the extrahepatic bile duct cyst, Roux-en-Y hepaticojejunostomy reconstruction and hepatic lobectomy. At the time of manuscript submission, no surgical intervention had been performed.

DISCUSSION

Choledochal cysts are rare congenital anomalies of the biliary tract characterised by cystic dilatation of the bile ducts and are associated with significant long-term morbidity. The widely accepted classification system is the Todani classification (1977), which categorises cysts according to the anatomical extent of biliary involvement. The Todani system categorises cysts into five major types according to the anatomical extent of biliary involvement [Table/Fig-3]:



[Table/Fig-3]: Diagrammatic representation of Todani classification of choledochal cysts [3].

Type I - Isolated extrahepatic bile duct dilatation (IA, IB, IC)

Type II - Extrahepatic bile duct diverticulum

Type III - Choledochocele

Type IVa - Combined intrahepatic and extrahepatic ductal dilatation

Type IVb - Multiple extrahepatic cysts

Type V - Isolated intrahepatic dilatation (Caroli disease) [1,2].

Type IVa cysts involve both intrahepatic and extrahepatic bile ducts and represent one of the more complex subtypes. These lesions are associated with an increased risk of complications, including recurrent cholangitis, pancreatitis, biliary cirrhosis, and malignant transformation [4-6].

Normal biliary anatomy consists of the confluence of the right anterior and right posterior sectoral ducts forming the right hepatic duct, which then joins the left hepatic duct to form the common hepatic duct. However, biliary anatomical variants are common and are reported in approximately 30%-40% of individuals [7]. Recognition of such variants is critical, as unrecognised aberrant ducts are a well-established cause of iatrogenic bile duct injury during hepatobiliary surgery.

Singh S et al., described a similar case and highlighted the importance of preoperative biliary mapping to prevent ductal injury [8]. Geraci G et al., reported successful surgical management of a choledochal cyst associated with an aberrant posterior hepatic duct and underscored the complexity of operative planning in such cases [9]. More recently, Boricha S et al., discussed the technical difficulties posed by anomalous biliary tree anatomy during cyst excision and reinforced the necessity of detailed preoperative imaging [10]. The findings in our case are consistent with these reports.

Although APBJ is frequently implicated in the pathogenesis of choledochal cysts due to pancreatic enzyme reflux, no such junctional anomaly was identified in this case. This observation supports the concept that cyst formation may occur even in the absence of demonstrable pancreaticobiliary maljunction [4].

Definitive management of type IVa cysts is surgical. Standard treatment consists of complete excision of the extrahepatic bile duct with Roux-en-Y hepaticojejunostomy. In cases with significant intrahepatic involvement, segmental hepatic resection or lobectomy may be required [6]. Early surgical intervention is recommended because the risk of malignancy persists even in asymptomatic individuals. In the present case, surgical excision with hepatic lobectomy was planned; however, surgery had not been performed at the time of manuscript submission.

The differential diagnosis of type IVa choledochal cyst includes Caroli disease (type V), primary sclerosing cholangitis, recurrent pyogenic cholangitis, obstructive biliary dilatation due to distal stricture or calculus, and cystic biliary neoplasms.

CONCLUSION(S)

This case underscores the pivotal role of MRCP in the comprehensive evaluation of choledochal cysts, enabling accurate diagnosis and classification according to the Todani system while simultaneously delineating associated biliary anatomical variants. Type IVa choledochal cysts should be carefully differentiated from other causes of intrahepatic biliary dilatation, and early surgical referral is recommended even in minimally symptomatic patients because of the potential for long-term complications, including malignant transformation.

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