

A RARE TYPE IV_a GIANT CHOLEDOCHAL CYST IN ADULT PATIENT: CASE REPORT

Jyotish Roy, Sudipta Saha, Bijaya Lakshmi Gayari, Debanu De

Department of Radiodiagnosis, Institute of Post-Graduate Medical Education and Research, Seth Sukhlal Karnani Memorial Hospital, Kolkata, India.

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ABSTRACT

Choledochal cyst is a rare congenital cystic dilatation of biliary ductal system. We report a 35 year male patient presented to the emergency with pain, fever, jaundice and upper abdominal swelling with history of similar attack in past. He was diagnosed as case of giant type IV_a choledochal cyst with massive dilatation of both intrahepatic and extrahepatic biliary tree with the help of ultrasonography and magnetic resonance cholangiopancreatography (MRCP). We will discuss about clinical feature and imaging characteristics of giant type IV_a choledochal cyst.

Key words : choledochal cyst, MRCP

Introduction

Abnormal, disproportionate congenital dilatation of biliary system known as choledochal cyst which may involve extrahepatic or intrahepatic biliary duct or both. Etiology is not known but close association with anomalous formation of pancreatic-biliary ductal system is reported. It is rare in western world but evidences of increase prevalence in Asia noted.¹ Symptoms varies with age, with more than 60% present in within first decades of life and occurs more frequently in female. Commonly presenting symptom in children are pain, jaundice and mass in upper abdomen. In adult symptoms may varies from vague upper abdominal pain to more severe complication. Type IV choledochal cyst is the second most common after type I, and divided into two type type IV involve both extrahepatic and intrahepatic. Type IV_b is defined as multiple dilatation of extrahepatic bile duct. We describe a giant type IV_a choledochal cyst in a adult patient with massive dilatation both intrahepatic and extrahepatic bile duct.²

Case Report

A 35 year old male patient attended to emergency of our hospital with pain, fever, jaundice and upper abdominal mass for last 2 month. There was past history of similar attack for last 1 year. On examination, patient had jaundice, fever and soft swelling in the right upper abdomen with tenderness.

Laboratory examination revealed total bilirubin 12.3 mg/dl; conjugated bilirubin 10.6mg/dl; unconjugated bilirubin 1.7 mg/dl; alkaline phosphatase 870 U/l; ALT 312 U/L; AST 246U/l.

Ultrasonography showed hugely dilated cystic lesion in both lobe of liver filled with low internal echo and multiple calculi. Common hepatic duct was dilated. On MRCP there was huge dilatation of IHBRs with multiple filling defects. Common bile duct (CBD) was also dilated. With these findings, diagnosis of giant type IV_a choledochal cyst with hugely dilated and tortous IHBR was confirmed.

Correspondence : Dr. Jyotish Roy
Department of Radiodiagnosis,
Institute of Post-Graduate Medical Education
and Research, Seth Sukhlal Karnani Memorial
Hospital, Kolkata, India.
Email: jyotishroy149@gmail.com

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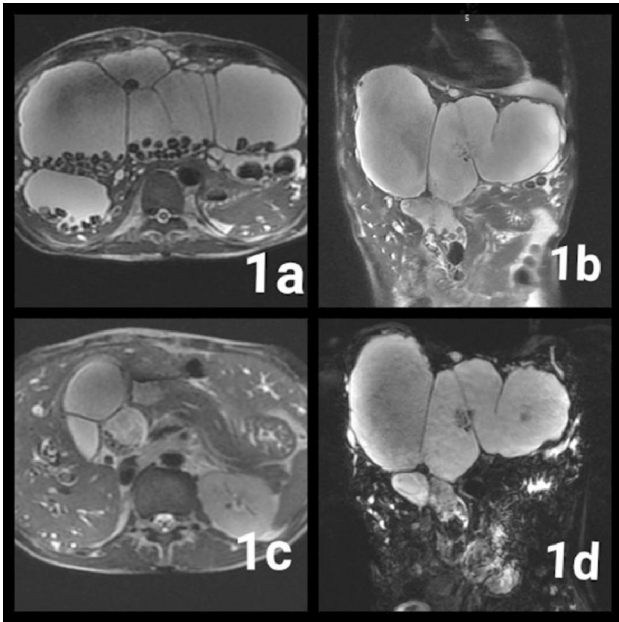


Figure 1: 1a. Axial T2-weighted image showing dilated bilobar IHBR with multiple T2 hypointense filling defects. 1b. Coronal T2-weighted image showing dilated bilobar IHBR and dilated CBD with multiple T2 filling defects. 1c. Axial T2-weighted image showing optimally distended gallbladder. 1d. 3D Rtr image showing dilated IHBR and dilated CBD.

Discussion

Choledochal cyst refers to rare, isolated or combined dilatation of extrahepatic and intrahepatic biliary system of congenital origin. The prevalence varies widely ranging from 1: 30000 - 500000 in western population to 1: 1000 in Asia.¹ Female are more affected than male and majority of the patient are present in childhood.

Alonso-Ley and colleagues³ describe classification of choledochal cyst in 1959 into 3 types which advanced to 5 types by Todani et al⁴ in 1977. Sixth type of choledochal cyst proposed in 1983 by Bode and Aust.² Type I is a dilatation of the extra hepatic bile duct which is further subdivided into 3 types. Type II is a single extrahepatic diverticulum. Type III is a dilatation of the intraduodenal portion of the bile duct. Type IV consists of two subtypes: IVa which is combined intra- and extrahepatic dilatation and IVb which is multiple extrahepatic cysts. Type V is also known as Caroli's disease and consists of cystic dilatation of the intrahepatic biliary system and associated with hepatic fibrosis. Type I is the most common type of all ages but prevalence of type IV more

common in adult population. The size of the cyst varies, and rarely exceeds 9 cm.⁵

The most widely accepted theory for occurrence of choledochal cyst postulates that an anomalous pancreatic biliary ductal junction (APBDJ) with long common channel is the primary abnormality. Others popular hypothesis includes ganglionosis, aganglionosis, congenital stricture with proximal biliary dilatation.⁶

Children present mainly with jaundice, pain and mass in upper abdomen. Adult mainly present with pain and complications of longstanding cysts such as recurrent cholangitis, pancreatitis and malignancies. Laboratory tests are not always helpful and it may reveals features of obstructive jaundice. On imaging, ultrasound is the best initials method for evaluation of biliary system and gallbladder, but it may not be accurately indentify that the cyst originate from the biliary system, especially in large cyst. Computed Tomography is not a best tools to evaluate biliary system.⁷ However invasive procedure like Direct contrast cholangiography is used to define extent of involvement and visualise the remaining biliary tree. Magnetic resonance cholangiography (MRCP) is a non-invasive technique and considered as "Gold standard" for confirming the diagnosis of choledochal cysts and defining extent of involvement pre-operatively.⁷


The extent of resection in type IVa choledochal cyst is controversial. It became very difficult to resect in case of bilobar IHBR dilatation. Partial extrahepatic cystectomy with biliary- enteric anastomosis is the treatment of choice .

We conclude that that giant type IVa choledochal cyst may present in adult with huge dilatation of intrahepatic and extrahepatic biliary system, making diagnosis very difficult especially in presence of complication. Use of MRCP is highly informative in definitive diagnosis and plan for surgical excision.

Conflict of Interest: Authors declared none.

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