Case Report

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Type IVA Choledochal Cyst in Adult: A Case Report

Surgery Section

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ABSTRACT

Choledochal cyst is a congenital disease of the biliary tract with extrahepatic and/or intrahepatic biliary tree dilatation. There is a female majority (4:1). Controlling biliary sepsis and curing pancreatitis are prerequisites of surgery. Here, a 35-year-old female patient, came with complaints of epigastric and right upper abdominal pain for three days. On examination per abdomen was soft, non distended, right hypochondriac tenderness was present, no guarding, bowel sounds were audible. Magnetic Resonance Cholangiopancreatography (MRCP) was done which showed fusiform dilatation of Common Bile Duct (CBD), Common Hepatic Duct (CHD), right and left hepatic duct, along with mild dilatation of central intrahepatic biliary radicals (4.8-5 mm)- suggestive of choledochal cyst type IVA. Hepatico-duodenostomy was done. The entire extrahepatic biliary tree should be removed when the choledochal cyst is diagnosed whether or not symptoms are present. Despite being congenital, the condition is typically discovered in adulthood as well. The treatment of choledochal cysts has evolved. The current standard of care is cyst excision and hepaticojejunostomy.

Keywords: Biliary tree, Biliary sepsis, Common bile duct, Hepatico-duodenostomy

CASE REPORT

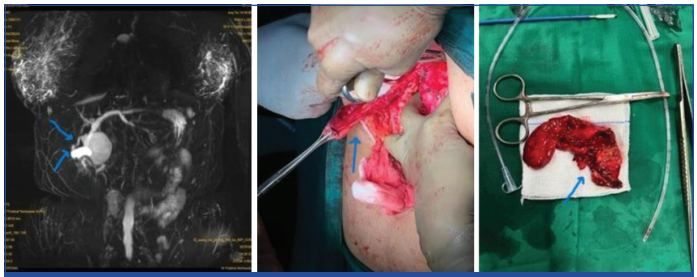
A 35-year-old female patient, came with complaints of epigastric and right upper abdominal pain for three days, which was continuous, low intensity, non radiating, was accompanied by nausea and frequent bouts of bilious vomiting mimicking biliary colic. There was no prior history of fever, jaundice, bowel or bladder issues, or similar complaints in the past, and no history of alcohol consumption, no prior medical or surgical conditions.

On general examination, patient's vitals were found stable. Per abdomen was soft, non distended, right hypochondriac tenderness present, no guarding, bowel sounds were audible. All biochemical analyses, including Liver Function Tests (LFTs), were within normal bounds [Table/Fig-1]. Abdominal sonography was suggestive of normal size gall bladder and wall thickness, no calculi was noted, and the CBD was normal in diameter. Pancreas showed a cystic lesion ~5.7*5.7 cm with a solid component at the head of the pancreas suggesting the possibility of a pseudocyst. Hence, MRCP was done for further evaluation.

Investigations	Values
Haemoglobin	10.70 g/dL
Total leukocyte count	4,600/microLt
Platelet count	339/microLt
Total bilirubin	0.55 mg/dL
Conjugated bilirubin	0.30 mg/dL
Unconjugated bilirubin	0.25 mg/dL
SGOT	40 U/Lt
SGPT	104 U/Lt
Alkaline phosphatase	110 U/Lt
Serum amylase	44 U/Lt
Serum lipase	82 U/Lt
Serum urea	18 mg/dL
Serum creatinine	0.66 mg/dL
Total protein	6.20 g/dL
Serum albumin	3.70 g/dL
Serology (HIV/HBsAg/HCV)	Non reactive
Prothrombin time/INR	12.01 seconds/1.02

[Table/Fig-1]: Biochemical investigations performed.
INR: International normalised ratio; SGOT-: Serum glutamic-oxaloacetic transaminase; SGPT: Serum glutamic pyruvic transaminase; HIV: Human Immunodeficiency virus; HBsAg: Hepatitis B surface antigen; HCV: Hepatitis C virus

MRCP [Table/Fig-2] showed fusiform dilatation of the CBD, CHD, and right and left hepatic duct, along with mild dilatation of central intrahepatic biliary radicals (4.8-5 mm)-suggestive of choledochal cyst type IVA [1]. Cystic duct was dilated at insertion of CBD. Heterogenous predominantly T2 hypointense filling defects were seen in the dilated CBD and left hepatic duct. She was taken-up for exploration. Choledochus was identified. It was extending from the supra-duodenal part of CBD up to the confluence of hepatic duct [Table/Fig-3]. It was excised and hepatico-duodenostomy was done [Table/Fig-4]. Distal stump of CBD was closed. There were no intra-operative or post-operative complications. Histopathological Examination (HPE) report of the specimen was suggestive of chronic cholecystitis with fibro collagenous cyst wall. Repeat Liver Function Test (LFTs) was within normal limits. She was discharged on post-operative day 10. She was advised for three monthly follow-ups with an ultrasound abdomen, but she didn't follow-up.



[Table/Fig-2]: MRCP showing Type 4A choledochal cyst-length 53 mm, diameter 36 mm (arrow). **[Table/Fig-3]:** Intraoperative photo of the gallbladder (arrow) along with the dilated CBD and CHD. **[Table/Fig-4]:** choledochus excised in toto along with the gallbladder arrow indicating. choledochal cyst. (Images from left to right)

DISCUSSION

Adults seldom develop choledochal cysts, a rare congenital cystic dilatation of the biliary tree. Vater A and Ezler C provided the initial description of it [1]. Choledochal cysts are uncommon in Western populations (1:100,000 to 1:150,000 people), however, they are far more common in Asian populations (approximately 1:13,000 individuals) Although choledochal cysts are most frequently diagnosed in children, 20% of individuals first seek treatment as adults [2,3].

A study conducted by Razafindrazoto CI et al., which showed type IVA choledochal cyst in an adult group of 23-year-old patient with female preponderance [4]. The case reported by de Albuquerque VVML et al., showed type IVA choledochal cyst in a 38-year-old female patient complained of severe abdominal pain for one month [5].

There are a number of theories that explain how choledochal cysts originate. The bile duct first becomes partially blocked, increasing proximal bile duct pressure and eventually causing extrahepatic segmental and intrahepatic component dilatation. The second explanation called the pathophysiological effects of activated pancreatic proteolytic enzyme reflux on the biliary tract wall is the foundation of Babbitt's theory [6]. Now-a-days, Anomalous Pancreaticobiliary Duct Union (APBDU) is a generally accepted hypothesis [7]. Along with the aforementioned theories, the literature identifies a number of additional causes, such as bile duct obstruction brought on by malfunction of the sphincter of Oddi, The terminal bile duct's autonomic innervation, atresia, stenosis, and fibrosis [8]. Classification of choledochal cyst most widely used was authored by Todani and his colleagues. Out of them, Types I and IVA account for more than 90% of the patients [1,9].

Jaundice, right hypochondriac discomfort, and a palpable mass make up the traditional triad of choledochal cysts. Children were found to have it more frequently than adults (85% versus 25%, respectively). Adults frequently complain of abdominal pain, which is what prompted the present case patient to seek care [10].

In spite of the fact that MRCP is currently the "gold standard" for staging choledochal cysts, ultrasound may be preferable as a preliminary assessment. In this case, initial ultrasonography evaluation for biliary colic was not suggestive of any biliary pathology and to avoid unnecessary radiation exposure in this young patient MRCP was chosen, which was suggestive of choledochal cyst. Diagnostic accuracy of MRCP is 98% in benign disease relatively high as compared to CT scan (82.86%) [11]. It is preferable due to its capability to analyse cyst structure, evaluate the degree, location, and type of bile duct dilatation, and detect Anomalous Pancreaticobiliary Duct Junction (APBDJ) as well as the fact that it is non invasive [12].

Choledochal cyst complications might include spontaneous cyst rupture, recurrent cholangitis, pancreatitis, and stone formation as a result of bile stagnation. Hepatolithiasis is more common in type IVA. The risk of malignant transformation (adenocarcinoma accounting to 73%-84%) is the main worry. A 90% of patients with malignancy also have type I or type IVA choledochal cysts [8].

Cholangiocarcinoma is rather uncommon in the general population. Patients with choledochal cysts, on the other hand, are thought to have a 6-30% probability of becoming malignant [9,13,14]. The kind, degree, and structure of the biliary tree must be accurately identified prior to surgery in order to plan the surgical strategy diseases of the pancreas and bile ducts. The preferred course of treatment is believed to be cholecystectomy along with complete excision of the extrahepatic component of the choledochal cyst, followed by Roux-en-Y biliary reconstruction. A hepaticoenterostomy can be performed using either a Roux-Y hepaticojejunostomy or a hepatico-duodenostomy. Elhalaby E et al., favoured it since it requires less time during surgery and does not require intestine anastomosis, but additional research is required to support this claim [15]. In addition, Wei MF et al., suggested employing the appendix, which has a vascularised pedicle, as a conduit between the hepatic ducts and the duodenum [16]. Surgery should be carried out as quickly as feasible, because risk of synchronous and metachronous cholangiocarcinoma increases with age.

CONCLUSION(S)

Thorough preoperative evaluation of cyst is necessary. The preferred course of treatment for the extrahepatic component of the condition is complete cyst removal, however, there is debate over how best to handle intrahepatic bile duct dilatations, particularly in the case of type IVA choledochal cysts.

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