

Giant Choledochal Cyst in Adult; a Rare Clinical Entity

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Abstract: Choledochal cyst is an aneurysmal dilatation of bile duct. Biliary cysts typically are a surgical problem of infancy or childhood; however, in almost 20% of patients, the diagnosis is delayed until adulthood. We report a case of choledochal cyst in a 42 year old female patient. She presented to the outpatient clinic with a history of lump abdomen, right upper quadrant abdominal pain & jaundice. The preoperative diagnosis was made by MRCP & it showed type Ia choledochal cyst. The patient underwent laparotomy & excision of cyst with reconstruction done and patient made early recovery. Early suspicion of this rare disease is important because surgical treatment is the only way to avoid dramatic complications of this disease. To the best of our knowledge, we report the largest choledochal cyst in adult.

Keywords: choledochal cyst, magnetic resonance cholangiopancreatography (MRCP), reconstruction

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I. Introduction

Choledochal cyst is an aneurysmal dilatation of the bile duct. It is a rare condition with an incidence ranges from 1:100000 to 1:150000 individuals in western countries to 1:13000 in certain parts of asia [1]. It is three to eight times more common in women than in men.[2]

Choledochal cysts are considered a premalignant condition requiring surgical intervention. They are commonly diagnosed in infancy but may present in adulthood. Here we present the biggest choledochal cyst reported in adults in literature in our knowledge till date.

II. Case Report

A 42 year old previously healthy female patient was admitted in our general surgery department through outpatient clinic, with a history of lump in right upper quadrant of abdomen since 6-7 months, which was insidious in onset & gradually progressing thereafter. It was associated with right upper quadrant abdominal pain since 2-3 months which was colicky in nature & moderate in intensity with no aggravating or relieving factors. Patient also noticed yellowish discoloration of body from last 2 months which started from eyes & then gradually progressed over whole of the body & associated with itching also. There was no history of TB\Diabetes\ Hypertension\ Asthma\ recent weight loss\ drug allergy\ surgical intervention in past. There was no significant family history. The patient was vegetarian by diet, non smoker, non alcoholic, with normal bowel & bladder habit and normal sleep pattern.

On general examination, patient was calm, cooperative & well oriented to time, place & person. Patient was well nourished & icteric. Pulse rate was 86 beats/min, regular with normal volume and character. Blood pressure was 130/80 mm/hg in right arm in supine position.

On inspection lump was seen in right upper abdomen of size approx. 12cm*10 cm with normal overlying skin & no visible pulsations or dilated veins. On palpation, lump of size approx 14cm*12 cm was palpable in right hypochondrium with smooth surface, cystic consistency, well defined margins & moving with respiration.



Fig. 1 preoperative image of patient

Blood investigations revealed a hematocrit of 44% , hb= 12.4 g/dl, tlc=7000/mm³, serum bilirubin(total\direct\indirect)= 16.8\12.2\4.6, serum ALP = 869. Renal function test, blood glucose, serum electrolytes, routine coagulation tests were all normal.

Definitive diagnosis was made on abdominal imaging studies. Ultrasound showed a fusiform dilatation of common bile duct with dilatation of intrahepatic biliary radicles & sludge in gall bladder.

MRCP confirmed the presence of large cystic lesion measuring 16.3cm*13.5cm*10.4cm in continuation with hepatic duct confluence extending upto head of pancreas s\o- type 1a choledochal cyst. There is upstream dilatation of intrahepatic biliary radicles. Gall bladder seen along the anterolateral aspect of cyst with cystic duct seen communicating with the cyst. CT whole abdomen also suggested cystic dilatation of CBD & central dot sign not visualised.

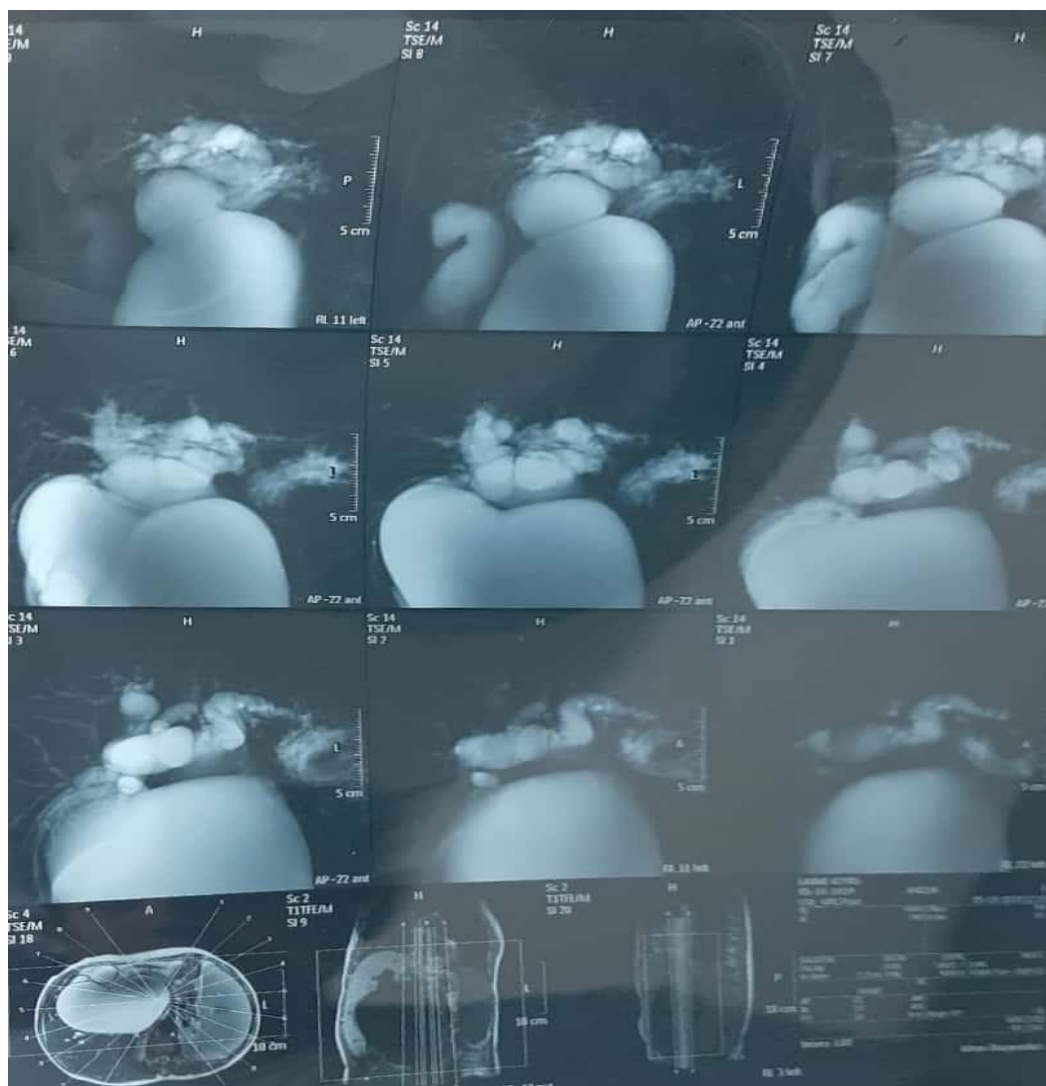


Fig.2 MRCP report

On laparotomy, an oval shaped cystic dilatation of CBD was present with cystic duct entering at upper part of cyst. Whole of the cyst was lifted from adjacent structures & excision of cyst along with gall bladder was done. Reconstruction of bile duct was done by means of hepaticojejunostomy with roux-en-y segment of jejunum about 30cm distal to duodenojejunal flexure joined to common hepatic duct above the cyst. No liver biopsy was performed.

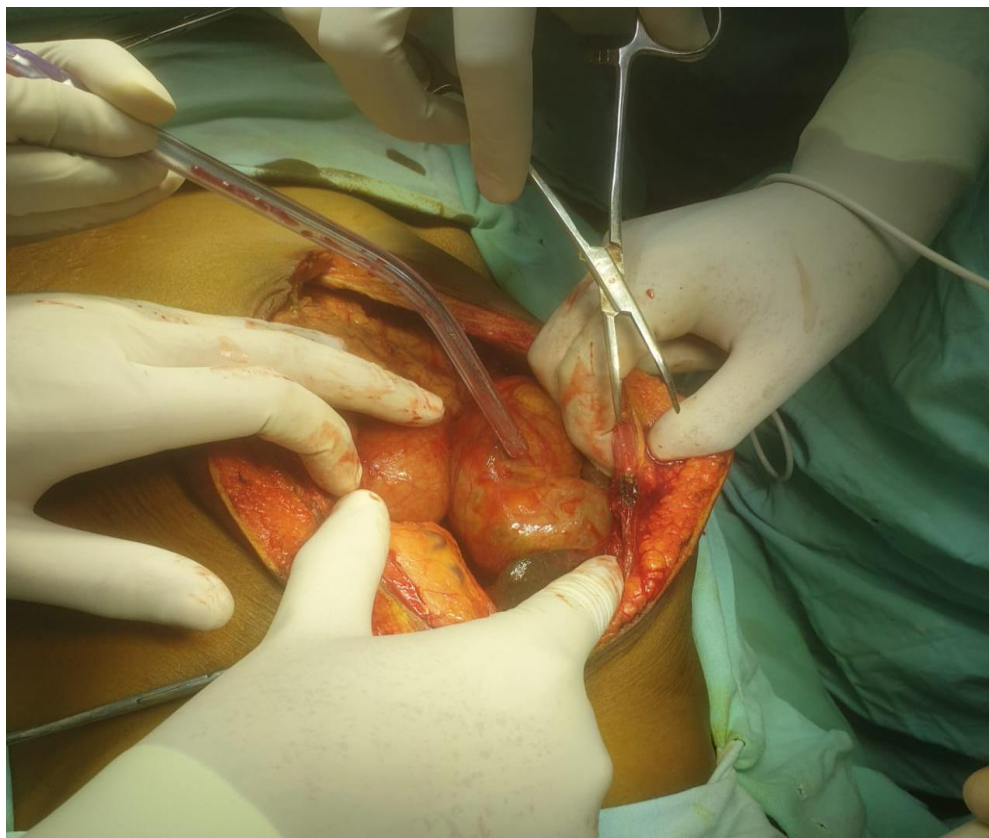


Fig.3 intraoperative image

After removing the specimen out of the abdomen , we measured it in both longitudinal as well as transverse dimension which was found to be 18 cm in longitudinal dimension & 14 cm in transverse dimension. The same was later on proved on histopathology report which gave the report of choledochal cyst 18cm in dimension.



Fig.4 specimen

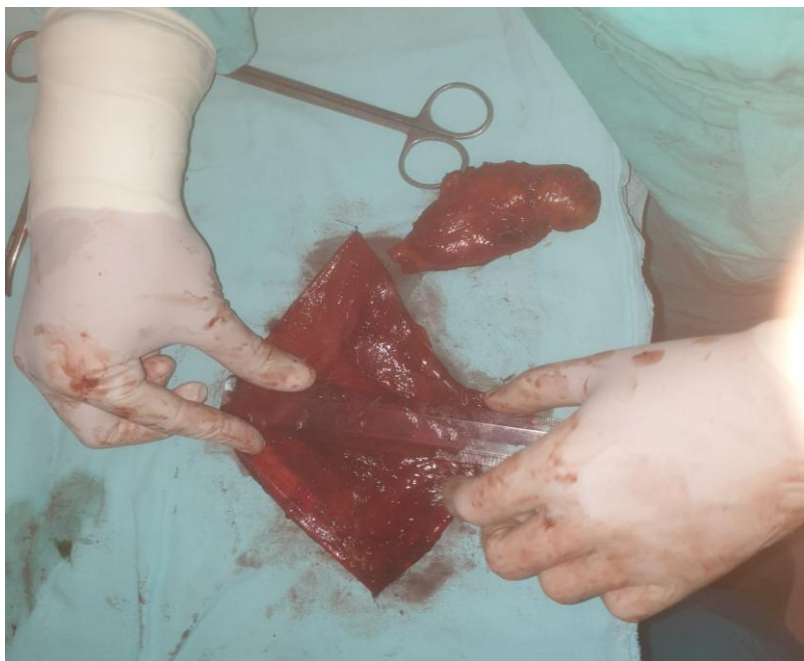


Fig.5 specimen

The patient tolerated the procedure well. She was kept in hospital for 7 days . She was allowed to take orally on fifth postop day & discharged after she started accepting orally well. The pathology confirmed choledochal cyst with no malignancy. Followup after 6 weeks , patient was well & no complications elicited.

III. Discussion

Although not proven, the commonly accepted theory of their pathogenesis relies on the presence of an anomalous pancreaticobiliary junction(APBJ). With APBJ, the pancreatic duct empties into CBD more than 1cm proximal to ampulla. APBJ is seen in up to 90% of patients with choledochal cysts .[1]

Choledochal cyst may present with abdominal pain, obstructive jaundice & a palpable mass in right hypochondrium. Although this classical triad is seen rarely, but was present in our case.[3]

Alonso-Lej et al published the first systematic description of choledochal cyst in 1953[4]. He classified choledochal cysts into three types which was further advanced by Todani et al and includes five major types with several subtypes [5].

Type 1- fusiform dilatation of extrahepatic biliary tree& these are most common type, seen in 75%-85% of cases[6]. In type1a cysts, gall bladder arises from choledochal cyst and extrahepatic tree appears dilated, type1b cysts have an isolated dilatation of most distal aspect of CBD&type1ccysts have smooth fusiform dilatation of CHD & CBD along with pancreaticobiliary malunion[1].

Type 2- saccular diverticulum off the common bile duct

Type 3- cystic dilatation of the intramural common bile duct within the wall of duodenum , also known as choledochocele

Type 4a- cysts involving intrahepatic & extrahepatic biliary tree

Type 4b- multiple cysts limited to extrahepatic biliary tree

Type 5- cysts involving intrahepatic ducts only, also known as caroli disease

Pathologically, cyst wall is composed of fibrous tissue with interspersed elastic & sparse smooth muscle fibres. The internal lining consists of columnar epithelium but often destroyed by inflammation & pressure of distending fluid. So, the choledochal cyst is devoid of epithelium & the wall is fibrous & fails to contract leading to poor emptying & significant bile stasis.

Ultrasound is the best initial investigation study & is diagnostic in many patients and it can help in detecting associated conditions like choledocholithiasis , intrahepatic biliary radicle dilatation, portal vein thrombosis, gall bladder wall thickness.

In most patients complimentary study is necessary including CT scanning & MRI abdomen which provides detailed information concerning relationship between lower end of CBD & pancreatic duct. MRCP helps to create complete cholangiogram. But distal bile duct is difficult to analyse by MRCP , so ERCP is more useful for defining distal biliary tree & pancreatic duct bile duct junction.[2]

Complications of choledochal cyst includes cystolithiasis, recurrent cholangitis, pancreatitis, cirrhosis, hepatic fibrosis & malignant changes.[7]

This anomaly is premalignant & incidence of malignancy ranges from 2.5%-26% [8]. The risk has been reported to be greatest in patients with intrahepatic biliary involvement [9]. The pathogenesis appears to be one of a field defect because the entire biliary tree is at risk, even in non dilated portion of biliary tract & hence, complete excision if benign choledochal cyst does not eliminate the risk of subsequent cholangiocarcinoma development [2].

Surgical management of choledochal cyst consists of resection of entire cyst & appropriate surgical reconstruction.

As in our case, huge type 1a choledochal cyst was there, we completely excised the cyst and biliary enteric anastomosis was done with the help of roux-en-y hepaticojejunostomy. But in some patients with repeated cholangitis & marked pericyclic inflammation, this disease may be best managed with resection of anterolateral aspect of the cyst followed by endocystic resection of the lining, leaving the back wall adjacent to portal vein in place, as reported by Lilly in 1977 [10], but we were able to excise entire cyst.

IV. Conclusion

The size of choledochal cyst varies and it rarely exceeds 9 cm. As the size of cyst is described by its extent & diameter, In our case, the cyst was extending from just distal to hepatic confluence upto head of pancreas (i.e 18cm in longitudinal dimension & 14cm in transverse dimension). The pathology confirmed the same. So, probably the case we encountered and presented may be the biggest choledochal cyst reported in adults till date.

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