

Type Iv-A Choledochal Cyst-A Case Report

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Abstract Choledochal cyst is a congenital cystic dilatation of the bile tract; it can be extrahepatic, intrahepatic biliary radicles, or both. It is a rare condition occurring more commonly in the developing countries (100 times more common in Japan) than in the west and these congenital lesions have a female bias (4 times more commonly than in males). We report a very rare case of Type IV-A (abnormally large-size) in a 15-year-old girl. She presented to the outpatient clinic with a history of recurrent pain and jaundice with fever for 2 months. The preoperative diagnosis was huge choledochal cyst about 20*10*11 cm in size. The patient made complete recovery after cyst-excision with Roux-en-Y hepaticojejunostomy. Early suspicion of this rare disease is important because surgical treatment is the only way to avoid the dramatic complications of this disease. In the case report we discuss the history, clinical features, investigations, complications and treatment of this large size choledochal cyst.

Keywords Choledochal Cyst, Rare, Large, Type IV A

1. Introduction

Choledochal cyst is an aneurysmal dilatation of the bile duct involving the extrahepatic biliary radicles, the intrahepatic biliary radicles, or both. It is a rare condition occurring more commonly in the developing countries (100 times more common in Japan) than in the west [1, 2, 3]. These congenital lesions have a female bias occurring in 4 times more commonly than in males [1, 2, 3, 4]. Presentation with the classic triad of pain, jaundice, and an abdominal mass occurs rarely that is only in 6% of the cases [1, 3, 4].

Todani in 1977 classified choledochal cysts into 5 types based on site of the cyst or dilatation [5]. Type IV-A is characterized by multiple dilations of both--the intrahepatic and extrahepatic biliary tree whereas type IV-B is characterized by multiple dilations of only the extrahepatic bile ducts. We describe a case of an unusual size of type IV-A choledochal cyst which showed massive cystic dilatation of the common bile duct (CBD).

2. History

A 15-year-old previously healthy girl was admitted into Civil Hospital of Ahmedabad, Gujarat with the complaints of recurrent pain and jaundice with fever for 2 months. A sudden severe localized pain first appeared about two months back. It was associated with high-grade, intermittent fever with chills and severe vomiting. It subsided by taking

antipyretics within 3 days.

She noticed a lump on her right upper abdomen, which had been gradually increasing in size for last 2 weeks. She developed yellow coloration of sclera and urine, which was relieved intermittently with the relief of episode of fever and abdominal pain. Thereafter she had several episodes of sudden, dull aching, localized right upper abdominal pain persisting for few hours and relieved spontaneously.

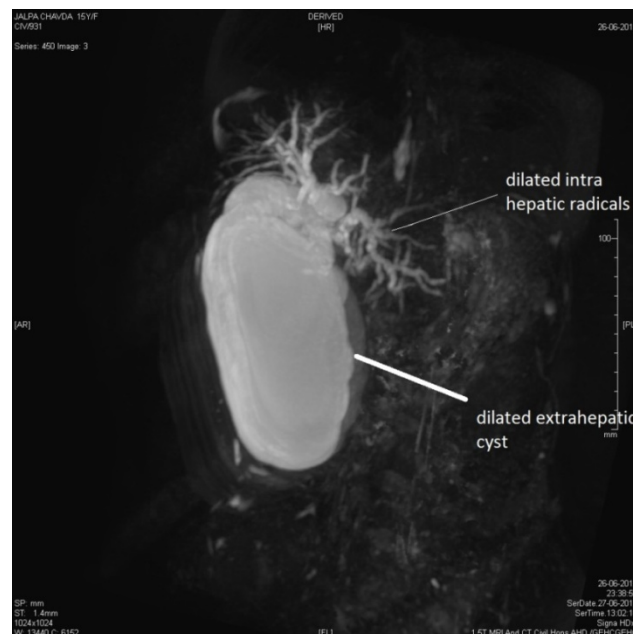


Figure 1. 3d Reconstruction of Mrcp

The patient she was found anemic and non-icteric. Clinical examination revealed a mass in the right hypochondriac, about 18 cm from right subcostal margin along midclavicular line, mildly tender, smooth, firm with

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Published online at <http://journal.sapub.org/surgery>

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rounded margin, dull on percussion and there was no bruit.

Investigation revealed hemoglobin of 8 gm/dl, total count of 7000/mm, and platelets count of 368,000/dl. Liver function test showed serum bilirubin of 0.8 mg/dl, serum glutamic-pyruvic transaminase (SGPT) of 90 u/l, and Alkaline Phosphatase of 915 u/l. Ultrasound showed grossly dilated intrahepatic, central biliary radicals and cystic duct of size 9*7 cm with fluid debris, internal echoes and multiple tiny calculi in right hypochondrium – suggesting type IV - A choledochal cyst (Figure 1).

The definitive diagnosis was made on the basis of magnetic resonance cholangiopancreatography (MRCP) which showed a cystic dilation of intrahepatic biliary radical (both right and left), common hepatic duct and common bile duct upto its distal end. Common bile duct measure 94 mm in transverse and 161 mm in cranio caudal – re-suggesting a Type IV-A choledochal cyst (Figure 2).



Figure 2. Coronal section of MRCP

On the basis of history, clinical examination and investigation, decision to do was choledochal cyst excision with Roux-en-Y hepatico-jejunostomy. Per-operatively findings were – huge choledochal cyst about 20*10*11 cm in size containing thick bilious fluid with grossly dilated gall bladder and dilated cystic duct with common hepatic duct. No distal opening of cyst was identified and no other abnormality in bowel loops identified (Figure 3).

Excision of the cyst was performed with en bloc removal of the gall bladder and cystic duct reconstruction of bile duct by means of Roux-en-Y hepatico-jejunostomy with jejunal loop about 25 cm from duodenal jejunal junction. The patient tolerated the procedure well; she was kept in the hospital for 8 days. There was no gaping and no discharge from wound; post-operative liver function test was normal on 3 months and six months.

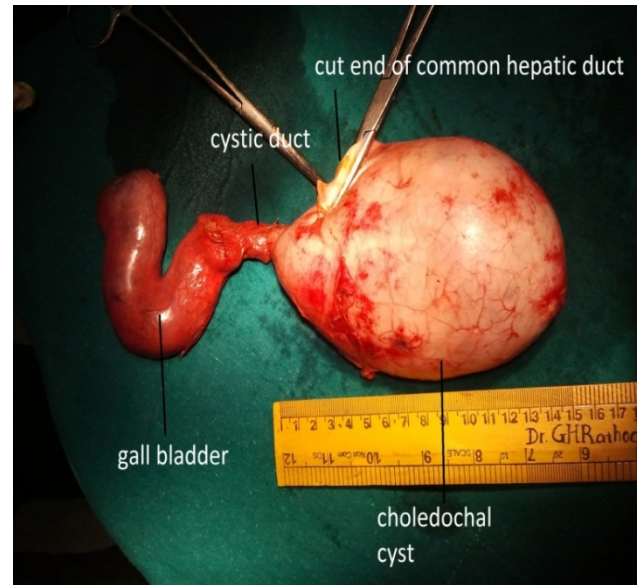


Figure 3. The specimen

3. Discussion

Choledochal cysts are congenital conditions involving cystic dilatation of bile ducts which may be due to the anomalous junction of the common bile duct (CBD) with the pancreatic duct which was first brought to light by Kozumi and Komanda in 1916[6]. But not until Babbitt in 1969 was this accepted universally[7].

An anomalous pancreatic bile duct junction (APBJ) is defined as the junction of the pancreatic duct with the CBD 1 cm or more proximal to where the CBD reaches the ampulla of Vater. The APBJ causes pancreatic secretions and enzymes to become activated which results in inflammation and weakening of the bile duct wall. Severe damage may result in complete denuding of the CBD mucosa.

Several types of choledochal cysts are recognized. Type I is the most common variety (80-90%) involving fusiform dilatation of a portion or entire CBD. Type II is isolated diverticulum protruding from the CBD. Type III or Choledochoceles arise from dilatation of duodenal portion of CBD or where pancreatic duct meets.

Type IV-A is characterized by multiple dilations of the intrahepatic and extrahepatic biliary tree whereas type IV-B has multiple dilations involving only the extrahepatic bile ducts. Type V or Caroli's disease involves cystic dilatation of intrahepatic biliary ducts. The size of choledochal cyst varies, and rarely exceeds 9 cm (in our case 11 cm)[8].

The classical presentation of choledochal cyst in children is jaundice, abdominal mass and abdominal pain[1, 3, 4], but very few patients (6%) present with classical triad as our patient presented[7]. The most common presentation is jaundice followed by pain and mass whereas in adults most common presentation is abdominal pain[3].

Transabdominal ultrasound—a widely available, cheap,

easily reproducible investigation is the best initial investigation for biliary tract. Our patient was initially screened with ultrasound abdomen for the jaundice and abdominal lump which was suspicious of type IV-A choledochal cyst. MRCP is the gold standard imaging for evaluation of choledochal cyst. In our case MRCP confirmed the presence of huge Choledochal cyst with intrahepatic as well extrahepatic components.

If untreated choledochal cyst produces liver damage, which progress to biliary cirrhosis and portal hypertension with all its manifestations[9]. Also it may cause other life threatening events like pancreatitis and cholangitis. Not only that the patient is also at very high risk for having cholangiocarcinoma. Fortunately our patient was diagnosed at very early age and none of these complications occurred.

Once diagnosed the choledochal cyst is treated surgically. Careful identification and protection of the orifice of the pancreatic are mandatory. Surgical treatment of the choledochal cyst is complete cyst excision followed by bilioenteric anastomosis.

Type I cysts are resected and biliary apparatus is reconstructed by hepatico-jejunostomy with a Roux-en-Y segment of jejunum. Type II cyst can be usually resected with the defect in the C.B.D primarily suture over a T-tube which is brought out through a separate incision in the duct. Type III cyst is approached through the duodenum by either a trans-duodenal sphincteroplasty or endoscopic sphincterotomy[10].

Rest of the types constitutes a difficult management problem. Patient with type IV-A, choledochal cyst are best treated with extra hepatic cyst excision and bilioenteric anastomosis which significantly reduces the chances of cholangiocarcinoma. If only left lobe is involved, then left hepatectomy may be done. The bilioenteric anastomosis is achieved using Roux-en-Y hepatico-jejunostomy. In our patient the dilated intrahepatic ducts were not localized to left lobe thus not been amenable to partial hepatectomy. Therefore decision to do extra hepatic cyst resection and bilioenteric anastomosis was done.

In conclusion, the classical presentations of choledochal cysts are quite rare. Also the occurrence of such large cyst of type IV-A is very uncommon. Initial screening can be done by means of ultrasound abdomen. If it is suggestive of such lesions, MRCP should be done as it is gold standard for

evaluation abnormal biliary tract anatomy. Surgical treatment by means of cyst excision is done by incorporate partial hepatectomy. The prognosis for most patients with cysts of the extra hepatic bile duct, appropriately resected and reconstructed is excellent. Post-operative occurrence of stricture is about 3%[3].

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