CASE REPORT

MANAGEMENT OF A GIANT TYPE IV A CHOLEDOCHAL CYST: CASE REPORT

Narendranath Nagoti[1], Amulya Chagalamarri[2], Dwarakanath Nagoti[3]

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ABSTRACT: Choledochal cysts are uncommon congenital anomalies of the biliary tree defined as abnormal, disproportionate, cystic dilatations of the biliary duct.(1) We report a 20 month old female child who presented with obstructive jaundice with recurrent episodes of cholangitis. She was diagnosed with a giant type IVA choledochal cyst with huge cystic dilatation of common bile duct (CBD) and multiple cystic dilatations of the intrahepatic biliary channels. The patient experienced complete recovery after the cyst was excised using LILLY technique and biliary reconstruction was done using Roux-en-Y hepatico jejunostomy at the level of the hilum. Here we discuss the clinical features and management of this giant type IVA choledochal cyst.

KEYWORDS: Choledochal cyst, hepaticojejunostomy, common bile duct, intrahepatic biliary dilatation.

ABBREVIATIONS: Common bile duct (CBD), Endoscopic Retrograde Cholangiopan-creaticography (ERCP), Intrahepatic Biliary Radical Dilatation (IHBRD)

CASE REPORT: A 20 month old female child was referred to our pediatric surgery department for recurrent attacks of fever and jaundice of 6 months duration, the present episode since 5 days. On examination, she had jaundice, fever and a large cystic swelling occupying the entire right upper abdomen with ill-defined borders. Laboratory investigations revealed total bilirubin 4.3 mg/dL, direct bilirubin 2.1 mg/dL, alkaline phosphatase 311 U/L, ALT 85 U/L and AST 26 U/L. The ultrasonographic examination demonstrated a huge cystic lesion below the liver with dilated intrahepatic ducts. Contrast enhanced CT scan disclosed a huge cystic dilatation 10x6 cm in close contiguity with the biliary tree extending from hilar region upto distal CBD level. CBD is not separately delineated from the lesion. Pancreas head was markedly compressed and pancreatic duct was stretched and displaced along the inner margin of the lesion.

A provisional diagnosis of giant type IVA choledochal cyst with enormous cystic dilatation of the extra hepatic CBD was made. As she was not fit for endoscopic retrograde cholangiopancreaticography (ERCP), she was taken up for the surgery after cholangitis subsided and after optimization of the coagulation and nutritional status.

On table a large thick walled cyst measuring 10x8 cms arising from the CBD was noted. The posterior wall of the cyst was densely adherent to the portal vein. Hence cyst was transversely opened; mucosectomy of the cyst was done to minimize the risk for injuring the portal vein and hepatic artery (LILLY technique). The extrahepatic component of the cyst was excised. There was no evidence of stricture between intra- and extra-hepatic ducts and intrahepatic duct dilatation was continuous to the dilated extra-hepatic duct, hence biliary-enteric flow was re-established through a wide mucosa to mucosa Roux-en-Y hepaticojejunostomy at the

level of the hilum. Post-operative period was uneventful, oral feeds were started on post op day 3 and the child was discharged on 10th day after sutures were removed.

CT SCAN IMAGES:

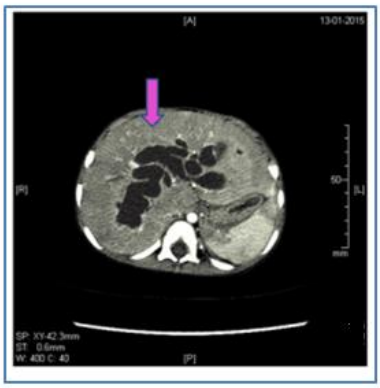

Fig. 1: CT scan image showing gross intrahepatic biliary dilation in both lobes of liver

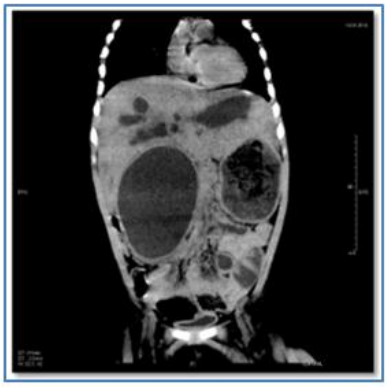

Fig. 2: CT scan showing gross cystic dilatation of CBD of liver

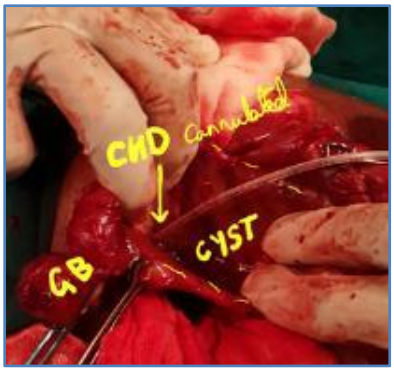

Fig. 3: Picture showing choledochal cyst was transversely opened and common hepatic duct was cannulated

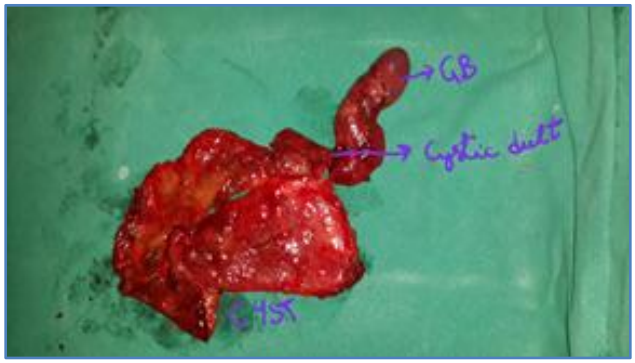

Fig. 4: Picture showing excised specimen of choledochal cyst (along with gall bladder)

INTRODUCTION: Choledochal cyst is a relatively rare congenital anomaly involving cystic dilation of various ducts of the biliary tree with an estimated incidence of 1 in 100,000 to 130,000 live births in western countries and is far more common in Japan.[2] More than two-thirds of cases are diagnosed in children younger than 10 years of age with a female predominance of 3 to 4:1.[3]

DISCUSSION: The exact etiology of origin of Choledochal cyst remains unknown and is thought to develop as a misarrangement of the embryonic connections in the pancreatic biliary ductal system as described by Babbitt in his "long common channel" theory of Pancreatico-Biliary Maljunction (PBM) anomaly in 1969.[4] As a result there is reflux of pancreatic juice into the biliary tree and destruction of the bile duct wall.

The most common type of choledochal cyst is Type I and accounts for more than 75% of all choledochal cysts, followed by type IV A. Type IVA cysts involve both intra- and extra-hepatic ducts and usually have strictures at the hepatic hilum.[5]

In children symptoms include recurrent abdominal pain (82%), nausea and vomiting (66%), mild jaundice (44%), an abdominal mass (29%), and fever (29%).[3] The classic triad of abdominal pain, jaundice, and abdominal mass occurs in less than 10% of patients. A large choledochal cyst, can lead to serious complications like cystolithiasis, cholangitis, pancreatitis, cirrhosis, hepatic fibrosis, portal hypertension, and development of cholangiocarcinoma.[6] The incidence of malignant transformation is reported to be 2.5–26% and is greater in patients with intrahepatic biliary involvement.[7]

Ultrasonography is the best screening method and can be used to detect the condition antenatally also. It clearly demonstrates intrahepatic biliary dilation and the state of the liver parenchyma. MRCP is the current "gold standard" in the imaging of choledochal cysts.[8] Intraoperative cholangiography is unnecessary if the entire biliary system has been delineated before cyst excision, but it should be used if the pancreaticobiliary ductal system has not been completely visualized.

Though internal drainage was performed as the standard operation for choledochal cysts, it increased the frequency of cholangitis, biliary stones, and the risk of malignant changes in the retained cyst or gallbladder.[9] Currently, the definitive treatment of choledochal cysts is to excise the whole extrahepatic bile duct and perform Roux-en-Y hepaticojejunostomy.

Before excision the anterior wall of the cyst is opened transversely. The posterior wall is visible from the inside, facilitating the dissection of the portal triad. If the cyst is extremely inflamed and the adhesions are very dense, mucosectomy of the cyst should be performed rather than attempting full-thickness dissection to minimize the risk for injuring the portal vein and hepatic artery (LILLY technique).[10]

To prevent postoperative pancreatitis or stone formation, the caudal CBD should be resected as close as possible to the pancreaticobiliary ductal junction. Severe IHBRD can be managed by segmentectomy of the liver, intrahepatic cystoenterostomy, or balloon dilatation of a stenotic lesion.[11] However, the incidence of late complications appears to be low and excessive surgical intervention may be unnecessary, except in specific cases. If IHBRD persists after definitive surgery, careful follow-up is mandatory.

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AUTHORS:

1. Narendranath Nagoti
2. Amulya Chagalamarri
3. Dwarakanath Nagoti

PARTICULARS OF CONTRIBUTORS:

1. Senior Resident, Department of General Surgery, NRIIMS, Visakhapatnam.
2. Associate Professor, Department of Obstetrics and Gynaecology, Andhra Medical College, Visakhapatnam.
3. Professor & HOD, Department of General Surgery, NRIIMS, Visakhapatnam.

NAME ADDRESS EMAIL ID OF THE CORRESPONDING AUTHOR:

Dr. Narendranath Nagoti,
403, M. K. Royale,
Opp. Grand Bay Hotel,
Visakhapatnam-530002.
E-mail: narendranath87@gmail.com

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