Case Report:
Ultra Sound Evaluation of Choledochal Cyst With Portal Hypertension

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Abstract: Choledochal cyst is an uncommon congenital cystic dilatation of the bile duct. The underlying etiology is believed to be an anomalous junction of the pancreatic duct and common bile duct (CBD) that allows free reflux of pancreatic enzymes into the CBD, weakening its wall. Portal hypertension is a rare complication of choledochal cyst. We report a case of choledochal cyst with portal hypertension confirmed by surgery.

Key Words: Choledochal cyst, Portal hypertension

Introduction:
Choledochal cyst is mainly seen in childhood although no age is exempt.(1) Some cases are diagnosed in adults accounting for 15 to 20%. With increase in advancement in technology, the incidence rate of choledochal cyst is increasing. Portal hypertension is an uncommon complication of choledochal cyst. The treatment of choledochal cyst complicated by portal hypertension has evolved from internal drainage of cysts to single stage excision of cyst with biliary-enteric anastomosis.

Case Report
A 7 month old female presented with complaints of yellowish discoloration of body since 2 days, intermittent whitish stool since 15 days and 2 episodes of blood mixed vomiting. There was no previous history of acute cholecystitis, pancreatitis, hematemesis or melena. Baby was of low birth weight (1.7 kg) delivered by LSCS. She cried immediately after birth. On examination, baby was sick, febrile with HR156/min and RR 48/min. Pallor (+) and icterus (++) was also present. Biochemical investigations: Haemoglobin- 6.4 gm/dl, Serum ALT-132.7 U/L, Serum AST- 145.5 U/L, Serum ALP -686.1 U/L , Total bilirubin 10.24mg%, Direct bilirubin 6.22 and indirect bilirubin 4.02mg%. Hepatitis B and C were non-reactive. Per abdomen examination showed firm, mildly enlarged liver with presence of caput medusae and visible tortuous veins. USG abdomen showed large cystic dilatation of CBD measuring 9cmx7cm compressing the portal vein with collaterals and multiple intrahepatic cystic dilatations of biliary channels. Few collaterals in periumbilical and epigastric region with moderate ascites were also seen. So the USG diagnosis of Choledochal cyst (Type IV A) with portal hypertension was made which was confirmed by surgery. Complete excision of the dilated extrahepatic duct followed by Roux-en-Y hepaticojejunostomy to restore the continuity was done. Patient’s hospital course was eneventful and she was discharged without any complications.

Fig 1: USG abdomen showing large cystic dilatation of CBD compressing the portal vein
The incidence of choledochal cysts has increased due to increase in pickup rate by means of advanced technology. Incidence in Asia is somewhat higher than in western countries. The reason for this geographical difference is still unclear.(2,3) There is also an unexplained female preponderance with female: male ratio commonly reported as 4:1. The most widely accepted hypothesis regarding etiology is an anomalous arrangement of the pancreaticobiliary ductal junction.(4,5) The triad of jaundice, right upper quadrant pain and a palpable subcostal mass is diagnostic but is not seen in all cases.

Choledochal cysts can be associated with biliary atresia, congenital hepatic fibrosis and cystic disease of the kidney especially renal tubular ectasia, sometimes combined with cortical and medullary cysts. Reported complications of choledochal cysts include secondary calculus formation, pancreatitis, biliary cirrhosis, cyst rupture with bile peritonitis, cholangitis, intrahepatic abscess, portal vein thrombosis and malignant transformation into cholangiocarcinoma. Todani et al (6) have classified choledochal cysts into five types:

- Type I: Fusiform cystic dilatation of extrahepatic CBD
- Type II: Eccentric fluid-filled cyst (diverticulum)
- Type III: Localised cystic dilatation of distal intramural segment of CBD
- Type IVA: Multiple intrahepatic and extrahepatic bile duct cysts
- Type IVB: Multiple extrahepatic bile duct cysts
- Type V: Multifocal saccular dilatation of IHBR (Caroli’s disease)

Ultrasoundography is preferred for initial evaluation. It reveals an anechoic cystic structure separate from the gall bladder that communicates with the hepatic ducts.(7,8) Differential diagnoses on ultrasound include other fluid filled structures in this region namely pancreatic pseudocyst, large right renal cyst, enteric duplication cyst and hepatic artery aneurysm. Hepatobiliary scintigraphy can also complement the diagnosis by showing late accumulation of radioisotope in the cystic structure.

Type II choledochal cysts may appear separate from the CBD as its neck may be narrow. Type III choledochal cysts (choledochocele) are rare and difficult to diagnose on US and CT but MRCP and cholangiography can reveal the typical cobra head appearance bulging into the duodenum. In patients with Type V choledochal cyst (Caroli’s disease), imaging reveals enhancing fibrovascular bundle of portal vein radicles completely surrounded by dilated bile ducts (central dot sign). MRCP and in particular cholangiography are best to show the communication of these cystic areas with the biliary tree. CT, MRI and cholangiography can accurately diagnose and classify choledochal cyst. MRCP is equivalent to ERCP in detecting and defining the morphology of choledochal cysts and in detecting the presence of anomalous union of the pancreatic and bile ducts.(9) Preoperative knowledge of the type and extent of choledochal cyst is important for surgical planning which in most cases involves excision of the choledochal cyst and biliary drainage by Roux-en-Y hepaticojejunostomy.

Caroli’s disease when localized to one hepatic lobe can be treated with hepatectomy, while diffuse involvement may necessitate liver transplantation.

Portal hypertension is a rare complication of long standing choledochal cyst manifested clinically as hepatosplenomegaly, jaundice, hematemesis, melena and ascites. Portal hypertension in patients of choledochal cyst may be due to extrahepatic biliary obstruction leading to secondary biliary cirrhosis, recurrent inflammation leading to portal vein thrombosis, direct compression of portal vein by choledochal cyst (our case) or associate congenital hepatic fibrosis in patients with Caroli disease. (5,10-12)

Conclusion:

Only few cases of Type (IVa) Choledochal cyst have been reported till now. Management issues of portal hypertensive patients are inadequately addressed, as its incidence is low and underlying causes variable. Management of choledochal cysts depends on the severity of liver disease in cases of cirrhosis of unrelated cause, while those with secondary biliary cirrhosis should be considered for surgical management. Endoscopic stenting may be considered as a temporary measure in high-risk cases.

Surgery of Type (IVa) choledochal cyst includes complete excision of the dilated extrahepatic duct, followed by a Roux-en-Y hepaticojejunostomy to restore continuity. Intrahepatic ductal disease does not require dedicated therapy unless hepatolithiasis, intrahepatic ductal strictures, and hepatic abscesses are present (in such instances, resection of the affected hepatic segment or lobe is performed).
Consent: Written, informed consent was obtained from the patient for the publication of this case report and accompanying images.

References: