

Decompensated cirrhosis as a late postoperative complication of choledochal cyst

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Sri Lanka Journal of Child Health, 2009; **38**: 38-39

(Key words: Choledochal cyst, cirrhosis, common bile duct, CBD)

Introduction

Choledochal cysts are congenital bile duct anomalies. They consist of cystic dilatation of the biliary tree involving extrahepatic biliary tree, intrahepatic biliary tree or both. Surgical intervention is the definitive management. Even after surgical intervention some may develop complications such as cholangiocarcinoma, stricture formation at site of anastomosis and intrahepatic bile duct stones.

Case report

A 7 year old boy presented with generalized oedema and jaundice of six months duration. His birth weight was 3.3 kg and he was born to non consanguineous parents. There was no history of neonatal jaundice. He was healthy up to the age of 4½ years when he developed intermittent obstructive jaundice and abdominal pain.

Investigations were carried out including ultrasound scan (USS) of abdomen and CT scan of abdomen. A choledochal cyst had been diagnosed and the child had undergone surgery at 4 years and 9 month of age.

After surgery child had been well up to 6½ years of age when he again developed obstructive jaundice with abdominal pain. Jaundice worsened progressively and he gradually developed oedema. During this period the child had a few bouts of haematemesis and melaena.

On examination, the child was oedematous, deeply icteric, and had pallor, leuconychia and clubbing. Abdomen was distended. Liver was hard in consistency, had an irregular surface and was non tender, extending 6 cm below the right costal margin; left lobe of the liver was easily palpable. Spleen was firm in consistency, extending 3 cm from left costal margin. There was marked ascites.

White cell count was $18.8 \times 10^9/l$ (N 80%). Haemoglobin was 6.3 g/dl. Liver function tests were as follows: AST 168 u/l (9-48), ALT 23 u/l (9-48), serum bilirubin, 249 $\mu\text{mol/l}$ (direct 165 $\mu\text{mol/l}$), prothrombin time 28 sec with INR 1.69, alkaline phosphatase 526 u/l (80-480). Total serum protein was 70 g/dl (64-83), albumin 19.3 g/dl (35-50) and globulin 50.7 g/dl. Serum cholesterol level was 1.7 mmol/l (3.6-5.7). Serum amylase level was 1336 IU/l (27-102). Serum alpha feto protein was 2.5 ng/ml (1-15).

USS of abdomen showed stenosis at anastomotic site of common bile duct (CBD) and jejunum. CT scan of abdomen showed an irregular, ill defined area of altered attenuation with mixed density segment of liver and intrahepatic bile duct dilatation suggestive of post surgical site stricture formation. HIDA scan showed hepatocyte dysfunction (impaired trace uptake) and impaired bowel activity. (no significant trace excretion up to 24 hours).

Discussion

Choledochal cyst has an incidence varying from 1:100,000-200,000¹. It is commoner in females with a 3:1 ratio. Choledochal cysts are more common in Asian than Western countries^{1,2}. Choledochal cyst was first reported in Sri Lanka in 1985³.

Pathogenesis of choledochal cyst is multifactorial. Anomalous junction between CBD and pancreatic duct can be demonstrated in 90-100 % of patients with choledochal cyst¹. This occurs when the pancreatic duct empties into the CBD more than 1 cm proximal to the ampulla. This abnormal union allows pancreatic secretions to reflux into CBD, where the pancreatic proenzymes become activated, damaging and weakening the bile duct wall. The second aetiologic theory is congenital weakness of the bile duct wall due to defects in epithelialization and recanalization of the developing bile ducts¹.

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(Received on 6 June 2008. Accepted on 25 June 2008)

Choledochal cysts are classified according to the anatomy of cyst^{1,2}. There are 5 major classes (type I – V) with sub classifications for types I and IV.

Type I: This represents 80-90% of choledochal cysts. There is saccular or fusiform dilatation of CBD involving entire duct or segment of duct.

Type II: These are isolated diverticula protruding from the wall of the CBD.

Type III: Cysts arise from the intraduodenal portion of CBD- Choledochocoele^{1,4}.

Type IV: Multiple dilatations involve intrahepatic and/or extra hepatic bile ducts.

Type V: Multiple dilatations involve only the intrahepatic bile ducts^{1,2}: - Caroli disease.

Clinical presentation of patients with choledochal cyst varies with age^{1,4}. Infants commonly present with jaundice, acholic stools and abdominal mass in the right upper quadrant which may be accompanied by hepatomegaly. After infancy, children typically present with intermittent biliary obstruction or recurrent episodes of pancreatitis. The triad of abdominal pain, jaundice and right upper quadrant abdominal mass is found in only 10-20% of cases^{1,5}. Complications include cholelithiasis, severe pancreatitis, hepatic abscess, cirrhosis and portal hypertension.

Laboratory investigations are non specific. White blood count is elevated with neutrophilia in the presence of cholangitis^{1,2}. Hepatocellular enzymes and alkaline phosphatase are elevated in most situations. Serum amylase and lipase levels are increased in the presence of pancreatitis. Elevated levels of amylase can be observed in biliary obstruction and cholangitis.

Imaging studies are more useful in diagnosis, ultrasonography being the investigation of choice¹. Choledochal cyst can be diagnosed via USS in the antenatal period as early as 12 weeks of gestation^{1,5}. Abdominal CT and MRI help to delineate the anatomy of lesions and surrounding structures, and define the presence and extent of intrahepatic ductal involvement. Magnetic resonance cholangio-pancreatography (MRCP) is valuable in defining the anomalous pancreatobiliary junctions¹.

If non invasive measures (USS, CT, and MRI) fail to delineate the anatomy, they should be supplemented by invasive procedures such as percutaneous transhepatic cholangiography (PTC) or endoscopic retrograde cholangiopancreatography (ERCP)¹.

Histological findings are supportive. Evidence of chronic inflammation is typically seen in cyst wall. In the liver, ductal fibrosis and portal oedema may be observed; changes of biliary cirrhosis may be seen in patient with long standing illness.

Treatment of choice is complete excision of cyst and construction of a biliary-enteric anastomosis to restore the continuity of the intestinal tract^{1,5}. If child presented with cholangitis broad spectrum antibiotics should be directed against common biliary pathogens, such as *E.coli* and *Klebsiella*¹.

Early postoperative complications include haemorrhage, wound infection, pancreatitis, ascending infection and bowel obstruction. Late postoperative complications include intrahepatic bile duct stones, stricture formation and cholangiocarcinoma¹. Cholangiocarcinoma is the most feared complication of choledochal cyst, incidence ranging from 9 – 28 %¹.

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