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TEACHING FILES (GRAND ROUNDS)

BILIARY PERITONITIS: RARE PRESENTATION OF SPONTANEOUSLY RUPTURED CHOLEDOCHAL CYST

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Clinical Problem:
A 6 months old girl presented with diarrhea and vomiting for 10 days and abdominal distension for 1 day along with oliguria. On examination, weight was 6 kg; child was dehydrated and had ascites with icterus and hepatomegaly. Other systems were normal. Investigations showed haemoglobin of 9.5 gm%, white cell count of 25800/cumm and adequate platelets on smear examination. Bilirubin was 12 mg% (direct of 5.2 mg%) with SGPT 56 IU/L, albumin of 1.8 gm/dl. Prothrombin time (PT) and Partial thromboplastin time (PTT) were 12.6 and 22.8 secs respectively. Ultrasonography (USG) abdomen with colour doppler showed mild hepatomegaly, contracted gall bladder and massive ascites with patent hepatic veins. CT abdomen and angiography confirmed same findings. On second day of hospitalization, child worsened with increased abdominal distension, poor peripheral circulation, tachycardia and altered sensorium. Paracentesis showed frank bile stained ascitic fluid. Ascitic fluid showed 3.0 gm/dl of proteins, 2080 cells/cumm (90% polymorphs, 10% lymphocytes) and 58 mg/dl of sugar. CRP was 25 mg/dl and blood culture and ascitic fluid culture revealed no growth. In view of biliary peritonitis, exploratory laparotomy was done that revealed a ruptured choledochal cyst that was resected and hepaticojejunostomy was done. Child was treated with ceftriaxone and clindamycin for 10 days along with dopamine for initial 3 days and subsequently on follow up after 20 days CRP was negative and her liver function tests showed serum bilirubin of 0.6 (direct-0.3), SGOT - 56 IU/L and SGPT - 34 IU/L.

What is the cause of rupture of choledochal cyst?

Discussion:
Choledochal cyst is a rare congenital anomaly of the bile ducts. It consists of cystic dilatation of the extrahepatic biliary tree, intrahepatic biliary radicles, or both.[1] The clinical presentation changes with age. Overt, dramatic signs and symptoms are more common in infancy. Jaundice, mass, and pain abdomen are the classical findings, especially in infancy.[2,3] Complications of choledochal cyst include pancreatitis, cholangitis, secondary biliary cirrhosis, spontaneous rupture of cyst, and cholangiocarcinoma.[4] Rarely, the patient may present with biliary peritonitis due to rupture of the cyst.[2] In most instances this apparently occurs spontaneously, but traumatic rupture has been documented, and rupture has also been reported during pregnancy or labour.[5] The exact cause of rupture is not definitely known. Distal obstruction is an important etiological factor along with primary weakness of duct wall. Trauma, association with pregnancy, anomalous pancreatico-biliary duct system, and continuous reflux of pancreatic juice in cyst weakening the wall have been implicated in few cases.[5,6,7,8] In our patient, there was diarrhea for 10 days preceding the peritonitis suggestive of ascending infection in the cyst leading to a rupture. Diarrhea and systemic infection has been reported as the cause of rupture of choledochal cyst.[9]

Onset of symptoms is usually acute with vomiting, abdominal distension, features of toxemia, signs of shock, with or without icterus. Similarly, our patient had icterus with ascites and ascitic tap revealed a biliary ascites. Presence of any fluid suggestive of bile should prompt a surgeon to evaluate the biliary tree as was done in our patient that revealed the choledochal cyst rupture. The recommended acute therapy for choledochal cyst rupture is peritoneal drainage, followed by complete cyst resection, cholecystectomy and Roux-en-Y hepaticojejunostomy reconstruction. Post hepaticojejunostomy complications includes cholangitis, intrahepatic bile duct stones, pancreatitis, stone formation in the intrapancreatic terminal choledochus or pancreatic duct, and bowel obstruction.[10] Our patient was therefore treated primarily as a surgical emergency with peritoneal drainage and a more de?nitive surgical procedure i.e. hepaticojejunostomy was accordingly done without any postoperative complication.

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