Choledochal cyst in children: 15-year experience

Choledochal cyst, a localized aneurysmal dilatation of the extrahepatic and/or intrahepatic biliary tree, is an uncommon cause of obstructive jaundice usually diagnosed in children, especially girls. We analyzed our experience with choledochal cyst in 27 children (21 girls) seen over a 15-year period (1990-2004).

The age at presentation ranged from 2 days to 12 years. Three patients were neonates (one of them was diagnosed antenatal at 28 weeks by ultrasonography), eight were below 5 years, twelve were between 5 and 10 years, and four were above 10 years of age. The commonest presenting feature was pain in abdomen (25 patients), followed by lump in abdomen (8). Jaundice was present in eight patients and history of jaundice in six. Five patients had vomiting. The classical triad of jaundice, mass and abdominal pain was seen in five patients.

Ultrasonography was diagnostic in 25 patients (92.5%). In one two-day-old neonate who presented with a large abdominal lump, the sonography diagnosis was intra-abdominal cyst; in the second patient the diagnosis on sonography was duplication cyst of duodenum. In ten patients CT was also done and confirmed the sonography findings.

Peroperative cholangiogram was done in all patients. Laparotomy revealed type I choledochal cyst in 25 cases and type IV in 2 cases. The gall bladder was distended, elongated and tortuous in five cases. In four cases, gallstones were also seen. Cultures revealed sterile bile in 24 cases, while in 3 cases staphylococci and Gram-negative organisms were isolated. Amylase levels in aspirated bile from the cysts were raised in 5 cases.

The surgical procedure performed was total cyst excision with reconstruction of the biliary tract by Roux-en-Y hepaticojejunostomy (23 cases). In four cases a palliative procedure (choleduodenostomy, cystojejunostomy, internal drainage) was done initially. The neonate with cystoduodenostomy had a transient attack of cholangitis. Six months later they were re-explored and excision of cyst with Roux-en-Y jejunostomy was performed. One patient died in the immediate postoperative period due to mismatched transfusion. During the follow-up period of 1 to 15 years (median 7) all patients were doing well and none developed jaundice.

The classic triad of jaundice, abdominal pain and mass, present in 13%-63% of unselected patients with choledochal cyst, is uncommon in children. Ultrasonography is the most cost-effective modality for the diagnosis of choledochal cyst. Only 17 cases with antenatal diagnosis had been reported till 2001. Choledochal cysts in children are predominantly type I cystic lesions, whereas type IV cysts are more common in adult patients. Cholangiocarcinoma is the most dreaded complication and is seen in 2.5%-15% of the cases, rarely before adolescence. In view of the high risk of cholangiocarcinoma, early excision of cyst and Roux-en-Y hepaticojejunostomy is the appropriate treatment of types I and II and the extrahepatic part of type IV biliary cysts. Two-stage procedure is indicated in infected cysts.

Roux-en-Y hepaticojejunostomy has potential disadvantages, viz., high risk of peptic ulcer, and fat malabsorption due to non-availability of bile in the duodenum and proximal jejunum. To overcome these, a number of alternatives have been described for reconstruction of the biliary tract with or without antireflux mechanism, viz., hepaticocodudenostomy with interposed jejunum loop, jejunum spur, intussusception with the jejunum, nipple valve at the lower end of anastomosis, and appendicular conduit.

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References