Giant choledochal cyst in infancy—A rare entity

Introduction

Dilatation of the biliary tree to variable extent and severity is called choledochal cyst (CDC). The prevalence is higher in Asian countries, especially Japan, and females are more affected than males in a ratio of 4:1.1,2 Eighty percent of CDC patients manifest clinically in childhood and are either detected on an antenatal scan or present later with jaundice and/or abdominal pain. The triad of pain, lump and jaundice is rare in children. Giant choledochal cysts (GCDC), defined as >10 cm in diameter, are uncommonly encountered in practice. The reports of GCDC that are described in the literature pertain to older children and adults. This report highlights that a GCDC can present even in infancy and without features of cholestasis.

Case report

A 5-month-old boy presented with failure to thrive and progressive abdominal distension since one month of age. There was no associated history of jaundice, fever or bowel complaints. The child looked emaciated and weighed 5.5 kg (<10th percentile for age). He was anicteric and mild pallor was noted. Abdominal examination revealed fullness on the right side. A soft, large, non-tender lump was palpable in the the right hypochondrium, extending to the right iliac fossa and umbilical region. The lump had a smooth surface and was continuous with liver dullness. Laboratory values were: Hb-10.9 g/dL, total bilirubin-0.7 mg/dL, SGPT-38 IU/L, SGOT-39 IU/L and serum alkaline phosphatase-1038 IU/L.

Ultrasound scan of the abdomen and contrast enhanced computed tomography scan revealed a 13 cm ×8.8 cm well-defined round to oval cystic structure occupying almost the entire abdominal cavity with thin peripheral enhancement and continuous with the right and left hepatic ducts. There was no appreciable intra hepatic biliary radical (IHBR) dilatation (Figures 1a & b).

The child underwent laparotomy and a GCDC was seen occupying the entire right side of the abdomen, reaching up to the pelvis with the ascending colon, duodenum and pancreas lifted antero-medially. The liver was grossly normal (Figure 2). After decompressing the contents (650 mL of greenish bile-tinged fluid) the cyst was completely excised. The common hepatic duct, proximal to the GCDC, was approximately 15 mm in diameter and the lower end of the cyst tapered before joining the pancreatic duct. After flushing the proximal ducts a standard

Figure 1: (a) CECT of abdomen in coronal plane showing the giant CDC, and (b): Axial section of the liver showing absence of IHBR dilatation.
Roux-en-Y hepatico-jejunostomy was performed. The postoperative recovery was uneventful. On follow-up the child is well and thriving.

Discussion

Very few reports of GCDC in infancy were found in the literature with the majority pertaining to adults. Tang et al in a series of 62 children with CDC described a child with jaundice and a cyst measuring 158 mm in the largest dimension, the age of the child was not mentioned though. A massive CDC presenting as an acute abdomen in a 5-month female infant was reported by Okoromah et al. It was initially mistaken as a pseudocyst of pancreas but later confirmed to be CDC by histopathological examination. A recent report by Yurttutan et al describes a 4-month old female infant with abdominal distension and cholestatic jaundice caused by a GCDC. The GCDC had a maximum diameter of 160 mm. The author believes that this is the largest GCDC reported so far in the literature. These scattered reports endorse the fact that this is a rare entity in children, more so in infancy.

The case reported here is unique since the child had neither jaundice nor any other features of cholestasis. This finding is consistent with the observation that a large choledochal cyst has low intra-cystic pressure and produces less cholestatic effects on the liver. This is probably also the reason for a grossly preserved liver with no IHBR dilatation in our case.

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Incidentally detected ectopic ampulla of vater in the antrum in a patient of colonic tuberculosis

Introduction

The ampulla of Vater is located in the posteromedial wall of the...