CASE REPORTS

HUGE CHOLEDOCHAL CYST

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INTRODUCTION

The first anatomic description of the choledochal cyst (CC) was presented by Vater in 1700's. Choledochal cyst is a rare abnormality usually found in infancy and childhood and occurs mainly in the females [1]. Presenting symptoms are age dependent with jaundice prevailing in childhood and abdominal pain in adults [2]. Outcome depends on early diagnosis, complete resection of the cyst and proper hepatico-enterostomy. Simple drainage procedures, leaving the cyst behind can lead to late complication, including development of carcinoma [1].

CASE REPORT

A one year old girl was brought to our department in Dec, 2003 with progressive abdominal distention, vague abdominal pain and intermittent low grade fever. There was no anorexia, jaundice or vomiting. Also there were no urinary or bowel complaints. Birth, developmental and family history was not significant

On examination she weighed 10Kg with average physique, but was pale. Abdomen was diffusely distended with prominent veins (fig. 1). Hemoglobin was 7gm/dl, with a total leucocyte count of 16000/cubic cm. Liver functions were slightly deranged and coagulation profile was within normal limits. Ultrasonography of abdomen showed about 18x20 cm sub hepatic cystic swelling separate from the liver. Hepatic texture was normal. CT scan abdomen revealed huge cystic swelling occupying most of the abdomen, displacing the gut. A diagnosis of choledochal cyst was suggested.

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Surgery was planned. Pre-operatively the patient was given blood, vitamin-k antibiotics and fluids. Abdomen was opened by supra-umbilical transverse incision. There was a fusiform (type-I) choledochal cyst measuring approximately 18x20cm is size (fig. 2). It was densely adherent to the surrounding structures, with colon plastered over its anterior wall and posteriorly to the portal vein .Its distal end was stenosed. Proximally cyst was extending up to the confluence of common hepatic duct (CHD). Cyst was excised in toto by Lilly's method in which intra-mural resection (mucosectomy) was carried out leaving behind only the leash of tissue adherent to portal vein. Retro colic Roux-en-y - hepaticojejunostomy was carried out.

Post-operative recovery was uneventful. Patient was discharged on 4th post-operative day with advice to continue oral antibiotics for 04 weeks. Patient is having regular follow up in our department and is symptom free 9 months after surgery. Histo-pathology report has confirmed choledochal cyst.

DISCUSSION

Choledochal cyst is the cystic dilatation of biliary tree. Girls are affected more than boys (ratio 4:1). There are five common anatomic forms of choledochal cyst with other variants. Type-I: It is the most common variant (constitutes 85-95% of the cases). It is the cystic or fusiform dilatation of common bile duct (CBD). Our patient belonged to this type. Type-II: diverticulum of CBD. Type-III: choledochoele, which is usually intraduodenal but occasionally is intra-pancreatic. Type-IV: is second most common. There is both extra and intra hepatic biliary dilatation. Type-V: there are multiple intra-

hepatic cysts with hepatic fibrosis (Caroli's disease).

The exact aetiology of CC is unknown. However, many theories have been offered to explain the pathogenesis of CC. Three of them have been widely accepted as causative factor, like congenital weakness of the wall of the CBD (primary ductal ectasia), distal CBD obstruction and long common channel theory (pancreaticobiliary maljunction-PBM). This PBM allows the reflux of pancreatic juices, containing enzymes like trypsin, lipases & amylases into the CBD, thereby weakening its walls by enzymatic destruction, resulting in inflammation, dilatation & cyst formation [3]. However this theory does not explain the aetiology of other types of CC. In our case we failed to demonstrate any PBM.

Clinical presentation varies according to age of patient, due to differences in their underlying pathology and can be divided into two groups. In infantile group, up to age of 6 months, the obstructive component predominates, so that the commonest presentation is jaundice. The so-called adult form of clinical presentation occurs in patients older than 6 months and usually older than two years of age. The presentation is much more subtle and confusing. In this group the usual presentation is vague abdominal pain with minimal jaundice. The classic triad of pain, mass and jaundice may be present but is uncommon. Our case only presented with a mass, and did not have any clinical jaundice. She also did not have gross loss of weight, in spite of the size of the mass.

In the past, the diagnosis of CC was considered to be very difficult and was usually based on upper gastrointestinal contrast study. Currently, abdominal ultrasonography (USG) is the mostly commonly used procedure. It is the best, non-invasive, screening method and in experienced hands, it is very accurate [4]. However, if the diagnosis is still, doubtful or the regional anatomy remains obscure, especially in older child, computerized

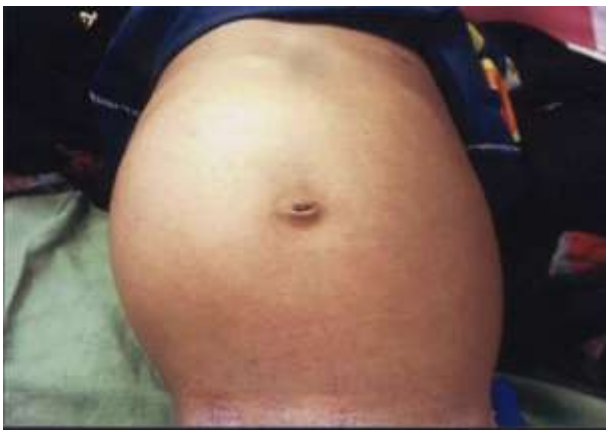

Fig. 1: View of the abdomen of the child, before operation.

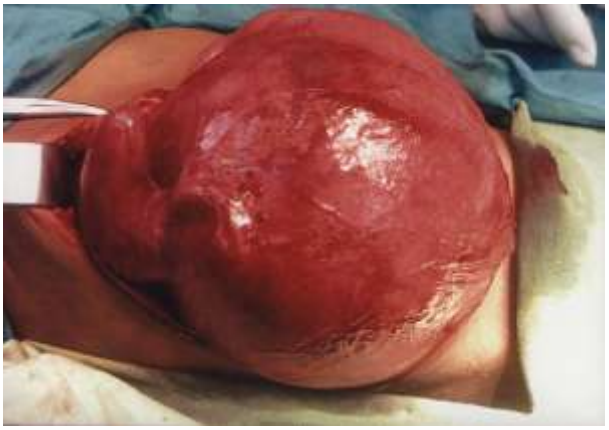

Fig. 2: Per-Operative view of the cyst.

tomography (CT) scan, endoscopic retrograde cholangiopancreaticography (ERCP), magnetic resonance imagining (MRI) technique and percutaneous transhepatic cholangiography (PTC) may have to be considered. ERCP has become a very useful technique especially in demonstrating an anomalous choledocho-pancreatic duct junction & long common channel [5,6].

The operative technique of CC has undergone many changes over the years. Internal drainage without cyst excision has been the most popular approach for many years, particularly cystoduodenostomy and the Roux-en –Y cystojejunostomy. These procedures are simple but the long-term morbidity from these procedures is high. This includes recurrent cholangitis [7], pancreatitis and malignancy (3-20%) from the retained

cyst wall [8]. Therefore, the treatment of choice is now considered to be complete excision of the cyst with internal drainage usually by a Roux-en-Y hepaticojejunostomy. If the pericystic inflammation is severe and obscures the anatomy, as in this young girl with a huge cyst, excision of the cyst by circumferential dissection can be difficult and hazardous. In this situation, we resorted to the intramural resection, leaving the back wall adjacent to the portal vascular structures in place, as described by Lilly [9].

Despite the good prognosis, careful long terms follow up of the patients is still required because late complications of cholangitis, pancreatitis and malignancy have been reported 20 to 30 years after surgery.

CONCLUSION

CC is rare congenital disease. Its correct diagnosis and prompt referral to a paediatric surgical center can improve the outlook for these young patients. In view of the high risk of cholangiocarcinoma, resection of the cyst is recommended. Patients who may have undergone internal drainage in the past still should undergo resection of the cyst. In view of complications occurring late in adult life, parents counseling is important.

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