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CHOLEDOCHAL CYSTS PRESENTING AS INTESTINAL OBSTRUCTION IN NEONATES- A DIAGNOSTIC DILEMMA?

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ABSTRACT

Choledochal cyst is a rare congenital condition, which presents as cystic or fusiform dilatation of the bile duct. The anatomic description and classification scheme for choledochal cystic disease was initially proposed by Alonso-Lej *et al.*, (1959) and later modified by Todani *et al.*,. The most frequently encountered type is type I cysts, accounting for approximately 85% to 90% of the lesions. Its usual presentation is with Mass abdomen, Jaundice and abdominal pain.

Keywords: Choledochal Cysts

INTRODUCTION

A 14 days old baby girl was admitted to us with a chief complaint of abdominal distension along with regurgitation of feed. Patient presented to us with complaints of vomiting and history of passing high colored urine since 4 days. She was passing stools in small quantities. On Clinical examination, Icterus was present till legs and abdomen was distended with sluggish bowel sounds and a mass was felt in the abdomen, which was soft in consistency around 7 cm in diameter. The birth weight of baby was 2.6 kg and abdominal girth 29 cm. No other abnormality was found in cardiovascular system, central nervous system or respiratory system. From the clinical examination the diagnosis of sub acute intestinal obstruction was made along with a cyst in the abdomen.

On investigating the patient; the haemoglobin was 16.7gm/dl, total leucocyte count was 16,000 and platelet count was 4.5 lacs. Bleeding time, clotting time and prothrombin time were normal. Renal function tests were normal. Serum Sodium was 131 mEq/l and serum potassium was 6.3 mEq/l. Liver function tests showed Total bilirubin to be 5.6 mg/dl, Direct bilirubin 2.2 mg/dl and indirect 3.4 mg/dl with alkaline phosphatase 294 U/L, SGOT 143 U/L and SGPT 67 U/L. Serial investigations of the patient revealed increase in Total bilirubin and indirect bilirubin to 12.4 mg/dl and 10.1 mg/dl respectively on 6th day of admission.

Ultrasound abdomen showed liver 5.4cm span with normal echo texture and intrahepatic biliary radicals were not dilated. Portal vein was normal and there was a presence of large abdominal cyst of size 8cmx8.3cmx6.9cm, which suggested being either a mesenteric cyst or enteric duplication cyst. On CT abdomen a large sharply marginated cystic lesion of size 7.6cmx6.4cmx7.9cm was seen occupying peritoneal cavity displacing the stomach and gut loops towards left. Superiorly the mass was extending upto the porta hepatis touching the liver and inferiorly going into the pelvis as shown in figure 1.The CT report suggested it to be omental cyst thus confusing the diagnosis further (Figure 2).

Another 20 days old male child presented to us with abdominal mass distension and regurgitation of feeds.

On examination the abdominal mass was around 3cm in diameter with icterus. The patient underwent routine tests which were normal except for the total bilirubin being 9.3 out of which direct was 0.7mg/dl initially and went out to be as high as 12 mg/dl and 1.0mg/dl respectively.

The ultrasound of the patient was done which described a mass in abdomen approx. 2cmx2cm with normal liver echotexture and IHBR not dilated with dilated gut loops. Then MRI of the patient was done and finding of fusiform dilatation of the CBD with maximum diameter 2.1 cm was seen. The Terminal part of CBD ended abruptly at the level of ampulla and proximally no dilatation of intrahepatic biliary radicals was seen and a diagnosis of choledochal cyst type 1a was made.

International Journal of Basic and Applied Medical Sciences ISSN: 2277-2103 (Online) An Open Access, Online International Journal Available at http://www.cibtech.org/jms.htm 2015 Vol. 5 (2) May-August, pp. 20-23/Aggarwal et al.

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Case 1 was taken up for surgery on day 28nd of life and intraoperatively was diagnosed to have choledochal cyst. Intraoperatively there were lot of adhesions present between the cyst and mesentery as well as between the cyst and small intestine. The liver surface was course and nodular. A Roux-En-Y-Hepaticojejunostomy was done in the patient.



Figure 1: CT Scan Showing Cyst

The operation went uneventful and the patient was shifted to Neonatal ICU for postoperative management.

Postoperative the patient's investigations were haemoglobin12.3gm/dl, total leukocyte count 8900/cumm. The Total bilirubin came down to 4.3 mg/dl and indirect 3.0 mg/dl. The abdominal distension was relieved and baby started taking oral feed on day 6 post operative. The bowel sounds returned to normal along with passage of normal colored stools.

The histopathological report of the resected tissue confirmed the diagnosis of choledochal cyst. The liver biopsy sent along showed presence of loss of architecture, presence of inflammatory cells consisting of neutrophils and few lymphocytes along with fibrosis. Liver cells showed presence of edema with periportal cholestasis. Nodule formation is also noted at occasional place. Appearances could be those of biliary atresia.



Figure 2: CT Scan Showing Cyst

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Figure 3: Intraoperative picture of resected choledochal cyst

The Case 2 was operated on day 24 of life and a Roux-en Y Hepaticojejunostomy was done. Intraoperatively cyst of 2.5cmx2cm was seen in relation to CBD with small gut loops and omentum firmly adherent to it. No evidence of any kink or band or adhesions of gut seen. The Patient was shifted to NICU postoperatively.

The patient recovered and his bilirubin levels dropped to normal and feed was started on 6^{th} day. The patient was discharged in a satisfactory condition with no complaints even on follow up.

Discussion

Choledochal cyst (a congenital abnormality of billary tree) is an aneurysmal dilatation of the bile duct involving extrahepatic, intrahepatic or both. It is a rare condition occurring more commonly in developing countries. Vater recorded the first description of a choledochal cystin 1723. Cysts were classified according to the Todani modification (Todani *et al.*, 1977) of the Alonso-Lej classification (Alonso-Lej *et al.*, 1959). Five types of choledochal cysts have been recognized in the intra and extra hepatic biliary tree (Evans, 1993). The most common type (type I) is a congenital cystic dilatation of the common bile duct without associated intrahepatic ductal dilatation. Type II is a diverticular malformation of the common bile duct. Type III is a choledochocele associated with an ampullary obstruction. Type IV malformation has multiple cysts of the intrahepatic and/or extra hepatic ducts. Type V has single or multiple intrahepatic cysts. Choledochal cysts in children have been predominantly type I cystic lesions. Type IV cysts is more common in adult patients (Chaudhary *et al.*, 1996). Our case was a type IA choledochal cyst with fusiform dilatation of the duct.

The etiology of choledochal cysts is controversial; however, the "common channel theory" is the most commonly accepted theory. This theory suggests that abnormal insertion of the bile duct into the pancreatic duct allows pancreatic enzymes to reflux into the bile duct. The pancreatic enzymes then cause inflammation, weakening, and eventual fibrosis of the bile duct. Distal obstruction then leads to progressive dilation of the biliary tree (Scudamore *et al.*, 1994).

The age of presentation is generally early, less than 6 months, but it has varied from being diagnosed antenatally to as late as 60yrs and above. It is associated with a triad of abdominal pain, jaundice and abdominal mass but only 10-20% patients present with these classical features. The disease is more common in females as compared to males. Generally regurgitation or vomiting is not a common symptom suggestive of choledochal cyst and is found in other causes of Intestinal obstruction. In case 1 there was compression of the gut and thus symptoms of regurgitation due to mass effect of the cyst whereas in case 2 gut loops were adherent to cyst and compressed by adhesions. The routine imaging modalities were also not helpful and choledochal cyst was not one of the suggested diagnoses. In both the cases it was the indirect fraction of bilirubin that was raised as compared to normally expected raise in direct bilirubin. This could be due to the age of presentation of both the patients and hence this caused a dilemma regarding the diagnosis as the features of obstructive jaundice were not present.

Thus through this article we want to create awareness that choledochal cysts can easily be missed out and should be kept as a differential diagnosis in a case of abdominal mass with obstruction in a neonate. We

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hope that this studywill prompt early recognition and attention to the potential complications of choledochal cyst in order to forestall morbidity and mortality.

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