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RESEARCH ARTICLE

SPONTANEOUS PERFORATION OF INFANTILE CHOLEDOCHAL CYST –A RARE PRESENTATION.

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Abstract

Spontaneous perforation is a rare complication of infantile choledochal cyst. We present a case of infantile choledochal cyst in a new born, which presented with abdominal distension, jaundice and acholic stools from 20 days of life. On evaluation was found to have focal dilatation of common bile duct and free fluid with septations. The infant underwent laparotomy and excision of the choledochal cyst followed by a Roux-en-y hepatico jejunostomy. It is important to follow up new born with choledochal cyst and intervene early with definitive management, for better outcome.

Introduction:-

Infantile choledochal cyst is a rare congenital abnormality of the biliary tract. They may present with unusual complications, besides their classical presentation with combinations of obstructive jaundice[1], abdominal pain and palpable mass reported in 7% of cases. Spontaneous rupture of choledochal cyst is anuncommon complication of infantile choledochal cysts, and may present with biliary peritonitis secondary to cyst perforationor rupture in 1 – 2 % of cases.

Case Report

A 28 days old girl, who was the second twin born late preterm by LSCS with a birth weight of 2.16 Kgs, presented with abdominal distension, jaundice and passage of acholic stools from 20 days of life. The child had an antenatal scan (Fig.1) done at 22 weeks with a cyst measuring 9.5 x 7.4 mm near the porta hepatis suggestive of Choledochal cyst. Post natal scan done on the 3rd day confirmed the findings of a fusiform cystic structure measuring 17 x 8.9 mm adjacent to portal vein suggestive of Type 1 Choledochal cyst, and the child was advised to follow up within a month.

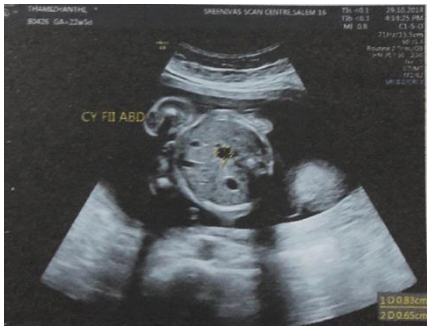

Fig 1:-Antenatal scan showing the cyst

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On physical examination the child appeared dehydrated and icteric. The abdomen was distended but soft, with bilateral reducible inguinal hernia and passage of acholic stools. On peritoneal tap there was golden yellow aspirate. The abdominal X-ray showed a diffuse haziness with few dilated bowel loops (Fig.2)

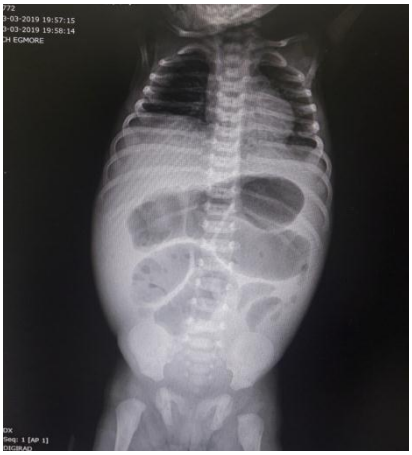

Fig 2:-X-ray chest and abdomen

Laboratory investigations revealed Hb – 12.7 gm%, TLC – 20,600/mm^3 (N62%, L26%), Total bilirubin – 9.8 mg/dL, Direct bilirubin – 5.4 mg/dL. Other biochemical and haematological investigations were within normal limits.An ultrasonography showed focal dilatation of CBD measuring 30mm x 15mm. There was free fluid with septations. No IHBRD, Liver echo-texture was normal. A provisional diagnosis of biliary peritonitis was made and management initiated.

At emergency laparotomy, there was bile filled peritoneal cavity especially staining the sub-hepatic space and the small bowel. The intestine was edematous but intact. There was a fusiform choledochal cyst with a 3x2 mm irregular edge perforation on its anterior wall (Fig.3) and two smaller perforations on the posterior wall. Since the child was haemodynamically stable a primary cyst excision with Roux-en-y hepatico-jejunostomy was done. The post-operative recovery was uneventful and the histological examination unremarkable.

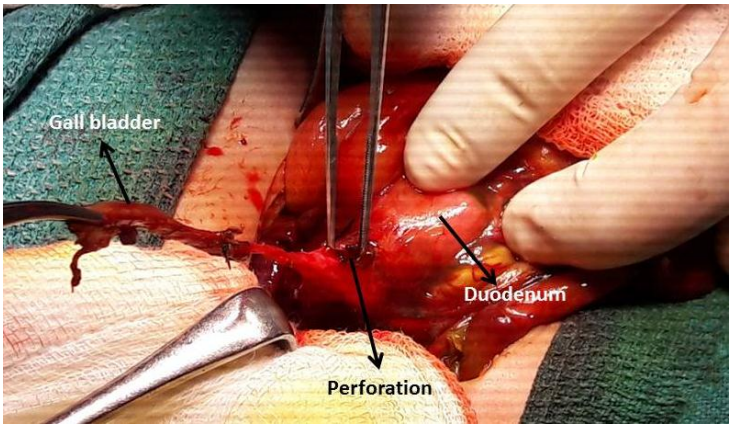

Fig 3:-Perforation of the choledochal cyst

Discussion:-

Spontaneous choledochal cyst perforation or rupture is a rare complication and can be a rare presentation. Patients with choledochal cysts can manifest clinical symptoms at any time during their life, with 80% of patients being symptomatic before the age of 10 years.

Abdominal pain, jaundice, and a palpable right upper quadrant abdominal mass is the classic triad described for patients with choledochal cyst[2]. The triad is only reported in about 20% of patients diagnosed. Two of the three symptoms are seen in two thirds of patients at the time of diagnosis. Some have applied a classification according to

age at presentation. The infantile form occurs before 12 months of age, and these patients tend to present with obstructive jaundice, acholic stools, and hepatomegaly similar to biliary atresia. The adult form occurs any time after 12 months of age and usually has a greater number of symptoms including fever, nausea, vomiting, and jaundice[4].
Perforation of the choledochal cyst is rare (1% to 12%) and thought to be due to a fragile cystic wall from inflammation, increased ductal pressure, or increased intra-abdominal pressure[3]. The site of rupture is at the low-flow region of the junction of the cystic and common bile ducts. These patients present with abdominal pain, sepsis, and peritonitis.

The etiology still remain obscure and the pathogenesis is a mix of congenital and acquired factors i.e. pancreaticobiliary malunion – pancreaticobiliary reflux and epithelial irritation, distal obstruction in the common pancreaticobiliary channel – anatomic or with inspissated protein plugs, mural immaturity and bile duct ischemia.
Ando K et al compiled the largest series of 13 cases of spontaneous rupture of choledochal cyst encountered in 30 years' experience of 187 cases[5]. All cases were below 4 yearsof age, the mean age was 22 months and the youngest was a month old. The case discussed is one of the youngest reported in English literature.

It is important to differentiate a spontaneous perforation of choledochal cyst from spontaneous perforation of the extra-hepatic bile duct[6] – a classic entity occurring exclusively in infants below 20 weeks of age, as the management is different.

Pre-operative imaging of the decompressed leaking cyst and proximal biliary tree in a sick infant is not necessary. The operative findings and subsequent imaging will clarify the pathology. Unlike a single, minute perforation seen in extra-hepatic bile duct perforations, spontaneous perforation of the choledochal cyst are grossly obvious of varying dimensions, single or multiple, usually on the anterior cyst wall and evenly sited along the extra-hepatic biliary tree.

Operation is the main stay of treatment. If the child is stable, a single stage cyst excision and Roux-en-y hepatico jejunostomy is done. If unstable, a temporary tube cholecystostomy or sub-hepatic drainage followed by cyst excision and biliary-enteric bypass later. The timing of operation as recommended iswithin 2 months, as this enables to differentiate from cystic type of biliary atresia. But many recommend it as early as possible, once child physiology is stabilised, because the grade of liver injury increases with age.

Conclusion:-

Infantile choledochal cyst needs regular follow up and monitoring to pick up complications like perforation, cholangitis and biliary peritonitis. They also need early definitive intervention to prevent secondary biliary injury and cirrhosis.

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