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Original Research Article

Spontaneous Perforation of Choledochal Cysts in Children, Presentation and Management, a Multi-Institutional Study

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Abstract:

Spontaneous perforation of the choledochal cyst is a disease entity in which the extrahepatic duct or the intrahepatic duct is perforated spontaneously without traumatic or iatrogenic injury [1]. The aim of the study was to analyse this rarity depending on clinical findings, diagnostic difficulty and optimum management plan in a limited resource set-up. A total of 15 patients presented with perforated choledochal cyst over a period of 7 years. There were 9 females and 6 males. All the children were infants. Children presenting with febrile illness having a documented choledochal cyst on ultrasonography with abdominal signs were evaluated for a perforation. Febrile illness with abdominal signs is the usual presentation. Jaundice is also very common. All patients were sick and in sepsis that required reasonable preoperative resuscitation. Malnourishment as suggested by anaemia and borderline hypoalbuminemia and on table finding of peritoneal contamination, unclear anatomy and oedematous bowel was seen in these children. An initial drainage procedure was done till the patient stabilised. We consider that spontaneous perforation of a choledochal cyst is not rare in infancy. We advocate an initial drainage procedure followed by a definitive surgery later once the patient stabilises.

Keywords: CDC(choledocal cyst), PBM (pancreatobiliary maljunction).

Introduction

Spontaneous perforation of the choledochal cyst is a disease entity in which the extrahepatic duct or the intrahepatic duct is perforated spontaneously without traumatic or iatrogenic injury [1]. The exact cause of Choledochal cyst perforation still remains uncertain; several mechanisms have been postulated to explain the spontaneous rupture of choledochal cyst.

The various theories proposed include congenital weakness of the bile duct, pancreaticobiliary anomalies including anomalous union of the pancreaticobiliary such as duct anomalous union of the pancreaticobiliary duct [2,3], pancreatitis, and distal obstruction of the bile duct because of inspissated bile, stones or stenosis, the presence of a diverticulum or abnormal gland of the bile duct wall, viral infection of the bile duct, tuberculosis, necrotizing enterocolitis, and birth trauma. Irritation of immature choledochal cyst wall from refluxed pancreatic juice [4] was considered as a cause for the perforation explained on the basis of the elevated serum amylase level in these patients.

Other authors consider obstruction of the distal CBD [5] along with congenital weakness of the cystic wall as the cause of spontaneous perforation of Choledochal cyst. Spontaneous perforation of the choledochal malformations is a rare complication in children. The diagnosis is often delayed because of its nonspecific presentation; hence, it is very rarely made preoperatively. In the present series, we focused on the diagnosis and management of a perforated choledochal cyst, in different pediatric centres of Union territory of Jammu and Kashmir. We consider that spontaneous

perforation of a choledochal cyst is not rare in infancy. The etiology of a perforation must be epithelial irritation of the biliary tract due to refluxed pancreatic juice caused by pancreatico-biliary malunion associated with mural immaturity due to infancy, rather than an abnormal rise in ductal pressure or congenital mural weakness at a certain point.[4]

Material and Methods:

All patients with spontaneous perforation of a choledochal cyst who were presented to three pediatric centres from January 2016 to December 2023 were included in the study. The aim of the study was to analyse this rarity depending on clinical findings, diagnostic difficulty and optimum management plan in a limited resource set-up. It was a retrospective observational study where patients presenting to the casualty department with biliary peritonitis between January 2016 and December 2023 were included. They were analysed with respect to symptomatology, laboratory parameters, radiology, emergency intervention with findings and definitive management plan. We analysed basic demographic characteristics, including sex, age, and body weight during the surgery, and clinical characteristics including signs and symptoms, laboratory markers, and radiologic findings at the initial visit. After screening the clinical features of perforated cases, we studied demographic characteristics; clinical findings, such as a presenting symptoms and signs; presence of fever and associated choledochalithiasis; start of diet; and hospital stay . Moreover, some laboratory results, including a white blood cell (WBC) count, C-reactive protein (CRP) level, and other parameters associated with biliary tract disease were analysed.

Results:

A total of 15 patients presented with perforated choledochal cyst over a period of 7 years. There were 9 females and 6 males. All the children were infants. The following were clinical symptoms of these patients.

Table 1:

Clinical Sign/Symptom	Patient Number
Jaundice	12
Abdominal pain	14
Abdominal distension	13
Vomiting	10
Fever	15
Shock	3

Table 2:

Laboratory parameters	Patient number
Anemia	8
Elevated Total leucocyte count	15
Elevated AST &ALT	15
Deranged coagulogram	3
Elevated serum amylase	10

Ultrasonography showed features of choledochal cyst perforation in all the 15 patients. Ultrasonography was highly specific as well as sensitive. Contrast enhanced CT scan was required in one patient. this patient had presented with shock.

MRCP was done and showed anamolous pancreatic maljunction in one patient. Serum amylase was raised in 10 patients.

Discussion

It is much commoner in Asia and highest incidence has been reported from Japan [6] Sex predilection for females with a female to male ratio of 3 to 4:1 is cited in all literature [7] and perforation is encountered in infantile period only.

The most accepted postulate was proposed in 1969 by Babbitt [8] He described proximal insertion of the pancreatic duct in to the common bile duct due to incomplete migration of the choledocho- pancreatic junction into the second part of duodenum. This PBM gives rise to a 'long common channel'. During embryonic development, PBM creates a low resistance pathway for reflux of pancreatic enzymes.

This, in association with an obstruction in distal insertion of the common channel causes cystic degenerative changes in the wall of CBD resulting in its fusiform dilatation. PBM, although proved to be the single most important aetiology, is difficult to delineate on radiology. Only one of our patients had a demonstrable PBM on MRCP.

Rupture of choledochal cyst should be distinguished from spontaneous bilary peritonitis in which usually there is a perforation at cystic duct and CBD junction, ascites is progressive and there

is no choledochal cyst in ultrasonography. This condition resolves with drainage only. The infantile form occurs in the first year of life. Perforation in a CDC is a very rare complication. The cognizance of this condition has mainly come from case reports. Our series is only second to the one reported by Ando et al in 1998 where they included 13 cases and documented its incidence to be around 7% [9] Fragile cyst wall from inflammation secondary to increased intraductal or intraabdominal pressure seems to be the instigating stimulus.

More incidences have been reported in abdominal trauma. Multiple subclinical episodes of cholangitis for longer duration causing degeneration of the ductal wall probably are the cause.

Children presenting with febrile illness having a documented choledochal cyst on ultrasonography with abdominal signs should be evaluated for a perforation. Febrile illness with abdominal signs is the usual presentation. Jaundice is also very common.

All patients were sick and in sepsis that required reasonable preoperative resuscitation. Malnourishment as suggested by anaemia and borderline hypoalbuminemia and on table finding of peritoneal contamination, unclear anatomy and oedematous bowel was seen in these children. An initial drainage procedure was done till the patient stabilized.

This appears to be a judicious and effective approach as indicated by resolution of dilatation of IHBR post operatively and largely asymptomatic course thereafter. Persistence of bulky pancreas may be idiopathic or indicative of prolonged but mild form of pancreatitis with irreversible morphological changes.

Management of a perforated CDC is challenging. Some advocate a laparotomy, lavage and external biliary drainage to tide over the crisis followed by definitive bilio-enteric reconstruction few weeks later.[9,10] Others recommend definitive surgery at the time of CDC perforation to avoid the morbidities of a second surgery and risk of failure of follow up on account of premalignant potential of the cyst.[19] We performed stage wise surgery in all children in view of patient profile, laboratory markers and intraoperative findings at the first operation. A drainage procedure was done initially. Since the perforation is focal, drainage stops after sometime and the child usually settles. In such cases we remove the drain, discharge the patient and plan a definitive surgery after proper optimisation. In cases were drainage persists drain is removed at the time of definitive surgery.

Conclusion

We consider that spontaneous perforation of a choledochal cyst is not rare in infancy. We advocate an initial drainage procedure followed by a definitive surgery later onese cce the patient stabilises.

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