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Case report

CASE OF CHOLEDOCHAL CYST PRESENTING AS PERFORATION ABDOMEN

Gobbur RH[1], *Baradol RV [2], Nyammannavar[3]

[1]Professor, [2]Asst Professor, [3]Asso Professor Dept. of Pediatrics, BLDEU's Shri B M Patil Medical College, Bijapur, Karnataka.

*Correspondence author email: ravikumar8901@gmail.com

ABSTRACT

Introduction: Choledochal cyst is an uncommon congenital anomaly of Hepoatobiliary system. It is a case of choledocal cyst presenting as perforation and biliary ascites which is rare in infants. **Clinical picture**: An infant was admitted at BLDE hospital with history of convulsion and breathlessness, on examination child had abdominal distension. Investigations like erect X ray abdomen done which revealed ascites with features of peritonitis, so exploratory laparotomy done which showed a choledochal cyst with perforation causing biliary ascites. **Treatment:** Child was treated in the pediatric intensive care unit for convulsion. Exploratory laparotomy done and the perforated choledochal cyst was sutured and drain placed in situ. The child improved from 2nd post operative day. **Conclusion:** Choledochal cyst can present as perforation, biliary ascites and peritonitis in infants. Therefore treatment should be oriented to this aspect also.

Keywords: Choledochal cyst, Bliary ascites

INTRODUCTION

Choledochal cysts are congenital bile duct anomalies. These cystic dilatations of the biliary tree can involve the extrahepatic biliary radicles, the intrahepatic biliary radicles or both. Cystic disorders of the bile ducts are rare entity but they are well defined malformations of pancreatic biliary systems involving intra hepatic bile ductile, extra hepatic or both. No strong unifying etiologic theory exists for choledochal cysts. The pathogenesis is probably multifactorial[1]. In many patients with choledochal cysts, an anomalous junction between the common bile duct and the pancreatic duct can be demonstrated. This abnormal union allows pancreatic secretions to reflux into the common bile duct, where the pancreatic proenzymes become activated, damaging and weakening the bile duct wall. Defects in epithelialization and recanalization of the developing bile ducts and congenital weakness of the ductal wall also have been implicated. The result is the formation of a choledochal cyst.

Since Vaten first described the entity in 1723, many cases have been reported worldwide. The incidence in the western population is 1:100000 to

1:150000 and in our population it is about 0.7% [2]. Choledochal cysts are three to four times more common in females than in males[3]. Their origin is unknown; however, they are considered congenital.

These anomalies can be classified using various systems. The most widely used subdivision of choledochal cysts are Todani's classification (Fig 1), which is a modification of the Alonso-Lej classification[4]

Generally the patient with choledochal cyst presents with cholestatic jaundice and abdominal pain. A palpable abdominal mass may be found in less than 20% of the cases. In adults, chronic and intermittent abdominal pain is the most common symptom. Children with choledochal cyst generally present with the triad of abdominal pain, abdominal mass and jaundice. Recurrent cholangitis and jaundice may also occur[5]. A choledochal cyst is rarely symptomatic, but should be considered if dilatation of the bile duct or the ampulla is demonstrated. Here we are presenting a case of congenital choledocal cyst presented with biliary ascites as a complication.

CASE

An infant, known case of seizure disorder was well controlled on anticonvulsants like phenytoin and valproate. This patient was brought into hospital with c/o fever, convulsions, cough, breathlessness and abdominal distension. On the 1st day, the patient was started on i.v. antibiotics like Ceftriaxone and Azithromycin, Inj Deriphylline was administered. Convulsions were well controlled on anticonvulsants. All routine blood and urine investigation along with chest X-ray were done. Haemogram showed neutriphilia with thrombocytosis. Haemoglobin was 12.8gm%. Bleeding time & clotting time were 2 & 4.45 min respectively. Serum sodium, potassium, chloride and calcium levels were 150meq/lit, 3meq/lit, 114 meq/lit and 9.5 mg/dl respectively. All reports were in normal range.

On 2nd day of admission there was progressive distension of the abdomen causing respiratory compromise. Emergency erects x-ray abdomen was taken which showed dilated bowel loops with features of peritonitis secondary to bowel perforation (Free air under the diaphragm). The child was taken for emergency explorative laparotomy. Intraoperatively choledochal cyst with perforation was found. Then the edges of perforation were sutured. 1 liter of bilious fluid was drained. Abdomen was closed with a drain in situ.

Baby required ventilator support for 2 days and was oxygen dependent for 7 days. Culture report was found to be sterile. The child was put on Piperacillin. As improvement was inadequate, Meropenem was started on the 4th postoperative day. The child showed improvement with a spontaneous eye opening on the 4th post operative day. Abdominal drain was removed on 5th Post operative day. RT is fed with milk was started on 6th POD. The child was afebrile, conscious and comfortable by 9th POD. RT feeds was gradually increased along with oral feeds.

DISCUSSION

This is the case report study of choledochal cyst presenting as abdominal perforation which is an uncommon complication and it is first reported by Weber in 1934.[11]

Choledochal cysts are rare abnormalities of the biliary tree. Choledochal cysts are generally classified using Todani's classification, which is as follows;

Classification by Todani and co workers[4] (as shown in figure no.1)

- I: Dilatation of hepatic and common bile duct (40% to 85%).
- II: Diverticulum of the common bile duct (2% to 3%).
- III: Intraduodenal common bile duct dilatation (1.4% to 5.6%).
- IV: Intra- and extrahepatic bile duct dilatation (18% to 20%).
- V: Intrathepatic bile duct dilatation (Carolis disease).

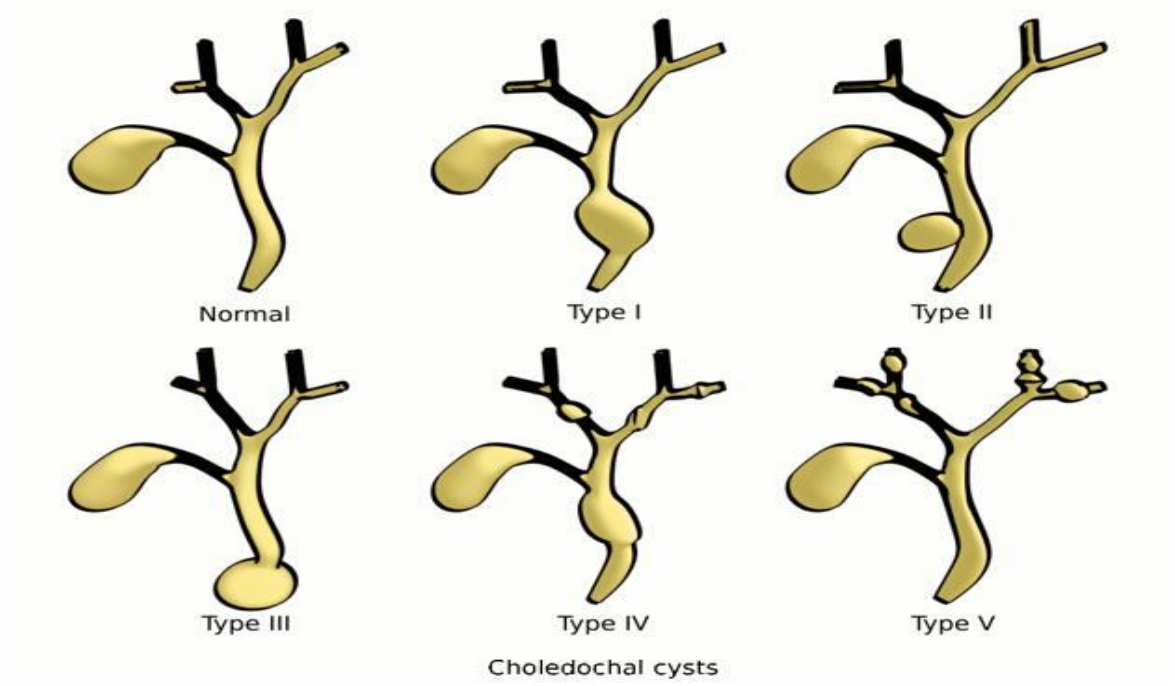

Fig: 1. Types of choledochal cyst (Classification by Todani and coworkers)

Patients with choledochal cyst commonly present with jaundice, abdominal pain and abdominal mass. Infants frequently present with jaundice and acholic stools. In early infancy, this may prompt a work-up for biliary atresia. In addition, infants with choledochal cysts often have a palpable mass in the right upper quadrant of the abdomen, accompanied with hepatomegaly. Children diagnosed after infancy typically have a clinical picture of intermittent biliary obstruction or recurrent bouts of pancreatitis[6].

The main diagnostic tool for detection of a choledochal cyst, especially in childhood, is ultrasonography. In adults, computer tomography can confirm the diagnosis; however, endoscopic retrograde cholangiography or magnetic resonance cholangiography is the most valuable diagnostic method and can accurately show cystic segments of the biliary tree[7].

Complications of choledochal cyst are mainly ascending cholangitis, intrahepatic bile duct stones, intrapancreatic terminal choledochal calculi, pancreatic duct calculus, bowel obstruction, cholangiocarcinoma, liver dysfunction and pancreatitis[8].

Surgery is the treatment of choice for a choledochal cyst. Complete excision of all cystic tissue is recommended because of the risk of recurrent cholangitis and the high risk of malignant degeneration[9]. Excision of the cyst and reconstruction of the biliary tree by choledochal/hepato-jejunostomy with a Roux-en Y-loop is the standard procedure. Another procedure includes excision of the distal common bile duct, excision of the common hepatic duct, dilatation of the intrahepatic bile duct, intraoperative endoscopy[10].

CONCLUSION

This case report study gives an idea of rare presentation of choledochal cyst as perforative abdomen with biliary ascites. So whenever a patient of abdominal perforation presents, choledochal cyst should keep in mind as a possible cause of perforation. The patient should be treated according to the cause of perforation.

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