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Sonographic Diagnosis of Choledochal Cyst in an Infant: A Case Report

[1] Dr. Md. Nazmul Islam, [2] Dr. Munshi Md. Arif Hosen, [3] Dr. Samira Sharmin, [4] Dr. Mohammad Simoon Salekin, [5] Dr. Afroza Naznin, [6] Dr. Farida Yasmin, [7] Dr. Lutfun Nahar

[1] Senior Medical Officer, Institute of Nuclear Medicine and Allied Sciences, Cumilla, Bangladesh
[2] MBBS, MD, Associate Professor, INMAS, Cumilla, Bangladesh
[3] MBBS, M.Phil, Principle Medical Officer, INMAS, Mitford, Bangladesh
[4] Senior Medical Officer, MBBS, INMAS, Satkhira, Bangladesh
[5] MBBS, M.Phil, Senior Medical Officer, INMAS, Mitford, Bangladesh
[6] MBBS, MD, Senior Medical Officer, INMAS, Mitford, Bangladesh
[7] MBBS, Medical Officer, 300 Bed Hospital, Khanpur, Narayanganj, Bangladesh

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Corresponding Author: **Dr. Md. Nazmul Islam**

Abstract

A choledochal cyst is traditionally considered as a cystic dilatation of the extrahepatic bile ducts. The exact etiology remains unknown; leading to postulation of multiple theories. Regional variations exist for describing the epidemiology of the disease condition. Most of the cases are diagnosed in early childhood. Differential diagnoses include hepatic cysts, duodenal atresia, gallbladder duplication and so on. Ultrasonography is the imaging method of choice. Regarding treatment, all cysts should be resected and bile flow should be restored. Studies suggest that a certain amount of these cysts turn into cancer; the prognosis of those who develop a malignancy is poor, with a 5-year survival rate of around 5%. Other complications could be ascending cholangitis, stricture and stone and sludge formation. To achieve the best results in the care and management of patients with choledochal cysts, it is essntial to form an interprofessional group of specialists including pediatric surgeons, pediatric gastroenterologists, pathologists, physiotherapists, nutritionists, oncologists and radiologists.

Keywords: Choledochal Cyst, Ultrasonography, Infant

Introduction

Choledochal cysts are congenital malformations of the biliary system and consist of cystic dilatation of the extrahepatic biliary ducts, intrahepatic biliary ducts or both. This condition most frequently affects the extrahepatic biliary ducts [1-3]. The classic symptoms in case of children are jaundice, abdominal pain and abdominal mass. Chronic and intermittent abdominal pain is the most common symptom in adults. Recurrent cholangitis and jaundice may also occur [1-3]. Initially, ultrasonography is the imaging modality of choice which is used to evaluate the choledochal cyst; whereas magnetic resonance cholangiopancreatography (MRCP) and endoscopic retrograde cholangiopancreatography (ERCP) are the most valuable diagnostic tools [4-6]. Complete excision of the cyst and reconstruction of the biliary tree by choledochal hepato-jejunostomy with a Roux-en-Y loop is the treatment of choice for both infants and children [1-2].

Case report

A female infant, aged 1 month, came to the outpatient department on the 08.08.24 and presented with jaundice, high colored urine and lack of appetite for the last 7 days. Physical examination revealed yellow coloration of skin and sclera. At this point, tt should be noted that, prenatal ultrasonography was found normal. Laboratory investigations done on 09.08.24 revealed, hemoglobin: 11.4 gm/dl; total count of WBC: 13000/cmm; total count of RBC: 3.94 million/cmm; hematocrit: 33%, platelet count: 378000/cmm, neutrophils: 68%; lymphocytes: 23%; serum total bilirubin: 18.17 mg/dl; direct bilirubin: 9.3mg/dl and indirect bilirubin: 8.87 mg/dl; C- reactive protein: 29 mg/L and blood group was B positive.

Abdominal ultrasonography performed on 10.08.24 illustrated: 1) Liver is normal in size with uniform parenchyma and normal echogenicity; 2) A cystic dilatation (5.8 x 5.0 mm) seen in CBD (Fig 1). Another cystic dilatation (6.2 x 5.0 mm) seen in left hepatic duct (Fig 2). Intrahepatic biliary ducts are just dilated at left side; these findings suggest choledochal cyst type- IVA. 3) GB is normal in size and contraction was seen after meal. Patient refused further investigation (MRCP) due to financial crisis.

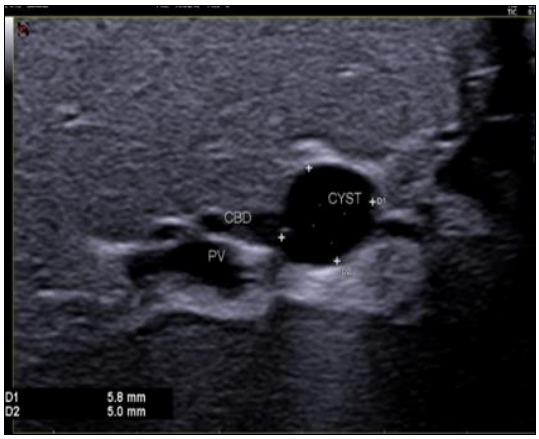

Fig 1: Cystic dilatation in CBD

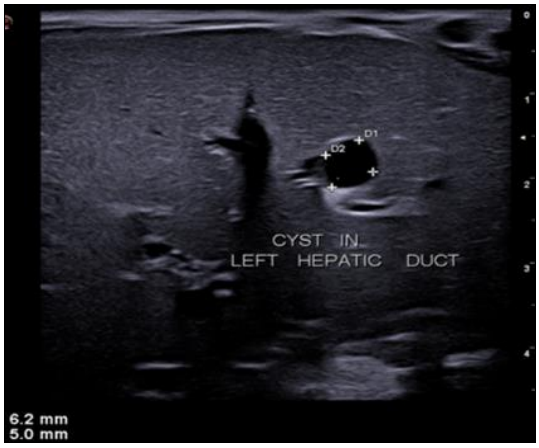

Fig 2: Cystic dilatation in left hepatic duct

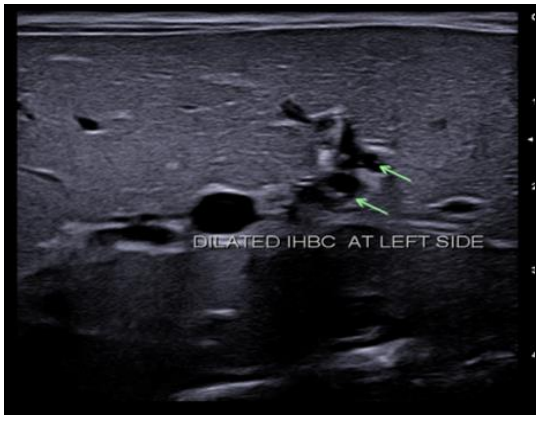

Fig 3: Just dilated left intrahepatic biliary ducts

Discussion

Choledochal cyst refers to a spectrum of congenital biliary tract abnormalities in which cystic dilatation of extrahepatic biliary tract with or without the dilatation of intrahepatic biliary tract. The incidence of choledochal cyst in western populations is 1 in 1,00,000-1,50,000 live births, which is higher in Asian populations ranging from 1 in 1,000-13,000 live births. Choledochal cysts are diagnosed in children and adults, although 80% cases are of infants and young children. In addition, choledochal cysts are two to four times more common in females than males. Alanso-Lej *et al.* classified choledochal cyst into three main types in 1959. Todani *et al.* modified this classification in 1977 into five main types with several subtypes [1-3, 7]. Type I choledochal cyst is the most common type; it is present in 80-90% of cases. This involves the dilatation of the entire common hepatic or common bile duct or of a few segments of both. Type I is further subclassified into IA, IB and IC [4, 7]. Type IA is the cystic dilatation of the common bile duct; type IB is the focal segmental dilatation of the common bile duct and type IC is the fusiform dilatation of both the common hepatic duct and common bile duct. Furthermore, type II is the true diverticulum in any segment of the common bile duct. Next, type III is the choledochocele, located within the duodenal wall at the pancreaticobiliary junction. Moreover, type IV choledochal cyst can be further subdivided into type IVA and IVB cysts depending on intrahepatic involvement; type IVA refers to extrahepatic biliary cystic dilatation with intrahepatic biliary cystic dilatation and type IVB refers to multiple extrahepatic biliary cysts without intrahepatic involvement. Lastly, type V is also known as Caroli's disease which refers to multiple cystic dilatations of intrahepatic biliary tree without evidence of extrahepatic cystic dilatation [1, 3, 4].

Symptoms are nonspecific and depend on the age at presentation. Majority of choledochal cysts are diagnosed in childhood and 80% cases are diagnosed within the first decade of life. The classical triad of jaundice, abdominal pain and upper abdominal mass is rare; however, jaundice is the main presenting symptom in children and abdominal pain in adults [3-5]. The complications of congenital choledochal cysts, namely are, obstetric jaundice, cholestasis with stone formation, recurrent cholangitis, malignant transformation, pancreatitis, portal hypertension and finally hepatic failure [4, 5]. Choledochal cyst can be diagnosed using multiple imaging techniques, for instance ultrasonography, computed tomography (CT) and magnetic resonance cholangiopancreatography (MRCP). These imaging techniques assess the extension of the cyst and associated complications or anomalies [4, 5]. Ultrasonography is widely used as a rapid, cost effective and reliable non-radiation technique. In our presenting case, ultrasonographic features included a well-defined cystic lesion in left hepatic duct and another well-defined cystic lesion in common bile duct.

Surgery is the treatment of choice for choledochal cyst. Currently most standard surgical procedure is total extension of the cyst and restore biliary enteric drainage into the duodenum via Roux-en-Y hepato-jejunostomy. Prognosis depends on early diagnosis and proper surgical treatment [4, 6].

Conclusion

Choledochal cysts are rare anomalies of the biliary tree and presentation in infant is very uncommon. The pattern of choledochal cyst presentation changes between childhood and adolescence. The diagnosis of choledochal cyst is based on patient's history, physical examination, laboratory findings and imaging modalities. Ultrasonography is a rapid,

cost effective, accurate and non-radiation method for early detection and is considered a very good diagnostic tool. Early diagnosis and proper surgical treatment results in better prognosis of the choledochal cyst.

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