Case Report

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Choledochal cyst presenting with neonatal intestinal obstruction: A case report

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KEYWORDS

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ABSTRACT

Background: Most patients with choledochal cysts are diagnosed during childhood. Therefore, the aim of this study is to report a case of early detected choledochal cyst in a neonate patient presenting with intestinal obstruction.

Case Presentation: A 5-day-old baby girl was brought to the hospital with a chief complaint of bilious vomiting. No history of jaundice was found. Ultrasound in the third trimester (37 weeks) showed an anechoic cyst with a smooth wall, no septum, with a size of 5.5x4.7x4.7 cm, suspected ovarian cyst. At 4 days old, MRCP showed dilated right and left Intrahepatic bile duct with a normal gallbladder, suspected as choledochal cyst Todani type 1. The patient was operated on at 7 days old and was discharged 15 days after surgery. Nine months after discharge, the patient was in good condition with no complaints of jaundice nor acholic stool and normal liver function test.

Conclusion: Prenatal diagnosis of choledochal cyst gave awareness to receive appropriate treatment. Bowel obstruction without jaundice could be an early symptom of a choledochal cyst in neonates.

INTRODUCTION

A choledochal cyst (CC) is a condition in which the biliary tract is dilated, causing an obstruction. CC can be found at any site within the biliary tree, yet the choledochal duct is the most frequent site. [1] The incidence of CC is more frequent in the Asian population than Western. [2] The exact pathogenesis of CC formation is unknown. However, there is a theory called Babbitt's theory. It explains how a choledochal cyst is formed. In some cases, a common duct can grow longer than usual causing the Oddi sphincter to fail to surround it completely. Thus, pancreatic secretion ought to backflow into the biliary tree. It potentially causes epithelial and mural layers injury due to the refluxing of proteolytic enzymes, leading to weakness, dilatation, and cyst formation. [1]

Most CCs are diagnosed during childhood. [2] However, some theories said that the development of CC starts from the fourth week of pregnancy when the ventral pancreas is shaped from the bilobed ventral pancreatic anlagen. [3] Its early presentation with signs of intestinal obstruction is unusual. Herein we present a case of choledochal cyst presenting in intestinal obstruction in a 5-day-old neonate.

CASE REPORT

A 5-day-old baby girl was brought with a complaint of bilious vomiting and abdominal distension without any history of yellowish discoloration of the skin. The patient was born full-term with a normal birth weight (2500 grams). An abdominal mass was diagnosed on prenatal scans. According to the physical examination, a solid palpable mass with clear margins, smooth surface, and venectasia was found in the right upper quadrant.

Prenatal ultrasonography (US) in the third trimester showed an anechoic cyst with a size of 5.5x4.7x4.7cm (Fig. 1A). Magnetic Resonance Imaging (MRI) done on the 4th day of life showed a cyst in the right upper quadrant with a clear and smooth border, with a size of 5.33x5.99x7.10cm, compressing the gallbladder, porta-hepatis, pancreas, stomach, duodenum, small intestine, and ascending colon (Fig. 1B). The Magnetic Resonance Cholangiopancreatography (MRCP) performed showed dilated right and left intrahepatic bile

ducts, suspecting a choledochal cyst, Todani type 1 (Fig. 1C).

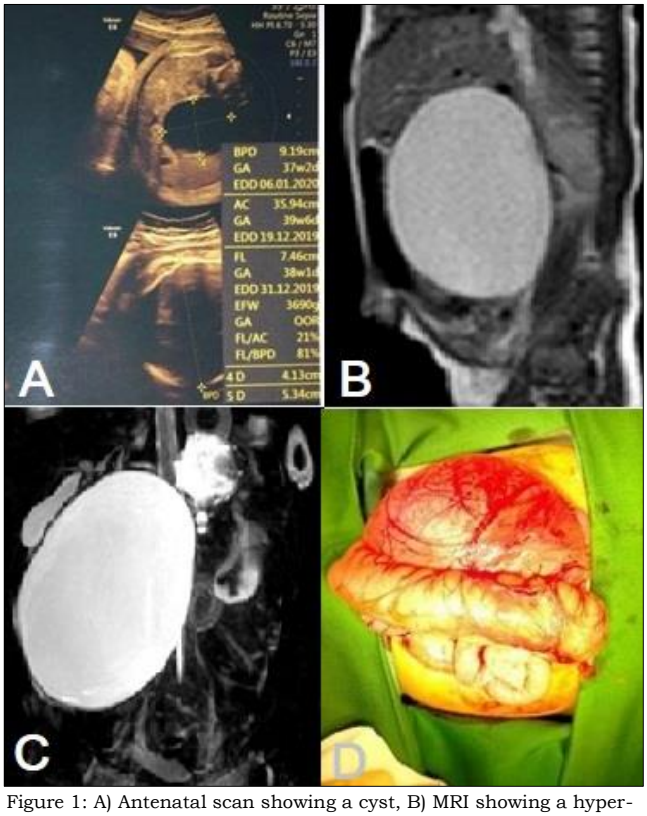

Figure 1: A) Antenatal scan showing a cyst, B) MRI showing a hyperintense cyst. C) MRCP showing CC type 1. D) Operative picture showing a huge cyst

Due to persistent vomiting a decision to operate the CC early was taken. An open cyst excision and Roux-en-Y hepaticojejunostomy were done on the 7th day of life. A CC with a size of 5x5x7cm was excised and a normal-sized gallbladder was found intraoperatively (Fig. 1D). The cyst was compressing the duodenum, which correlated with clinical symptoms. The patient was discharged 15 days after surgery. After nine months of observation at the outpatient clinic, the patient is doing fine.

DISCUSSION

Despite an increased detection of prenatal intra-abdominal cysts, prenatal diagnosis of choledochal cysts (CC) remains challenging. The differential diagnosis of CC such as hepatic cysts, duodenal duplication cysts, mesenteric or omental cysts, intestinal duplication, gallbladder duplication, and ovarian cysts must be ruled out. [4]

CC in our patient manifested with abdominal mass and bilious vomiting, which indicated compression of the cyst on the duodenum, while previous studies found many infants with CC were asymptomatic. [5] The presentation and certainly the treatment are usually delayed even in antenatally diagnosed cases. [1] In our case, the patient presented early owing to the compression effects of the CC on the duodenum thus warranted an early intervention.

Ultrasonography is one of the fundamental diagnostic modalities for CC as it shows the contour and location of the CC, the condition of the proximal ducts, vascular anatomy, and hepatic echotexture. [4] The prenatal US in this case showed a cyst without a specific location, suspected as an ovarian cyst. Previous studies stated that abdominal cysts can be diagnosed as early as the late first trimester of gestational age but more often in the second trimester. [3,6] The patient subsequently underwent postnatal MRI and MRCP which located the cyst more precisely and showed more characteristics of the cyst. This was expected as the accuracy of MRI and MRCP can be up to 96-100%. [3,7]

Bowel obstruction in cases with CC is more commonly found as a complication of surgery. It is likely caused by post-operative bowel adhesion or excessive length of the Roux loop. [8] Intestinal obstruction as a complication of CC itself is rarely reported. A previous literature review in 2020 found 17 cases of CC with bowel obstruction due to intestinal malrotation between 1982 and 2022 with ages ranging from 1 day to 15 years old. [9]. In this case, bowel obstruction was found as a clinical symptom due to the compression effects of the large-sized cyst.

In this patient, the surgery was done on the 7th day of life due to obstructive symptoms. Few studies recommended surgery within the first month of life to prevent the risk of progressive liver fibrosis. [2,10,11] A similar patient was reported by Agrawal et al in 2015; the patient was a 14-days old baby girl who came with complaints of abdominal distention and regurgitation of feed. The patient underwent an operation on the 28th day of life. Intraoperatively the cyst had adhesions with the mesentery and small intestine as a cause of obstructive symptoms. CC excision and Roux-En-Y- Hepaticojejunostomy were proven curative.[12]. We performed the surgery in the first week of life without any major challenges; this may alter the surgical approach in the future, especially in the infantile variety of CC that usually had a risk of the early development of hepatic fibrosis.

Even though with the excision of CC, the prognosis is good (5-year survival rate: 95.5%), long-term complications including malignancy can occur if the procedure is not done meticulously. [2,13,14] The patient was followed up for 9 months after discharge resulting in no sign of cholangitis, pancreatitis, bowel obstruction, and a need for reoperation, indicating that the excision and reconstruction went well. However, prolonged follow-up is required to look for long-term complications. [10,15,16]

To conclude, the presentation of a prenatally diagnosed case of choledochal cyst with compressive bowel symptoms is a rarity and warrants an early intervention that too was proven safe in the index case.

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