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Pediatric

Cystic biliary atresia: A distinct clinical entity that may mimic choledochal cyst

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ABSTRACT

Cystic biliary atresia (CBA) is a relatively uncommon but clinically significant variant of biliary atresia. The presence of a cyst in the hepatic hilum on imaging in an infant with cholestasis supports the diagnosis of CBA, but can also be seen in patients with a choledochal cyst—the main differential diagnosis in patients with CBA. The reported case outlines the clinical presentation and imaging findings in a patient with surgically confirmed and treated CBA and emphasizes the importance of distinguishing CBA from choledochal cyst at diagnostic imaging given the disparate timing and type of surgical treatment necessary for successful management of these distinct entities.

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Introduction

Cystic biliary atresia (CBA) is a relatively uncommon but notable variant of biliary atresia. It is defined as cystic change in an otherwise obliterated biliary tract [1] and accounts for approximately 5%-10% of biliary atresia cases [2]. The presence of a cyst within the hepatic hilum and cholestasis may also be seen in infants presenting with choledochal cyst, which is the main differential diagnosis when these patients present. Noninvasive imaging plays a vital role in the diagnosis of these biliary abnormalities and may help distinguish between CBA and choledochal cyst—a noteworthy distinction considering

the disparate treatments for these 2 conditions. This report describes a case of CBA and emphasizes the noninvasive imaging findings supportive of the diagnosis.

Case report

We present a 3-month-old baby referred to our institution with cholestasis and acholic stools. The patient was noted to have an elevated total bilirubin (18 mg/dL) while in the newborn nursery that down trended to 14 mg/dL with phototherapy

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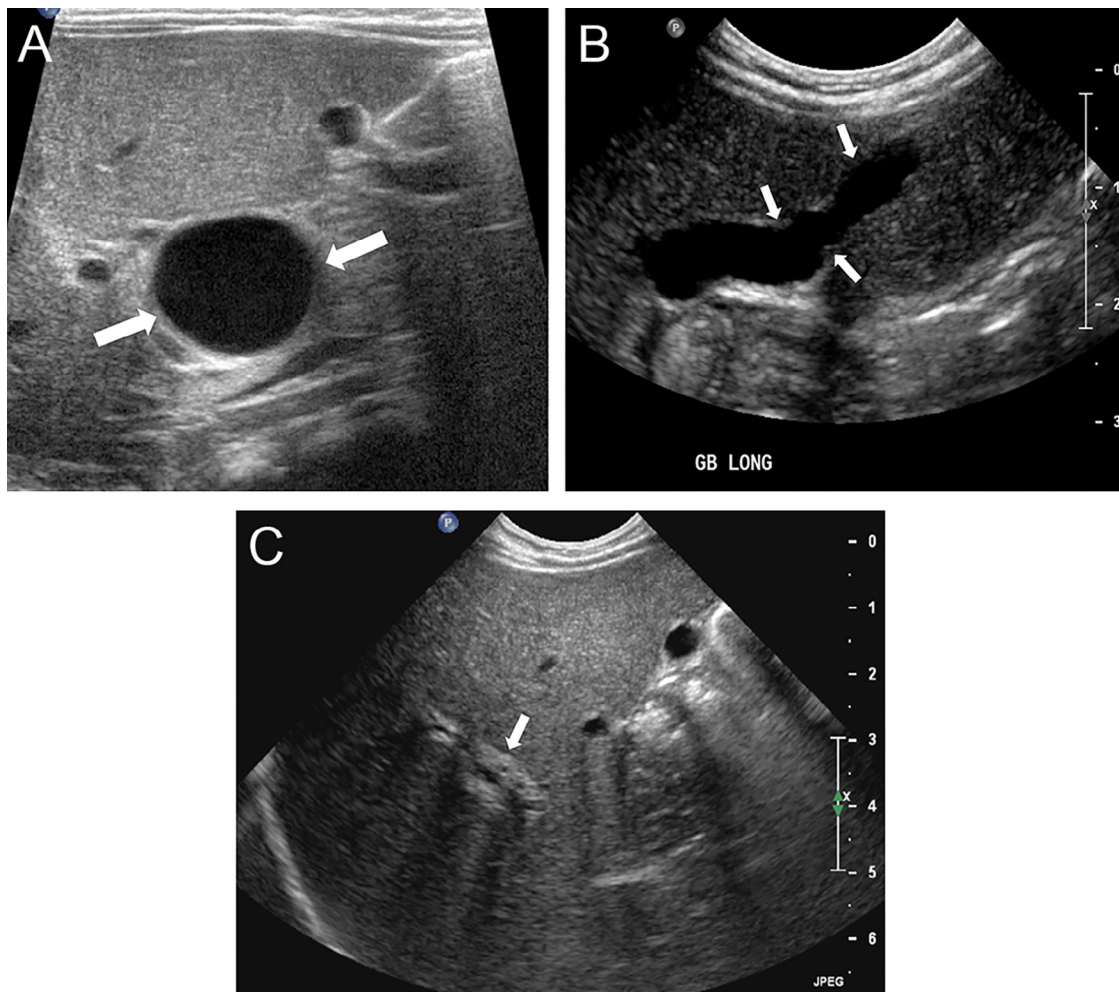


Fig. 1 – (A) Transverse grayscale sonographic image in the porta hepatis shows a round cyst (arrows) with anechoic internal fluid. (B) Longitudinal grayscale sonographic image demonstrates an elongated gallbladder measuring greater than 1.5 cm in length that has irregular walls and luminal narrowing (arrows). The distal aspect of the gallbladder was intrahepatic. (C) Transverse grayscale sonographic image of the right lobe of the liver reveals abnormal echogenic thickening (>4 mm) along the anterior wall of the right portal vein (arrow), the “triangular cord sign.” Note the absence of intrahepatic biliary duct dilation.

before discharge. At her 2-month well-child check, the patient remained jaundiced with an elevated total bilirubin of 7.2 mg/dL (normal < 2 mg/dL) and direct bilirubin of 3.4 mg/dL (normal < 1 mg/dL).

Ultrasound obtained at the outside institution revealed dilation of the extrahepatic common bile duct thought to be due to a choledochal cyst. Upon initiation of care at our institution, the patient had a repeat ultrasound performed that showed a cyst located within the hepatic hilum (Fig. 1A). A normal extrahepatic common bile duct could not be identified. None of the intrahepatic bile ducts were dilated, and no stones or sludge was present in the abnormally dilated segment of the common bile duct. The gallbladder was partially intrahepatic, elongated, and exhibited irregular walls (Fig. 1B). Close inspection of the hepatic hilum revealed abnormal and echogenic thickening along the right anterior wall of the right portal vein (Fig. 1C) consistent with a triangular cord sign. The constellation

of findings was most consistent with CBA, but choledochal cyst could not be entirely excluded.

Subsequently, a percutaneous cholangiogram was performed to definitively assess patency of the intra- and extrahepatic bile ducts. The gallbladder was percutaneously accessed, a flexible catheter was advanced into the cyst, and approximately 2 mL of light green serous fluid was aspirated. Anteroposterior fluoroscopic image (Fig. 2) obtained during hand injection of water-soluble iodinated contrast material demonstrated the distal segment of the irregular gallbladder and cystic duct communicating with an approximately 2-cm round cyst in the hepatic hilum. There was no communication with the intrahepatic bile ducts or with any additional extrahepatic bile ducts or bowel, confirming a diagnosis of CBA. The patient underwent Kasai portoenterostomy 48 hours after diagnosis (94 days of age), and total bilirubin is trending down on follow-up laboratory analysis.

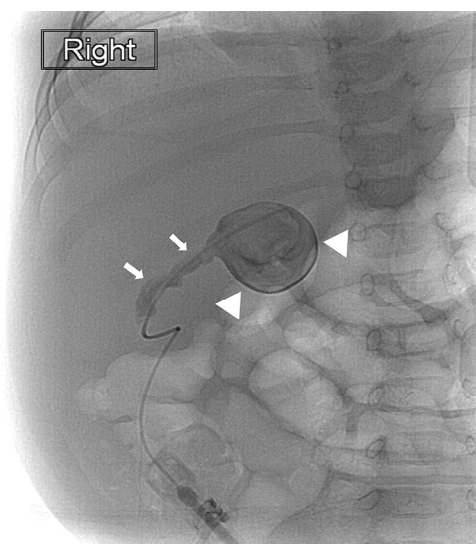


Fig. 2 – Anteroposterior fluoroscopic image of the right upper quadrant with catheter access to the porta hepatis cystic structure (arrow heads) through the abnormal distal gallbladder and cystic duct (arrows). Note the absence of filling of the intrahepatic and remaining extrahepatic biliary ducts, confirming the diagnosis of cystic biliary atresia.

Discussion

Biliary atresia is a progressive, fibro-obliterative disease of the intra- and extrahepatic bile ducts in infancy. Children usually present with biliary obstructive symptoms in the first 3 months of life. The overall incidence is low, but it occurs worldwide and affects an estimated 1 in 8,000-18,000 live births [3]. The etiology and pathogenesis of biliary atresia remains unknown but many theories have been proposed. Generally, biliary atresia is categorized into 2 forms—congenital and acquired. The congenital form is less common (20%) and linked to syndromic associations (ie, biliary atresia splenic malformation). The acquired form is more common and may be the end result of viral trigger and complex interactions between innate and adaptive immune responses [4]. Biliary atresia has been classified based on the most proximal level of biliary obstruction: type I (~5% of cases)—luminal patency as far as the common bile duct and proximal cystic biliary duct; type II (~2% of cases)—luminal patency to the level of the common hepatic duct; type III (~90% of cases)—the most proximal part of the extrahepatic biliary tract within the porta hepatis is entirely solid [5].

CBA is an uncommon but significant variant of biliary atresia that upon initial assessment both clinically and sonographically can have a very similar appearance to a choledochal cyst—the main differential diagnosis in these patients. However, distinguishing between these entities is of paramount importance as the type and timing of surgical intervention is quite disparate.

Prior studies have investigated ultrasound findings that can help distinguish between CBA and choledochal cyst. Kim et al. described findings in 28 patients, including intrahepatic bile

duct dilation observed in all patients with choledochal cyst and only 5% of patients with CBA, a normal sized (defined as gallbladder length of 1.5 cm or greater) or distended gallbladder in all patients with choledochal cyst in distinction to the atretic (defined as gallbladder length less than 1.5 cm) or irregularly elongated gallbladder (defined as gallbladder length 1.5 cm or greater with a thick wall and irregularly compromised lumen) in CBA, and an overall cyst size that was larger in choledochal cysts than that seen in CBA [6]. More recently, Zhou et al. described ultrasound findings in 23 patients, corroborating the intrahepatic bile duct and gallbladder findings by Kim et al. Additionally, Zhou et al. evaluated for the presence of the triangular cord sign (defined as thickness of the anterior echogenic wall of the right portal vein just proximal to the right portal vein bifurcation site of greater than 4 mm) which was identified in all but 1 CBA patient (11 of 12 patients) and none of the choledochal cyst patients (0 of 11 patients) [7]. The absence of gallstones and sludge within the cyst at the hepatic hilum supports the diagnosis of CBA over choledochal cyst [8].

Noninvasive imaging beyond ultrasound (ie, computed tomography and magnetic resonance imaging) has a relatively limited role in the diagnosis of biliary atresia and its variants. However, assessment with phenobarbital-enhanced hepatobiliary scintigraphy has proven useful in biliary atresia diagnosis in some studies [9], although the merit of this modality specifically in assessment and diagnosis of CBA has not been explored to our knowledge. Ultimately, the diagnosis of CBA can be confirmed by percutaneous or intraoperative cholangiography, revealing obliteration of the intrahepatic bile ducts and a lack of communication of the cystically dilated extrahepatic bile duct with the intestinal lumen.

Notably, response to surgery in the cystic variant of biliary atresia appears to correlate with age at surgery, just like other variants of biliary atresia; thus, earlier surgical intervention results in an improved outcome as judged by clearance of cholestasis [2]. Infants with CBA tend to be younger at presentation, and a delay in treatment with portoenterostomy beyond 70 days of age was associated with poor long-term survival with the native liver [10]. In distinction to the early intervention with Kasai portoenterostomy necessary for treatment of CBA, patients with choledochal cyst may undergo somewhat comparatively delayed surgical intervention to allow for patient growth, helping to optimize surgical outcome with cyst resection and hepaticojejunostomy.

In conclusion, the case presented demonstrates multiple findings of CBA and nicely summates supportive imaging features that can lead one to consider this important diagnosis. Considering the differences in treatment, it is incumbent on the radiologist to be aware of this relatively rare but important variant of biliary atresia. With accurate diagnostic evaluation, the patient with suggestive clinical presentation, hepatic hilar cyst, and supportive ultrasound findings can be appropriately guided to cholangiogram and, ultimately, timely and appropriate surgical management.

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