Radiology Case Reports 13 (2018) 415–418

Available online at www.sciencedirect.com

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Pediatric

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

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ARTICLE INFO

*Article history:*
Received 28 September 2017
Accepted 19 January 2018
Available online 9 February 2018

*Keywords:*
Pediatric
Cystic biliary atresia
Choledochal cyst

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.



## 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

Competing Interests: The authors have declared that no competing interests exist.

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https://doi.org/10.1016/j.radcr.2018.01.025

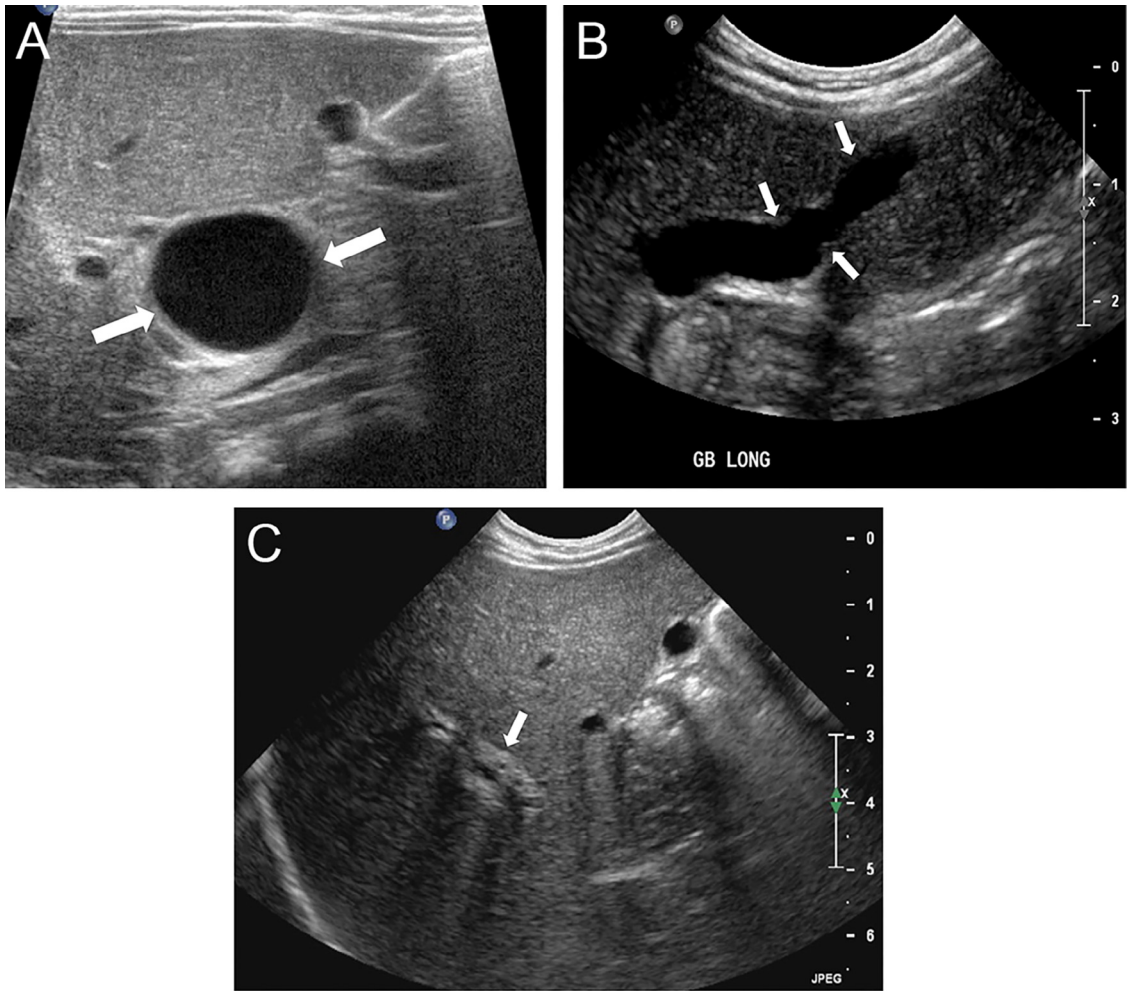


**Fig. 1 – (A) Transverse grayscale sonographic image in the porta hepatis shows a round cyst (arrows) with anechoic internal fluid. (B) Longitudinal grayscale sonographic image demonstrates an elongated gallbladder measuring greater than 1.5 cm in length that has irregular walls and luminal narrowing (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.