Extrahepatic biliary atresia with choledochal cyst: Prenatal MRI predicted and post natally confirmed: A case report

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Abstract

Extrahepatic biliary atresia (EHBA) is an uncommon cause of neonatal jaundice. Antenatal Magnetic Resonance Imaging (MRI) diagnosis of EHBA has not been published to the best of our knowledge till date. EHBA with cystic component is likely to be mistaken for choledochal cyst. A case that was antenatally predicted and postnatally confirmed by surgery and histopathology is being reported. All imaging signs are analyzed herewith. Imaging helps in the prediction of EHBA and also helps in early postnatal surgical referral which in turn improves the results of Kasai's portoenterostomy.

Key words: Choledochal cyst; extrahepatic biliary atresia; ghost triad; prominent hepatic artery; prenatal MRI; USG

Introduction

The diagnosis of extrahepatic biliary atresia (EHBA) was always delayed and done by exclusion, peroperative cholangiogram, and histopathology. Recent advances in imaging have led to increased antenatal prediction of EHBA, particularly on high-resolution maternal ultrasonography.[1]

Prenatal MR studies enhance fetal diagnosis,[2] with many reports of antenatal diagnosis of choledochal cysts (CDCs) on ultrasound (USG)[3] and Magnetic Resonance Imaging (MRI) studies[4] published. Amongst these, to the best of our knowledge, biliary atresia (BA) has not been reported.

We report a case of EHBA with terminal cystic dilatation of CBD predicted on antenatal and postnatal imaging with operative and histopathologic confirmation.

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Case Report

A 25-year-old primigravida presented to our hospital at 22nd week of gestation for target scan Targetted imaging for fetal anomalies (TIFFA). Fetal biometric parameters were normal. Liquor was adequate. Anatomical survey revealed a longitudinally oval, avascular anechoic cyst measuring 14 × 9 mm in the right upper abdomen, separate from duodenum, showing continuity with hepatic hilum superiorly without identifiable gall bladder (GB). No other abnormalities were detected. Presumptive diagnosis of CDC was made.

Fetal MRI was performed 2 weeks later on 1.5T Essenza (Siemens, Erlangen, Germany) to investigate the correct anatomical location of the cyst and its relationship with adjacent organs to assist with prognostication and treatment planning for the perinatal period.

T2-weighted Half-Fourier acquisition single-shot turbo spin-echo (HASTE, TR/TE 900/86 ms, 4-mm slice thickness) sequences were obtained in the axial, coronal, and sagittal planes with respect to the fetus. Sequences were monitored in real time and manipulated to determine optimal imaging planes during the scan, as fetal orientation is unpredictable and often changes during acquisitions. In addition to

the standard orthogonal planes, oblique planes were acquired for demonstrating the relationship to surrounding structures.

MRI showed a 14 × 9 mm ellipsoid [Figure 1A] homogenous cystic lesion at liver hilum hyperintense on T2W images. Axial images revealed the cyst posterior to the duodenum [Figure 1B]. The cyst was longitudinally oval, coursing posteroinferiorly from hepatic hilum appreciated on sagittal images [Figure 1C]. Intrahepatic ducts were not dilated; CBD was not visualized. Coronal images revealed hyperintense cord-like structure above the cyst, showing communication with it [Figure 1D]. The normal morphology of fetal GB was not visualized in the inferior surface of liver in right upper quadrant. The spleen was normal. Inferior vena cava (IVC) showed normal course. Liver showed normal signal intensity and contour.

The imaging characteristics on USG and MRI are presented in Table 1.

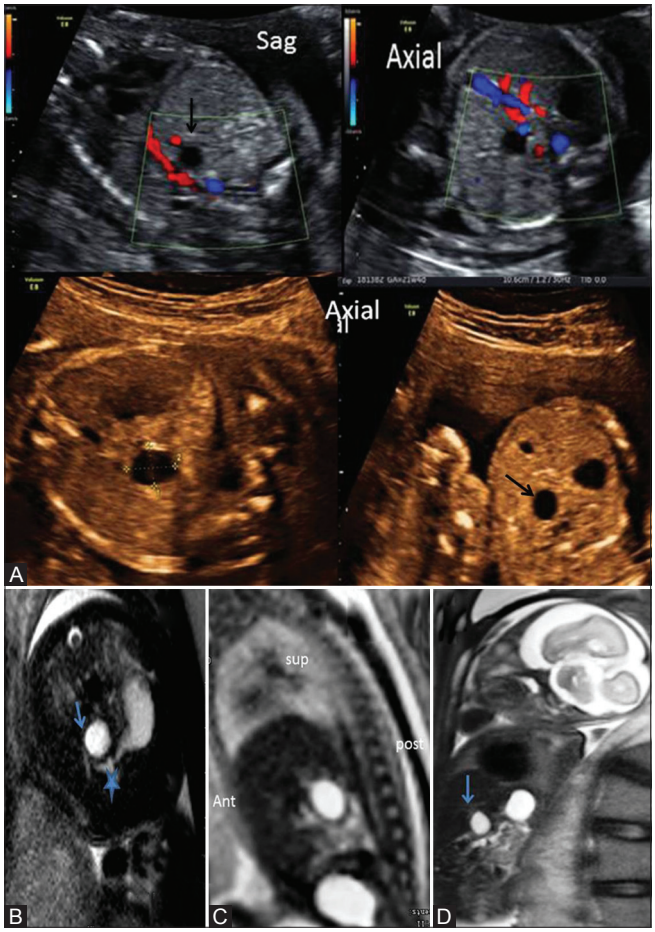

Figure 1 (A-D): (A) USG: Cystic lesion (14 × 9 mm) at porta hepatis (arrow) separate from duodenum without identifiable gall bladder. There was no intrahepatic ductal dilatation. (B-D) MRI: Axial images reveal the cyst (arrow) posterior to the duodenum (asterisk) (B). On sagittal images, the cyst is coursing posterior to the midabdomen (C). On coronal images, hilar structures were replaced by hyperintense cord-like structure above the cyst (arrow) (D)

Follow-up using serial USG scans in pregnancy revealed static size of the cyst with persistent ghost GB. Interval growth was normal till term.

A female infant weighing 2.9 kg was delivered by cesarean section at 39 weeks.

Postnatal USG, on day 3 after a 4-h fast, revealed liver extending beyond subcostal margin; echotexture was homogenous with normal surface contour. Intrahepatic ducts were not dilated. There was an echogenic area anterior to the portal vein at porta and measured 3.2 mm [Figure 2A]. A well-defined, longitudinally oval anechoic cyst was present at the hilum ~14 × 9 mm. The cyst size, echogenicity, and morphology were the same as seen in prenatal imaging [Figure 2B]. Maximum width and length of GB was 11 × 2 mm, with irregular, compressed lumen s/o ghost GB [Figure 2C]. Hepatic artery was prominent, running parallel to the right portal vein, and measured 1.7 mm. Main portal vein (MPV) diameter was obtained at the same level, which measured 3 mm. The ratio of HA/MPV diameter was >0.5 (>0.45 increase) [Figure 2D]. Hepatic subcapsular flow appeared subjectively increased. Spleen measured 3.8 cm. No varices were present. No free fluid was noted.

The imaging characteristics at porta are presented in Table 2.

Liver function tests (LFT) revealed increased γ-glutamyl transferase (831 U/L), elevated alkaline phosphatase

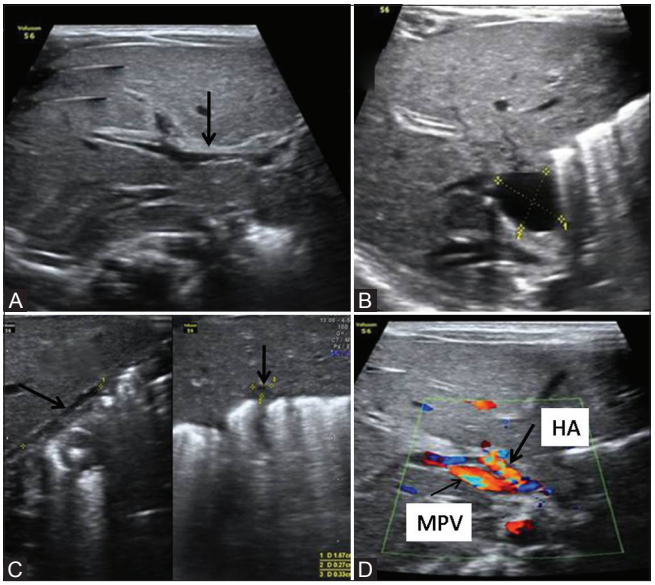

Figure 2 (A-D): (A) Echogenic area anterior to the portal vein at porta (arrow):triangular cord sign. (B) A well-defined, longitudinally oval anechoic cyst present at the hilum ~14 × 9 mm. No sludge in the cyst (C). (C) Maximum length and width of GB was 11 × 2 mm with irregular compressed lumen (arrow) (s/o ghost GB). (D) Prominent hepatic artery (arrow) seen running parallel to the right portal vein and measured 1.7 mm. The ratio of HA/MPV diameter was >0.5 (>0.45 increased)

(698 U/L), conjugated hyperbilirubinemia (2.8 mg/dL), and normal serum glutamate pyruvate transaminase (SGPT) 10 U/L, serum glutamate oxaloacetate transaminase (SGOT) 29 U/L, albumin, and International Normalised Ratio [(INR). Hepatobiliary scintigraphy was performed on day 24 which showed prompt and good uptake by the liver and absent tracer excretion into the intestines in 24-h delayed images. GB and extrahepatic ducts were not visualized [Figure 3].

A diagnosis of extrahepatic cystic biliary atresia (CBA) was made based on imaging.

The child was explored with on day 31. Operative findings included firm brownish liver, fibrotic porta inclusive of non-visualized right/left hepatic duct, common hepatic duct, and proximal CBD [Figure 4A]. GB was small and did not have a lumen. A cystic structure was seen at the site of terminal part of CBD. The cyst was going behind the duodenum and ending blindly. No communication was seen with the pancreatic duct. Corresponding schematic representation is shown in Figure 4B and was classified as type 3 BA according to Japanese classification system.

Excision of fibrotic hilar structures including cyst *en masse* with a Kasai's portoenterostomy was performed with ~30 cm of Roux-en-Y jejunal loop. Histopathologic examination confirmed the diagnosis of BA. Bilirubin returned to 1 mg by the 11th postoperative day. The child is aged 2 months now and doing well.

Discussion

Table 1: Prenatal imaging features

Feature	US	MRI
Cyst	14×9 mm, oval, anechoic, separate from duodenum	14×9 mm specific to biliary tree Coursing posterior and inferior to duodenum
Gall bladder	Not visualized	Not visualized
Hyperechoic/ intensity at porta	Not visualized	Present

MRI: Magnetic resonance imaging

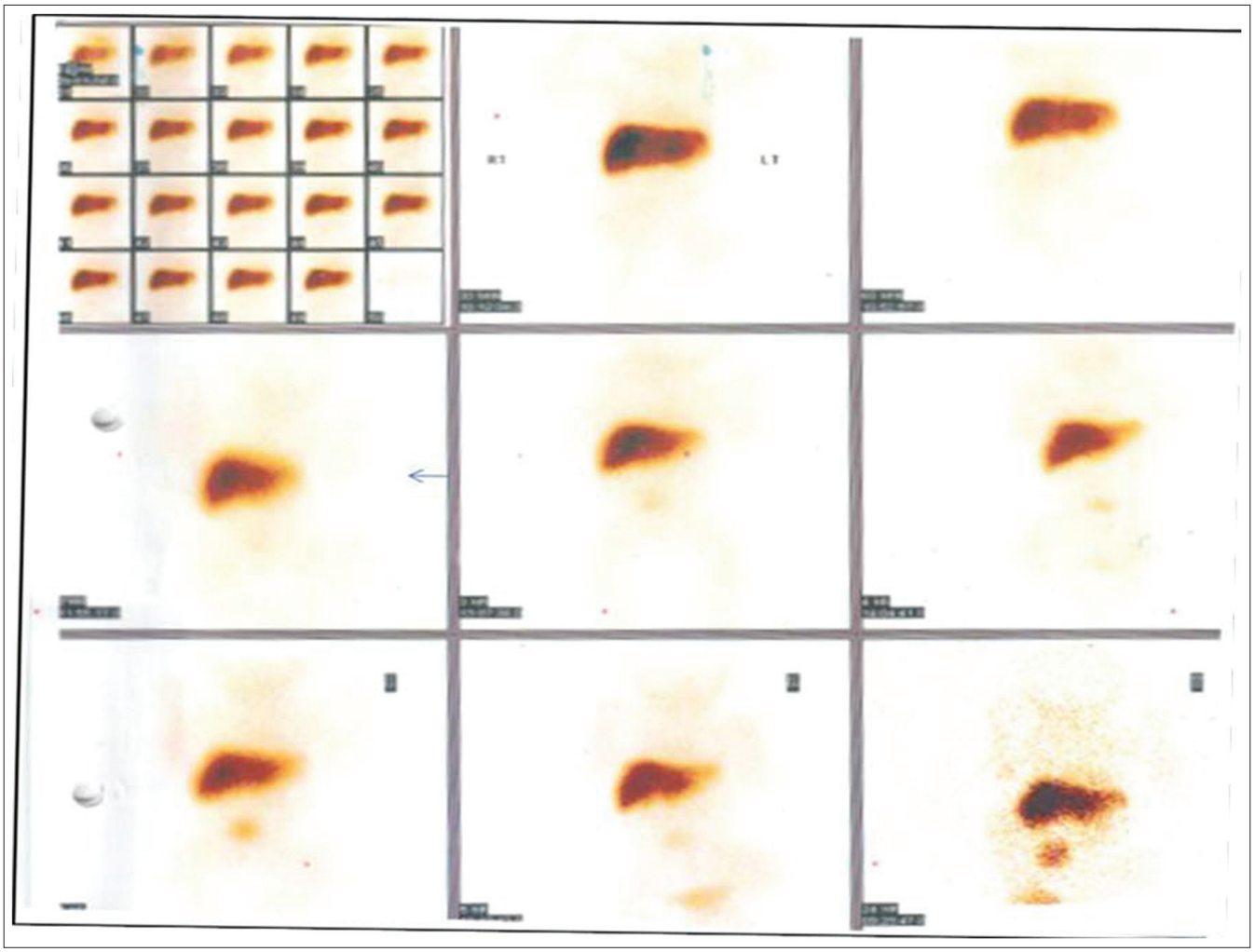

Figure 3: Hepatobiliary scintigraphy showed prompt and good uptake by the liver and absent tracer excretion into the intestines in 24-h delayed images. Gallbladder and extrahepatic ducts were not visualized

Table 2: Postnatal USG features

Finding	Description	Remarks
Cyst	Constant in size, echogenicity as compared to prenatal scans	Cyst associated with biliary atresia [Figure 2B]
Echogenicity anterior to porta	Present and measured 3.2 mm	Triangular cord sign [Figure 2A]
Gall bladder	Small, 11×2 mm (length×width), with irregular compressed lumen	Atretic gall bladder [Figure 2C]
Hepatic artery	Measured 1.7 mm	Prominent [Figure 2D]
Hepatic artery/MPV ratio	0.5	Increased [Figure 2D]
Increased subcapsular flow	Increased	Subjective

MPV: Main portal vein, USG: Ultrasound

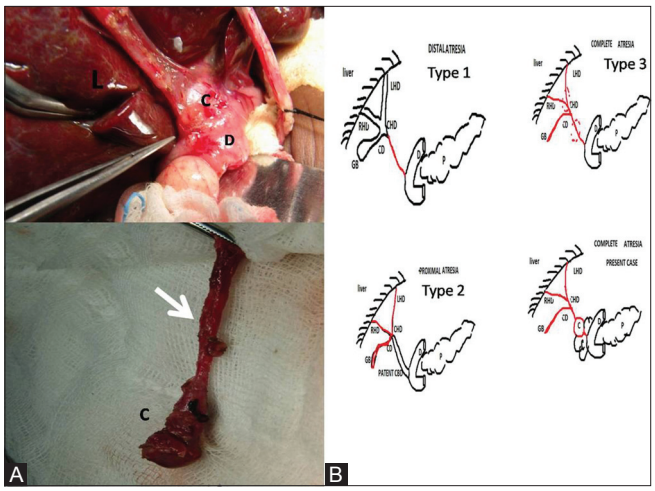

Figure 4 (A-B): (A) Operative findings included firm brownish liver (l), fibrotic porta inclusive of non-visualized right/left hepatic duct, common hepatic duct, and proximal CBD (arrow). Cystic structure was seen at the site of terminal part of CBD (C), going behind the duodenum (D), and ending blindly. (B) Corresponding schematic representation showing Japanese classification system with our case classified as type 3 atresia of entire ductal system (arrow) with terminal dilatation of CBD (C), ending blindly and being engulfed by pancreatic tissue (p)

CBA is an uncommon variant of BA in which prognosis may be relatively favorable but liable to misdiagnosis as CDC. It is classified into three types according to the Japanese classification system based on the atretic segment of bile duct into type 1 (distal atresia), type 2 (proximal atresia), and type 3 (complete atresia).[5] The cyst can occur anywhere along the atretic segment and is proximal to atresia in types 1 and 2 and distal to it in type 3.

A thorough literature search revealed less than 100 cases of EHBA associated with CDCs reported till date[5] of which majority were type 1-EHBA (76%) and only 21% were type 3-EHBA as in our case, classified according to Japanese pathological classification system.[5] The line diagram depicts our case.

No single preoperative investigation can diagnose EHBA with 100% accuracy.[6] The mainstay of confirmatory diagnosis in most of the published reports was surgical exploration and intraoperative cholangiogram.[6] Zhou *et al.* have demonstrated the accuracy of USG diagnosis to be 98-100% and evaluated objective differential characteristics of CBA and CDCs.[7] Triangular cord depicting fibrotic hilum in BA has been described on USG, MRI.[8] The collaborative signs include GB signs, hilar signs, hepatic artery signs, and miscellaneous signs (increased hepatic subcapsular flow) on USG.[7]

Ghost GB is an indirect sign of unfilled atretic GB in EHBA described by Zhou *et al.* as GB length <19 mm, lack of a smooth mucosal lining with an indistinct wall, and irregular or lobular contour.[7] Hilar cyst in BA represents fibrotic remnant of the CBD, explaining the constant size and echogenicity.[7] Prominent hepatic artery has been a well-known association with BA, making it a possible collaborative imaging sign.[7]

The present case showed triangular cord and all the collaborative signs described on postnatal USG as presented in Table 2.

Prenatal diagnosis ensures early surgery and better outcomes.[2,7]

On prenatal imaging, fetal GB presents as a right-sided pear-shaped structure on the inferior surface of liver and is consistently demonstrated from 18 weeks of gestational age onward. Before 27 weeks, the signal intensity of fetal bile is exclusively hyperintense on T2W, corresponding to T1W hypointensity. Non-visualization of fetal GB with cyst at porta is diagnostic of CBA on prenatal imaging.[9]

In our case, the normal shape of GB was not visualized in its expected location, thus confirming ghost GB. Thus, prenatal MRI can improve the pick-up rate of EHBA and also differentiate between CBA and CDC. To the best of our knowledge, this is the first report of prenatal MR predicted CBA. Non-visualization of GB and cyst specific to biliary tract on prenatal MRI are suggestive of BA.

Bhatnagar *et al.*[10,11] have reported CDC with BA, but the present case differs in that the BA existed both proximally and distally. Abnormal pancreas was evident in this case peroperatively where the terminal cystic structure was engulfed by pancreas.

Kasai's portoenterostomy has been the universally accepted initial surgical modality to establish biliary drainage.[12] The present case also underwent Kasai's portoenterostomy after the excision of fibrotic extrahepatic biliary ducts. Early surgery, as evident in this case that was operated <8 weeks of life, has yielded satisfactory initial biliary excretion leading to normal serum bilirubin

levels.

Imaging helps in the prediction of EHBA and also helps in early postnatal surgical referral, improving the results of Kasai's portoenterostomy.

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