Multimodality Imaging of Congenital Variants in the Gallbladder

Pictorial Essay

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Abstract: Congenital variants of the gallbladder development are infrequent and sometimes represent an asymptomatic imaging finding. In this case series, we want to present multimodality imaging findings that permit the diagnosis. Ultrasound, multidetector computed tomography, and magnetic resonance images are included.

The proper recognition of these infrequent congenital benign entities avoids unnecessary complementary examinations and misdiagnosis. Although ultrasound represents the most commonly used initial diagnostic tool, the increased use of magnetic resonance imaging and the wide use of multidetector computed tomography make it crucial to know how these entities are seen.

A retrospective review of cases of congenital variants of gallbladder through different imaging modalities was performed in our computer archives. Representative images were selected. Agenesis, hypoplasia, septated, duplicated, intrahepatic, and left-sided gallbladder are described.

Better understanding and recognition of congenital anomalies in gallbladder images will avoid misdiagnosis.

Key Words: congenital gallbladder anomalies, congenital gallbladder variant, ultrasound, magnetic resonance, congenital biliar disease

(*Ultrasound Quarterly* 2019;35: 195–199)

BACKGROUND AND PURPOSE

Variants of the gallbladder development are infrequent and sometimes represent an asymptomatic imaging finding. In this pictorial essay, we present multimodality imaging findings that allow for an accurate diagnosis. It is possible to study the gallbladder by means of different imaging techniques: ultrasound (US), multidetector computed tomography (CT), and magnetic resonance imaging (MRI). These congenital entities are detected initially by US, which is an excellent method for the initial study of the gallbladder.[1]

Received for publication July 26, 2018; accepted October 24, 2018.

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DOI: 10.1097/RUQ.0000000000000423

Every radiologist during his/her professional career can detect congenital gallbladder pathology in his/her practice. Proper recognition of these infrequent congenital benign entities avoids unnecessary complementary examinations and misdiagnosis such as neoplastic or inflammatory-infectious gallbladder pathology.

Although US represents the most commonly used initial diagnostic tool, the increased use of MRI and the wide use of CT make it crucial to know how these entities are seen.

In adults, the average length of the gallbladder is 7 to 10 cm; width, 2 to 3.5 cm; volume, 30 to 50 cc; and wall thickness, 2 to 3 mm.[2,3] The gallbladder is located in a fossa on the anterior lower surface of the liver in the plane between the left and right hepatic lobes. The gallbladder wall is composed of 4 layers: mucosa, lamina propia, muscularis propia, and serosa; the gallbladder has no muscularis mucosae or submucosa.[2]

There are different congenital variants of the gallbladder. They may be characterized by location, size, number, and configuration.[3,4] The gallbladder is normally scanned in the right upper abdominal quadrant, but it can be located in another position.[4] In abnormal location, the intrahepatic or left-sided gallbladder is uncommon. In anomalous development, agenesis, hypoplasia, and duplication of the gallbladder are rare. Multiseptate gallbladder is very uncommon.[3] The most frequent congenital anomalies are variations in the gallbladder location.[3]

MATERIALS AND METHODS

A retrospective review of cases of congenital anomalies of the gallbladder through multimodalities imaging was performed in our computer archives (US, CT, and MRI). Pediatric and adult cases were included. Representative images were selected to highlight key imaging features including agenesis, septate, and duplicated gallbladder imaging, and a review of the different congenital variants is discussed.

Agenesis of the Gallbladder

Agenesis is the absence of the gallbladder in patients without history of cholecystectomy. It is present in 1 of 6 of cases of biliary atresia. The isolated absence of the gallbladder and the cystic duct is rare. The estimated incidence described is 10 to 65 per 100,000, with female patients more frequently affected.[5] The pathogenesis is related to the embryogenesis of the

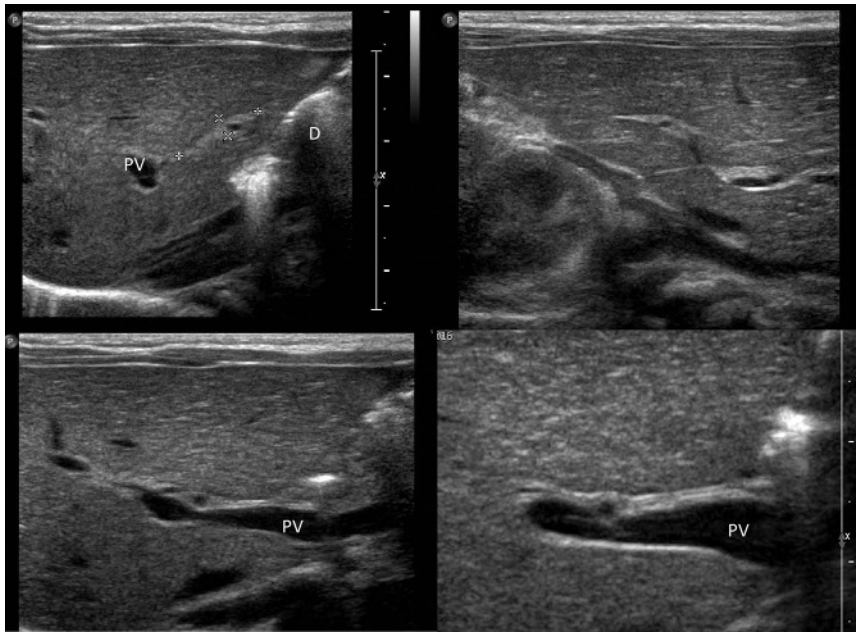

FIGURE 1. Gallbladder agenesis. A 10-day-old newborn child with neonatal hyperbilirubinemia. Abdominal US showed gallbladder agenesis with a normal intrahepatic and extrahepatic biliary tree. The US only depicted a hyperechoic pseudonodular image with a 2-mm cyst inside, without recognizing a structure resembling a gallbladder (dashed circle). D, duodenum; PV, portal vein.

gallbladder and the cystic duct, which do not sprout from the common bile duct.[5]

Patients present with symptoms (colic abdominal pain, dyspepsia) and may be asymptomatic or have association with other fetal anomalies such as horseshoe kidney, Klippel Feil syndrome, or bowel malrotation. There is a high incidence of choledocholithiasis.[6]

In gallbladder agenesis, using US or MRI, we observed the absence of the normal gallbladder in the fossa or in abnormal location (Fig. 1). Magnetic resonance cholangiopancreatography

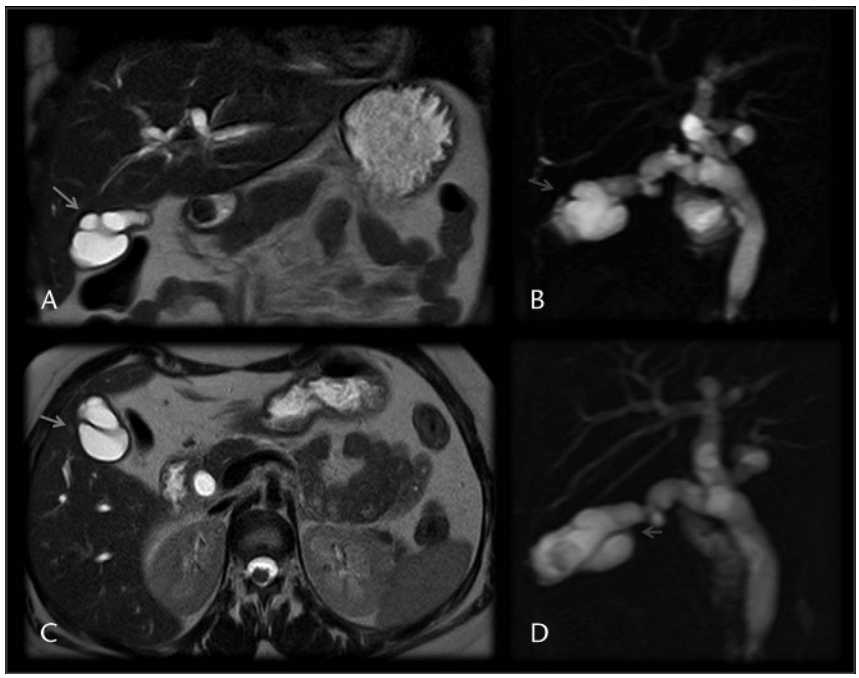

FIGURE 2. Magnetic resonance imaging cholangiogram depicting partial septate gallbladder. The sequences show multiple incomplete septa, which are connected to each other. Partial septa are best evaluated in cholangiography sequences B–D. A, T2 haste cor. C, T2 haste ax. B and D, Cholangiography 3-dimensional sequence.

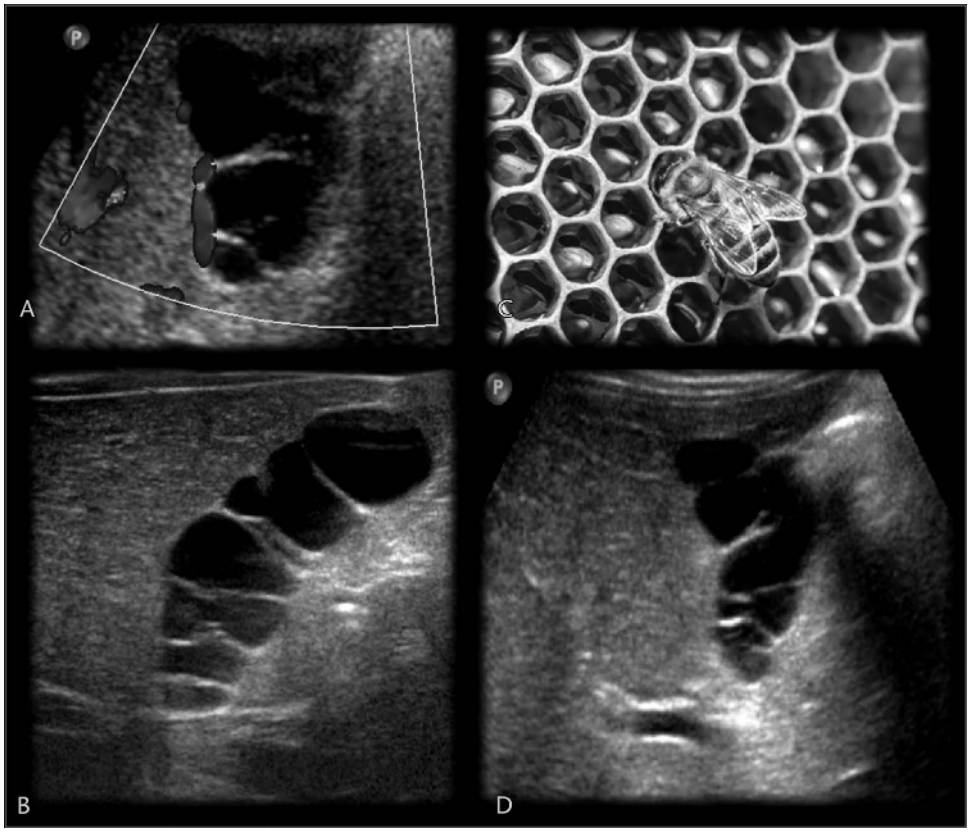

FIGURE 3. Abdominal US in a newborn child depicting a complete multiseptated gallbladder as an incidental finding, resembling a "honeycomb" pattern image.

is considered the best imaging modality in the study of gallbladder agenesis suspicion, especially for detecting other possible anomalies of the biliary tract.[5]

Agenesis of the gallbladder can be misinterpreted as cholecystitis with cystic duct obstruction or as a scleroatrophic gallbladder, therefore leading to unnecessary surgery.[3] A history of cholecystectomy or ectopic position of the gallbladder should be excluded before outlining this diagnosis. For symptomatic patients, conservative treatment and smooth muscular relaxant are proposed.[5]

Gallbladder Hypoplasia

There are very few reports of gallbladder hypoplasia in comparison to agenesis, which is more commonly reported.[7] In embryology, the hypoplastic gallbladder can be produced by an incomplete development of gallbladder bud or a defect of recanalization of solid primordium.[7]

Association between hypoplastic gallbladder with different conditions such as cystic fibrosis, cholangitis, neonatal hepatitis, and biliary atresia is described.[8] Around 33% of patients are symptomatic.[7]

In gallbladder hypoplasia, US shows a contracted, collapse, or small gallbladder.[7] Magnetic resonance cholangiopancreatography can show an ectopic gallbladder, anatomical variants, or biliar malformations and is also more accurate in the diagnosis of gallbladder hypoplasia.[9] In imaging, a postprandial contracted gallbladder must always be ruled out. In

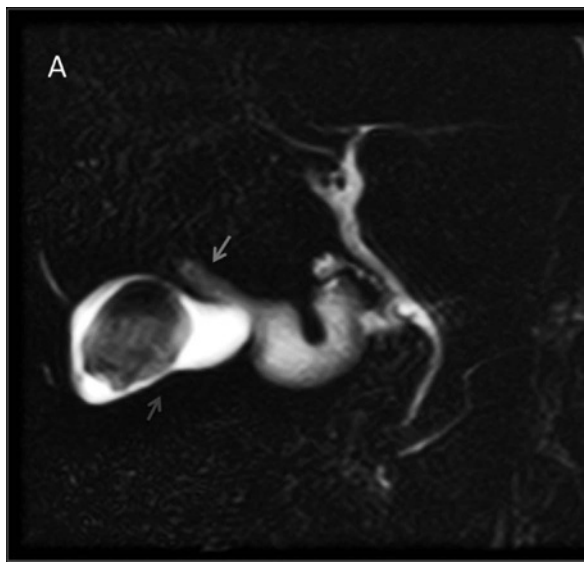

FIGURE 4. "Y" gallbladder configuration. Magnetic resonance imaging cholangiogram performed as a presurgical study to rule out choledocholithiasis in a middle-aged woman with US gallstone diagnosis. The US showed no subtle abnormalities other than gallstones. Three-dimensional MRI cholangiogram depicts 2 independent gallbladders (arrows) with a common infundibulum and cystic duct.

the differential diagnosis of rudimentary gallbladder, the choledochal cyst has to be considered.[10]

There are no specific treatment guidelines for patients with gallbladder hypoplasia.[7] Conservative management includes smooth muscle relaxants with the options of sphincterotomy as a second choice. Studies have shown that symptomatic patients were cured after laparoscopic cholecystectomy.[10]

Intrahepatic Gallbladder

It is a rare condition where the gallbladder is partially or completely embedded or incorporated within the hepatic parenchyma.[11] Ectopic position of the gallbladder is rare. The estimated incidence of this condition is 0.1% to 0.7% and includes intrahepatic, retrohepatic suprahepatic, supradiaphragmatic, within the falciform ligament or abdominal wall musculature, retroperitoneal, left-sided, and intrathoracic locations.[11]

In imaging, the intrahepatic gallbladder is located in anomalous position, different from the normal one, surrounded by the liver.

Intrahepatic gallbladder is usually associated with biliary stasis because of inefficient emptying leading to cholelithiasis, which is a presentation in more than half of patients.[12] This location makes the diagnosis of cholecystitis clinically difficult. It may present as a hepatic abscess secondary to a perforated gallbladder, cholecystitis, or choledocholithiasis.[5]

It is very important to have a preoperative diagnosis to avoid hepatobiliary injuries during cholecystectomy. Imaging including US can easily identify this condition, but MRI cholangiopancreatography is considered a gold standard imaging tool for all biliary tract anomalies.[11]

Left-Sided Gallbladder

Left-sided gallbladder is the anomaly where the gallbladder is located to the left of the ligamentum teres.[13] It is a rare anatomical variation with a prevalence of 0.1% to 1.2%.[13,14] It can be associated with situs inversus. There are 3 anatomical types of left-sided gallbladder: situs inversus, left-sided ectopic gallbladder, and right-sided ligamentum teres accompanied by failure in the right lobe development.[13]

Despite being located on the left side of the liver, it usually presents with right upper quadrant pain when being symptomatic.[14] It may also present with pain radiating to the right flank and a positive Murphy sign. Therefore, signs and symptoms could be misleading, and the diagnosis might be missed.[13]

In imaging, a true left-sided gallbladder is defined as a gallbladder located to the left side of the ligamentum teres.[13] These gallbladders are situated under the left lobe of the liver between segments III and IV or on segment III to the left of the falciform ligament.[15]

On US, they present as a cystic mass in front of the pancreas with a narrow neck that connects to the bile duct.[16] In CT and MRI, the gallbladder is not seen in the normal location, which is in the plane of the interlobar fissure of the liver. Magnetic resonance imaging cholangiography is very useful in assessing the connection of the gallbladder with the bile duct, especially in a preoperative setting.[17]

Left-sided gallbladders have been reported to be associated with portal vein anomalies, biliary system anomalies, and segment IV atrophy.[13]

Multiseptate Gallbladder

Septa of the gallbladder can be either partial or complete. It is a rare condition, with only small series or case reports in the literature, with one literature review from 2011 that described 44 cases, of which 30% were children.[18]

Multiseptated gallbladder is a rare anomaly, having a multichambered or multiloculate lumen with multiple septa, creating a "honeycombed" appearance,[4] probably due to incomplete vacuolization of developing gallbladder bud (Figs. 2, 3).

In US and MRI, the gallbladder cavity is divided by thin septa in different loculations.

Differential possibilities of the multiseptated appearance of the gallbladder on US include the following: multiseptated gallbladder, hyperplastic cholecystosis, cholecystitis, and possibly polypoid cholesterolosis.[19]

Duplicated Gallbladder

Gallbladder duplications are rare, with a described incidence of 1 in 3 to 4000 and a higher prevalence in men.[3]

In imaging, a duplicated gallbladder can present as bilobed, "Y" shaped, or "V" shaped. Bilobed gallbladders are 2 completely divided gallbladder cavities with a single common cystic duct. The external contour of the gallbladder can be normal, and the division between each cavity may be imperceptible. A prominent gallbladder fold or Phrygian cap may be a mimic.[20]

In the fully duplicated gallbladder, its external contour may or may not be normal; 2 separated gallbladders will have their own cystic duct and cystic artery.[20] Cystic ducts may have a common insertion point to the bile duct (Y-type, Fig. 4), or they can be separated with a distant insertion into the extrahepatic bile duct (V-type).

It may be easier to distinguish between 2 gallbladders if there is sludge, cholelithiasis, or vicariously excreted contrast, only in one of the gallbladders. The duplicated gallbladder may become inflamed with either independent involvement of 1 of the 2 lumens, or both lumens may be involved.[3]

In the differential diagnosis, we can consider a folded gallbladder, choledochal cyst, Phrygian cap, gallbladder diverticulum, focal adenomyomatosis, or fibrous or vascular bands.[3]

CONCLUSION

A better understanding and recognition of congenital anomalies in the gallbladder through multimodality images will avoid misdiagnosis and unnecessary complementary examinations. The radiologist should know how to recognize these entities.

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