Gallstones with Multiple Biliary Hamartomas: A Case Report

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AUTHORS' CONTRIBUTIONS

Lianhai Zhao performed the literature search and drafted the manuscript. Aiyan Qiu, Pingan Wang, Dong Xue critically revised, and gave final approval to the draft. All authors have read and agreed to the published version of the manuscript.

DISCLOSURES

The authors declare no conflict of interest.

CONSENT

Did the author obtain written informed consent from the patient for submission of this manuscript for publication? Yes. Images were obtained with patient permission accordingly to current National Privacy Regulations.

HUMAN AND ANIMAL RIGHTS

Not applicable.

ABSTRACT

Multiple biliary hamartomas (MBH), also known as Von Meyenburg complexes (VMC), are rare congenital disorders characterized by multiple cystic biliary dilatations without significant gender or age predilection. Due to limited reported cases and a lack of standardized guidelines, the management of these patients remains a clinical challenge. However, MBH may mimic other hepatic tumors and carry a potential risk of malignant transformation. Clinicians should recognize this rare entity, understand its clinical and imaging features, and differentiate it from other cystic hepatic lesions to avoid unnecessary overtreatment. This article retrospectively analyzes the clinical data of one case of MBH and reviews relevant literature.

CASE REPORT

BACKGROUD

Multiple biliary hamartomas (MBH) or Von Meyenburg syndrome (VMC), first described by von Meyenburg in 1918, are rare benign malformations composed of multiple well-defined duct-like structures lined by biliary epithelium and surrounded by fibrous stroma. These lesions arise from remodeling of the primitive ductal plate during embryogenesis and are typically small, non-communicating cysts resembling a "starry sky" pattern on imaging. Most cases are incidental findings during abdominal imaging or autopsy, with a reported prevalence of 0.69% (up to 3% in some autopsy series). Diagnosis relies on ultrasonography (US), computed tomography (CT), magnetic resonance imaging (MRI), and magnetic resonance cholangiopancreatography (MRCP), which reveal multiple irregular lesions approximately 10mm in diameter. We report a case of a patient with gallbladder stones and MBH who underwent laparoscopic cholecystectomy with intraoperative biopsy and pathological confirmation.

CASE REPORT

A 59-year-old male presented with right upper quadrant abdominal pain and fever for 3 days. CT revealed cholecystitis with gallstones. Contrast-enhanced MRI demonstrated a hepatic "starry sky" pattern. Physical examination showed right upper quadrant tenderness and a positive Murphy's sign. Laboratory tests revealed elevated total bilirubin (36 μmol/L; normal <28 μmol/L), high-sensitivity C-reactive protein (129.12 mg/L; normal <8 mg/L), and serum amyloid A (300 mg/L; normal <10 mg/L). Complete blood count, liver function tests, alkaline phosphatase, blood urea nitrogen, creatinine, electrolytes, and coagulation profiles were within normal ranges. Abdominal imaging showed multiple small, round, hypodense nodules on CT and non-enhancing hypointense nodules on contrast-enhanced MRI. Magnetic resonance cholangiopancreatography showed multiple small, T2-weighted, high-intensity cystic nodules in the liver without biliary duct communication, forming a "starry sky" appearance (Figure 1). Laparoscopic cholecystectomy

was performed, during which multiple dark-purple nodules of varying sizes on the liver surface were observed. The nodules had smooth surfaces. Among them, a biopsy of a 2×2 cm nodule from the left hepatic margin confirmed hamartomatous changes (Figure 2). Two-year follow-up MRI and MRCP showed no significant changes (Figure 3). We will continue with the subsequent follow-up.

DISCUSSION

Etiology and demographics

Multifocal biliary hamartomas, also known as Von Meyenburg complexes, were first described in 1918 and result from ductal plate malformation of the smallest intrahepatic bile ducts during embryonic development [1]. Clinically asymptomatic, these lesions are typically incidentally detected during abdominal surgery or autopsy, with reported prevalence rates ranging from 0.69% to as high as 3% in autopsy series [3].

Clinical and imaging findings

With advancements in imaging technology, MBH can now be diagnosed non-surgically. Radiographically, they typically manifest as multiple well-defined nodules measuring 1 mm to 1.5 cm, occasionally presenting as a solitary large mass [4]. On ultrasound, biliary hamartomas may appear hypoechoic or anechoic, rarely hyperechoic. CT demonstrates multiple rounded, small hypodense nodules without enhancement following contrast administration [5]. MRI reveals hypointensity on T1-weighted images and hyperintensity on T2-weighted images compared to hepatic parenchyma, though less intense than simple fluid. The hyperintensity of intraductal fluid is attenuated by hypointense fibrous tissue. Gadolinium-enhanced MRI shows slow, homogeneous hyperintensity on T1-weighted sequences. Unlike metastases, prolonged echo times on T2-weighted images further enhance lesion conspicuity. MRCP demonstrates normal intrahepatic ducts without direct communication with lesions, often described as a "starry sky" appearance [6,7]. Classic imaging findings strongly support the diagnosis of Von Meyenburg complexes; however, liver biopsy is indicated in cases of diagnostic uncertainty, particularly in patients with extrahepatic malignancies [8].

Classification

There is a classification of biliary hamartomas based on the degree of hamartoma consistency/biliary dilatation, Class 1, predominantly solid pattern with narrow bile channels; Class 2, intermediate pattern; and Class 3, marked cystic dilatation of bile ducts withinthe lesions[9].

Differential diagnoses

MBH, polycystic liver disease (PCLD), Caroli disease (CD), congenital hepatic fibrosis (CHF), and choledochal cysts(CCs) are collectively termed fibrocystic liver diseases [10]. These conditions should be considered in the differential diagnoses of biliary hamartomas. In contrast to PCLD and biliary hamartomas, the cystic lesions in Caroli disease demonstrate demonstrable communication with the biliary tree. The diagnosis of PCLD requires the presence of over 20 cystic lesions exceeding the size of hamartomas, with magnetic resonance cholangiopancreatography (MRCP) serving as the preferred imaging modality [3]. Key diagnostic criteria for choledochal cysts include segmental or fusiform dilatation of the bile ducts, or the presence of cysts contiguous with the biliary system, after exclusion of definitive mechanical obstruction [11]. The hallmark imaging features of congenital hepatic fibrosis predominantly involve hypertrophy of the left lateral hepatic segments and caudate lobe, atrophy of the right hepatic lobe, splenomegaly, intra- and extrahepatic biliary duct dilatation with coexisting cystic and solid lesions (regenerative nodules), periportal thickening, and associated hepatic/renal cysts [12].

Treatment, prognosis, and follow-up

Biliary hamartomas represent developmental anomalies rather than true neoplasms, warranting no specific therapy upon diagnosis. However, rare case reports document potential malignant transformation into cholangiocarcinoma, particularly with imaging evidence of lesion enlargement or density changes (solid to cystic/dilated) [13]. Given this risk, long-term follow-up via imaging (especially MRCP) is recommended. Notably, CA 19-9 should not be used as a diagnostic marker, as persistent elevations have been reported in non-malignant cases [14,15].

CONCLUSION

In summary, biliary hamartomas are rare benign biliary malformations with characteristic MRI features. Often incidentally detected during imaging for unrelated abdominal conditions, they may mimic hepatic metastases, microcysts, or other cystic lesions. Improved imaging modalities have significantly increased diagnostic accuracy. While inherently benign, their potential for malignant transformation into cholangiocarcinoma necessitates ongoing vigilance.

TEACHING POINT

MBH is a rare congenital disorder predisposing to malignant transformation. Recognition of its imaging features facilitates accurate diagnosis and treatment planning.

QUESTIONS

Question 1: Which imaging modality best demonstrates the "starry sky" pattern characteristic of MBH?

A. Ultrasonography (US)

B. Computed Tomography (CT)

C. Magnetic Resonance Imaging (MRI)

D. Magnetic Resonance Cholangiopancreatography (MRCP) (applies)

E. Positron Emission Tomography (PET)

Answer 1: Magnetic Resonance Cholangiopancreatography (MRCP). [The "starry sky" pattern arises from non-communicating hepatic cysts best visualized on MRCP.]

Question 2: What distinguishes MBH from polycystic liver disease (PCLD) on imaging?

A. MBH cysts are larger (>2 cm).

B. PCLD involves fewer than 10 cysts.

C. MBH cysts communicate with the biliary tree.

D. PCLD typically has >20 cysts. (applies)

E. MBH shows hyperintensity on T1-weighted MRI.

Answer 2: PCLD typically has >20 cysts. [MBH involves smaller, scattered cysts, whereas PCLD features numerous large cysts.]

Question 3: What histological feature defines biliary hamartomas?

A. Malignant transformation to hepatocellular carcinoma.

B. Dilated bile ducts lined by cuboidal epithelium with fibrous stroma. (applies)

C. Communication with intrahepatic bile ducts.

D. Absence of inflammation.

E. Cystic lesions filled with serous fluid.

Answer 3: Dilated bile ducts lined by cuboidal epithelium with fibrous stroma. [Pathological hallmarks of MBH include ductal plate malformations and fibrosis.]

Question 4: Why is long-term imaging surveillance recommended for MBH?

A. Risk of spontaneous hemorrhage.

B. Potential malignant transformation to cholangiocarcinoma. (applies)

C. High likelihood of cyst enlargement.

D. Risk of portal hypertension.

E. Association with pancreatic cysts.

Answer 4: Potential malignant transformation to cholangiocarcinoma. [Rare but documented cases of MBH progressing to cholangiocarcinoma necessitate follow-up.]

Question 5: Which clinical finding is most consistent with MBH?

A. Elevated CA19-9 levels.

B. Jaundice and pruritus.

C. Incidental discovery on imaging. (applies)

D. Abdominal pain and hepatomegaly.

E. Portal vein thrombosis.

Answer 5: Incidental discovery on imaging. [Most MBH cases are asymptomatic and detected incidentally during imaging.]

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FIGURES

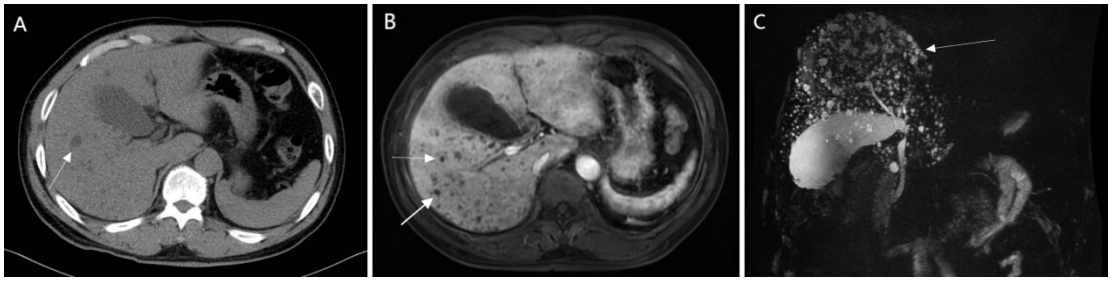

Figure 1: 59-year-old male with Multiple biliary hamartomas (MBH) in liver. (2022)
1A: CT reveals multiple round, small, low-density nodules.
1B: CE-MRI shows multiple small, low-density, non-enhancing nodules within the liver.
1C: Coronal MRCP image depicts the typical "starry sky" configuration of MBH in the liver, due to the presence of multiple, small, high signal cystic lesions scattered throughout hepatic parenchyma, not communicating with the non-dilated intra- and extrahepatic biliary tree

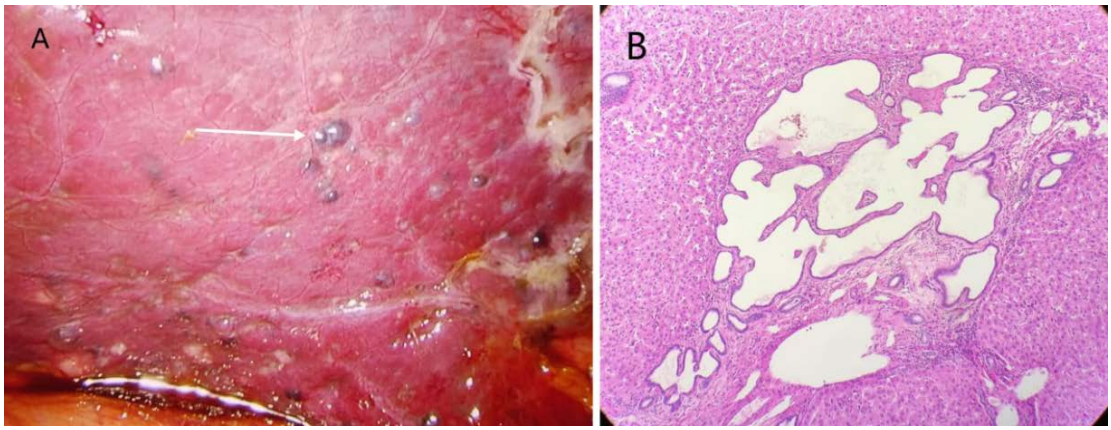

Figure 2: 59-year-old male with Multiple biliary hamartomas (MBH) in liver. (2022)
2A: Multiple dark purple nodules of varying sizes (ranging from millimeters to centimeters) are scattered on the liver surface, with smooth surfaces. Their dark purple color suggests possible blood stasis or tissue changes, distinguishing them from normal liver tissue.
2B: Pathological images show multiple irregularly dilated biliary duct-like structures lined by uniform cuboidal epithelial cells, embedded in a dense, grayish-white, tough fibrous stroma. These findings are consistent with biliary hamartoma (magnification, ×100).

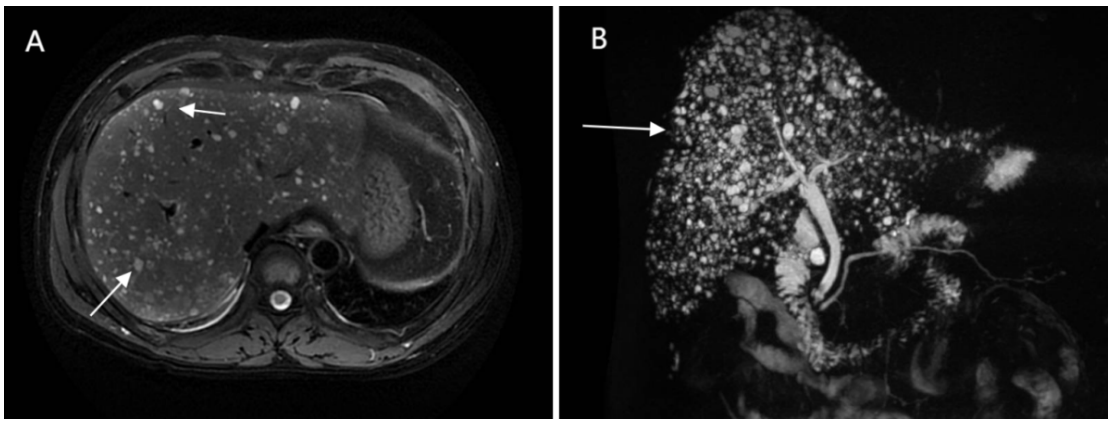

Figure 3: 59-year-old male with Multiple biliary hamartomas (MBH) in liver (2024). Postoperative two-year follow-up MRI (3A) and MRCP (3B) showed no significant changes compared to the previous studies two years ago.

KEYWORDS

Biliary hamartoma; fibrocystic liver diseases; Polycystic liver disease; Von Meyenburg syndrome; choledochal cysts