



SPOT THE DIAGNOSIS: IMAGE CHALLENGE

A Patient with Liver Lesions

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CLINICAL VIGNETTE

A 57-year-old woman presented to the emergency department with one week of cough and subjective fever. Review of systems was otherwise negative. A chest radiograph, followed by computed tomography (CT) of the chest to clarify a potential lesion, showed no pulmonary abnormalities. However, multiple liver lesions were incidentally identified (Figure 1). The patient had no abdominal pain or gastrointestinal symptoms; liver chemistry tests were normal.

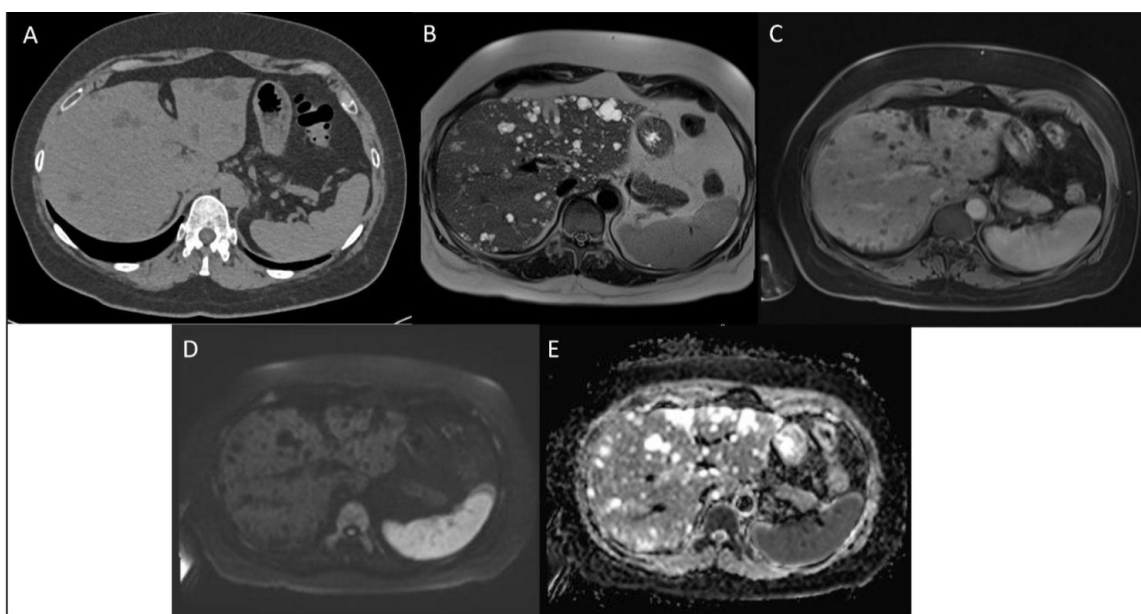


Figure 1. Axial imaging demonstrates numerous small, diffusely distributed hepatic lesions. **(A)** Non-contrast CT image demonstrating numerous hypoattenuating hepatic lesions. **(B)** T2-weighted magnetic resonance imaging (MRI) with diffuse, nodular and cystic, homogeneous (without internal septations or mural components) hyperintense hepatic lesions (up to 19 mm) without enhancement on subsequent **(C)** post-contrast T1-weighted MRI images, which also did not reveal clear communication with the primary biliary tree or biliary ductal dilatation. No restricted diffusion was observed when correlating **(D)** diffusion-weighted imaging (DWI) with **(E)** apparent diffusion coefficient (ADC) maps.

The imaging findings are most consistent with which of the following diagnoses?

- A. Biliary hamartomas
- B. Caroli disease
- C. Liver metastases
- D. Liver abscesses
- E. Liver cysts

ANSWER

A. Biliary hamartomas

DISCUSSION

Biliary duct hamartomas, also known as Von Meyenburg complexes (VMCs), are common yet rarely diagnosed benign malformations of the ductal plate, composed of dilated, disorganized small intrahepatic bile ducts within a fibrous stroma.¹⁻² They are found in 5.6% of adult autopsies, without sex or age predilection, and are strongly associated with fibropolycystic disease, such as autosomal dominant polycystic kidney disease (ADPKD).² Although 97% of livers with ADPKD have VMCs, ADPKD accounts for only 11% of patients with at least one VMC, suggesting most arise from a separate genetic, inflammatory, or ischemic process.²

VMCs are typically smaller than 1.5 cm, though lesions may vary in size within the same liver, and are usually incidental imaging findings without liver biochemistry abnormalities; symptomatic presentation is rare.³⁻⁵ Because they most often present as multiple hepatic lesions, they may be mistaken for metastatic disease; concurrent non-malignant CA 19-9 elevation can compound this, limiting its discriminatory value.^{1,5} Other mimics include hepatic abscesses, simple or polycystic liver cysts, peribiliary cysts, and Caroli disease.^{1,3} On MRI, VMCs are characterized by innumerable small lesions distributed throughout both lobes with T2-weighted hyperintensity, termed the "starry sky" pattern (Figure 1B).

The lesions appear hypointense on T1-weighted images and show no central enhancement following gadolinium administration, although a thin peripheral rim or an enhancing mural nodule is common and should not be mistaken for a metastatic deposit (Figure 1C).^{1,4} Absence of communication with the rest of the biliary tree distinguishes VMCs from Caroli disease and is best demonstrated on magnetic resonance cholangiopancreatography (MRCP).³⁻⁴

Why Not the Other Choices?

- **Caroli disease (B):** The lesions lack direct communication with the primary biliary tree, a key feature of Caroli disease.
- **Liver metastases (C):** The lesions demonstrate a degree of T2 hyperintensity greater than is typically seen with hepatic metastases. In addition, the absence of restricted diffusion and contrast enhancement makes metastatic disease less likely.
- **Liver abscesses (D):** Liver abscesses typically demonstrate restricted diffusion, peripheral rim enhancement, and surrounding inflammatory change. Patients commonly also have right upper quadrant pain and systemic signs of infection, which were absent in this case.
- **Liver cysts (E):** Simple hepatic cysts may also appear T2-hyperintense and non-enhancing. However, they are typically fewer in number, larger, and more variable in size. The presence of numerous small, cystic lesions diffusely distributed throughout the hepatic parenchyma favors biliary hamartomas.

Owing to variation in the relative proportion of ducts to fibrous stroma, VMCs are not invariably cystic and may appear solid or mixed solid–cystic.^{1,5} They are benign lesions requiring no specific surveillance; while histopathologic series have described transition to small-duct-type intrahepatic cholangiocarcinoma, a true precursor role remains unproven.^{1,3}

Recognition of these characteristic imaging findings can help establish the diagnosis and also avoid unnecessary invasive pursuits, with biopsy reserved for indeterminate cases.^{1,3} In this asymptomatic patient, the diagnosis rested on characteristic imaging findings reported by the radiologist. The largest lesion measured 19 mm, above the size typically described, but the constellation of MRI findings, together with the absence of interval change on serial imaging, supported the final diagnosis.⁴⁻⁵ Histopathologic confirmation was not obtained; the patient remains well on clinical follow-up.

CONFLICT OF INTEREST STATEMENT

The author declares no conflict of interest.

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ETHICS STATEMENT

The authors obtained verbal informed consent, witnessed and documented in accordance with institutional guidelines, for publication of medical information and images.

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