BILIARY HAMARTOMATOSIS: THE DIAGNOSTIC CHALLENGE BEHIND AN INCIDENTAL FINDING.

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Abstract

Multiple biliary hamartomas, or Von Meyenburg complexes, are uncommon benign malformations of the intrahepatic bile ducts, typically asymptomatic and identified incidentally. Their principal diagnostic challenge is differentiation from small multifocal hepatic metastases.

We report the case of a 62-year-old woman presenting with cholestatic-pattern elevation of liver enzymes, pruritus, and chronic liver disease with significant fibrosis. Cross-sectional imaging demonstrated several subcentimeter hepatic lesions with arterial-phase hyperenhancement, accompanied by persistently elevated CA 19-9 levels. Histological examination of a liver biopsy established the diagnosis of multiple biliary hamartomatosis.

Although CA 19-9 may be chronically elevated in the absence of malignancy, isolated cases of progression to intrahepatic cholangiocarcinoma have been documented in patients with underlying chronic liver disease, thereby supporting individualized surveillance strategies. Overall, biliary hamartomatosis is associated with a favorable clinical course.

Keywords: biliary hamartomas, hepatic metastases, chronic liver disease.

Introduction

Multiple biliary hamartomas, also known as Von Meyenburg complex, are rare benign malformations that originate from the embryonic ductal plate.[1]. They are usually diagnosed incidentally on imaging studies, as most patients remain asymptomatic[2].

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In some cases, they may cause elevated transaminases, pruritus, or be associated with chronic liver disease, and may lead to portal hypertension. Additionally, an association with hepatic and renal polycystic disease has been described[2].

The main differential diagnosis is multiple small liver metastases. The prognosis, however, is usually favorable.[2,3].

Clinical case

A 62-year-old woman with a personal history of hypothyroidism and hypercholesterolemia, occasional alcohol consumption, and under follow-up since 2020 for hypertransaminasemia with a cholestatic profile and pruritus. Chronic liver disease with significant fibrosis was documented (Fibroscan: 18.8 kPa). During the etiological workup, multiple subcentimeter-sized hepatic FLL were identified that were hyper-enhancing in the arterial phase and exhibited nonspecific behavior, with no apparent radiological signs of malignancy; additionally, the CA 19-9 marker level has been around 200 U/mL since the start of the workup. Finally, a liver biopsy was performed, revealing histological findings consistent with multiple biliary hamartomas associated with nodular regenerative liver changes.

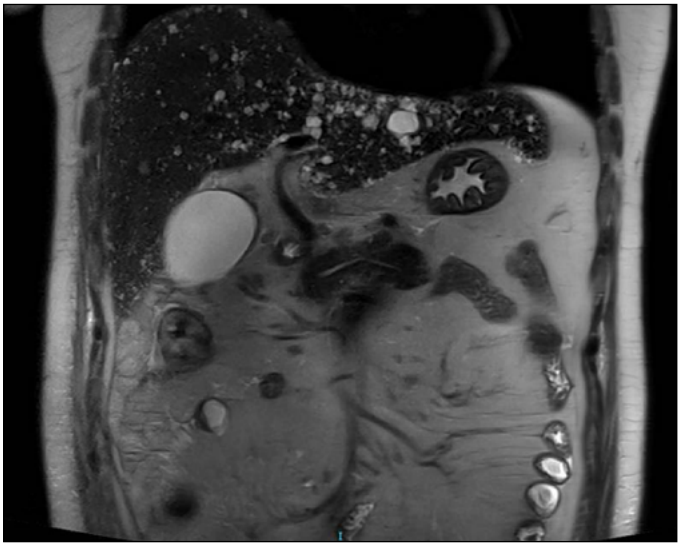

Figure 1. ***Multiple hyperintense lesions in the arterial phase.***

Discussion

Biliary hamartomas can be located in any segment of the liver, although they tend to occur more frequently in the peripheral regions. They are usually small, measuring less than 1.5 cm in most cases[1,2]. In their evaluation, the differential diagnosis must be considered against multiple cystic lesions, small, diffusely distributed metastases, infiltrative-pattern hepatocellular carcinoma, disseminated microabscesses,

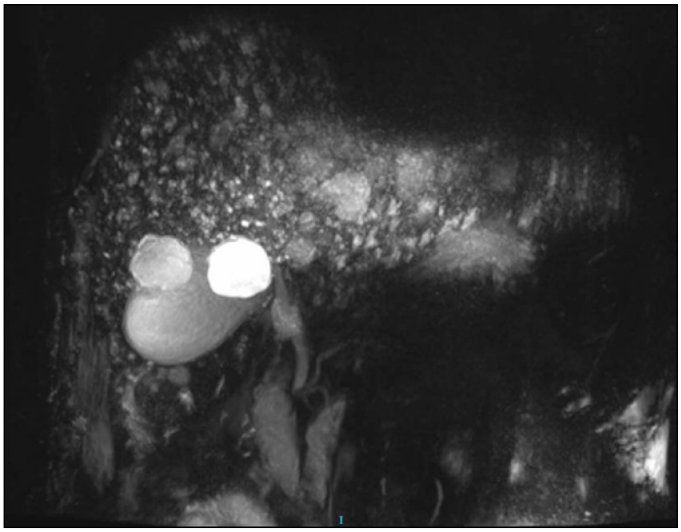

Figure 2. ***MR Cholangiography.***

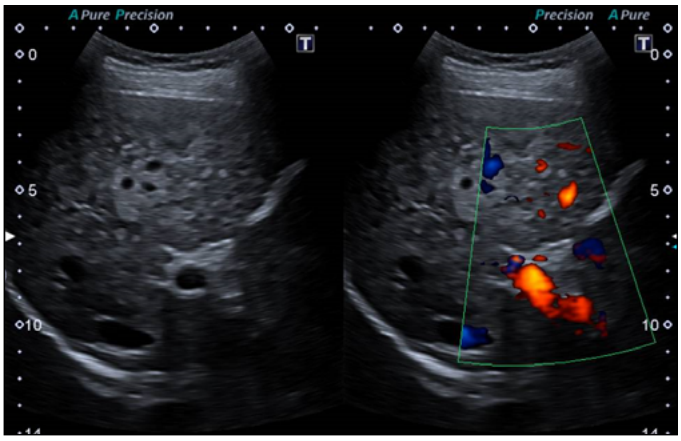

Figure 3. ***Space-occupying lesions on the liver with irregular morphology.***

and multiple nodular steatosis.[2,3]. On computed tomography, they appear as hypodense areas. On gadolinium-enhanced magnetic resonance imaging, they are characterized by T1-hypointense and T2-hyperintense signals.[1,2].

The CA 19-9 tumor marker is nonspecific and may remain persistently elevated in patients with hamartomatosis without indicating malignancy. However, exceptional cases of transformation into intrahepatic cholangiocarcinoma have been described in the literature, particularly in patients with underlying chronic liver disease[3], which warrants individualized clinical follow-up in certain patients.

In general, biliary hamartomatosis does not require specific treatment, except in symptomatic patients (e.g., right upper quadrant pain or pruritus) or in those in whom a complication is suspected. The prognosis is usually favorable, although it is prudent to maintain clinical surveillance in patients with associated advanced liver disease[3].

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Bibliography

1. Pech L, Favelier S, Falcoz MT, Loffroy R, Krause D, Cercueil JP. Imaging of Von Meyenburg complexes. Diagn Interv Imaging. 2016;97(4):401-409. Frenette C, Mendiratta-Lala M, Salgia R, Wong RJ, Sauer BG, Pillai A. ACG clinical guideline: focal liver lesions. Am J Gastroenterol. 2024;119(7):1235-1271.

2. Monteiro de Barros J, Stell D, Bracey TS, Mavroeidis VK. Diffuse liver hamartomatosis (diffuse von Meyenburg complexes) mimicking hepatic metastases on a background of previous cancer. Ann R Coll Surg Engl. 2020;102(1):e1-e4.

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