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Case Report

The Starry Sky Liver: An Appearance of Von Meyenburg Complex, a Benign Lesion with Potential Malignancy

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Abstract

Biliary hamartomas are rare congenital anomalies of bile duct development, usually discovered incidentally. They often present as multiple lesions on the liver surface, resembling metastatic lesions.

We report a case of a 45-year-old woman with no significant comorbidities who presented with recurrent hepatic colic episodes and gastroesophageal reflux. An ultrasound revealed a diffuse heterogeneous liver echotexture. Biliary MRI (Magnetic Resonance Imaging) confirmed the diagnosis of von Meyenburg complex.

Our case aims to discuss the imaging diagnosis of these rare benign hepatic lesions while highlighting the potential for malignancy-an often overlooked feature-underscoring the importance of follow-up.

Keywords: *Multiple Biliary Hamartomas (MBH); Von Meyenburg Complexes (VMCs); MRI (Magnetic Resonance Imaging)*

Introduction

Multiple biliary hamartomas (MBH), also known as Von Meyenburg complexes (VMCs), are congenital malformations resulting from ductal plate dysgenesis [1]. They are classically described as benign liver pathologies, often detected incidentally. However, there has been ongoing debate in the literature regarding their potential malignancy [2]. Their prevalence is approximately 3% at autopsy [3], whereas the prevalence on imaging is less than 1%, as most hamartomas are smaller than 5 mm and often go undetected.

This article reports a case of von Meyenburg complex in a 45-year-old female patient, highlighting the rarity of such cases, the imaging findings that can resemble metastases, and the potential for malignancy, which is currently being debated in the literature.

Case Report

A 45-year-old woman with no significant comorbidities presented to our department with recurrent hepatic colic and gastroesophageal reflux. She also reported passing dark-colored urine and clay-colored stools without any history of fever, loss of appetite, or unintentional weight loss.

On physical examination, her vital signs were stable. She was anicteric, and the abdominal palpation revealed mild tenderness in the right hypochondrium, with the rest of the examination being normal.

The biological tests showed hepatic cytolysis at twice the normal level, without biological cholestasis or inflammatory syndrome, and the rest of the tests were normal. Serological tests for hepatitis B and hepatitis C were negative.

An abdominal ultrasound was performed, which showed a diffuse heterogeneous liver echotexture. No dilation of the intra- or extra-hepatic bile ducts was noted, and the gallbladder was normal without any signs of lithiasis.

Given the persistent pain and the abnormal enzymatic activity in the biological tests, a biliary MRI was requested to investigate the suspicion of biliary lithiasis migration. MRI (Figure 1 and 2) revealed multiple scattered, bilateral microcystic lesions in the liver, non-communicating with the bile ducts, giving a "starry sky" appearance, with no localized lesions suggestive of malignancy. There was no dilation of the intra- and extra-hepatic bile ducts, and the gallbladder had a thin wall and showed no signs of lithiasis.

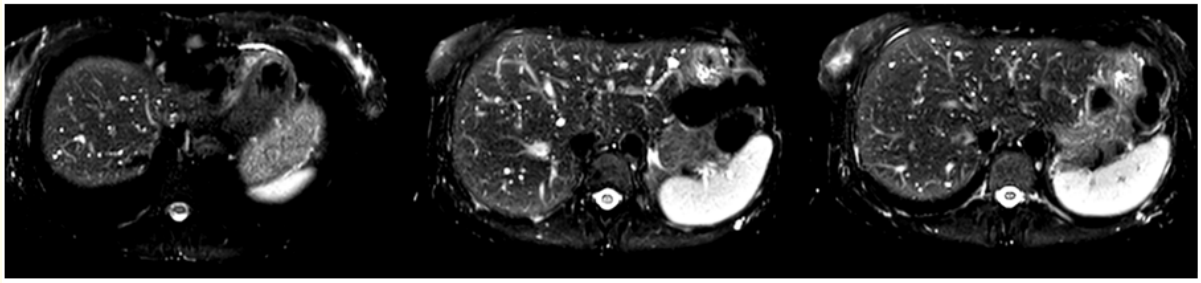

Figure 1: *Axial T2 SPAIR (SPectral Attenuated Inversion Recovery) images showing the multiple small, high signal T2 hepatic lesions confirming their cystic nature, scattered throughout hepatic parenchyma.*

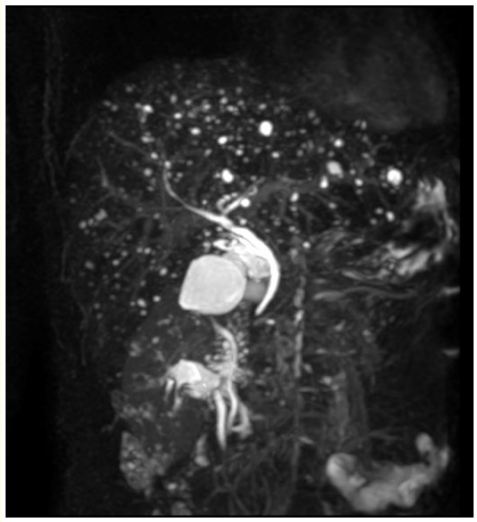

Figure 2: *Coronal MIP MR-cholangiopancreatography image revealing the typical "starry sky" appearance of multiple biliary hamartomas in the liver due to the presence of small multiple scattered cysts throughout the liver with no communication with the non-dilated intra- and extrahepatic biliary tree.*

The exploration was completed with a biliopancreatic endoscopic ultrasound, which suggested the presence of microlithiasis in the gallbladder, with no lacunar images observed in the main bile duct.

The evolution was marked by a significant clinical and biological improvement under symptomatic treatment, with a reduction in pain and normalization of the liver function tests. The patient is now asymptomatic and is being followed up on an outpatient basis.

Discussion

Von Meyenburg complexes were first described in 1918 by von Meyenburg. They consist of multiple biliary micro-hamartomas [3]. The origin of these lesions is congenital, related to a developmental anomaly of the ductal plate [4]. It appears to be a disorder in the remodeling of the small bile ducts that develop from the ductal plate, which should normally give rise to the peripheral intrahepatic bile ducts.

These bile ducts should normally connect with the biliary system of endodermal origin, which develops from the foregut.

Because this disease typically does not cause clinical symptoms, it is often discovered incidentally during imaging studies. The prevalence of von Meyenburg complex is approximately 0.6% in 2,000 biopsies [1].

These lesions can occur in isolation or be associated with fibro polycystic diseases, such as Caroli's disease or congenital hepatic fibrosis [3,4]. Von Meyenburg complex is therefore considered part of the spectrum of congenital cystic liver diseases, which also includes congenital hepatic fibrosis and Caroli's disease.

The sonographic findings of the von Meyenburg complex are variable, including multiple, small, hyperechogenic images with poorly delimited margins, with or without- posterior reverberation, or even hypo-echogenic images with a "target" pattern and well-delimited margins [6,7]. As regards the number of hamartomas in imaging studies, they may present as unimodular lesions, but most frequently, they are multinodular [5,6]. Such characteristics lead to further investigation, particularly in the presence of some known tumors, considering their similarity with metastases. In the literature, several case reports and series have described an association between the von Meyenburg complex and cholangiocarcinoma [8]. As a result, the benign nature of the von Meyenburg complex is now being questioned, and it is considered a potential risk factor for the development of cholangiocarcinoma [2].

In our case, Von Meyenburg complexes were found incidentally. Although this condition is a benign liver pathology, it can sometimes clinically mimic liver metastases or metastatic cholangiocarcinoma. This highlights the importance of pathological confirmation to rule out malignancy before considering treatment for any metastatic cancer.

Conclusion

Multiple biliary hamartomas (MBH) are a rare cause of multiple benign hepatic lesions. The condition is also known as von Meyenburg complexes, multiple bile duct hamartomas, or biliary micro hamartomas. It is usually an asymptomatic condition, which is usually found incidentally. It is important to differentiate them from other causes of multiple liver lesions, particularly metastases.

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