Multiple biliary hamartomas (Von Meyenburg complexes): Imaging findings and clues for differential diagnosis

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Learning objectives

The purpose of this review is to show all the radiologic findings of Multiple Biliary Hamartomatosis (Von Meyenburg complexes) on Ultrasound, CT and Magnetic Resonance, emphasizing the diagnostic clues.
Background

Biliary hamartomas, also called biliary microhamartomas or Von Meyenburg complexes (VMC), are benign malformations of the intrahepatic bile ducts, due to the failure of embryonic involution. They are part of the spectrum of ductal plate malformations as Caroli’s disease, Hepatorenal Polycystic disease and Congenital Hepatic Fibrosis.

Although there are only a few cases reported in the literature, the incidence seems to be higher than thought, approximately 5.6% based in a study of 2843 autopsies.\(^1\)

**PATHOLOGY:**

At gross examination, VMC are well circumscribed uniform greyish nodules ranging from 1mm to 15mm (generally <10mm), diffusely scattered throughout the hepatic parenchyma with a predilection for subcapsular locations. Microscopically, VMC are well-defined irregular cystic dilations of bile ducts, containing sometimes a small amount of inspissated bile, and embedded in a fibrocollagenous stroma.

Although they have been classically considered benign lesions, associations with some malignancies, especially with cholangiocarcinoma have been described. Although the incidence of this association is considered to be very low (there are less than 20 cases reported in the scientific literature), several recent anatomopathological studies seem to have demonstrated transitional foci showing varying degrees of dysplasia adjacent to cholangiocarcinoma areas.\(^2\) These different degrees of dysplasia would represent the progression from a normal VMC to a malignant lesion (which still retains some features from the hamartomas) in what some authors call adenomatous neoplastic transformation.\(^3\)

The increased risk of malignancy associated with VMC has been attributed to bile stasis and prolonged exposure of the hepatic parenchyma to potential carcinogens.

**CLINICAL RELEVANCE**

The main diagnostic problem with multiple biliary hamartomatosis (MBH) to the radiologist however, is its differentiation with other pathological processes especially with metastatic liver disease. This problem is accentuated when there is primary neoplastic history and above all, when we evaluate the isolated findings of only one imaging modality. Thus, for example, is frequent infer in a report, that the findings are suggestive of corresponding to
a metastatic liver or to a Polycystic Liver disease taking into account only the Ultrasound or MR findings respectively.
Imaging findings OR Procedure details

Ultrasound:

Classically, US findings have been described as either hyperechoic or hypoechoic small nodules, typically measuring less than 10 mm (Fig. 1 on page 8, Fig. 2 on page 8, Fig. 3 on page 9). In our patients, they mostly were hyperechoic. As for the anechoic lesions, since the MBH is associated with the presence of liver and kidney cysts that, on the other hand are not uncommon in the general population, it becomes difficult if not impossible, to secure with any imaging modality, that they are hamartomas instead of cyst. However, this dilemma has no clinical significance. Usually, those measuring more than 10mm are considered cysts.

Lesions may show a comet-tail artifact (Fig. 4 on page 10, Fig. 5 on page 10, Fig. 6 on page 11), although it is also common to see this sign in the absence of a clear underlying lesion, probably due to its small size.

Another common finding is the visualization of a diffusely heterogeneous liver (Fig. 7 on page 12). This appearance resembles that of the chronic liver disease or even the metastatic liver, is due to the presence of a great number of small ill-defined, irregular in shape lesions (mostly hyperechoic) scattered along the liver parenchyma. In these cases, the use of high frequency transducers or the zoom function, are useful to identify the individual lesions (Fig. 8 on page 13).

CT:

CT shows multiple hypodense small nodules uniform in size and scattered throughout both lobes, without central or peripheral enhancement after intravenous administration of iodinated contrast material (Fig. 9 on page 14, Fig. 10 on page 14). Although there are described in the literature some cases of apparent peripheral rim-like enhancement (also after Gadolinium administration on MR), histopathology studies have correlated this pseudo-enhancement with the compressed liver parenchyma surrounding the lesions.4

MR:

On MR, lesions are hypointense compared with liver parenchyma on T1-weighted images and markedly hyperintense on T2-weighted images (Fig. 11 on page 14). As was the case with the iodinated intravenous contrast, they do not show any kind of enhancement after administration of gadolinium.
**MR Cholangiography** (MRC) is very useful in evaluating MBH, showing multiple tiny "cystic" lesions scattered through both lobes with normal appearance of the extrahepatic and intrahepatic bile ducts. There is no communication with the biliary tree allowing us to differentiate it from Caroli’s disease.

MRC has higher sensitivity than CT scans and MRI, detecting a greater number of lesions and defining their morphology even better (Fig. 12 on page 15).

A recent study that correlated the histological findings of 11 patients retrospectively with the findings encountered in previous MR studies, has provided a sign not described until now, the "**mural nodule sign**". According to the authors of this study, 10 of the 11 patients showed in the wall of at least one of the cystic images, a small mural nodule isointense with liver parenchyma in T1 and T2-weighted images that enhanced after gadolinium administration. They correlated this finding in the histological study with an endoluminal polypoid projection of connective tissue.

However we didn’t identify this sign in any of our patients. There is the possibility that these projections correspond to the fibrous stroma which surrounds the hamartomas, adopting a pseudo-polypoid appearance, due to the known irregular shape of the VMC.

**KEYS FOR THE DIFFERENTIAL DIAGNOSIS**

**Liver metastasis**: Previous history of extrahepatic malignancy. Lesions are ill defined on CT. They tend to be less homogeneous in size and often show a pattern of enhancement after administration of IVC, including peripheral enhancement in cystic metastasis.

**Hepatic cysts**: Anechoic with posterior acoustic reinforcement in ultrasound, tend to be variable in size and number. If they measure more than 15 mm, they are unlikely hamartomas. They are better defined than the hamartomas on CT and MR.

**Caroli's disease**: Multiple cystic images of varying sizes, demonstrating communication with the intrahepatic biliary tree (especially on MRC) and with a central enhanced nodule (**central dot sign**).

**Liver microabscesses**: history of immunosuppression, infectious or painful symptoms. "**Target sign**" on US.
- **Peribiliary cysts**: Multiple cystic dilations of the peribiliary glands. Cystic lesions located in the hilum and intrahepatic portal branches region.
Fig. 1: Diffusely heterogeneous liver appearance due to the presence of multiple poorly defined nodules less than 10mm, mostly hyperechoic. Larger anechoic nodules were considered as hepatic cysts.

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**Fig. 2:** US magnified scan of the same patient using zoom function, showed multiple hyperechoic nodules.

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**Fig. 3:** US scan of a different patient showing a heterogeneous liver parenchyma due to the presence of multiple ill-defined tiny hypoechoic nodules.

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**Fig. 4:** Multiple typical "comet-tail" echoes in MBH.

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Fig. 5: Comet tail-echoes as an incidental finding in a patient with liver metastasis (not shown).

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**Fig. 6:** Small hyperechoic nodules with associated "comet-tail" artifact in both liver lobes.

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Fig. 7: US scan of the liver in a patient with MBH and no history of liver disease or known malignancy, showing a diffusely heterogeneous appearance not being possible to clearly individualize the lesions (apart from several hepatic cysts).

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Fig. 8: Two ultrasound images of the left liver lobe in an asymptomatic patient with MBH, by using a 4 MHz transducer (a) and 8 MHz transducer (b). In the first case, a high suspicious heterogeneous liver echo texture can be seen. When using the high frequency probe, the appearance changes allowing us to clearly identify hyperechoic small nodules (arrows).

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Fig. 9: Plain CT images (a) and after administration of IVC in arterial phase (b) and in portal phase (c). Note the presence of multiple small nodules scattered throughout both lobes, hypodense in all series and with no enhancement after intravenous contrast administration. The two larger lesions visualized in segment II of the left liver lobe (arrows), were shown anechoic on US, being considered hepatic cysts. The rest of lesions, were found hyperechoic.

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Fig. 10: Plain CT (a) and contrast-enhanced CT in arterial phase (b) and portal phase (c). Multiple ill-defined hypodense small nodules are seen in all phases. They didn´t show any enhancement. The lesions are distributed along both lobes, being most abundant in the left liver lobe. Note the presence of bilateral renal cortical cysts. This patient ultrasound findings are shown in Figure 8.

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**Fig. 11:** Axial T1-weighted MR image (a), shows multiple small hypointense nodules throughout both liver lobes. On axial (b) and coronal (d) T2-weighted MR images, the lesions are strongly hyperintense. They do not show any kind of enhancement after gadolinium administration (c). Several lesions measuring more than 1cm and showing similar features are seen. These latest were described as hepatic cysts due to its anechoic appearance on US.

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**Fig. 12:** MR Cholangiography of previous patient shows no communication between the multiple "cystic" lesions and the biliary ducts. The nodules are more apparent and numerous than on T1 and T2-weighted images.

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Conclusion

Multiple biliary hamartomatosis is a more common entity than was thought, and in most cases can and should be diagnosed reliably without the need of biopsy, by the joint assessment of the findings on ultrasound, CT and MRI.

We should consider as likely this diagnosis in the presence of the following findings: *Heterogeneous liver echo texture due to multiple scattered nodules, mostly measuring less than 10mm, and similar in size, poorly defined hyperechoic or hypoechoic with or without comet tail artifact, shown hypodense on CT, hypointense on T1-MR and hyperintense on T2-MR with no enhancement after administration of intravenous contrast and not connected to the biliary tree (MRC).*

However, we think that some isolated findings would be sufficient to infer the suspicion of HBM:

- Multiple *comet-tail* echoes on ultrasound.

- Multiple "cystic" lesions without communication with the biliary tree, uniform in size and generally lesser than 1 cm, seen on MR or MRC, in the absence of infectious signs.

- Multiple tiny *hypodense* nodules homogeneous in size, with a scatter distribution and with no enhancement after contrast on CT, shown as *hyperechoic* on US in patient with no history of primary neoplasm.

It seems prudent however to follow up patients who still show typical findings, and/or present a history of extrahepatic malignant tumor. Liver biopsy would be considered for patients with history of primary neoplasm, who don’t show all the typical findings of HBM.
References


