PICTORIAL ESSAY | Abdominal imaging

# Von Meyenburg Complexes (biliary hamartomas): Imaging with US , CT and MRI.

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## ABSTRACT

**Purpose.** The purpose of this study is to describe the radiological characteristics of Von Meyenburg complexes, a generally benign condition that may be confused with other entities and to emphasize on its relationship with other diseases.
**Methods.** We studied 21 patients over a period of 10 years aged between 21 and 54 years old which were evaluated with Ultrasound(US) and/or Computed Tomography(CT),Magnetic Resonance Imaging(MRI) modalities.
**Results.** In 16 of our patients Von Meyenburg Complexes were found exclusively, in 3 patients in combination with simple cysts, in one patient in combination with peribiliary cysts and in the last one in combination with peribiliary cysts and ADPKD (autosomal dominant polycystic kidney disease).
**Conclusions.** The knowledge of the pathology and radiological appearance of Von Meyenburg complexes contributes to the establishment of correct diagnosis and the avoidance of unnecessary interventions.

KEY WORDS | Von Meyenburg Complex, biliary hamartomas , Imaging,MRI, CT , US

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## Introduction

Von Meyenburg complexes (VMCs) or multiple biliary hamartomas are small (<10 mm) benign liver lesions which represent cystic dilated bile ducts, lined by a single layer of cuboid epithelium and surrounded by abundant fibrous stroma,which sometimes is hyalinized [1].

The lumen of the dilated bile ducts of VMCs may contain clear bile, inspissated bile, polypoid projections ,in the lumen or islands of connective tissue.

Their pathogenesis is attributed to ductal plate malformations involving the smaller interlobular bile ducts in the late phase of embryologic development of the intrahepatic bile ducts(a factor arresting or perturbing the remodeling of the ductal plates) [2, 3].The large number of bile ducts profiles and the continuity with the portal tracts suggest a pollard willow abnormality in the branching pattern of the peripheral portal vein ramifications [2].

Their prevalence is around 5,6% in autopsy series[4]. The biliary hamartomas are well- circumscribed, irregular or round structures which can be found into the liver, especially in the subcapsular area,in continuity with or merged into the portal tracts [5,6]. They may contain sclerotic arteries or be devoid of vessels [5].

We estimated 21 patients with VMCs with US, CT and MRI.

CT scans were obtained by a SIEMENS Somatom Perspective 16 machine with a 2mm scan width before and after intravenous contrast material administration. MRI images were obtained using a 1,5 Tesla PHILLIPS machine.

From these 21 patients16 revealed only VMCs, 1 patient had also peribiliary cysts, 3 patients had also simple liver cysts and the last patient had simple cysts, peribiliary cysts and ADPKD (autosomal dominant polycystic kidney disease) (**fig.1**). 3 patients had a biopsy and histological results confirmed the diagnosis of VMCs.

Ultrasound examination of the liver was negative in 11 cases and in the rest of the cases it was very difficult to identify the accurate number of the lesions. In 3 cases the

VMCs appeared as echogenic small dots with and without the comet-tail artifact (**fig.3**). In 3 patientsVMCs reveal a cystic appearance with posterior enhancement (**fig.4**) (**fig.6**) .

CT exams of the liver were able only in 6 cases to detect the lesions with low sensitivity (**fig.8**).

MRI exams especially with T2-weighted sequences and MR Cholangiography were better both in the visualization and in the confirmation of the exact number of lesions (**fig.2**).

## Discussion

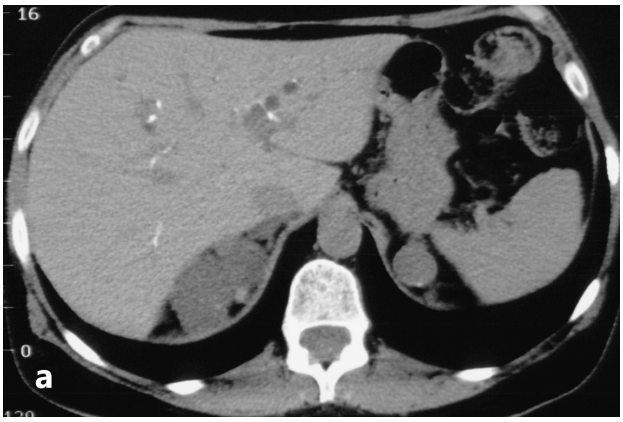
Von Meyenburg Complexes (VMCs) are ductal plate, non cancerous malformations of the liver that are usually diagnosed on imaging studies. There has been described association of VMCs with autosomal dominant polycystic kidney disease (ADPKD), simple liver cysts, peribiliary cysts and pancreatic cysts [2,4,7].

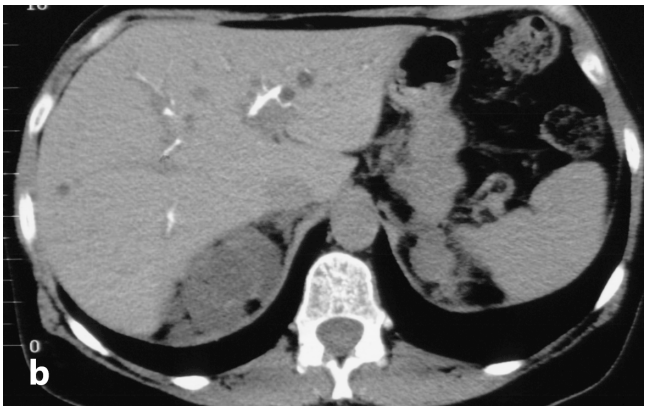
The malignant transformation of biliary hamartomas to intrahepatic Cholangiocarcinoma is rare [8,9,10]. This has been found to occur due to gradual transition from VMCs to hyperplastic or adenomatous lesions and then progress to malignant transformation [11].

There is heterogeneity in ultrasound imaging findings that actually reflects the histological features, including the amount of fluid or inspissated bile of the dilated ducts and the fibrous stroma surrounding them [12]. The multiple biliary hamartomas may appear as micro-nodules , hypo- or hyperechoic depending from the material inside the lumen of the dilated bile ducts [13,14,15]. There has been described a typical imaging finding of multiple comet-tail echoes [16], representing the posterior enhancement of the tiny cystic lesions. Furthermore, some of the hyperechoic lesions are found to be cystic when magnification is applied[13].

On plain CT , VMCs appear as multiple round or irregular hypoattenuating lesions in both hepatic lobes [12,13,17]. In most of the cases there is no enhancement of the lesions after intravenous administration of contrast medium (poor vascularity) , but the biliary hamartomas become more clearly visible [13].However homogeneous enhancement has been reported, possibly concerning lesions with prevalent fibrous stroma [18,19].

VMCs are well-circumscribed round or irregular lesions which tend to be hypointense on T1-weighted images and show markedly high intensity on T2-weighted images, due to the fluid- containing ductules [13, 19, 20,21,22].In some cases the high T2 signal is attenuat-



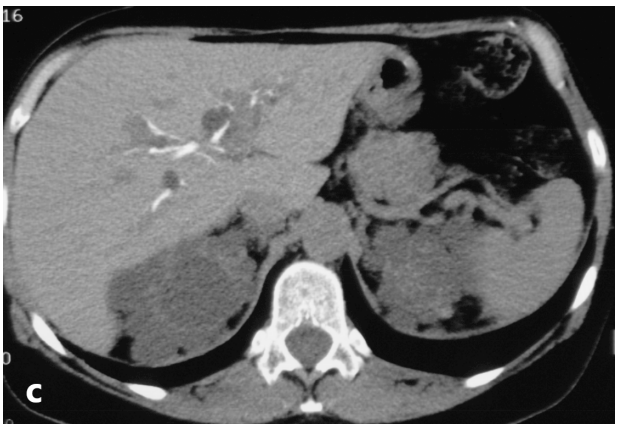
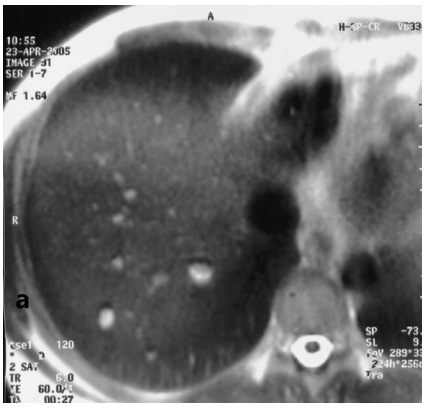
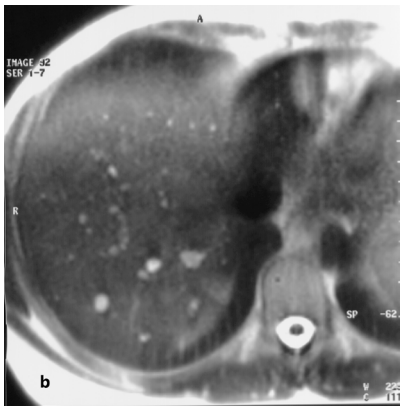
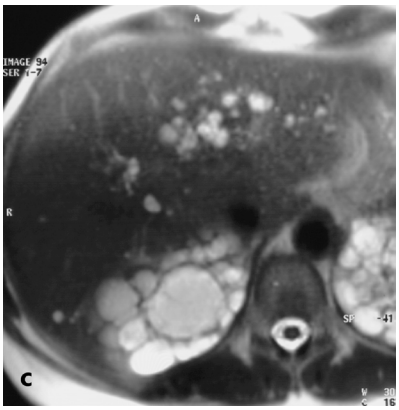




***Figure 1***
*(a,b,c) axial post-contrast (cholangiographic contrast media) CT images which reveal the peribiliary cysts, simple cysts and cysts in the kidneys (ADPKD)*



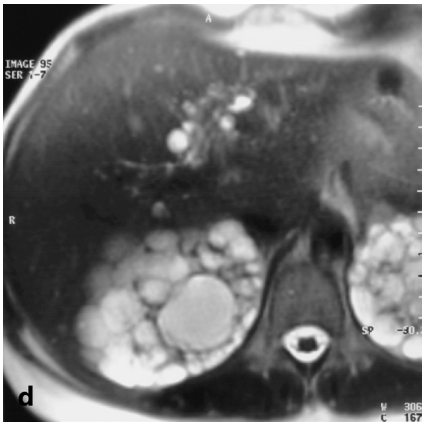




***Figure 2***
*(a,b,c,d) axial T2WI and MR Cholangiography which clearly depicts the VMCs, peribiliary cysts, simple cysts and the cysts in the kidneys (ADPKD).*

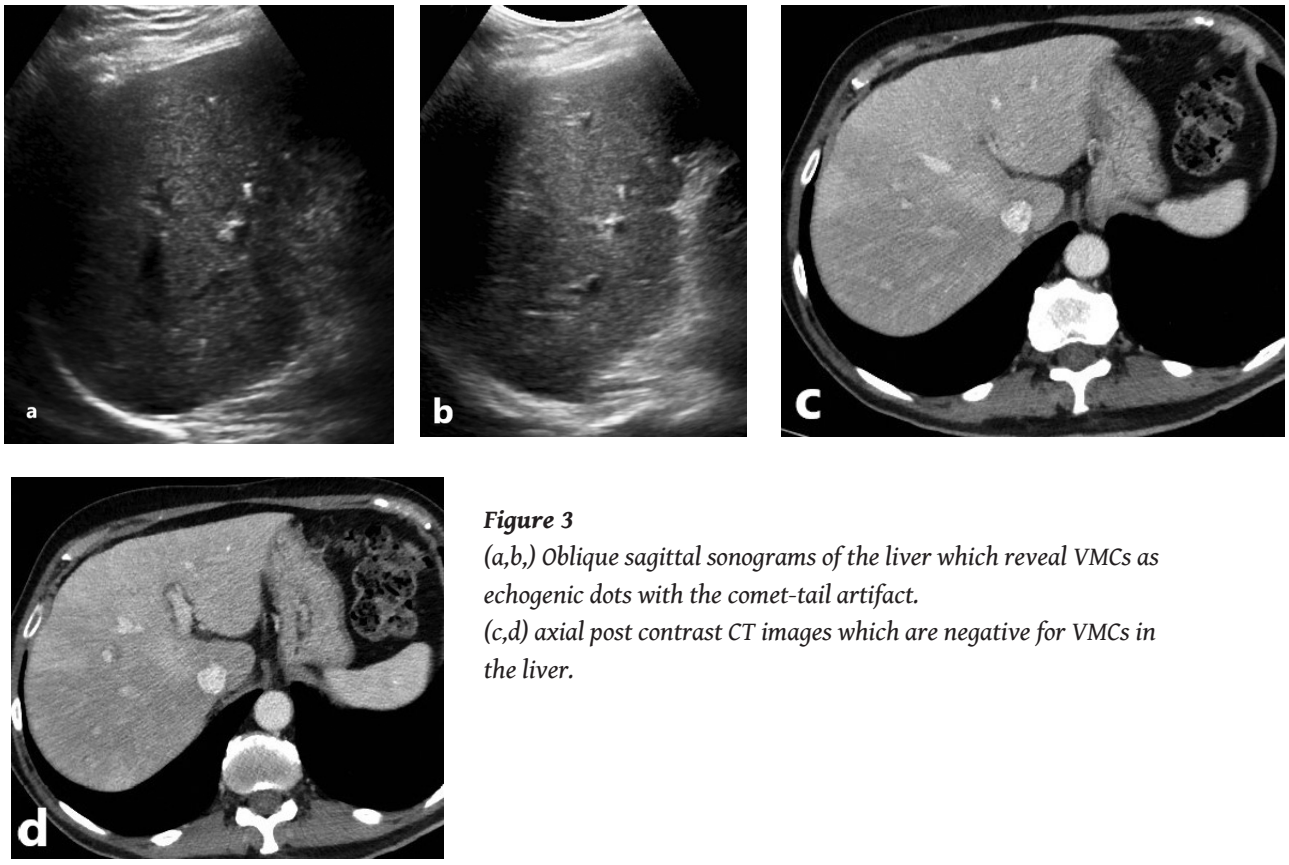



***Figure 3***
*(a,b,) Oblique sagittal sonograms of the liver which reveal VMCs as echogenic dots with the comet-tail artifact.*
*(c,d) axial post contrast CT images which are negative for VMCs in the liver.*

ed by the low signal intensity of the fibrous tissue [20]. Where the echo time increase at T2- sequences , the signal intensity increases and becomes similar to cerebrospinal fluid[19,21,22]. On diffusion- weighted sequences , biliary hamartomas mimic cystic lesions ,as they have a high apparent diffusion coefficient [23].

MR Cholangiography has been considered to be highly sensitive in demonstrating both the biliary tree and the cystic liver lesions, including their relationship with the ducts [24,25]. Additionally, MRCholangiography displays more lesions and delineates better their shape than conventional MRI , because of its higher contrast resolution [13,24 ] **(fig.7)**.There are no abnormalities affecting the biliary system and there is no obvious communication of the VMCs with the biliary system [13, 22].

Sometimes ,on T2-weighted images and MR Cholangiography the image of multiple VMCs have the appearance of a <<starry sky>> [26[ **(fig.5)**.

On T1- weighted images after administration of gadolinium there has been described different patterns ranging from no enhancement [12, 13 ] to homogeneous enhancement [19].A thin regular rim of early dynamic enhancement on early dynamic images that persists on late images may be observed and represents compressed liver parenchyma and inflammatory cells surrounding the hamartomas[24, 27]. Finally, a small enhancing mural nodule has been described ,which correlates histopathologically with an endocystic polypoid projection of collageneous supporting tissue, and is considered specific for VMCs [22]. The nodule appears isointense to liver parenchyma on T1- weighted images ,of intermediate signal on T2-weighted images and shows enhancement in all phases , including the delayed ones . The administration of contrast material with biliary excretion does not aggregate in hamartomas [23] **(fig.9)** **(fig.10)**.

The VMCs must primarily be distinguished from small metastases, especially in patients with a known malignancy. The lesions found in patients with biliary hamartomas are relatively uniform and less than 1 cm in size . In addition, they show varying degrees of enhancement after intravenous administration of contrast medium. Even when the VMCs show a peripheral rim of enhancement, it is thin and regular, as opposed to liver metastases, in which thicker, irregular and progressive

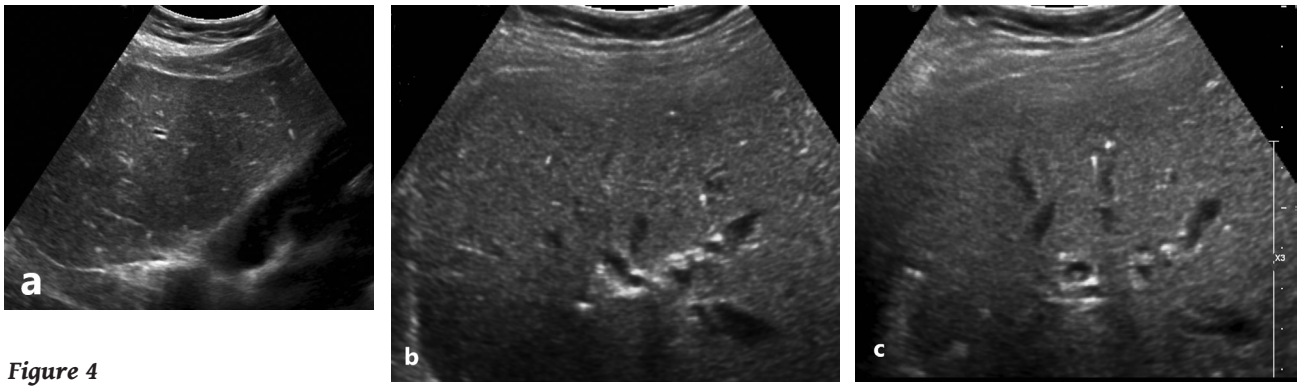



***Figure 4***
*(a,b,c) oblique transverse sonograms which reveal many VMCs as echogenic dots with posterior comet-tail artifact.*

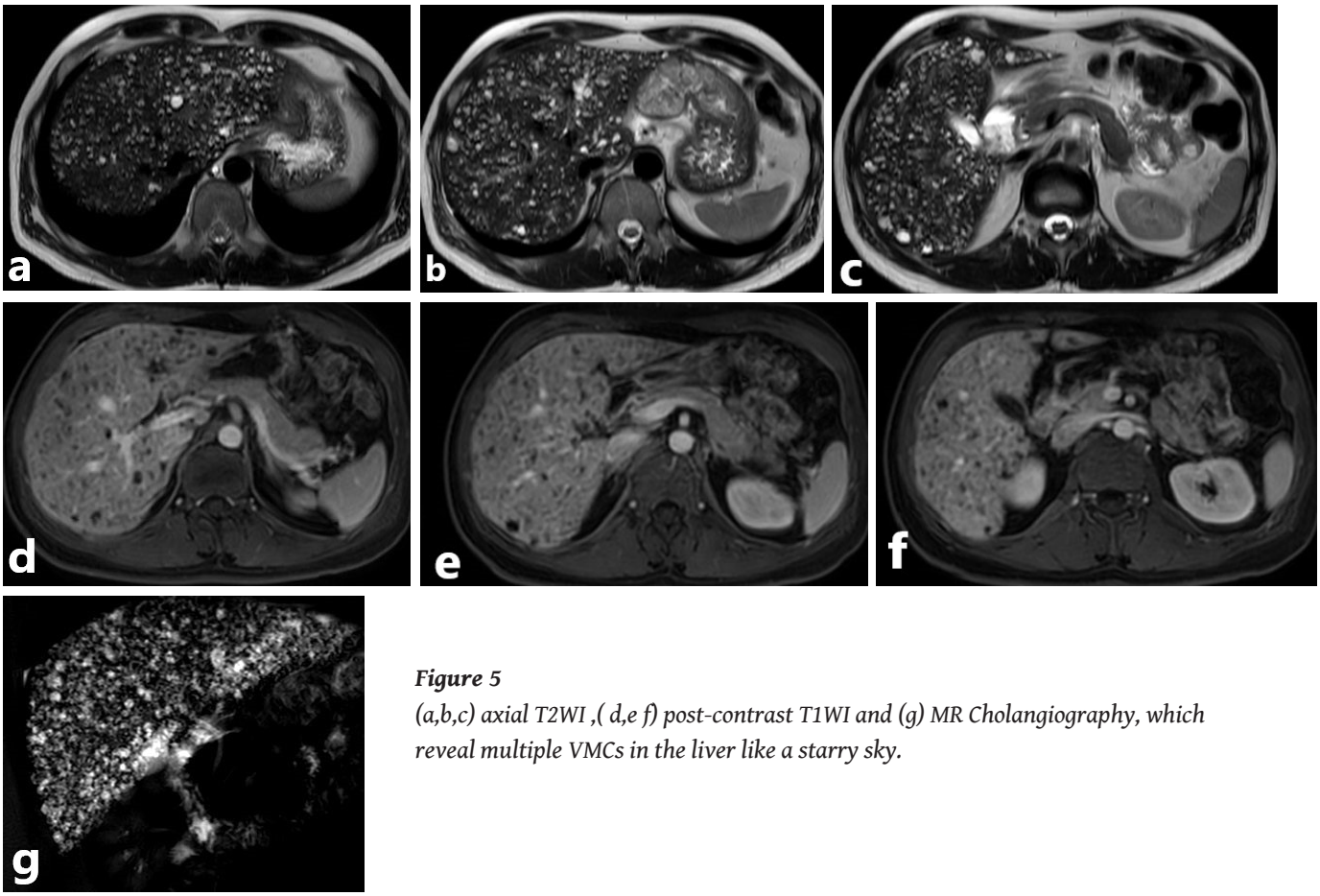



***Figure 5***
*(a,b,c) axial T2WI ,( d,e f) post-contrast T1WI and (g) MR Cholangiography, which reveal multiple VMCs in the liver like a starry sky.*

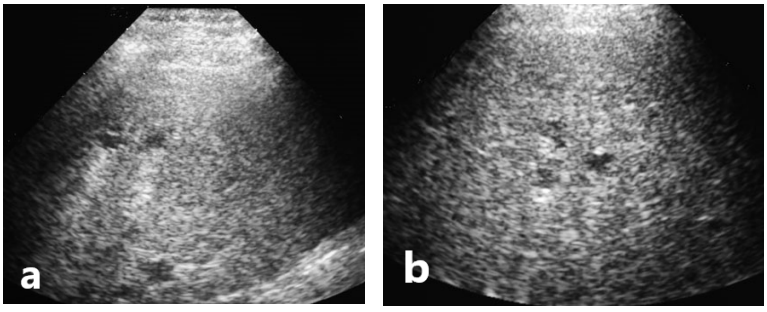



***Figure 6***
*(a,b) oblique transverse sonograms which reveal some VMCs with cystic appearance.*

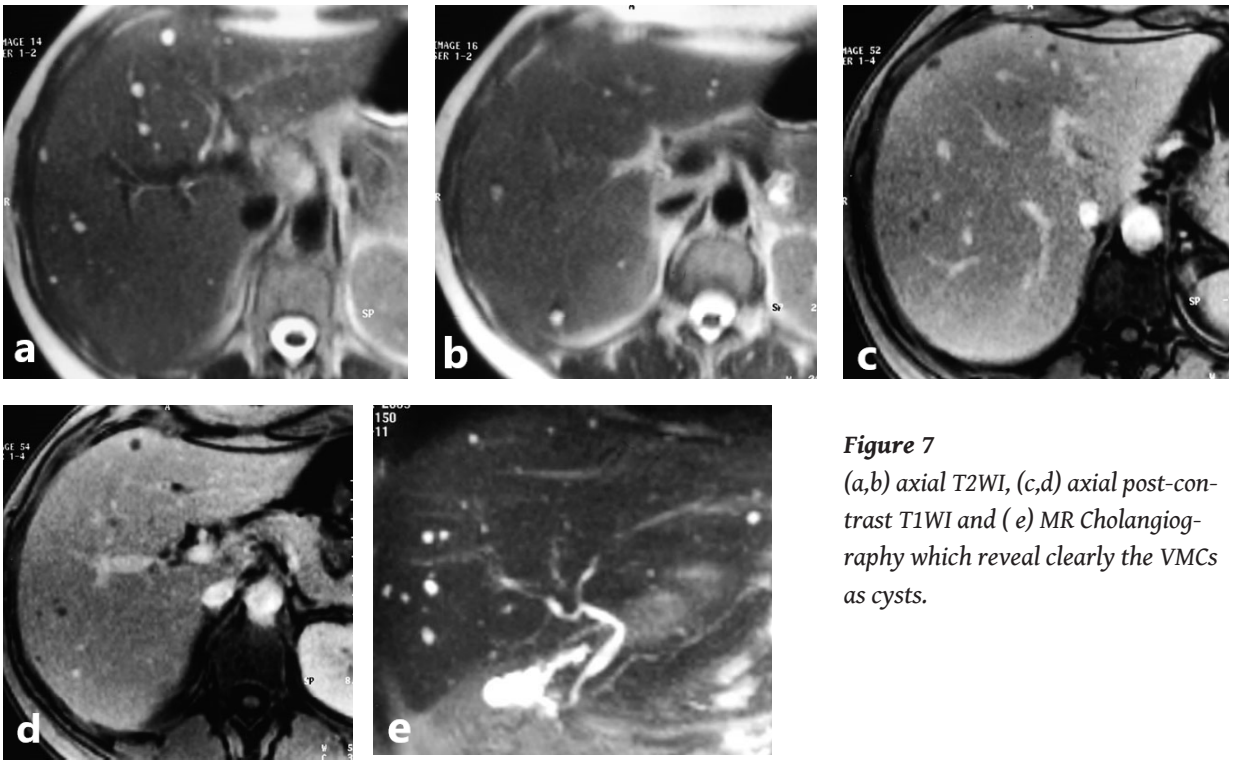



***Figure 7***
*(a,b) axial T2WI, (c,d) axial post-contrast T1WI and ( e) MR Cholangiography which reveal clearly the VMCs as cysts.*

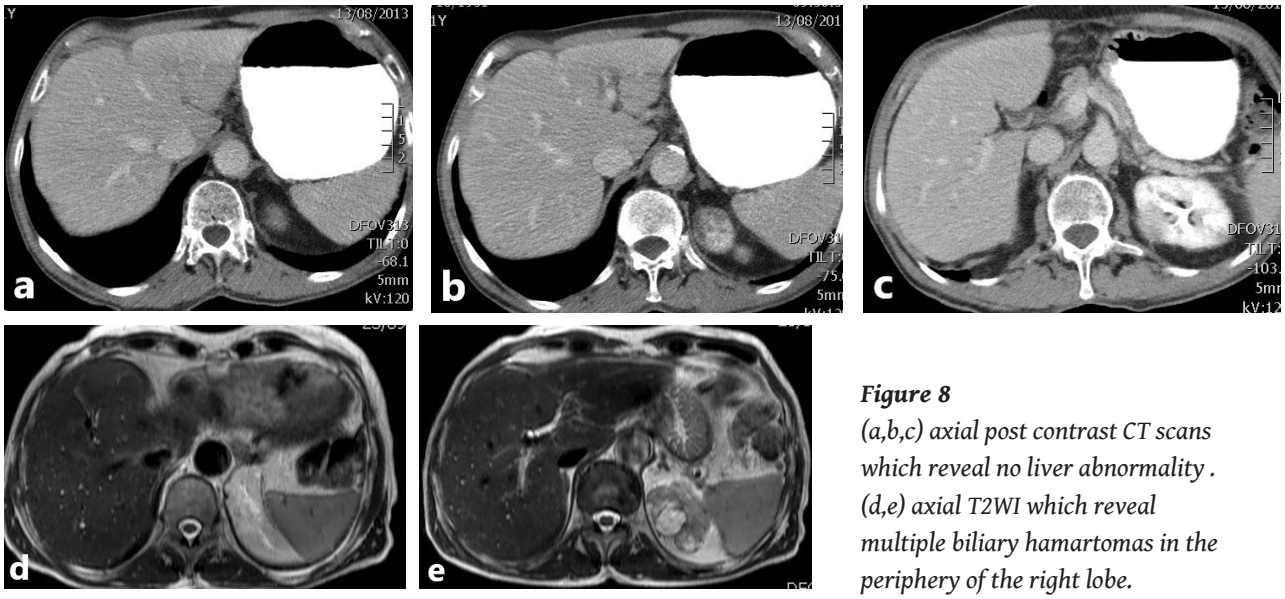



***Figure 8***
*(a,b,c) axial post contrast CT scans which reveal no liver abnormality . (d,e) axial T2WI which reveal multiple biliary hamartomas in the periphery of the right lobe.*

centripetal enhancement is observed. On T2-weighted images, the biliary hamartomas show hyperintense signal , which helps differentiate them from metastases. The latter are not visible when small on MR Cholangiography, unlike VMCs.Also unlike metastasis on T2-weighted images with a longer echo time, VMCs reveal an increased signal intensity.

In conclusion, diffusion-weighted MRI may be useful, as biliary hamartomas have a high apparent diffusion coefficient.

The differential diagnosis also includes multiple microabscesses, but the clinical context is important. Pa-

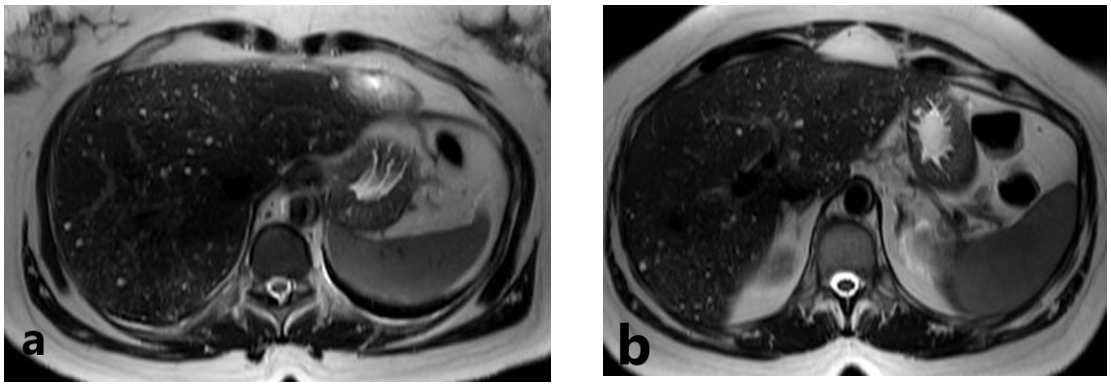



***Figure 9***
*(a,b) axial T2WI which reveal many VMCs*

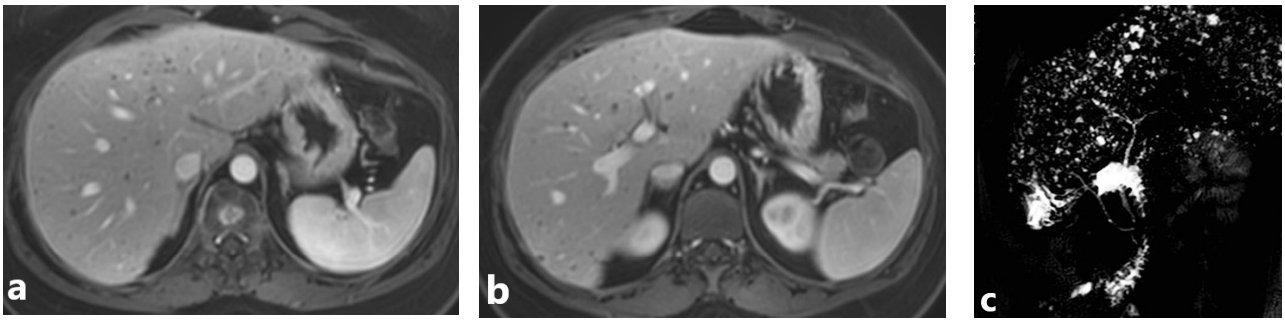



***Figure 10***
*(a,b) axial post contrast T1WI. VMCs have a cystic appearance without enhancement after contrast medium administration.*
*(c) MR Cholangiography which clearly reveal multiple biliary hamartomas.*

tients are almost always critically ill or even immunosuppresssed with a swinging fever and right upper quadrant abdominal tenderness. The abscesses show diffusion restriction on MRI and "target" appearance on US. Other conditions from which VMCs must be differentiated are: multiple hepatic cysts (there are lesions that are larger than 1 cm, round in shape,do not enhance and sometimes coexist with polycystic kidney disease), peribiliary cysts (they are located exclusively in the hepatic hilum and along the larger bile ducts), dilated bile ducts, diffuse primary hepatocellular carcinoma (usually in cirrhotic patients), Caroli's disease (the "cysts" communicate with the biliary tree at MR-Cholangiography and demonstrate enhancing "central dot sign" , which corresponds to the intraluminal portal vein branch).

In our study we found out that the T2-weighted sequences and MR Chongiography were the most valuable in the imaging of the VMCs.

## *Conclusion*

VMCs are benign liver malformations and their correct diagnosis can be made when the typical imaging findings are present without histological confirmation, thanks to the higher resolution and the advance of the imaging techniques. **R**

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## *Conflict of interest*

The authors declared no conflicts of interest.

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