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Multiple Biliary Hamartomas: The ‘‘Von Meyenburg Complexes’’

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ABSTRACT

Multiple biliary hamartomas are rare benign malformations of the intrahepatic bile ducts first described by von Meyenburg in 1918. This due to the developmental malformation of the interlobular bile duct in the liver during the embryonic period, leading to the formation of cyst-like structures of different sizes. Imaging has an important role in the diagnosis and differential diagnosis. A correct diagnosis might be obtained when typical imaging findings are present even without a histological confirmation.

Keywords: Liver cyst; Biliary hamartomas; Magnetic resonance cholangiopancreatography

CLINICAL HISTORY

A 48 year old female, with no medical history present for mild pain of the upper right quadrant.The clinical examination found no abnormalities. Laboratory tests showed no liver dysfunction, with normal levels of alanine aminotransferase, alkaline phosphatase, and bilirubin and a normal international normalized ratio.The patient was referred to the Department of Radiology and Abdominal ultrasound was performed and showed two cystic hypochoic lesions with no cloisons or bourgeons and multiple hyper and hypoechoic areas in the liver. Magnetic Resonance Cholangiopancreatography (MRCP) was also performed revealing two cystic lesions multiple small diffuse liver lesions of variable size (< 15 mm) hyperintense on hypointense on T1- weighted images and T2-weighted images with no enhancement after intravenous administration of gadolinium. In addition, MR cholangiography showed multiple small cystic liver lesions scattered through both liver lobes, with normal appearances of the intrahepatic and extrahepatic bile ducts. There was no communication between the lesions and the draining bile ducts, creating a “starry sky” appearance. These findings were consistent with multiple biliary hamartomas or “von Meyenburg complexes”.

IMAGING FINDINGS

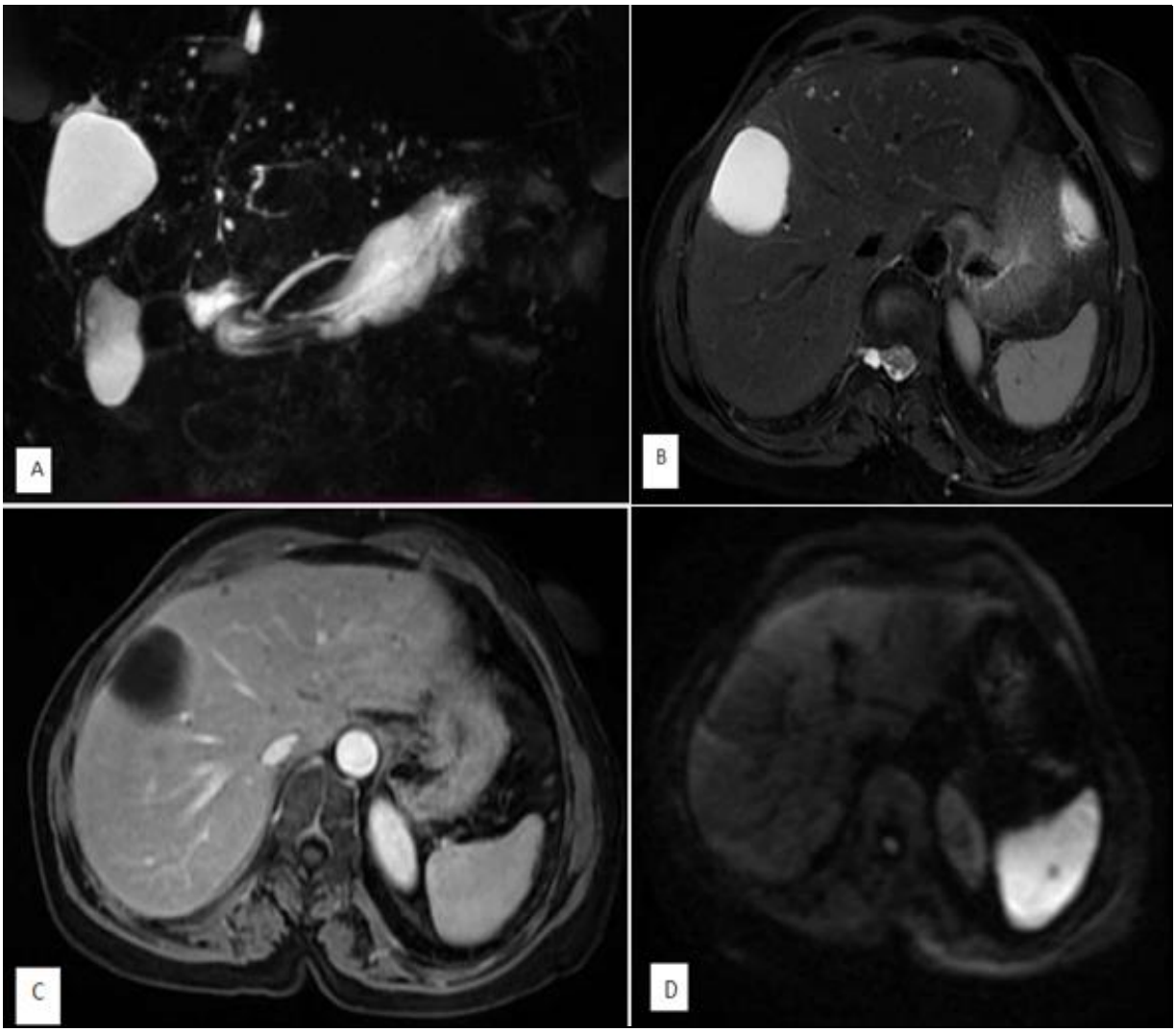

Figure A: MRCP shows no communication of the lesions with the biliary duct.

Figure B: T2-weighted MR image show multiple hyperintense lesions.

Figure C: T1-weighted MR image reveals multiple hypointense lesions of the liver and of variable size without enhancement after intravenous administration of gadolinium.

Figure D: DWI MR image showing no diffusion restriction.

FINAL DIAGNOSIS

Multiple biliary hamartomas

DISCUSSION

Biliary hamartomas also called as von Meyenburg complexes (VMCs) are benign liver malformations, which occur in about 0.6 %-2.8 % of the general population.[1] They belong to the spectrum of fibropolycystic liver diseases.[1] Biliary hamartomas histologically contain locale bile duct like structure aggregation, varying degree of bile ducts dilatation, and lining bile duct epithelium on the background of abundant connective tissue which are surrounded by dense fibrous compartments,[2] it usually manifests as small gray-white nodules scattered throughout the liver or several liver segments. Biliary hamartomas are rare, clinically asymptomatic, and diagnosis is usually incidental.[3] Imaging has an important role to confirm the diagnosis. CT scans can detect multiple, nodular and irregular cystic liver lesions, smaller than 15 mm, with low attenuation and no enhancement on contrast injection, this phenomenon might correlate to the poor vascularity of VMCs described on histology.[4] MRCP has proven to be highly sensitive in detecting intraand extrahepatic bile duct anomalies and cystic lesions of the liver as well as their relationship with bile duct system, the lesions appears as hypo-intense on T1-weighed MRCP shows no communication of the lesions with the biliary duct. T2-weighted MR image show multiple hyperintense lesions. DWI MR image showing no diffusion restriction. T1-weighted MR image reveals multiple hypointense lesions of the liver and of variable size without enhancement after intravenous administration of gadolinium images and hyper-intense on T2-weighed images when compared with surrounding liver parenchyma. Differential diagnosis for the imaging appearance of (VMCs) includes metastases, microabcesses, liver cysts, dilated bile ducts, biliary adenomas, and caroli's disease. At first, some authors claimed that the definitive diagnosis of VMCs was confirmed by liver biopsy, and the imaging finding were not specific, however with the use of advanced imaging modalities and long-term imaging follow-up, some authors pointed out that it might be possible to make a correct diagnosis of VMCs by imaging.[4]

VMCs are benign liver malformations that may mimic microabscesses, liver cysts, dilated bile ducts and biliary adenoma. Also, caution must be taken not to misinterpret these lesions as metastases in the context of a known extrahepatic malignant lesion. However, the large number of lesions, the predominantly small size of the lesions, the MRI features, and most of all, the presence of tiny mural nodules seen on MRI imaging with nodular enhancement are highly characteristic and considerably increase the accuracy of diagnosis.[3,5]

AUTHOR'S CONTRIBUTIONS

All authors contributed to this work. All authors have read and approved the final version of the manuscript.

CONFLICTS OF INTEREST

The authors declare that there are no conflicts of interest regarding the publication of this paper.

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