von Meyenburg's complex in a patient with primary malignancy: role of MRI

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Abstract von Meyenburg's complexes are benign liver malformations histologically constituting cystic dilatations of the bile ducts, surrounded by abundant fibrous stroma. Appearance of these lesions on MRI is characteristic and a fine needle aspiration or biopsy can be avoided and judicious management planned. We report a case of esophageal carcinoma who had suspicious lesions in the liver on CT scan, which after MRI were confirmed as biliary hamartomas.

Keywords Biliary hamartoma · von Meyenberg complex · Magnetic resonance imaging

Introduction

von Meyenburg's complexes (VMC; biliary hamartomas) of the liver are rare benign malformations that originate from embryonic bile ducts that fail to involute [1]. They are incidentally encountered at autopsy or surgery and are usually asymptomatic. Appearance on MRI is sparingly described due to their uncommon occurrence and rare presentation [2]. We describe the MRI appearances of a case of biliary hamartomas in a patient with esophageal carcinoma and its role in helping it differentiate from metastasis.

Case report

A 61-year-old male presented with one month history of dysphagia to solid and liquid foods, with occasional vomiting. A barium swallow examination was done which showed irregular, ulcerated growth in the mid-thoracic esophagus with luminal narrowing. Subsequent upper gastrointestinal endoscopy confirmed the growth and the biopsy taken showed squamous cell carcinoma. As a part of pre-operative work up, CT scan was done. It showed asymmetrical circumferential wall thickening involving mid-thoracic esophagus (Fig. 1A). Liver showed multiple tiny hypodense lesions in the right lobe and in segment 4 (Fig. 1B). As these lesions were suspicious for metastasis, MRI was done for further characterization of the lesion. It showed diffusely scattered lesions, less than 1 cm in size, in both lobes of the liver, which were hypointense on T1 weighted image and hyperintense on T2 weighted image (Fig. 2). The thick slab MRCP image showed these lesions studded in the liver parenchyma with no obvious biliary communication. The imaging appearance was characteristic and a diagnosis of VMC was made. The patient was operated and is enjoying good health after surgery.

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Fig. 1 Contrast enhanced CT scan, coronal reconstructed image (A) and axial image (B). There is asymmetric wall thickening of the mid-thoracic esophagus (arrow). The liver shows multiple tiny hypodense lesions (arrow heads)

Fig. 2 Axial T1 weighted (A) and T2 weighted (B) MR images show multiple tiny lesions hypointense on T1 and hyperintense on T2 weighted images

Discussion

von Meyenburg's complexes are uncommon malformation of the bile ducts, with a prevalence of 0.69 to 2.8% [3]. They are seen scattered throughout the liver parenchyma as tiny cystic structures usually less than 1.5 cm in diameter without any communication with the biliary tree [4]. Grossly, they are seen as grayish-white or black nodules with irregular outline; microscopically, they are seen as dilated, tortuous channels lined by single layer of cuboidal epithelium, surrounded by fibrous stroma [3]. The lesions are congenital and develop as a result of non-involution of embryonic bile ducts. There is also association between VMC and polycystic liver or kidney disease [5].

On ultrasonography, these lesions may be seen as hypoechoic or hyperechoic nodules, sometimes with comet tail artifact [6, 7]. This variation in appearance is due to the presence of cystic lesions, fibrous stroma and crowded

interfaces. The lesions are seen as hypodense nodules on a non-contrast CT scan, which mostly show no enhancement on post-contrast images [6, 7]. They are usually less than 1.5 cm in size and have irregular margins, helping in differentiation from hepatic cysts [4].

Appearance on MR imaging is characteristic, though sporadically described. The lesions are tiny and multiple and are hypointense on T1 weighted image and hyperintense on T2 weighted sequences [2]. The MRCP picture gives a typical appearance of multiple tiny cysts scattered throughout liver parenchyma, without any biliary communication [8]. On post-gadolinium images, the lesions show homogeneous or rim enhancement. The presence of a mural nodule showing enhancement is specific for VMC [2]. The features on MRI are characteristic in most cases and a histological diagnosis is not needed [6].

The most important differential diagnosis to be considered is metastatic lesions in a patient with primary malignancy. Sonography and CT scan does not usually differentiate

between the two. MRI is very helpful as VMC shows the characteristic imaging features [7]. A follow up imaging, in addition, can confirm VMC, obviating the need for biopsy. Another differential is polycystic liver disease, which can be differentiated by the cysts having > 1.5 cm size, smooth outline and association with renal cysts. In our case, MRI helped us make a proper diagnosis which was vital before a surgical excision of the tumour could be done. The typical appearance obviated the need for fine needle aspiration or biopsy.

In conclusion, the MRI and MRCP appearance of VMC are characteristic. In a patient with primary malignancy and suspicious liver lesions on sonography or CT scan, MRI helps in providing a diagnostic clue and thus avoids a biopsy and allows for judicious patient management.

Conflict of interest The authors do not have any disclosable interest

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