Case study

Intrahepatic multicyctic biliary hamartoma: presentation of a case report and magnetic resonance imaging / magnetic resonance cholangiopancreatography findings

Abstract : Biliary hamartomas, known as von Meyenburg complexes (VMCs), are benign liver malformations. They are histologically characterized by cystic dilated bile ducts surrounded by numerous fibrous stromal elements measuring up to 5 mm in diameter. Incidental detection of VMCs by autopsy is difficult. Detection of VMCs by imaging is also difficult because of their asymptomatic nature and small size and also rarity. Moreover, they are easily confused with metastatic diseases of the liver, especially in imaging .

A 39-year-old man presented to our hospital with a 6-month history of recurrent nonspecific abdominal pain. Abdominal ultrasonography (US) revealed multiple cystic lesions in the liver. The diagnosis of metastases was suggested. However, final diagnosis of VWCs was confirmed by magnetic resonance imaging and magnetic resonance cholangiopancreatography .This case report highlights the routine differential diagnosisof biliary microhamartomas by magnetic resonance imaging and magnetic resonance cholangiopancreatography.

Key words : biliary microhamartomas, magnetic resonance imaging (MRI), magnetic resonance cholangiopancreatography(MRCP)

Introduction

Biliary hamartomas, known as von Meyenburg complexes (VMCs), are benign liver malformations. They are histologically characterized by cystic dilated bile ducts surrounded

by numerous fibrous stromal elements [1,2] measuring up to 5 mm in diameter. Incidental detection of VMCs by autopsy is difficult. Detection of VMCs by imaging is also difficult because of their asymptomatic nature and small size [3]. VMCs are also rare. Moreover, they are easily confused with metastatic diseases of the liver, especially in imaging [4].

Therefore, an understanding of the imaging traits of VMCs is needed to establish a list of differential diagnoses, which will decrease the need for methods such as biopsy or laparotomy [5]. We herein report a case of VMCs and describe the routine diagnostic magnetic resonance imaging (MRI) and magnetic resonance cholangiopancreatography (MRCP) findings of biliary microhamartomas.

Case report

A 39-year-old man presented to our hospital with a 6-month history of recurrent nonspecific abdominal pain. Physical examination findings were unremarkable. Laboratory examination results were normal with the exception of a slight elevation of gamma-glutamyl transferase (142 mg/dL; reference range, 0–55 mg/dL). His mother has history of biliary hamartoma Patient has no alarm symptoms . Abdominal ultrasonography (US) revealed multiple cystic lesions in the liver that appeared similar to metastases. Subsequent MRI showed multiple small cysts that were hypointense on T1-weighted images (Fig. 1a, b) and hyperintense on T2-weighted images; they were scattered in the liver parenchyma (Fig. 2a, b). MRCP showed small cysts distributed uniformly within the contour of the liver, creating a “starry sky” configuration (Fig. 3a, b).

The patient was diagnosed with multiple VMCs based on the typical MRI features. Verification using these imaging techniques within the 6-month follow-up confirmed the diagnosis of VMCs.

Discussion

A VMC is a benign congenital malformation of the biliary duct. It was first defined in 1918 by von Meyenburg [6]. They originate from embriyonic bile ducts that fail to involute .Biliary hamataromas are rare , clinically asymptomatic ,and diagnosis is usually incidental.Techical advances in radiologyhave made them easily detectable ,providing more accuracy rate diagnosis to avoid biopsy, which should be performed for confirmation of diagnosis when ,in doubt [7).

Although jaundice and portal hypertension may be caused by a mass effect, patients are usually asymptomatic [8]. VMCs may be single or multiple, with sizes ranging from 1 to 15 mm [1]. Because of the small size of the lesions, an ultimate description is difficult to attain.

The prevalence of VMCs by autopsy ranges from 0.6% to 2.8% [9]. Histologically, the lesions include disorganized and dilated bile ducts and ductules surrounded by fibrous stroma [10]. US imaging shows hypoechoic, hyperechoic, or mixed heterogenic echoic structures [1,3,4]. The multiple comet-tail sign is considered to be a specific US finding of VMCs [3]. Additionally, lesional echogenicity might be related to the number and size of dilated bile ducts and the degree of fibrosis [10]. In contrast, enhanced computed tomography shows that VMCs are usually of low attenuation with irregular margins. Most reported cases have suggested that VMCs do not demonstrate contrast enhancement [3,10]. On MRI, VMCs are

defined as hypointense on T1 and hyperintense on T2 compared to the surrounding liver parenchyma [1,10].MRCP can alsıo help the differantion of VMCs from liver metastases,polycysticdiseaseand Caroli Diseasae, requiring the admistration of intravenous gadolinium.Contrast enhancement is seen metyastatic lesions and Caroli Disease , and lack of communication the biliary tree can be observed in the later [11,12]

Although VMCs are benign, some reports have described hepatic malignancies with a background of VMCs, including hepatocellular carcinoma and cholangiocarcinoma [13]. VMCs are rare and usually only seen as multiple small nodules. They are sometimes confused with metastatic liver disease, microabscesses, diffuse primary hepatocellular carcinoma, biliary cysts, or Caroli disease [1,6,9].When it is diagnosed patients require monitoring because malignant transformation to hepatic cholangiocarcinoma(The use of Ca 19-9 to diagnose malignant transformation should be discouraged , since persistent elevation of this tumor marker has been described with multıple biliary hamartoma without malignancy (Our patient has no alarm symptoms and no elevation of Ca 19-9 [14,15) In case of alarm symptoms or elevation of tumor marker , perform MRCP. If a suspicious lesion is found consider biopsy ın our case .

There was no significant lesion and elevation of tumor marker after 6 months of follow-up.

Conclusion

The use of various imaging modalities with follow-up has proven helpful for the diagnosis of VMCs. A correct diagnosis is easier to reach when typical imaging findings are present. Otherwise, histological verification may be needed.

Consent Disclaimer:

As per international standard or university standard, patient's consent has been collected and preserved by the author.

References

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Figure1A: T2-weighted three-dimensional magnetic resonance cholangiopancreatography images (coronal plane). Multiple hyperintense cysts with scattered placement are observed in the liver parenchyma, the largest diameter reaching about 2 cm. No significant association between the cysts and biliary ducts is present.

Figure 1b: T2-weighted three-dimensional magnetic resonance cholangiopancreatography images (coronal plane). Multiple hyperintense cysts with scattered placement are observed in

the liver parenchyma, the largest diameter reaching about 2 cm. No significant association between the cysts and biliary ducts is present.

Figure2a :T1-weighted contrast-enhanced axial fat-suppressed sequences. (a, b) Multiple hypointense cysts, the largest of which is 2 cm in diameter, are observed in the liver parenchyma without contrast enhancement.

Figure 2b :T1-weighted contrast-enhanced axial fat-suppressed sequences. (a, b) Multiple hypointense cysts, the largest of which is 2 cm in diameter, are observed in the liver parenchyma without contrast enhancement.

Figure 3a :Multiple hyperintense cysts in the liver parenchyma. (a) Coronal-plane T2-weighted sequence, (b) axial fat-suppressed T2-weighted sequence

Figure 3b: Multiple hyperintense cysts in the liver parenchyma. (a) Coronal-plane T2-weighted sequence, (b) axial fat-suppressed T2-weighted sequence.

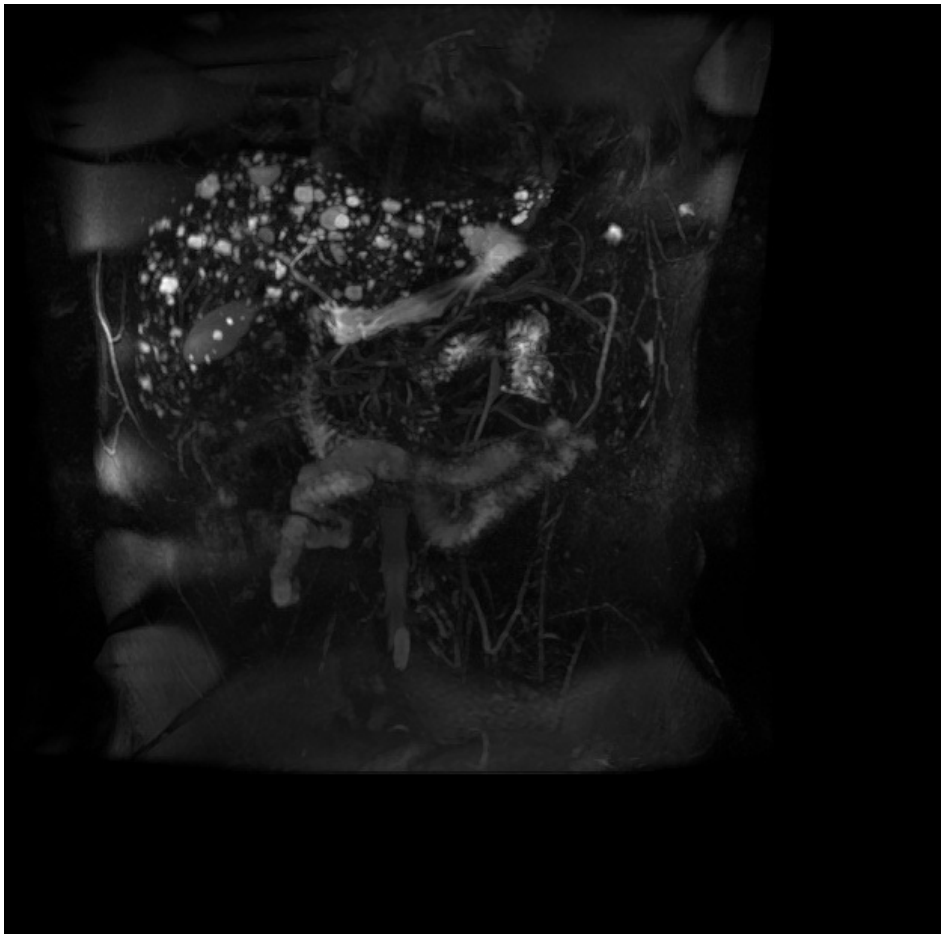

Figure1A

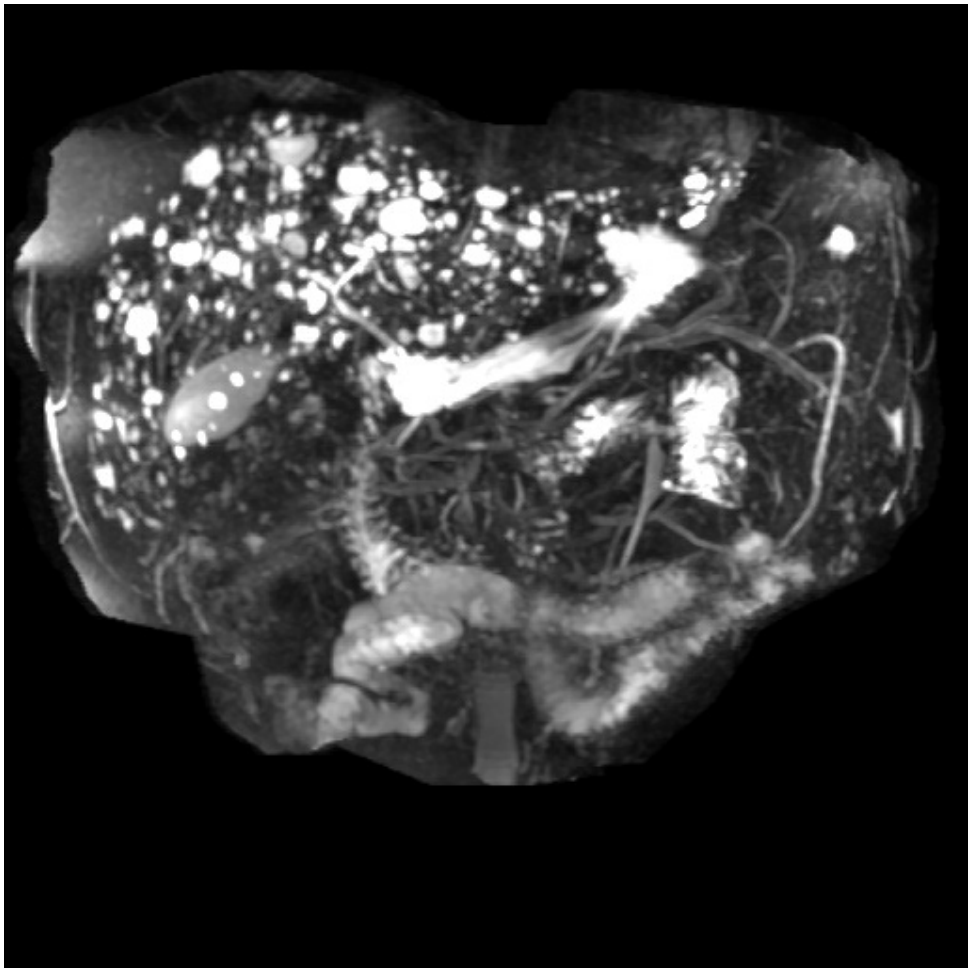

Figure 1b

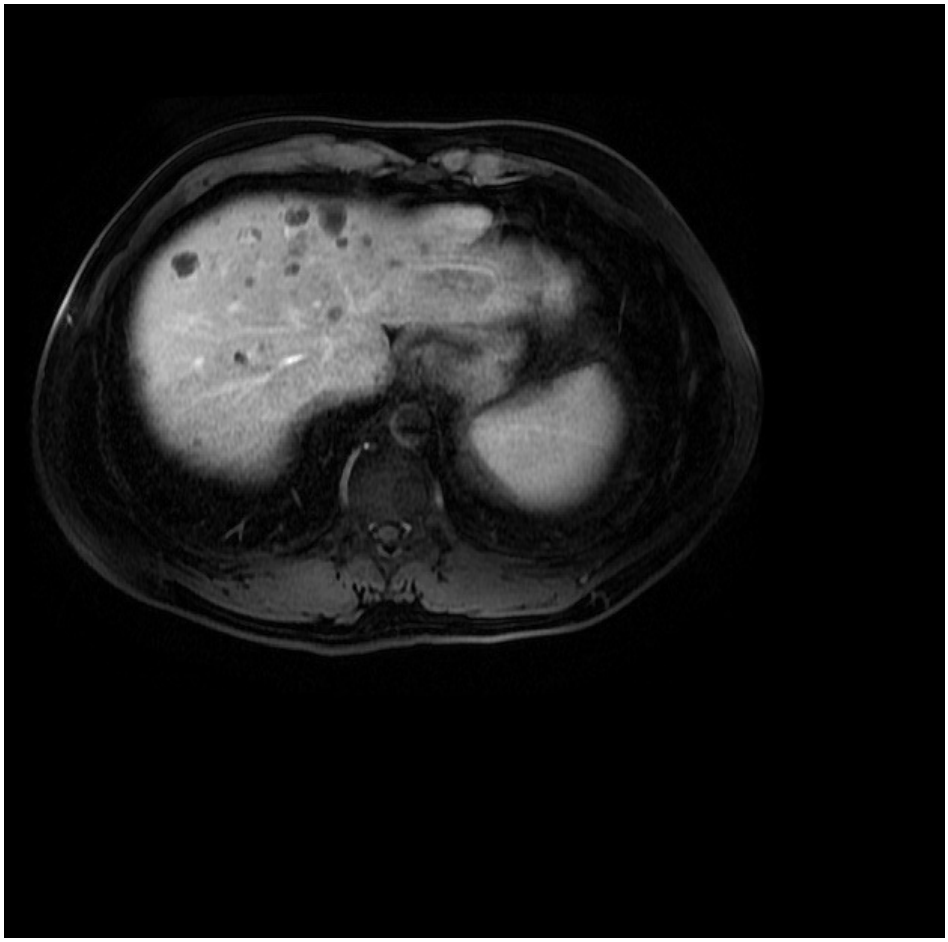

Figure2a

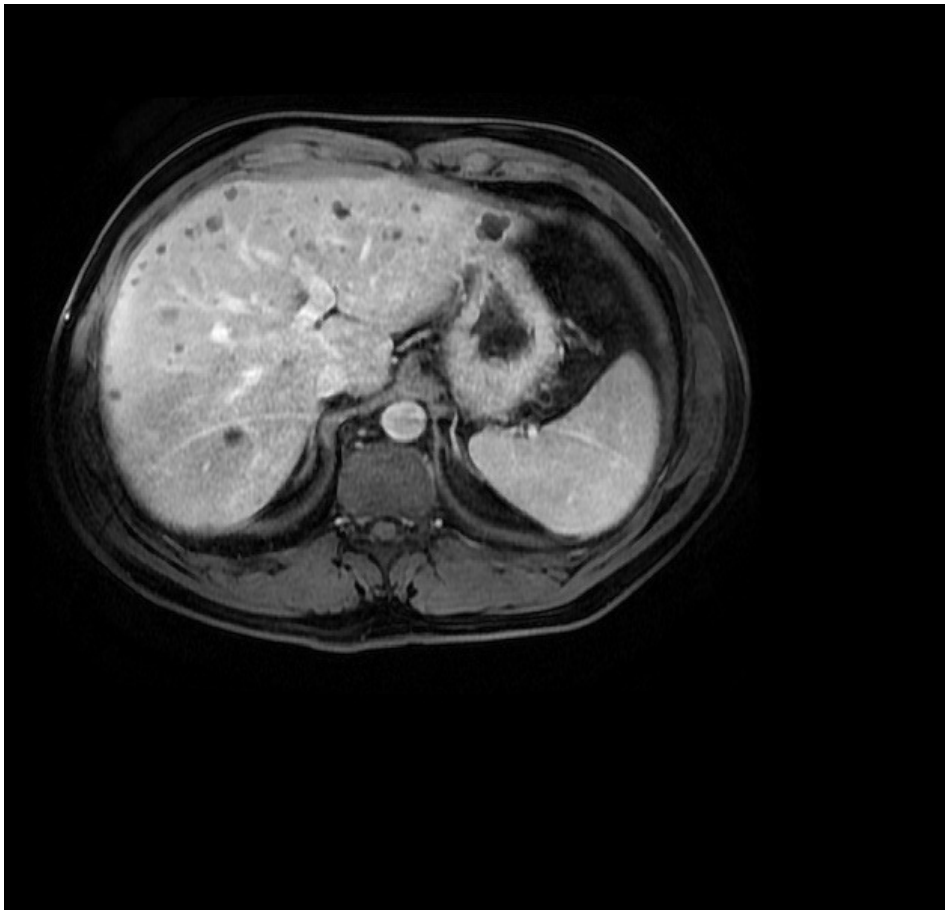

Figure 2b

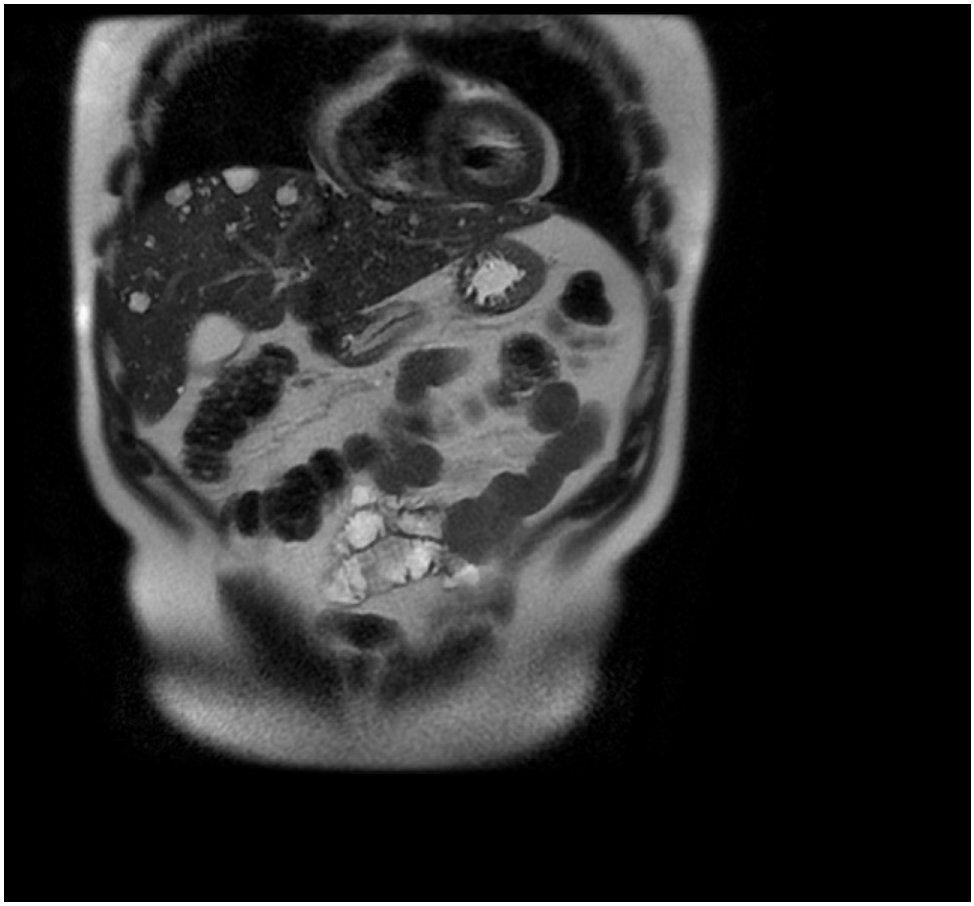

Figure 3a

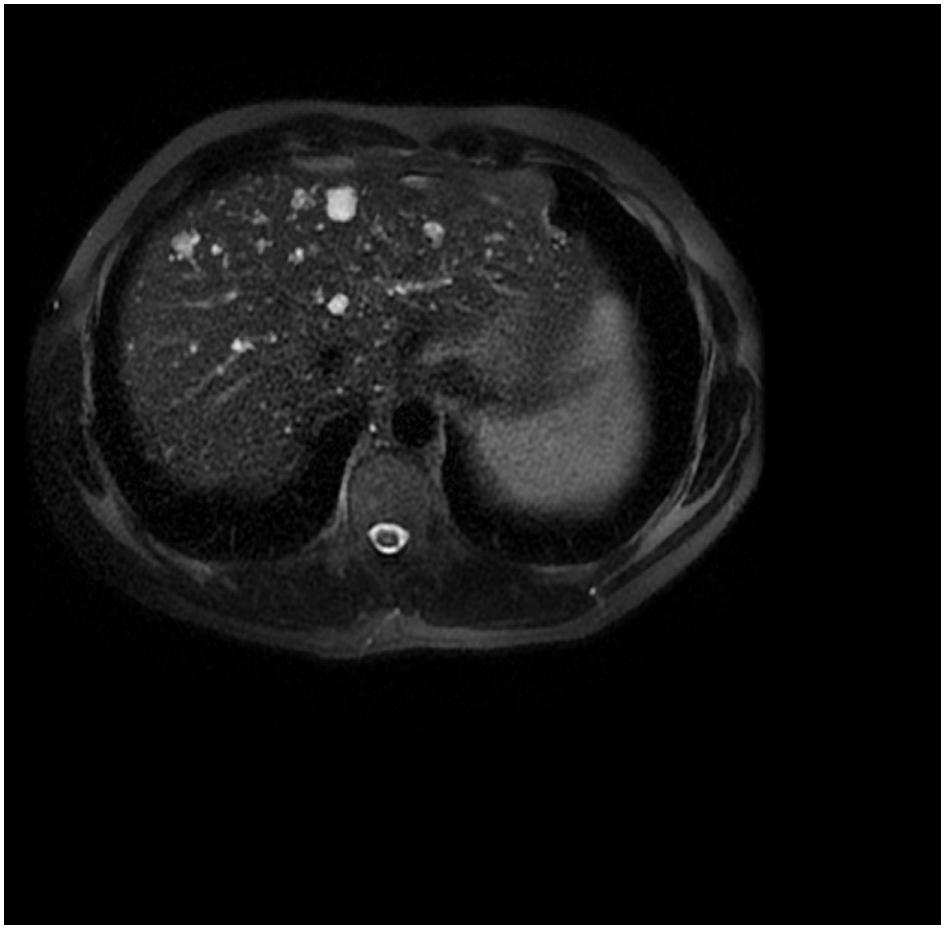

Figure 3b