Case study

Intrahepatic multicystic/ biliary hamartomas: 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 the rarity. Moreover, they are easily confused with metastatic diseases of the liver, especially on 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, the final diagnosis of VMCs was confirmed by magnetic resonance imaging and magnetic resonance cholangiopancreatography.

This case report highlights the routine differential diagnosis of biliary hamartomas by magnetic resonance imaging and magnetic resonance cholangiopancreatography.

Key words : biliary hamartomas, 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 measuring up to 5 mm in diameter [1,2]. 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 lesions of the liver, especially on 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 hamartomas.

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).Tumor markers were normal. His mother has a history of biliary hamartomas. Patient has no alarm symptoms and has no weight loss Body mass index was normal .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.

After 6 months of follow-up, the lesions remained stable.

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 embryonic bile ducts that fail to involute . VMCs are ductal plate malformations. Ductal plate malformations include different polycystic liver and kidney diseases, Caroli disease and Caroli syndrome, congenital hepatic fibrosis, and biliary atresia. VMCs may be isolated or associated with one or several of these malformations.Biliary hamataromas are rare , clinically asymptomatic ,and diagnosis is usually incidental. Technical advances in radiology have made them easily detectable ,providing more accuracy rate diagnosis to avoid biopsy, which should be performed for confirmation of diagnosis when ,in doubt [7]. Von Meyenburg complexes are one of the polycystic liver diseases, characterized by bile duct hamartomas. These cysts come from the biliary tract but the cysts do not communicate with them. Because of asymptomatic course, the lesions usually are confirmed in the course of diagnosis for another reason. It is not possible to define the entire diagnosis based ultrasonography imaging, as cyst could mimic metastasis, micro-abscesses and multiple focal nodular lesions. Because of the small size of the lesions (0.5-15 mm), computed tomography-may be also inconclusive .On the basis of magnetic resonance imaging (MRI) and cholangio-MRI we can determine the diagnosis of the complexes. Liver biopsy is obligatory in case of suspicion of a neoplastic process. These complexes do not require treatment, but a long-term follow-up is indicated because of the risk of cholangiocarcinoma development in a patient with von Meyenburg complexes.Although jaundice and portal hypertension may be caused by a mass effect, patients are usually asymptomatic [8].

The prevalence of VMCs on 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 signs are 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]. Sonographic findings of VMCs vary and are not very specific. Liver parenchymal echotexture often appears heterogeneous and coarse. VMCs appear as multiple micro-nodules, either hypo- or hyperechoic These micronodules are often very tiny and may show comet-tail artifacts, which explains why they are difficult to differentiate from aerobilia and from intrahepatic stones [6,9,12]. Variations in imaging findings may be explained by the difference in number and size of the dilated bile duct (hypoechoic lesions), and by the different density of the fibrous tissue surrounding them (hyperechoic) This explains why on sonography VMCs can be confused with liver metastases, micro-abscesses, biliary stones or fibrosis[5]

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]. They are difficult to characterize due to their small size, often below the centimeter. It is impossible to exclude the possibility that the lesions are small metastases, in particular in a patient with known primary neoplasm [13].On MRI, VMCs are defined as hypointense on T1 and hyperintense on T2 compared to the surrounding liver parenchyma [1,10]. VMCs are often irregular in shape with well-defined margins. On diffusion-weighted MRI, they mimic cystic lesions. On heavily T2-weighted sequences, the contrast with liver parenchyma is more marked, and the signal intensity is identical to that of the cerebrospinal fluid [9,12]. Because of a high contrast resolution, MR cholangiography reveals more VMCs and highlights those that are smaller [12,15]. MR cholangiography also makes it possible to see if there is any communication between VMCs and the biliary tree. Intra and extrahepatic bile ducts look normal [6,14]. On T1-weighted MR images obtained after intravenous administration of gadolinium chelate, VMCs may display different patterns. They can show no enhancement [6,9] or display a thin, regular rim of enhancement on early dynamic images that persist on late images . This enhancement correlates with compressed liver parenchyma that surrounds the lesions [5]. Finally, in a recent study, a small enhancing mural nodule was observed in 9/11 patients, correlating at histopathologic examination with polypoid projection [14]. VMCs do not communicate with the intrahepatic bile ducts. The administration of contrast medium that has biliary excretion does not result in a change of the signal inside VMCs unlike inside saccular dilatations observed in Caroli disease. To date, MRI is considered the best imaging tool to assess VMCs. MR cholangiography sequences and, more generally, heavily T2-weighted sequences are essential for differential diagnosis

MRCP can also help the differentiation of VMCs from liver metastases,polycystic disease and Caroli disease, requiring the admistration of intravenous gadolinium.Contrast enhancement is seen metastatic lesions and Caroli Disease , and lack of communication the biliary tree can be observed in the later [16]

Although VMCs are benign, some reports have described hepatic malignancies with a background of VMCs, including hepatocellular carcinoma and cholangiocarcinoma [17]. 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 they are diagnosed, patients require monitoring because of the risk of 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 multiple biliary hamartomas without malignancy[18,19]. In case of alarm symptoms or elevation of the tumor marker , perform MRCP. If a suspicious lesion is found consider a biopsy.

Conclusion

VMCs are not so rare imaging findings in everyday practice and are easily recognizable and differentiated from other intrahepatic conditions by MRI and MR cholangiography. Once diagnosed, they may be present in more complex pathologies and have a potential for malignant transformation.VMCs could easily be considered as minor malformations. Although it is impossible to perform genetic screening for diffuse VMCs or regularly monitor patients with VMCs.

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

Consent:

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

Ethical approval: NA

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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.

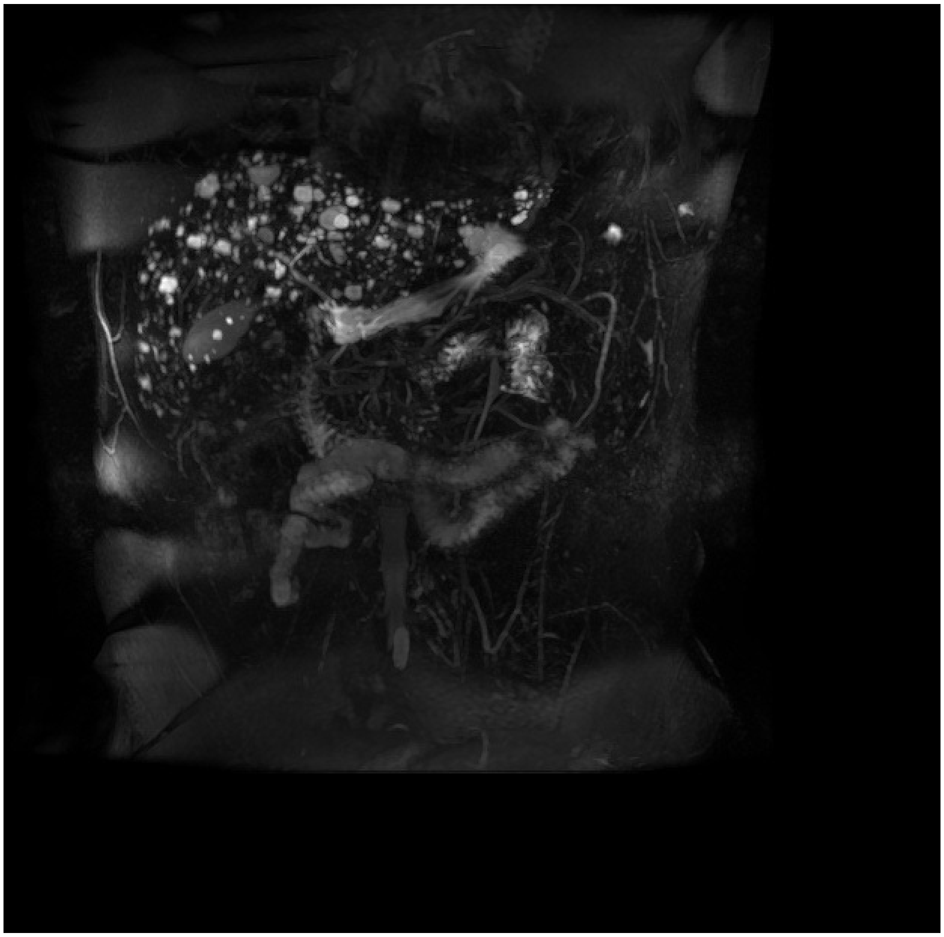

Figure1A

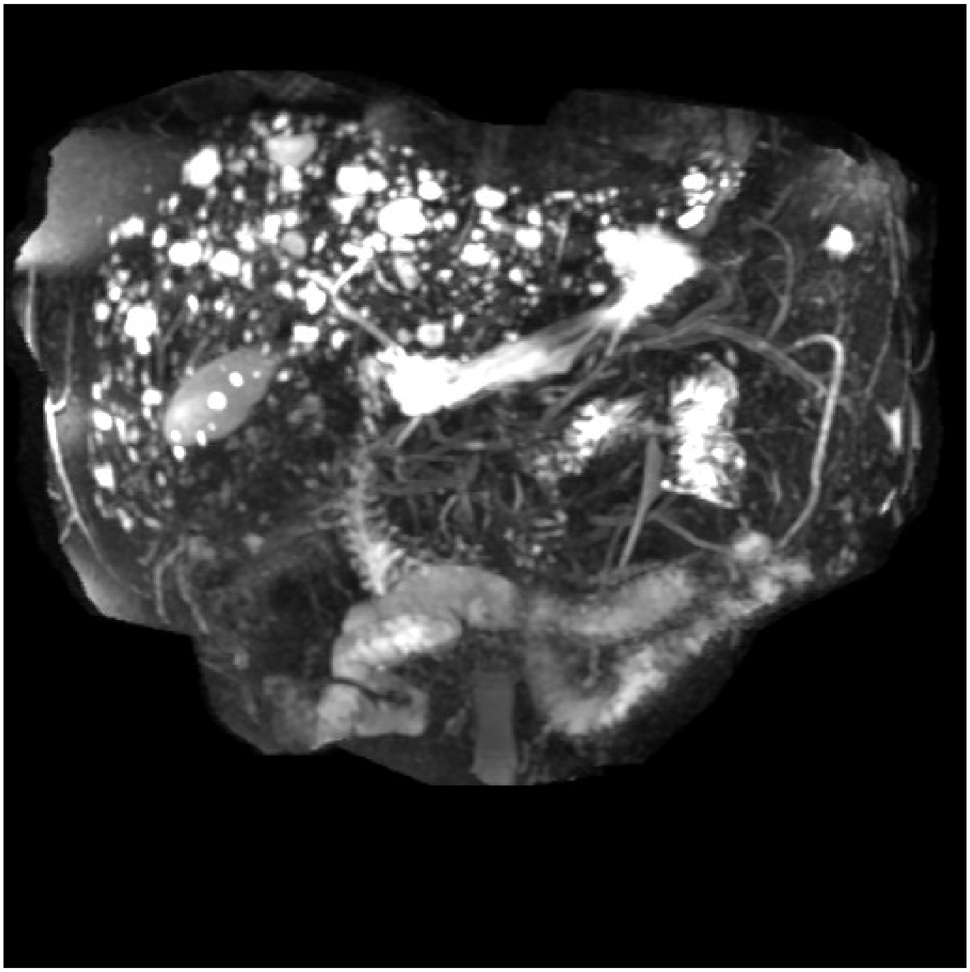

Figure 1b

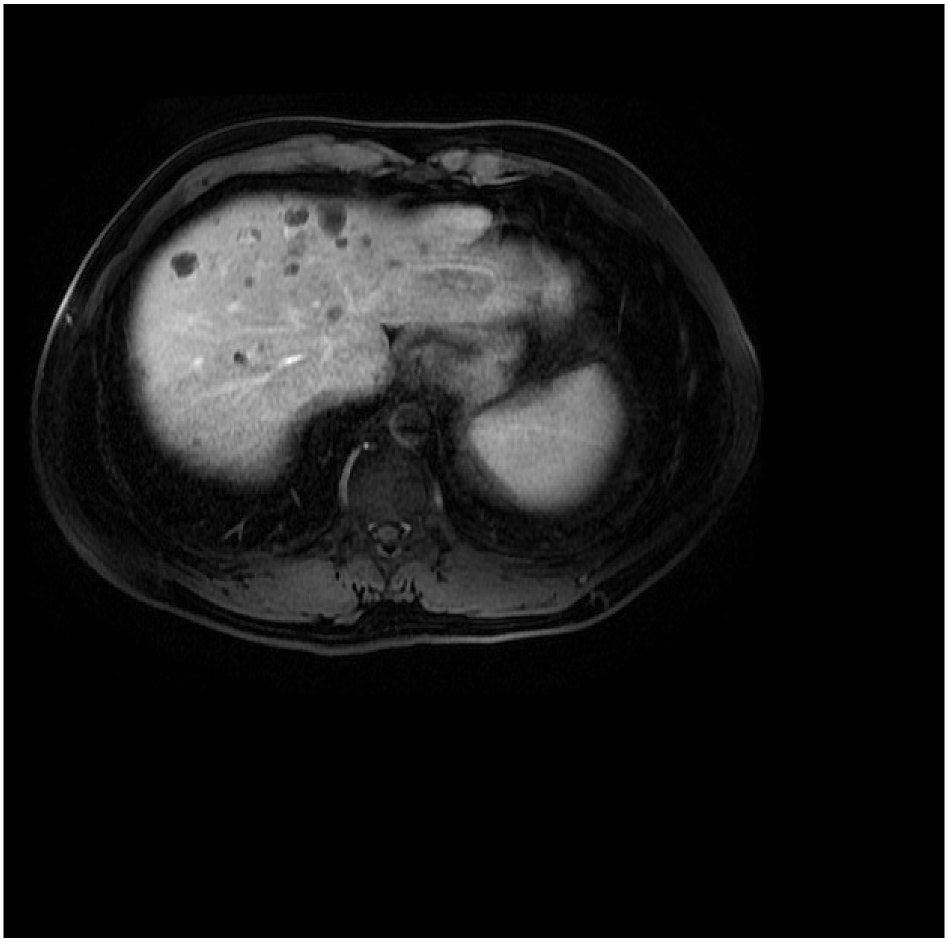

Figure2a

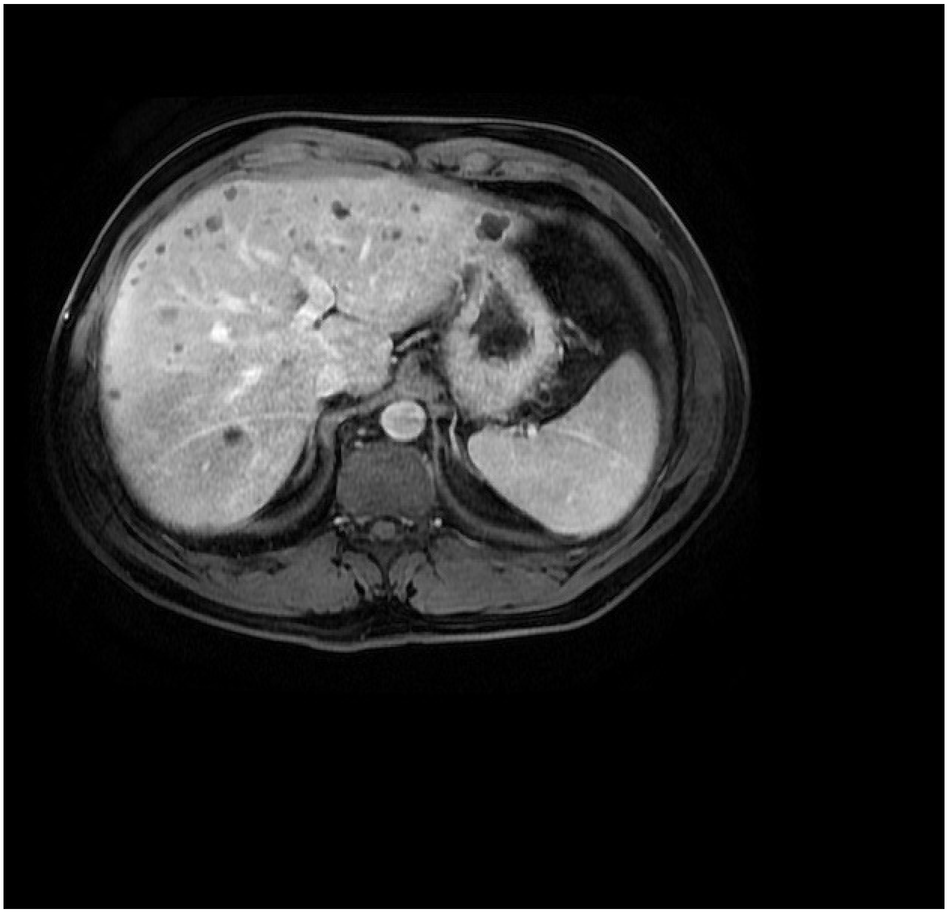

Figure 2b

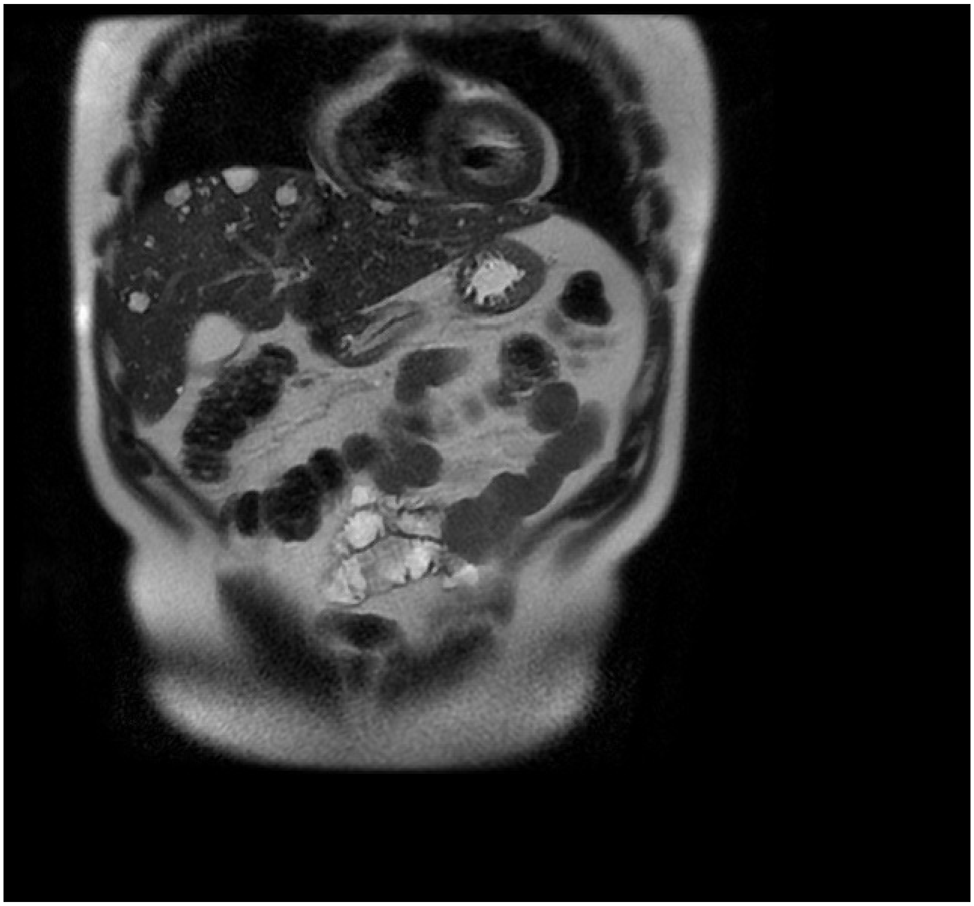

Figure 3a

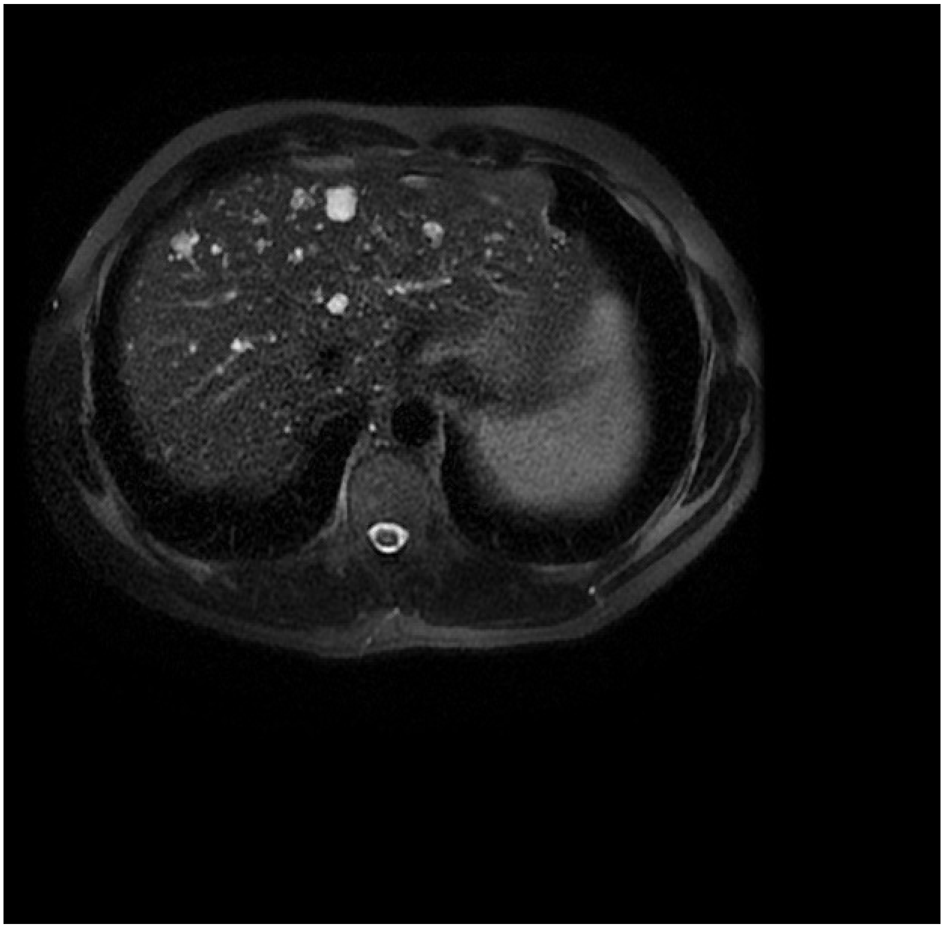

Figure 3b