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Von Meyenburg Complexes Associated with Polycystic Kidney Disease and The Endocystic Mural Nodule Sign: A Case Report

Polikistik Böbrek Hastalığının Eşlik Ettiği Endokistik Mural Nodül Bulgusu Olan Von Meyenburg Kompleksi: Olgu Sunumu

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Abstract

Von Meyenburg complexes (VMCs) are one of the rare, congenital, fibropolycystic benign diseases of the liver. We report a 40-year-old woman with characteristic imaging findings for diagnosis of VMCs associated with polycystic kidney disease. Multiple micro-nodular lesions, many of which were cystic, were detected on ultrasound (US). There were echogenic mural nodules and comet tail artifacts in their internal structures. In magnetic resonance imaging (MRI), the mural nodular enhancement and starry sky appearance were observed. The presence of endocystic mural nodules in both US and MRI, as well as the starry sky appearance that shows VMCs, are very important findings for the diagnosis of this condition.

Keywords: Von Meyenburg complex, Bile duct, Polycystic kidney, Endocystic nodule, Starry sky appearance.

Özet

Von Meyenburg kompleksleri (VMC'ler), karaciğerin nadir görülen doğuştan, fibropolikistik iyi huylu hastalıklarından biridir. Polikistik böbrek hastalığı ile ilişkili VMC'lerin tanısı için karakteristik görüntüleme bulguları olan 40 yaşında bir kadın olguyu sunuyoruz. Ultrasonografik (US) görüntülemede çoğu kistik yapıda olan çok sayıda mikronodüler lezyon tespit edildi. İç yapılarında ekojenik mural nodüller ve kuyruklu yıldız artefaktları vardı. Manyetik rezonans görüntülemede (MRG), mural nodüler kontrastlanma ve yıldızlı gökyüzü görünümü gözlemlendi. Hem US hem de MRG'de endokistik mural nodüllerin varlığı ve VMC'leri gösteren yıldızlı gökyüzü görünümü bu durumun tanısı için çok önemli bulgulardır.

Anahtar Kelimeler: Von Meyenburg kompleksi, Safra kanalı, Polikistik böbrek, Endokistik nodül, Yıldızlı gökyüzü görünümü.

Introduction

Von Meyenburg complexes (VMCs), also known as multiple biliary hamartomas, are defined as bile duct plate malformations. VMCs may be isolated or they may be accompanied by other ductal plate malformations, such as Caroli's disease/syndrome, congenital hepatic fibrosis, and polycystic liver and kidney disease. They are usually asymptomatic and detected in autopsy series during laparotomy or by routine imaging

methods. Magnetic Resonance Imaging (MRI) findings are sufficient for the diagnosis of this condition [1,2]. In this paper, we present the imaging features of VMCs associated with polycystic kidney disease using ultrasound (US) and MRI.

Case Report

A 40-year-old female presenting with abdominal pain lasting for a few weeks was referred to our department of radiology for an ultrasound (US). Her physical examination revealed no pathological features. Laboratory examinations showed liver enzymes, alkaline phosphatase, gamma glutamyl transferase, bilirubin and tumor marker values within the normal range. There was no family history of liver or bile duct malformation.

Imaging Findings

The abdominal US was performed using a MyLab™X7 scanner and a 2-5 MHz convex transducer (Esaote, Genova, Italy). There were hyperechoic and hypoechoic multiple lesions located in all segments of the liver, with dimensions below 13 mm. Comet tail artifacts and an endocystic mural nodule were detected in the lesions (Figure 1A, B). The abdominal MRI was undertaken using an Optima 1.5T scanner (General Electric, United States). The MRI pulse sequences included axial T2-weighted fast spin echo [repetition time (TR) = 1900, echo time (TE) = 84 msec; ETL=15; field of view = 340 mm; matrix size = 256 x 256] and fat-saturated axial T1-weighted three-dimensional spoiled gradient echo (3D SPGRE)-breath hold (TR = 3.8 and TE = 1.8 msec; flip angle = 12°; matrix size = 256 x 256) MRI images before contrast and 30, 60 and 90 seconds and three minutes after the intravenous administration of 0.2 mL/kg of a gadolinium-based contrast material (Gadobutrol, Gadovist®, Bayer). In addition, the patient was undergoing magnetic resonance cholangio-pancreatography (MRCP) using thick-slab fast spin echo breath-hold (TR/TE = 2900/1260 msec) and three-dimensional sequences.

On the axial T2-weighted images, there were multiple well-circumscribed hyperintense subcapsular lesions along the periphery of both lobes of the liver. The measurement of the lesions ranged from 1 to 13 mm (Figure 1C). The MRCP of the abdomen also showed multiple hyperintense lesions of 1-13 mm diameters that were not connected to each other. There was no dilatation of the intrahepatic and extrahepatic bile ducts or connection between the lesion and the bile duct. These lesions had a starry sky appearance, which is a characteristic MRCP finding for the diagnosis of VMCs (Figure 1D). The pre-contrast T1-weighted image showed multiple low-signal lesions, some of which had the endocystic polypoid projection of fibrous stroma located in the periphery of the lesion (Figure 2A). The post-contrast T1 weighted images revealed rim-like contrast enhancement in the lesions, and there was contrast filling in the portal vein adjacent to the lesion. Moreover, the gadolinium-enhanced T1-weighted image demonstrated the enhancement of a mural nodule (Figure 2B, C). Based on the characteristic imaging findings, the patient was diagnosed with VMCs. In addition, microcysts were found in both kidneys on post-contrast T1 weighted images, indicating that VMCs were associated with polycystic kidney disease (Figure 2D).

Discussion

VMCs, first described in 1918, are rare benign liver malformations that have been associated with other ductal plate malformations, including Caroli's disease, congenital hepatic fibrosis, and polycystic liver and kidney disease [1,3]. Sasaki et al.[4] found MUC1 apomucin, an oncofetal antigen, in the biliary epithelial cells of VMCs and polycystic kidney disease, and they noted that this antigen was not present in normal livers. Furthermore, mutations in the polycystic kidney and liver disease gene 1 (PKDH-1) were reported to cause polycystic kidney disease, which is a kind of ductal plate malformation. However, these mutations have also been identified in VMCs [5]. In our case, since VMCs and polycystic kidney disease occurred together, it was concluded that there was a strong relationship between these two diseases.

VMCs are histopathologically well-described disease and their imaging findings have been described in recent case reports. MRI is

considered to be the best method for the diagnosis of VMCs [3].

Typically, using MRI, there is hypointensity on the T1-weighted image and significant hyperintensity on the T2-weighted image. The fluid in the biliary duct has been reported to have a high signal while fibrous stroma in its periphery has a low signal [3]. In our case, in addition to these findings, we detected a mural nodule in the lesion with a polypoid projection. The gadolinium-enhanced T1-weighted image showed the enhancement of the mural nodule and also revealed the peripheral rim-like contrast of other lesions. Tohmé-Noun et al.[3] reported that in 10 of 11 (91%) cases, mural nodular contrast was due to the endocystic polypoid projection of fibrous stroma. The authors found that this mural nodule was also isointense on the pre-contrast T1 weighted image and had intermediate signal intensity on the T2-weighted image. Furthermore, after contrast injection, they noted an endocystic enhancement of the mural nodule in most cases (91%). Similarly, in our case, an isointense mural nodule was detected on the T1-weighted image, which showed contrast enhancement. In MRCP, these lesions were diffuse with a starry sky appearance and were not connected to bile ducts [6].

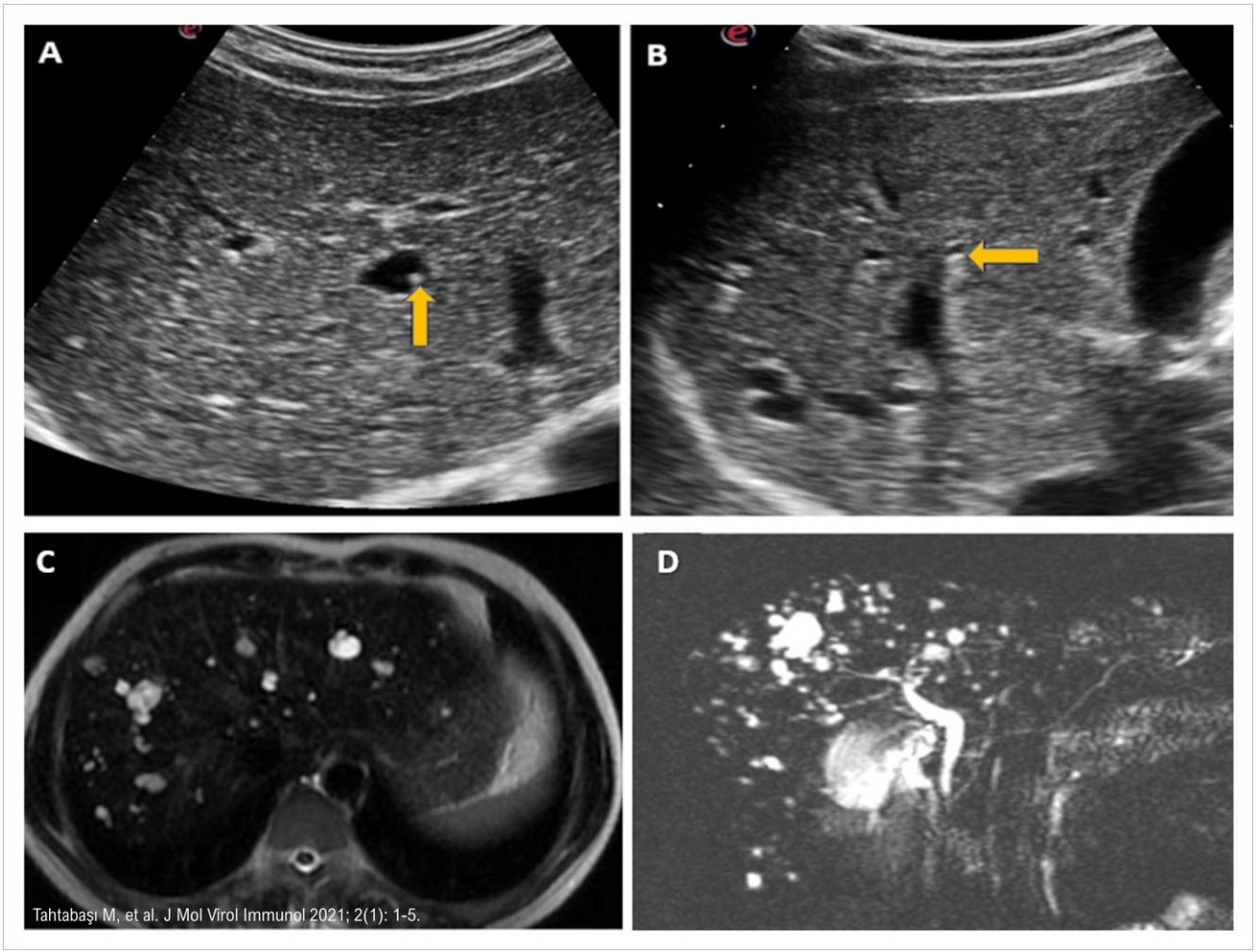

Figure 1. (**A**) The abdominal US showing the hyperechoic mural nodule (arrow), and (**B**) echogenic foci with large posterior comet-tail artifacts (arrow), (**C**) Axial T2-weighted and (**D**) three-dimensional MRCP maximum intensity projection image showing a starry sky appearance of well-circumscribed, multiple hyperintense lesions.

The US appearance of VMCs is not very specific and these malformations mostly present as hyperechoic or hypoechoic micronodular lesions in the liver. Within these micronodules, comet-tail artifacts may also be seen but it is not easy to distinguish from an intrahepatic stone or

aerobilia [1,7]. In the current case, we observed the typical comet tail artifacts and a mural nodule in the lesions. Recent studies have shown that VMCs are at risk of malignant transformation and is a precursor to colangiocarcinoma [1,3,8]. VMCs are benign liver malformations which might imitate microabscesses, liver cysts, peribiliary cysts, dilated bile ducts, and biliary adenoma [3,6]. It should also not be confused with the metastasis of known primary malignant lesions. Nevertheless, the presence of multiple lesions, their predominantly small size, and most importantly, the presence of tiny mural nodules with nodular enhancement on MRI imaging are characteristic findings that markedly increase the accuracy of diagnosis [3].

In conclusion, VMCs are rare and mostly detected incidentally. They can be diagnosed by imaging methods, such as MRI and US. The characteristic MRI and MRCP findings are essential to distinguish VMCs from other cystic liver diseases. In suspected cases of VMCs, clinicians should also carefully investigate the presence of other accompanying ductal plate malformations.

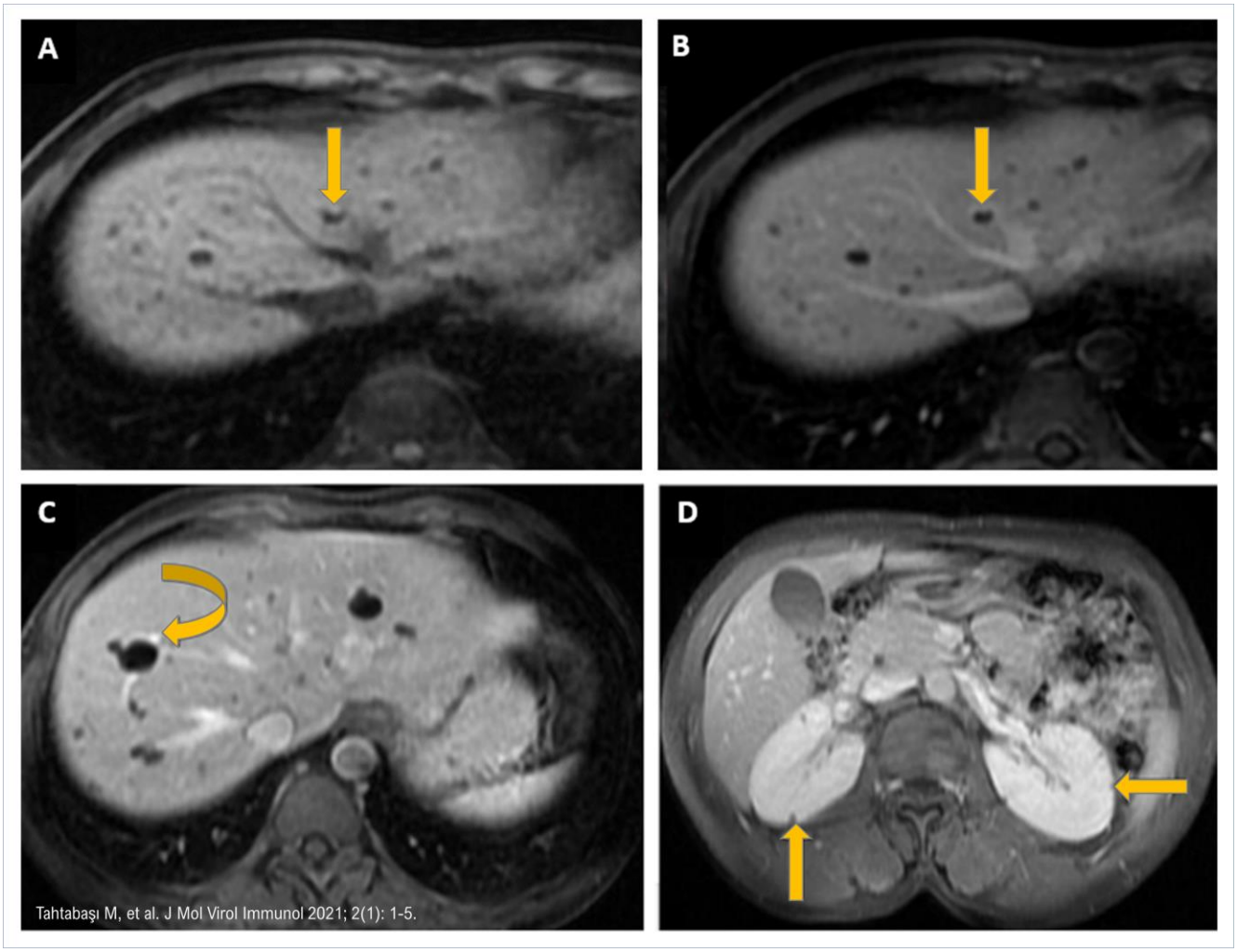

Figure 2. (**A**) T1-weighted MRI image showing multiple low-signal lesions in the liver and the endocystic polypoid projection of fibrous stroma (arrow), (**B**) gadolinium-enhanced T1-weighted image demonstrating the enhancement of the mural nodule and peripheral rim-like enhancement (arrow). (**C**) Post-gadolinium T1-weighted image revealing peripheral enhancement of the lesions with contrast filling in the portal vein adjacent to the lesions (curved arrow), and (**D**) several microcysts (arrows) seen in the cortex of both kidneys.

Abbreviations: VMCs: Von Meyenburg complexes (VMCs), MRI: Magnetic resonance imaging, US: Ultrasound, TR: Time to repetition, TE: Time to echo, ETL: Echo train length, 3D SPGRE: Three-dimensional spoiled gradient echo, MRCP: Magnetic resonance cholangiopancreatography, PKDH-1: Polycystic kidney and liver disease gene 1.

Authors' contributions: MT and SE wrote and revised the manuscript. EK and EC revised the manuscript and is the article guarantor. MT, SE and EC revised the manuscript. EK provided scientific support and valuable advice. All authors proofread the manuscript and revised it critically. All authors read and approved the final manuscript. **Consent for publication:** Informed consent was obtained for this case report. **Conflict of interest:** The authors declare that there is no conflict of interest. The authors alone are responsible for the content and writing of the paper. **Financial disclosure:** There is no financial support to this study.

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