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Conclusion: Von Meyenburg complexes can have atypical presentation and have been associated with hepatobiliary malignancy.

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CASE REPORT

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Keywords: Adult polycystic kidney disease, Cholangiocarcinoma, Multi-nodular cystic lesions, Von Meyenburg complexes

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INTRODUCTION

Von Meyenburg complexes (VMCs) are uncommon, benign cystic dilatations of the intrahepatic bile ducts surrounded by fibrous stroma [1, 2]. Generally, the size of VMCs is less than 15 mm and they are formed due to ductal plate malformations that occur during embryonic development [3]. The diagnosis of such lesions is usually incidental as patients are usually asymptomatic and have normal liver function. We present a case where VMCs were incidentally diagnosed when evaluating renal cysts in a patient.

CASE REPORT

A 74-year-old African-American male presented with difficulty urinating and abdominal pain. During that admission, he was diagnosed with acute exacerbation of chronic kidney injury and renal ultrasound showed renal cysts up to 3.2 cm on the right and 4.1 cm on the left

kidney. Magnetic resonance imaging scan of abdomen with contrast showed innumerable bilateral renal cysts consistent with polycystic renal disease while the images of the liver revealed multiple areas of heterogeneous T2 hyperintensity with diffusion abnormalities.

Subsequently, an MRI triple phase contrast imaging of the liver reported wedge shaped perfusion abnormalities with corresponding hypervascular areas (Figure 1A–B). There was also central linear branching areas of non-enhancement on the portal venous phase of the imaging which were concerning for thrombosis of the small portal vein branches or an infiltrative disease. The patient's liver enzyme levels were normal. Further hypercoagulable and liver serological workup was negative. After consultation between physicians and the patient, it was decided that the bleeding risk associated with anticoagulation was high so such treatment was not given. His renal function recovered to baseline with medical management and he was discharged with plan of outpatient nephrology and gastroenterology (GI) evaluation and management.

The patient was seen in GI clinic where a follow-up CT scan triple phase contrast imaging of the liver (Figure 2) showed patchy, heterogeneous enhancement throughout the arterial phase but no diagnosis was reached. Subsequently, the patient underwent liver biopsy which showed grade 2 mild to moderate inflammatory activity and stage 2 peri-portal fibrosis. Foci of bile duct proliferation within collagenous fibrosis were seen which were consistent with bile duct hamartoma (Von Meyenburg complex) (Figure 3A–C). The patient's liver enzymes remained normal. Subsequent imaging of liver did not show any new pathology. Patient continues to be monitored with ultrasound of liver, liver function laboratory tests as well as follow-up in GI clinic every six months.

DISCUSSION

Von Meyenburg complexes (VMCs), or bile duct hamartomas are benign liver lesion that have a prevalence of 0.69–2.8% in the population [4]. VMCs are associated with autosomal dominant polycystic kidney disease (ADPKD), Caroli disease and congenital hepatic fibrosis [5–7]. Our patient was diagnosed with ADPKD which has a prevalence 11% in adults with at least one VMC and 40% in those with four or more such lesions in a large study involving autopsies [5]. The same study found the overall prevalence of VMCs in livers on autopsy to be 5.6%.

Von Meyenburg complexes are generally asymptomatic and do not cause liver enzyme abnormalities, although there have been cases where patients have presented initially with abdominal pain [6]. Our patient was asymptomatic at presentation and diagnosis.

An MRI scan is superior to both ultrasound and CT scan to diagnose these lesions where VMCs are often seen

Figure 1: Magnetic resonance imaging scan of abdomen with and without contrast – Focal hypervascular areas described as hyper-intense on T2 imaging and bright on diffusion weighted images with persistent nonspecific diffusion abnormalities throughout the liver. (A) T2 weighted MRI image, and (B) Diffusion weighted MRI image.

Figure 2: Computed tomography scan with and without contrast (liver protocol)– Arterial phase consistent with patchy, heterogeneous enhancement throughout the liver.

Figure 3: (A–C) Liver biopsy - Grade 2 mild to moderate peri-portal fibrosis. Foci of fibrosis and bile duct proliferation are consistent with bile duct hamartomas (Von Meyenburg complex). (A) H&E stain, x100, (B) H&E stain, x400, (C) Masson's Trichrome stain, x400.

as irregular T2 hyperintense cystic nodules throughout the liver without communication of these lesions with the biliary tree [6]. Most VMCs appear with hypointense T1 and hyperintense T2 MRI imaging [2]. Our patient's MRI showed similar though non-specific characteristics of the lesions. However, some sources also note that it can be difficult to definitively diagnose VMCs by MRI as T1-weighted sequences can show both enhancement and no enhancement [6]. The presence of cystic lesions with surrounding fibrous stroma and congested interfaces may account for the variation in appearance [2].

A liver biopsy is required if lesions have atypical characteristics and remain undiagnosed on MRI scan [6]. Since there was concern for an infiltrative process, liver biopsy was performed and definitively diagnosed VMCs in our patient.

There have been reports of development of cholangiocarcinoma and hepatocellular carcinoma (HCC) in patients with VMCs [6, 8, 9]. The exposure of the biliary epithelium to chronic cholestasis is thought to lead towards neoplastic transformation. Although there are currently no guidelines for follow-up of patient with VMCs, such patients could be followed per guidelines for patients at risk of cholangiocarcinoma and HCC.

CONCLUSION

Von Meyenburg complexes (VMCs) are uncommon lesions of the liver. Medical imaging, particularly MRI, is usually diagnostic. Our patient had an atypical presentation of VMCs on MRI and required liver biopsy for diagnosis. Hepatobiliary malignancy has been reported in patients with such liver lesions so regular follow up including medical imaging and liver function tests could be done after VMCs have been diagnosed in a patient.

Author Contributions

Nikhil S. Kadle – Acquisition and analysis of data, Drafting the article, Final approval of the version to be published

Hammad Liaquat – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Saad P. Shaheen II – Acquisition of data, Revising it critically for important intellectual content, Final approval of the version to be published

Luis S. Marsano – Substantial contributions to conception and design, Revising it critically for important intellectual content, Final approval of the version to be published

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

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