Journal of Gastrointestinal Cancer (2019) 50:972–974
https://doi.org/10.1007/s12029-018-0151-y

CASE REPORT

Von Meyenburg Complexes Mimicking Diffuse Metastatic Liver Disease

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Published online: 6 August 2018

Introduction

Von Meyenburg complexes (VMCs), also known as multiple biliary hamartomas or biliary microhamartomas, are rare benign developmental malformations consisting of small cystic dilatations of the intrahepatic bile ducts [1]. Their reported prevalence is 0.6% and 0.9–5.6% in needle biopsy and autopsy series respectively [2, 3]. In most cases, VMCs are asymptomatic and are detected incidentally at laparotomy, during autopsy, or on imaging studies performed for unrelated conditions. Their clinical significance lies in the fact that they may mimic metastatic liver disease [4], on both imaging studies and even on gross examination.

Case Presentation

A 49-year- old male was referred to our department for elective cholecystectomy for symptomatic uncomplicated gallstone disease. His medical history and physical examination findings were unremarkable whereas routine hematological and biochemical investigations were within normal limits.

Ultrasonography that had been performed at another institution showed a normal-sized gallbladder with multiple tiny gallstones and increased liver echogenicity. During laparoscopic cholecystectomy, multiple whitish nodules were detected scattered throughout both liver lobes (Fig. 1). Since the patient was asymptomatic and the preoperative biochemical investigations were normal, the diagnosis of Von Meyenburg complexes was considered. A liver biopsy was, however, performed because a complete imaging investigation had not been performed preoperatively. Histological examination findings were suggestive of Von Meyenburg complexes consisting of multiple moderately to highly dilated bile ducts, surrounded by fibrous stroma. Many of the ducts were irregular or angulated and contained intraluminal bile (Fig. 2). The postoperative course was uneventful. The patient is asymptomatic 14 months after surgery. He is currently being followed up on a regular basis.

Discussion

VMCs were initially described by von Meyenburg in 1918 as benign liver malformations of the embryonic ductal plate [2, 5]. They are multiple white or grayish nodules measuring up to 1 cm in diameter that are scattered throughout both liver lobes, predominantly in the subcapsular and periportal regions, not communicating with the biliary tree [1, 2]. They may be associated with Caroli disease, Caroli syndrome, and polycystic liver and kidney disease [1, 3].

Histologically, VMCs are characterized by well defined, irregular, or round in shape cystically dilated bile ducts that are lined by a single layer of cuboidal cells embedded in abundant fibro-collagenous stroma [2]. Bile stained granular material may be present [1, 6].

Due to the differences in size and number of the dilated bile ducts, variable imaging findings have been reported. On ultrasonography, VMCs may appear as multiple small hyper- and hypoechoic micronodules with comet tail echoes [2], while on CT, they may appear as multiple hypoattenuating lesions with a round or irregular shape that are more clearly visible after the administration of contrast material [1].

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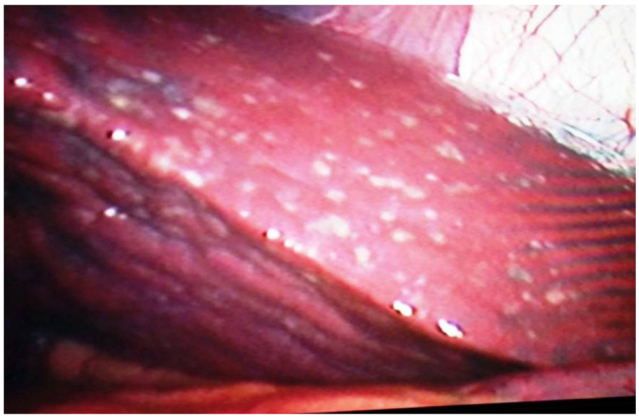

Fig. 1 Intraoperative photo of Von Meyenburg complexes. Multiple whitish nodules scattered throughout the liver surface

Magnetic resonance imaging (MRI) is the best imaging modality for the diagnosis of VMCs. They may appear as multiple lesions with low signal intensity on T1-weighted images and markedly high signal intensity on T2-weighted images. MRI can also detect the absence of communication between VMCs and biliary tree [1, 2].

The most important differential diagnoses of VMCs include liver metastases, liver microabscesses, intrahepatic stones, Caroli disease, and peribiliary cysts [1].

Liver biopsy for the definitive histological diagnosis of VMCs should be performed when the imaging findings are not characteristic, or in cases with a known history of malignancy.

VMCs have been associated with an increased risk of developing cholangiocarcinoma of the liver. Malignant transformation of VMCs to intrahepatic cholangiocarcinoma (iCC) has been reported in 35 cases in the literature so far [5]. However, this association should be further evaluated in a larger number of patients.

The histological transition from VMCs to iCC may be due to genetic progressive mutations on key oncogenes reinforcing the precancerous potential of VMCs [1].

Bhalla et al. [6] reviewed 86 hepatectomy specimens with the diagnosis of iCC and found a morphological association between iCC and VMCs in 35% of the cases. These cases exhibited a gradual neoplastic progression from VMCs to biliary dysplasia and then to iCC. The authors reported that the VMC-associated iCCs were well to moderately differentiated, with unique morphological features and relatively less aggressive with low T stage at presentation, compared to non-VMC-associated iCCs. It was finally suggested that VMCs may become neoplastic serving as an in situ carcinoma lesion to transform to iCC [6].

Follow-up examinations in patients with VMCs are necessary especially in patients with a known history of extrahepatic malignancies [2] and because of the reported increased risk of developing iCC.

In conclusion, Von Meyenburg complexes are a rare clinicopathologic entity resulting from a developmental defect of the ductal plate. Their clinical significance lies in the fact that they mimic other liver lesions particularly diffuse liver metastases. Liver biopsy is necessary to obtain a definitive diagnosis when imaging features are not characteristic. Long-term follow-up of the patients is necessary because VMCs have been postulated to progress to cholangiocarcinoma in some cases.

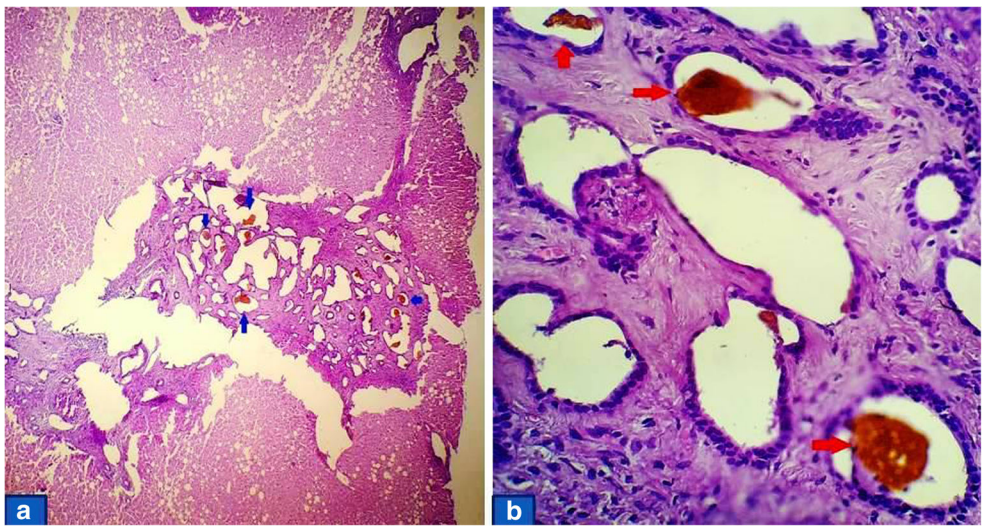

Fig. 2 Histopathological findings of VMC. **a** Low-power photomicrograph from a liver biopsy, displaying among the hepatocytes a non-encapsulated, sharply outlined cluster of moderately to highly dilated bile ducts, surrounded by fibrous stroma. Many of the ducts are irregular or angulated and contain intraluminal bile (blue arrows) (hematoxylin and eosin × 40). **b** High-power photomicrograph showing the dilated ducts, some of whom contain intraluminal bile (red arrows). Ductal epithelium is comprised of monolayered cuboidal to columnar cells, without atypia (hematoxylin and eosin × 400)

Compliance with Ethical Standards

Conflict of Interest The authors declare that they have no conflict of interest.

References

1. Pech L, Favelier S, Falcoz MT, Loffroy R, Krause D, Cercueil JP. Imaging of Von Meyenburg complexes. Diagn Interv Imaging. 2016;97:401–9.
2. Zheng RQ, Zhang B, Kudo M, Onda H, Inoue T. Imaging findings of biliary hamartomas. World J Gastroenterol. 2005;11:6354–9.
3. Redston MS, Wanless IR. The hepatic von Meyenburg complex: prevalence and association with hepatic and renal cysts among 2843 autopsies[corrected]. Mod Pathol. 1996;9:233–7. Erratum in: Mod Pathol 1996;9:803
4. Elsoueidi R, Mularz SJ, Richa EM. Bile duct hamartoma mimicking metastatic cholangiocarcinoma. J Gastrointest Cancer. 2017;48:87–8.
5. Kim HK, Jin SY. Cholangiocarcinoma arising in von Meyenburg complexes. Korean J Hepatol. 2011;17:161–4.
6. Bhalla A, Mann SA, Chen S, Cummings OW, Lin J. Histopathological evidence of neoplastic progression of von Meyenburg complex to intrahepatic cholangiocarcinoma. Hum Pathol. 2017;67:217–24.