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*Case Report*

# A CASE REPORT OF VON MEYENBURG COMPLEXES

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**ABSTRACT**

Background: The von Meyenburg complexes (VMC) are a rare clinicopathological entity. They are usually asymptomatic and are presented as multiple small-sized cystic liver lesions. Ultrasonography, CT and MRI are used for diagnosis.

Case report: A 74-years old female patient with previous oncologic surgeries was admitted to our department with clinical, laboratory and imaging findings of acute cholecystitis and cystic lesions in the liver. A laparoscopic cholecystectomy with liver biopsy was performed. Liver histology revealed bile duct hamartomas (von Meyenburg complexes).

Conclusion: VMC represent a diagnostic problem in patients with oncologic history because they are difficult to be distinguished from liver metastases.

**Key words:** von Meyenburg complexes, bile duct hamartomas, cholecystitis

## INTRODUCTION

The von Meyenburg complexes (VMC) are defined as multiple benign bile duct hamartomas. They are a rare clinical finding that is presented as multiple small-sized (<1.5 cm) cystic liver lesions which affect both lobes. (1) VMC are usually discovered incidentally and are generally asymptomatic but occasionally may be presented with non-specific abdominal symptoms such as diffuse abdominal pain and discomfort. (2) Imaging methods used for diagnosis are ultrasonography, computed tomography (CT) and MRI. (3) VMC may represent a diagnostic challenge in patients with known primary malignancy or in such that underwent oncologic surgery. (4)

Here, we present an interesting case of the von Meyenburg complexes in a patient operated for acute cholecystitis who was diagnosed with multiple liver lesions preoperatively and underwent two oncologic surgeries in the past.

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## CASE REPORT

A 74-years old female patient was admitted to the Department of General surgery of University Hospital "St. George" with complaints of right upper abdominal quadrant pain with irradiation to the right

shoulder, nausea, vomiting. Physical exam revealed no fever, slight tachycardia, cicatrices from thyroidectomy and left mastectomy, right upper abdominal tenderness with Murphy sign positive. The patient informed about arterial hypertension as comorbidity. She underwent a total thyroidectomy for thyroid cancer 10 years ago and a left mastectomy for breast cancer 3 years ago. The patient refused to have adjuvant radio- and chemotherapy after both oncologic surgeries and had no clinical or imaging follow-up for the period before the current admission. Relevant laboratory findings at the time of hospitalization were following: HGB - 157 g/l; RBC - 5.33;WBC – 13.21; PLT – 217; glucose - 6.7 mmol/l;CRP – 15 mg/l; LDH 569 U/l; t. protein – 62 g/l; PT – 77.4 %. Ultrasonography findings were hepatomegaly and tense and enlarged gall bladder with multiple stones in the lumen. CT scan revealed calculous cholecystitits, multiple cystic lesions with round and irregular shape in both liver lobes, biggest with size of 5.5/2.8 cm; several cystic lesions in both kidneys, biggest with size of 4.6/3.7 cm; **(Figure 1 A, B)** A laparoscopic cholecystectomy was performed, Mascagni's lymph node was harvested for histologic examination and a pinch biopsy from a liver lesion in IVb segment was performed. The results from the histological analysis were following: 1. Chronic hypertrophic cholecystitis with cholesterolosis; 2. Lymph node with reactive lymphadenitis and sinus histiocytosis; 3. Liver tissue sample with bile duct hamartomas (von Meyenburg complexes). **(Figure 2 A, B).** The postoperative period was uneventful and the patient was discharged in 3 days.

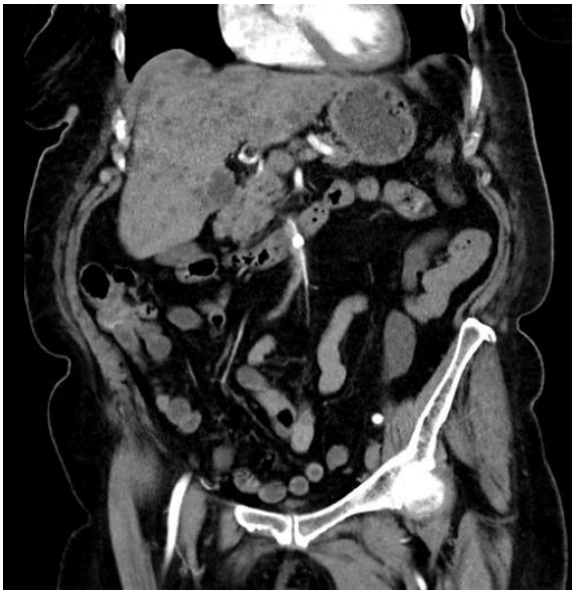

**Figure 1A.** Multiple cystic lesions in both liver lobes

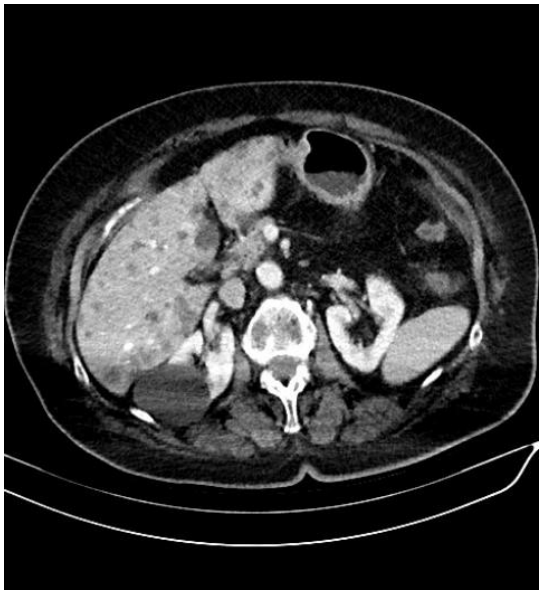

**Figure 1.**B Multiple cystic lesions in both liver lobes and a right kidney cyst

## DISCUSSION

The von Meyenburg complexes or bile duct hamartomas are described for the first time in 1918. (5) They are usually found incidentally in autopsies or surgical procedures. The prevalence rate of this pathology in adults is reported to be from 0.7 to 5.6%. (6) VMC are ductal plate malformations which may be isolated or associated with liver and kidney cystic diseases, Caroli syndrome, congenital hepatic fibrosis and biliary atresia. (7, 8)

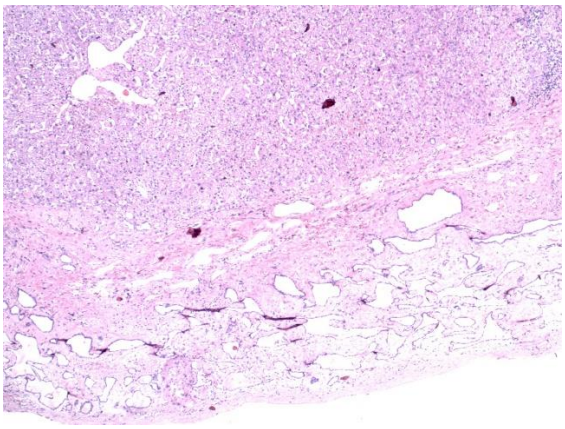

**Figure 2A.** Liver parenchyma with groups of dilated bile ducts that consist of uniform cells without atypism. The ducts are surrounded by fibrous stroma. (von Meyenburg complexes)

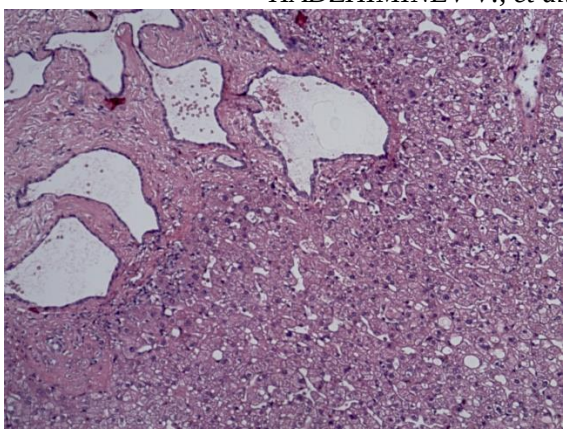

**Figure 2B.** Dilated bile ducts with fibrous stroma (von Meyenburg complexes)

Usually VMC are asymptomatic but rarely may be presented with abdominal pain and symptoms of recurrent cholangitis and severe portal hypertension. (2, 9)

Histologically, VMC present multiple, greyish, nodular liver lesions with small size varying from 1 to 30 mm. VMC are characterized microscopically as small cystic dilated bile ducts that are lined by single layer of cuboidal epithelium cells. They are round or irregular in shape and are embedded in abundant fibrous stroma. Usually, VMC affects both liver lobes predominantly in subcapsular and periportal areas and do not communicate with the biliary tree. (10-12)

On ultrasonography VMC appear as multiple micro-nodules, either hypo- or hyperechoic that may show comet-tail artifacts. (13) The CT-appearance of VMC consists of multiple hypoattenuating lesions round or irregular in shape with no enhancement that are more clearly visible after intravenous administration of iodinated contrast material. (14, 15) On ultrasonography and computed tomography, VMC could hardly be differentiated from small metastases or intrahepatic stones. This may represent a diagnostic challenge in patients with known primary malignancy or in such that underwent oncologic surgery. MRI and MRCP are considered as the best imaging modalities for the assessment of VMC. In all the cases where the diagnosis is doubtful, a liver biopsy should be performed. (16, 18)

In the case of VMC which we reported, the main symptomatology was the acute cholecystitis. Preoperatively, liver lesions were not detected on ultrasonography, but were seen on abdominal CT. Initially they were considered as metastases evaluating the patient's oncological history (two previous oncologic surgeries without any adjuvant radio- and chemotherapy and with no follow-up performed by surgeon or oncologist). However, several renal cysts detected on the CT correspond to the relation of VMC with other cystic diseases cited in the medical literature. (6, 7) It should be noted that some of the liver lesions were larger in size (up to 5.8 cm) than typical for VMCs. When performing the laparoscopy, macroscopically it was also impossible for us to exclude liver metastases. The final diagnosis von Meyenburg complexes was made after a careful histological analysis of the liver biopsy sample.

## CONCLUSION

Von Meyenburg complexes are a rare pathologic entity. Their clinical, imaging, and macroscopic features make them

difficult to be distinguished from liver metastases, especially in patients with previous oncologic history. Liver biopsy and histological analysis play a crucial role in this disease.

**Abbreviations**

VMC – von Meyenburg complexes
CT – computed tomography

**REFERENCES**


1. Redston MS, Wanless IR. The hepatic von Meyenburg complex:prevalence and association with hepatic and renal cysts among 2843 autopsies [corrected]. *Mod Pathol;* 9:233—7, 1996.
2. Davidoff S, Kim S, Friedman B. von Meyenburg complexes (bile duct hamartomas). *Clin Gastroenterol Hepatol*. 4:A26, 2006.
3. Pech L, Favelier S, Falcoz MT, Loffroy R, Krause D, Cercueil JP. Imaging of Von Meyenburg complexes. *Diagn Interv Imaging.*; 97(4):401-409, 2016. doi:10.1016/j.diii.2015.05.012
4. Nagano Y, Matsuo K, Gorai K, Sugimori K, Kunisaki C, Ike H, et al.Bile duct hamartomas (von Mayenburg complexes) mimickingliver metastases from bile duct cancer: MRC findings. *World JGastroenterol*; 12:1321—3, 2006.
5. von Meyenburg H. Uber die Cystenleber. *Beitr Pathol Anat;* 64: 447-532, 1918.
6. Salo J, Bru C, Vilella A, Gin#s P, Gilabert R, Castells A, et al. Bile duct hamartomas presenting as multiple focal lesions on hepatic ultrasonography. *Am J Gastroenterol.* 87: 221-223, 1992.
7. Desmet VJ. Pathogenesis of ductal plate abnormalities. *MayoClin Proc;* 73:80—9, 1998.
8. Drenth JPH, Chrispijn M, Bergmann C. Congenital fibro-cystic liver diseases. *Best Pract Res Clin Gastroenterol*; 24:573—84, 2010.
9. Yoshida S, Kurokohchi K, Ueno T, Yoshino M, Shimada M, Masaki T. Hepatic von Meyenburg complex: A trigger of severe portal hypertension. *Liver Int*.; 29(4):614-615, 2009. doi:10.1111/j.1478-3231.2008.01903.x
10. Lev-Toaff AS, Bach AM, Wechsler RJ, Hilpert PL, Gatalica Z, Rubin R.The radiologic and pathologic spectrum of biliary hamartomas. *Am J Roentgenol*; 165:309-13, 1995.
11. Röcken C, Pross M, Brucks U, Ridwelski K, Roessner A. Cholamgiocarcinoma occurring in a liver with multiple Bile Duct Hamartomas (von Meyenburg complexes). *Arch Pathol Lab Med;* 124:1704–6, 2000.
12. Karahan OI, Kahriman G, Soyuer I, Ok E. Hepatic von Meyenburg complex simulating biliary cystadenocarcinoma. *Clin Imaging;* 31: 50–3, 2007.
13. Zheng R-Q, Zhang B, Kudo M, Onda H, Inoue T. Imaging findingsof biliary hamartomas. *World J Gastroenterol*; 11:6354—9, 2005.
14. Mortelé KJ, Ros PR. Cystic focal liver lesions in the adult:differential CT and MR imaging features. *Radiographics*; 21:895—910, 2001.
15. Wohlgemuth WA, Böttger J, Bohndorf K. MRI, CT, US and ERCP inthe evaluation of bile duct hamartomas (von Meyenburg com-plex): a case report. *Eur Radiol*; 8:1623—6, 1998.
16. Tohmé-Noun C, Cazals D, Noun R, Menassa L, Valla D, VilgrainV. Multiple biliary hamartomas: magnetic resonance featureswith histopathologic correlation. *Eur Radiol*; 18:493—92008.
17. Semelka RC, Hussain SM, Marcos HB, Woosley JT. Biliary hamar-tomas: solitary and multiple lesions shown on current MRtechniques including gadolinium enhancement. *J Magn ResonImaging*; 10:196—201, 1999.
18. Kim HK, Jin SY. Cholangiocarcinoma arising in von Meyenburgcomplexes. *Korean J Hepatol*; 17:161—4, 2011.