

Von Meyenburg Complexes: a starry sky

D. ZENG, Y. WAN

Department of Gastroenterology, Guangzhou Panyu Central Hospital, Guangzhou, China

Abstract. – The von Meyenburg complex (VMC) comprises of the bile duct hamartomas (BHs). VMCs are benign biliary malformations that originate from the disorganization of the small intrahepatic bile ducts. VMCs typically cause no symptoms or disturbances in the liver function; thus, in most instances, they are diagnosed incidentally. However, on some occasions, they are difficult to differentiate from other liver lesions, in particular from small liver metastases; imaging studies are usually noncontributory, and biopsy is necessary for a definite diagnosis. We describe the case of a 61-year-old man who presented with abdominal pain.

Key Words:

Von Meyenburg complex, Liver cystic lesions, Multicystic biliary hematoma, Benign biliary malformations.

changes with mild glandular atypia. Computed tomography (CT) indicated that it may be a bile duct hamartoma (Figure 1A), and magnetic resonance imaging (MR) was recommended. Hence, an MRI was performed and the presence of multiple cystic lesions, T2-weighted, hyperintense cystic nodules in the liver, with no biliary duct communication, assuming a “starry sky” appearance (Figure 1B and 1C); typical of VMCs, were confirmed. Ultrasound-guided fine-needle aspiration cytology was performed, which revealed proliferating and disordered small bile duct-like structures (Figure 1D), with lumens of different sizes and cystic dilation, lining a single layer of squamous epithelium, and no atypical nuclei. No specific treatment was initiated, and at a follow-up visit several months after presentation, the imaging findings remained unchanged.

Introduction

The von Meyenburg Complex (VMC) was first described and discovered by von Meyenburg in 1918. This disease is extremely rare. It has been reported in literature that it commonly occurs in children, especially children under 2 years of age, and it is even rarer in adults. The VMC is a rare benign tumor-like lesion in the liver^{1,2}. This is due to the developmental malformation of the interlobular bile ducts in the liver during the embryonic period, leading to the formation of cyst-like structures of different sizes.

Case

A 61-year-old Chinese man presented with distention and melena for 10 days. On examination, tenderness beneath the xiphoid process and right upper quadrant was appreciated; his carcinoembryonic antigen(CEA) was 5.27 ng/ml and his serum hyaluronic acid level was 129.30 ng/ml. Gastric endoscopy showed multiple stomach ulcers, hence, malignant transformation was not excluded, although the pathological results suggested that it was consistent with ulcerative

Discussion

VMC is a rare benign tumor of the liver, which is more common in infants and children under 2 years of age and is rare in adults. VMC usually manifests as small gray-white nodules scattered throughout the liver or several liver segments²⁻⁴. Pathological manifestations include local bile duct-like structure aggregation, varying degrees of bile duct dilation, and lining bile duct epithelium on the background of abundant connective tissue compartments, which are surrounded by dense fibrous compartments⁵. The disease has no typical clinical manifestations or signs. It is mostly accidentally discovered during imaging or autopsy. Occasionally, symptoms are mostly caused by coeliac disease. The diagnosis and differential diagnosis of VMC rely mainly on imaging and pathological examinations. Plain CT showed multiple low-density cystic lesions in the liver, and there was no enhancement during the enhanced scan. The literature mostly believes that the ultrasound and CT findings of intrahepatic bile duct hamartoma are not specific, but they can be used

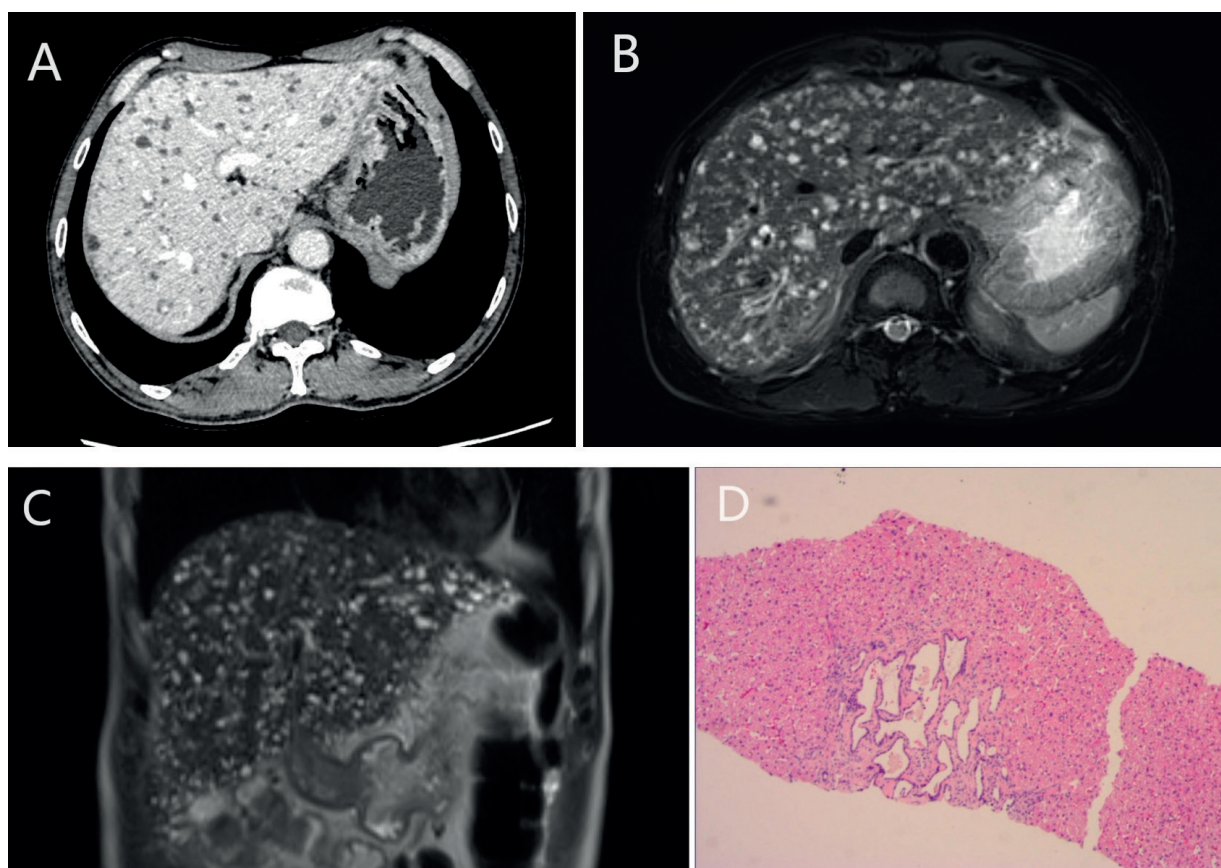


Figure 1. A 61-year-old Chinese man presented with distention and melena for 10 days **A**, the liver is full of small low-density foci, about 10×12mm, some of the foci are slightly enhanced after enhancement, and some have no enhancement. **B**, and **C**, T2-weighted magnetic resonance imaging showed to be hyperintensities and scattered throughout the liver which creating a “starry sky” appearance. **D**, pathology of the liver showed proliferating benign bile ducts.

to diagnose VMC. On plain MRI plain scan, the T1WI lesions showed a signal lower than the liver parenchyma, and they were multiple and distributed along the vascular and bile duct tree. The T2WI lesions were clearly displayed, showing obvious cystic hyperintensity, like a “starry sky”⁶. In the enhanced scan, the arterial phase lesions were not enhanced and showed a low signal; the portal vein phase lesions were also not enhanced, showing a clear boundary between the low signal and enhanced liver parenchyma, showing multiple spot-like low signal nodules. Pathological examination showed that the frequently occurring foci were mostly located in the portal area, and the bile ducts were mostly cystic dilatation, the cyst wall was composed of bile duct epithelial cells, the bile duct contained bile, and collagen fibers wrapped around the bile duct⁷⁻⁹. VMC is a benign disease that is mostly based on follow-up observations. For a single VMC, it has little effect

on the surrounding liver tissue, and the prognosis after surgical resection is good, and there are no reports of malignant transformation.

Conclusions

Some patients with diffuse VMC often have repeated liver function abnormalities, and liver transplantation is recommended.

Conflict of Interest

The Authors declare that they have no conflict of interests.

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