Von Meyenburg complex (hamartoma of the bile duct) mimicking liver metastases

Fon Majenburgov kompleks (hamartom žučnog kanala) sličan metastazama u jetri

Filip Vukmirović*, Irena Tomašević Vukmirović†, Mihailo Vukmirović‡

*Department of Pathology, †Department of Radiology, ‡Department of Cardiology, Clinical Center of Montenegro, Podgorica, Montenegro

Abstract

Introduction. Hamartomas of the bile duct (von Meyenburg complex) are benign lesions of the liver that may mimic liver metastases. Histologically, they consist of cystic dilatations of the bile duct, encompassed by fibrous stroma.

Case report. We reported a 68-year-old male patient in whom ultrasonic and MSCT appearance suggest multiple liver metastases. The diagnosis of von Meyenburg complex was made after histopathologic examination of liver biopsy.

Conclusion. Von Meyenburg complex in an uncommon entity which should be taken into consideration as a differential diagnosis of liver metastases.

Key words: hamartoma; bile ducts; diagnosis, differential; diagnostic techniques and procedures; biopsy.

Introduction

Von Meyenburg complexes (VMCs) is a benign liver lesion. This lesions consists of bile ducts which are incorporated in the connective stroma. Some of biliary ducts are cystically dilated. The incidence of VMCs is age-dependent. Redston et al. 1 reported that this lesion is found in approximately 1% in children and in about 5–6% in adults. Patients often, have no symptoms and the lesion is usually discovered accidentally. Röcken et al. 2 and Jain et al. 3 suggest to possible malignant transformation of these lesion. Von Meyenburg complexes are small lesions, and they are often not detected on radiological examinations, including ultrasound, computed tomography (CT) and magnetic resonance imaging (MRI) 4–7. Its clinical importance is that it may mimic malignant liver disease. We presented a case of VMCs which preoperatively suggested hepatic metastases.

Case report

A 68-year-old male patient with multiple hepatic lesion which ultrasonographic and multisliced computed tomography (MSCT) appearance suggested multiple liver metastases was accepted for surgical exploration and liver biopsy. The patient vomited for a month and lost weight. During surgery numerous whitish irregular lesions of various sizes scattered in the hepatic surface imitating metastatic deposits were noted through both liver lobe and all liver quadrants. Exploration of the abdominal cavity showed no pathological changes nor peritoneal carcinomatosis. Liver biopsy was done and three samples taken for analysis. Tissue was of brown-yellow-gray color and medium-firm consistency. Histological analysis demonstrated multiple lesions (Figure 1) composed of biliary ducts incorporated in fibrotic tissue (Figure 2), with cystic dilatations of the same intrahepatic biliary ducts (Figure 3) suggestive hamartomas of the bile duct or von Meyenburg complex.
Discussion

Hamartomas of the bile duct named von Meyenburg complex are rare lesions, usually characterized by multiple small nodules located near the Glisson's capsule. The size of nodules is usually a few millimeters. VMCs are usually asymptomatic and being accidentally discovered during laparotomy or autopsies. Multilocular occurrence is possible although they are rarely spread throughout the whole liver, as it was observed in the presented patient. According to data from the literature VMCs can be in association with Caroli’s syndrome, congenital hepatic fibrosis, autosomal dominant polycystic renal disease, cholangiocarcinomas and cholangitis.

VMCs can be misdiagnosed on ultrasonography with interpretation that is as metastatic hepatic disease. The definitive diagnosis of this lesion was confirmed by biopsy of the liver. The ultrasonographic findings of the von Meyenburg complex are diversiform lesions which include hyperechogenic and hypoechoic nodules with poorly or well limited margins. Nagano et al. reported that magnetic resonance cholangiography is the best method for examination of hamartomas of the bile duct. This method makes possible to distinguish the different forms of bile duct abnormality, such as sacular dilatation of the biliary system (Caroli disease) and ductal cystic dilatation. This method also makes possible to detect the presence of cholangiocarcinoma or diffuse liver metastases.

Histologically, VMCs include dilated small bile ducts, surrounded by fibrous stroma. Microscopically, they are characterized by abundant fibrous stroma with cystic dilatations of the bile duct which can be of various sizes, and sometimes with associated periductal glands.

In the literature we have not encountered studies in a larger number of cases describing the malignant potential of these lesions. Xu et al. in two case reports described association of von Meyenburg complex with cholangiocarcinoma, for commented it as lesion which carries an increased risk for the development of cholangiocarcinoma.

Except for cholangiocarcinoma, VMCs can be associated with hepatic cysts, polycystic renal disease and cholangitis. Owing to the association between these diseases, Vitule et al. emphasized that in cases of adult polycystic disease, screening for VMCs is very important.

Conclusion

Von Meyenburg complexes are an important differential diagnosis of liver metastases which also includes other benign liver lesions, like hemangiomas, adenomas or infectious lesions. Due to the presence of liver metastases it is of great importance for therapeutic decision making in patients with malignant diseases, distinguishing metastatic and benign changes that mimic them. As VMCs are small, usually less than 5 mm in size, they can escape preoperative radiologic diagnostics. The macroscopic appearance of VMCs can mimic liver metastasis as demonstrated in our reported patient.
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