A Pitfall in the Diagnosis of Unresectable Liver Metastases: Multiple Bile Duct Hamartomas (von Meyenburg Complexes)

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Abstract
Von Meyenburg complexes (VMC) are a cluster of benign liver malformations including biliary cystic lesions, with congenital fibrocollagenous stroma. This rare entity can mimic multiple secondary hepatic lesions. We report a case of a 56-year-old woman who had multiple liver lesions 12 years after operation for breast cancer. Biopsy of the hepatic lesion confirmed the diagnosis of VMC. Preoperative discovery of multiple gray-white nodular lesions scattered on the surface of the liver should not always contraindicate curative liver resection. The diagnosis of VMC should be known and confirmed with liver biopsy.

Case Report
A 56-year-old woman was operated in 1996 for breast cancer. Ten years later, levels of serum tumor marker CA15-3 progressively increased until 2N although other serum tumor markers (α-fetoprotein, carcinoembryonic antigen, carbohydrate antigen 19-9) were normal. Axial contrast-enhanced multislice CT scan showed a solitary and hypovascular 4-cm lesion located in the right lobe of the liver (segment VII). Liver MRI found this lesion in segment VII to be associated with multiple and small lesions with high signal intensity scattered throughout the liver especially in the subcapsular area in T2-weighted images (fig. 1a, b). After contrast injection, the multiple masses showed irregular progressive enhancement from arterial phase through delayed phase. Gastroscopy and coloscopy did
not find new tumors and PET scan showed hyperfixation only on the hepatic lesion in segment VII. The diagnosis of a single liver metastasis of breast cancer was suspected although its characteristics were unusual (hypovascular lesion). The diagnosis of von Meyenburg complexes (VMC) was also suspected preoperatively, but the presence of multiple secondary hepatic lesions could not be dismissed because of breast cancer history. Percutaneous biopsy of the largest hepatic lesion found fibrosis but no malignant infiltration and could neither confirm the diagnosis of metastasis of breast cancer nor the diagnosis of VMC. Since this tumor had unusual characteristics, we performed laparoscopic assessment to obtain histological proof of malignancy. During laparoscopy, multiple gray-white nodular lesions (about 0.5–1 cm in diameter) were found on the surface of the liver (fig. 1c). A liver biopsy was performed because we feared multiple liver metastases (contraindication of curative resection). Histological examination confirmed the diagnosis of multiple benign VMC (biliary hamartomas) encompassed by major fibrous stroma. After conversion to laparotomy, right hepatectomy was performed and the pathological findings of the specimen confirmed the liver metastasis of the breast cancer in segment VII (without argument for cholangiocarcinoma) associated with multifocal, irregularly dilated tinged bile ducts. The patient was discharged from the hospital 9 days after surgery without complications.

Discussion

VMC is a cluster of benign liver malformations and is among the most common congenital liver lesions. When these lesions are multiple and deep, they can simulate secondary metastases. The diagnosis of VMC should be known and confirmed by liver biopsy. Preoperative discovery of multiple gray-white nodular lesions should not always contraindicate curative liver resection. The diagnosis of VMC is suspected based on imaging studies, but histological examination is necessary to confirm the diagnosis. The presence of multiple secondary hepatic lesions in the setting of breast cancer should raise suspicion for metastases, but the diagnosis of VMC should be considered in cases where the lesions are multiple and deep. The differential diagnosis includes cholangiocarcinoma, metastases, and multifaceted intrahepatic lymphocytic lesions. The presence of multiple secondary hepatic lesions is a contraindication to curative surgery. The diagnosis of VMC should be considered in cases where the lesions are multiple and deep. The diagnosis of VMC is confirmed by histological examination of liver biopsy specimens. The presence of multiple secondary hepatic lesions is a contraindication to curative surgery.
Fig. 1. **a** Axial T2-weighted liver MRI showing numerous and small lesions with high signal intensity scattered throughout the liver especially in the subcapsular area of the right liver lobe. **b** Coronal MR cholangiopancreatography demonstrating no communication between the cystic lesions and the normal biliary system. **c** Specimen of right hepatectomy showing gray-white nodular lesions (about 0.5 cm in diameter) scattered on the surface of the liver. **d** Specimen of right hepatectomy showing gray-white nodular lesions throughout the liver parenchyma associated to a large lesion (liver metastasis) with retraction of the liver capsule. **e** Microscopic view showing multiple bile ducts with slightly dilated lumens embedded in the collagenous stroma (hematoxylin and eosin, ×100).
References


