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CASE REPORT

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Preoperatively undetected solitary bile duct hamartoma (von Meyenburg complex) associated with esophageal carcinoma

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Abstract Bile duct hamartomas, also known as von Meyenburg complexes, are benign neoplasms consisting of cystic dilatation of the bile duct surrounded by fibrous stroma. We report a rare case in a 60-year-old man who presented with coexistent von Meyenburg complex and esophageal carcinoma. Preoperative computed tomography did not reveal any liver tumors. Intraoperatively, a small lesion was discovered in segment III of the liver; the lesion was suspected to be a solitary liver metastasis from the esophageal carcinoma. Partial resection of the liver was performed, and pathological findings revealed bile duct hamartoma (von Meyenburg complex). As von Meyenburg complexes are small cystic lesions located throughout the liver, and as they do not present characteristic imaging findings, their preoperative morphological diagnosis and differential diagnosis from liver metastasis is extremely difficult. In conclusion, von Meyenburg complex should be considered in the differential diagnosis of intrahepatic cystic neoplasms.

Key words Bile-duct hamartoma · von Meyenburg complex · Esophageal carcinoma · Liver metastasis

Introduction

Von Meyenburg complexes, which were initially described by von Meyenburg in 1918,[1] are benign neoplasms of the liver that, pathologically, consist of cystic bile duct dilatation surrounded by fibrous stroma. The incidence of these lesions (also known as benign bile-duct hamartomas) is approximately 5%–6% in adults.[2] Because von Meyenburg complexes are small lesions, ranging from 0.1 to 1.0 cm in diameter,[3] their precise diagnosis by radiological imaging is difficult.[4,5] In particular, when detected intraoperatively, the differential diagnosis between von Meyenburg complex and metastasis from a primary tumor is difficult. The coexistence of von Meyenburg complexes with esophageal carcinoma is extremely rare. Only two such cases have been reported in the medical literature. We present the case of a Japanese patient with a preoperatively undetected solitary von Meyenburg complex associated with primary esophageal carcinoma.

Case report

A 60-year-old man with dysphagia was admitted to our hospital in February 2007. Although he had suffered from atrial fibrillation and chronic heart failure 2 years previously, his physical examination was unremarkable. His serum levels of hepatobiliary enzymes and tumor markers, including carcinoembryonic antigen (CEA) and squamous cell carcinoma (SCC), were within the normal ranges. Upper gastrointestinal scope showed a type III tumor in the upper thoracic esophagus. Histological examination of the biopsy specimen showed SCC consisting of well-differentiated squamous cells. Gastroenterography showed esophageal stenosis with irregular walls from the upper thoracic esophagus over a length of 5 cm. Preoperatively, liver metastasis was not detected by abdominal ultrasonography (US), computed tomography (CT; Fig. 1), or magnetic resonance imaging (MRI). Clinical staging by the tumor-node-metastasis (TNM) classification indicated a T3N2M0, stage III tumor. Therefore, the patient underwent an esophagectomy.

Intraoperative findings on laparotomy showed a preoperatively undetectable nodule on the surface of segment III

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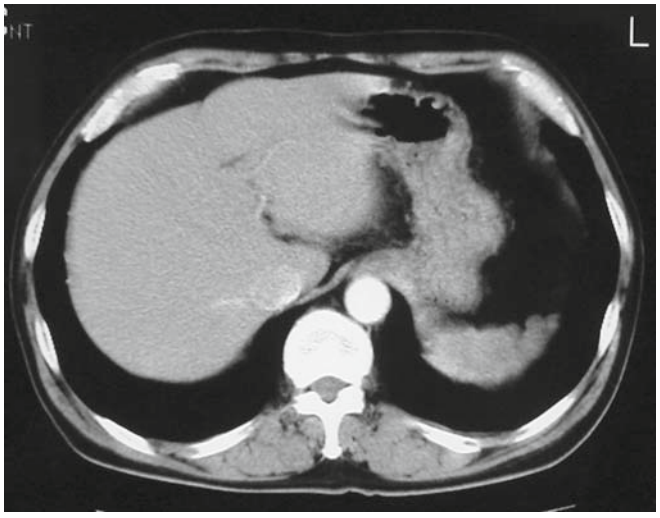

Fig. 1. Preoperative enhanced computed tomography (CT). No focal lesions can be detected in the liver

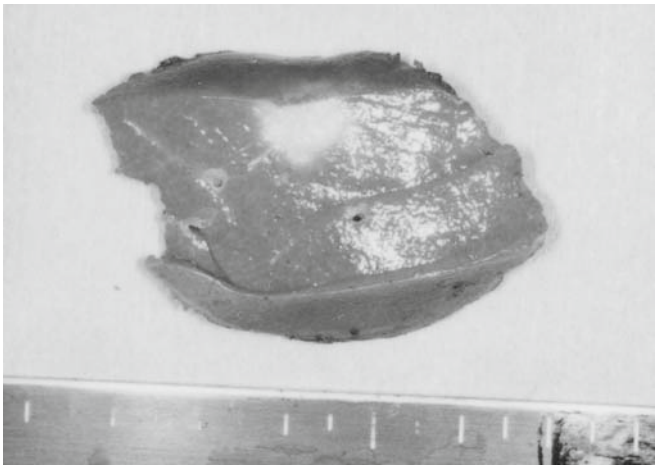

Fig. 3. The resected specimen shows a regularly circumscribed, hard, whitish tumor (1 cm in diameter) on the surface of the liver

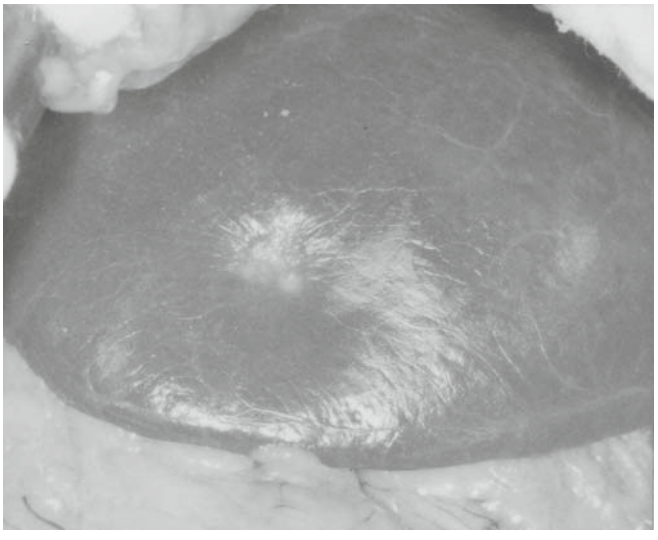

Fig. 2. Intraoperative findings on laparotomy show a grayish-white nodule on the surface of segment III of the liver

of the liver (Fig. 2). The nodule was a grayish-white lesion approximately 1 cm in diameter that resembled cholangiocarcinoma or liver metastasis from the esophageal carcinoma. To confirm the diagnosis and for curative resection, enucleation of the liver tumor was performed. The cut surface of the resected specimen showed a regularly circumscribed, hard, whitish tumor that was 1 cm in diameter (Fig. 3). Pathological findings of a frozen section of the surgically resected specimen showed multiple dilated bile ducts with surrounding fibrous stroma (Fig. 4a), features that are characteristic of benign bile-duct hamartoma (von Meyenburg complex). The patient's postoperative course was uneventful, and he was discharged from the hospital 35 days after the surgery.

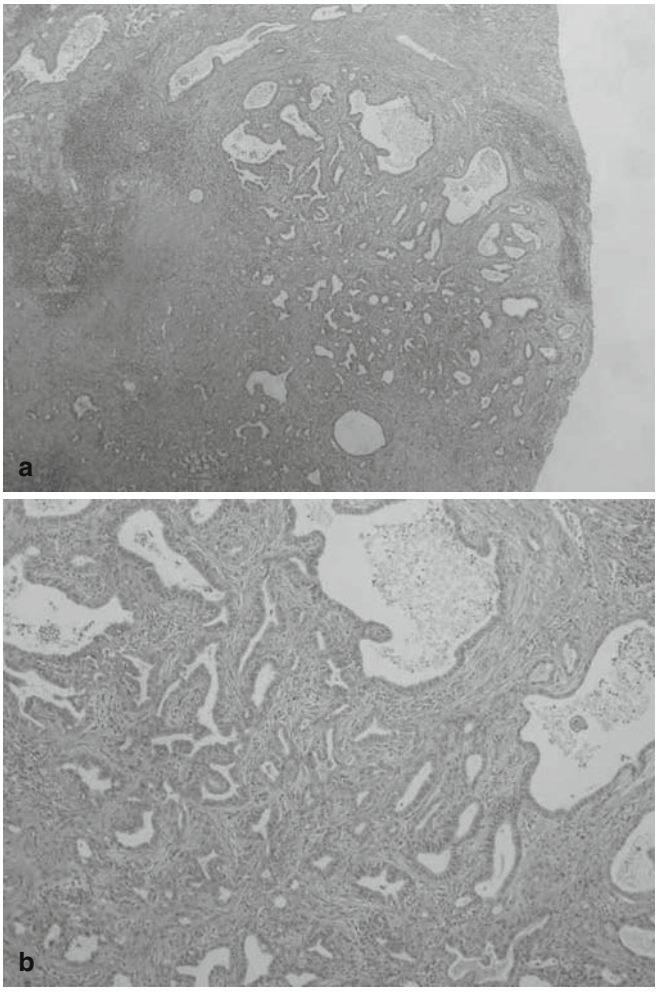

Fig. 4a,b. Histological findings of the liver tumor. **a** Multiple bile ducts with slightly dilated lumens surrounded by fibrous stroma are shown. **b** Dilated bile ducts lined by a single layer of cuboidal epithelial cells with no cytological abnormalities are shown. **a** and **b** H&E, **a** ×40; **b** ×100

Discussion

As stated above, von Meyenburg complexes (bile-duct hamartomas), which were first described in 1918 by von Meyenburg,[1] are benign liver neoplasms that include biliary cystic lesions, which, together with congenital hepatic fibrosis, cause ductal plate malformations.[3] The present patient had an intraoperatively detected liver lesion suspected to be a solitary liver metastasis from esophageal carcinoma, and the lesion was diagnosed as von Meyenburg complex based on the pathological findings of the resected specimen. Fritz et al.[5] reported two cases of intraoperatively detected von Meyenburg complexes mimicking diffuse liver metastases arising from esophageal carcinoma, and Nagano et al.[6] reported a case of von Meyenburg complexes mimicking liver metastases from bile duct cancer. Therefore, it is believed that the differential diagnosis between von Meyenburg complexes and liver metastasis is extremely difficult. Some series have reported malignant transformations of von Meyenburg complex, such as cholangiocarcinoma.[7,8] The mechanism by which the bile duct epithelium is transformed to cholangiocarcinoma is unclear. One mechanism may be that long-term cholestasis of cystic bile containing carcinogenic substances causes malignant transformation of the bile duct epithelium;[9] however, a pathogenetic association between von Meyenburg complexes and cholangiocarcinoma remains controversial. Further, the pathogenetic relationship between von Meyenburg complex and esophageal cancer is unclear. The coexistence of von Meyenburg complex and esophageal cancer, as in our patient and as reported by Fritz et al.[5] may be merely an incidental complication.

The prognosis of patients with metastatic esophageal carcinoma remains poor. Distant metastases from esophageal carcinoma are commonly observed in the lung, liver, and bones. In particular, in Japan, the incidence of liver metastases from esophageal carcinoma is 12.1%. Curative resection of the primary esophageal cancer is impossible in patients with liver metastases; therefore, liver tumor lesions should be identified and diagnosed as precisely as possible. It is possible to preoperatively diagnose liver metastases by CT and MRI with high sensitivity and specificity. The sensitivity of these diagnoses ranges between 74% and 85%.[10] However, when the liver lesions measure less than 1.5 cm in diameter, the detection of metastatic lesions is difficult and false-negative results of liver metastases may be obtained.[10] Therefore, it is difficult to precisely diagnose small lesions of the liver. In patients with correctly diagnosed solitary von Meyenburg complex, no treatment is required. However, von Meyenburg complex should be preoperatively distinguished from other liver lesions, such as simple cyst, cystadenoma, inflammatory pseudotumor, liver abscess, Caroli's disease, cystadenocarcinoma, and cholangiocarcinoma, by using radiological techniques. In particular, the ability of a von Meyenburg complex to mimic liver metastasis is of clinical importance.

Von Meyenburg complexes of the liver are usually detected during laparotomy or autopsy.[2] The incidence of von Meyenburg complex at autopsy is 5%–6%, but clinically, this disorder is very rare. It is believed that morphological diagnosis is difficult because typical von Meyenburg complexes are small lesions.[2,11] Even if preoperatively detected by CT or MRI, differentiating between metastases and von Meyenburg complex is difficult because von Meyenburg complexes generally appear as small cystic lesions located throughout the liver and they do not have characteristic findings on US and CT scans. US scans show multiple hyper- and hypoechoic lesions less than 1 cm in diameter with multiple comet-tail echoes, and CT scans show multiple tiny hypodense lesions with no enhancement.[12] T1-weighted images show multiple small lesions with low-signal intensity, and T2-weighted images show high-signal intensity.[12] MR cholangiography (MRC) can aid in the diagnosis of von Meyenburg complexes only by detecting multiple hyperintense nodules that have no communication between von Meyenburg complexes and the intrahepatic bile ducts;[6] these nodules have been described as "seeming like flowers blooming." In the present patient, the solitary liver lesion was located on the surface of segment III of the liver and was approximately 1.0 cm in diameter; it could not be preoperatively detected by CT and MRI. Retrospectively, we examined the preoperative CT and MRI scans, but the tumor was not detected.

In conclusion, as the existence of liver metastases affects therapeutic decision-making in malignant diseases, von Meyenburg complexes are important for the differential diagnosis of liver metastases. The von Meyenburg complexes are usually small cystic lesions for which preoperative morphological diagnosis is difficult. Intraoperative pathological diagnosis of biopsy specimens of the tumor may be helpful for an accurate diagnosis.[5] If a solitary tumor, such as that in the present patient, is detected intraoperatively, the tumor should be surgically removed and a pathological diagnosis using the frozen section should follow.

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