CASE REPORT

Rare liver tumor: symptomatic giant von Meyenburg complex

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Abstract
von Meyenburg complexes are hamartomas that arise from intra-hepatic bile ducts. Symptomatic lesions are uncommon and giant lesions are rare. When encountered, they should be excised because there are reports of malignant change in large, symptomatic lesions. We report a case of a symptomatic giant von Meyenburg complex.

INTRODUCTION
von Meyenburg complexes (VMCs) are hamartomas that arise from intra-hepatic bile ducts (BD) [1]. Reports of symptomatic VMCs are uncommon and giant lesions are rare. We report a case of a giant, symptomatic VMC that created a diagnostic dilemma.

CASE REPORT
A 55-year-old woman complained of worsening epigastric pain and vomiting on a background of early satiety and unquantified weight loss for ~1 year. There was no history of chronic medical illnesses, medication use or alcohol intake. The abdomen was soft and non-tender, but a vague mass was present in the left upper quadrant.

Gastroscopy suggested extrinsic compression of the gastric fundus but the mucosa was normal in appearance. Abdominal ultrasound revealed the presence of a solitary cystic mass in the left upper quadrant that measured ~9 x 10 x 11 cm (Fig. 1). Despite computed tomography scanning (Figs 2 and 3) and magnetic resonance imaging (MRI) (Fig. 4), the origin of the cyst could not be determined because it was intimately related to stomach, liver and spleen. No other lesions were present in the chest or abdomen. Hepatitis B surface antigen, liver function tests and tumor markers (CA 19–9, CEA and Alphafoetoprotein) were all within normal limits.

A preoperative diagnosis could not be cemented. Therefore, this patient was taken for exploratory laparoscopy. At laparoscopy, the cyst was noted to arise within segments II/III of the liver and was not attached to stomach or spleen. A laparoscopic left lateral segmentectomy was performed to excise the lesion following oncologic principles (Fig. 5). The postoperative recovery period was uneventful and she was discharged within 48 hours.

Ex-vivo examination revealed that the cyst was filled with congealed gelatinous material and surrounded by a thick wall measuring 3 cm in maximal thickness. Histologic examination confirmed that the cyst wall was composed of loose collagenous tissue lined by a single layer of cuboidal cells in keeping with biliary type epithelium (Fig. 6). The adjacent liver parenchyma contained several ectactic BD with focal branching. The surrounding stroma was densely hyalinized with a mild lymphocytic infiltrate and dilated lymphatic channels (Fig. 7). The overall histologic picture was consistent with a solitary non-neoplastic cyst arising within a bile duct hamartoma.
In 1918, a Swiss pathologist, Hanns von Meyenburg, described intra-hepatic cysts that were formed by clusters of BD. This lesion, the VMC, is now recognized to be a hamartoma arising in the intra-hepatic BD. It is theorized that this lesion results from failure of embryonic BD to involute. The persistent ducts dilate when bile becomes inspissated, eventually forming macroscopic cysts. The resultant biliary stasis may lead to intra-ductal precipitation of cholesterol, leading to dilated mature BD with reactive peri-ductal fibrosis. These classic microscopic features were present in our case.

Macroscopically, VMCs usually appear as multiple, nodular lesions beneath Glisson’s capsule, ranging in diameter from 0.5 to 1.5 cm. Giant VMCs are exceedingly rare, accounting for only 0.4% of all cases. The lesion in our patient was 11 cm in diameter, much smaller than the largest lesion reported, that was 21.6 cm at its widest diameter. However, it was comparable to the giant complexes encountered in medical literature, reported to be 9.8 cm in average diameter.
VMCs are uncommon, occurring in 5% of unselected adults at autopsy [2]. Since most are asymptomatic, they are diagnosed less often in vivo where the incidence ranges from 0.4% to 2.8% [3, 4]. They tend to be commoner in patients with polycystic disease [2, 4] and have not noted gender predilection.

The majority of these lesions remain asymptomatic [4]. When they become clinically evident, patients may experience vague abdominal pain from hemorrhage or cholangitis [2–4]. In modern practice, most are incidentally detected at abdominal imaging [5]. Often, VMCs may create a diagnostic dilemma because they appear similar to hepatic metastases [5, 6]. Similarly in our case, a preoperative diagnosis could not be cemented.

On ultrasonography, they appear as small intra-hepatic lesions with mixed echogenicity [5]. They often appear as target lesions, with central hyper-echogenicity due to cholesterol crystals precipitating out of solution within the dilated BD [6]. On CT scans, VMCs appear irregular with low attenuation areas that do not enhance normally with contrast [5].

The most accurate modality to diagnose VMCs is probably a MRI with gadolinium [5, 6]. It can also assist in distinguishing Caroli’s disease, intra-hepatic cholangiocarcinomas and hepatic metastases [5, 6]. On MRI, they typically appear as hyperintense, irregularly delineated cysts that do not communicate directly with the biliary tree [6].

There have been reports of neoplastic transformation of VMCs. When Melnick [4] retrospectively evaluated the results of 70 autopsies in persons with known polycystic liver disease, they reported benign neoplastic transformation of VMCs in two cases (3%). Malignant transformation to cholangiocarcinoma has been reported uncommonly in few other reports [3, 7–10] and tends to occur more commonly with giant complexes [3, 10].

Röcken et al. [7] reported the largest series of cholangiocarcinomas arising on a background of VMCs. Interestingly, they reported four cases—all of which were males in the seventh decade of life [7]. There were associated malignancies in 75% of cases, with two patients having co-existent colorectal carcinoma and one co-existing hepatocellular carcinoma [7]. By examining multiple histologic specimens, Xiu et al. [8] and Röcken et al. [7] were able to independently demonstrate gradual transition from hyperplasia-dysplasia-neoplastic change in multiple foci on the background of VMCs. Although many consider VMCs as benign hamartomatous lesions, Röcken et al. [7] and Xiu et al. [8] have suggested that giant VMCs should be considered potentially premalignant, demanding excision and surveillance.

Clinicians should be aware of the diagnosis as they may occasionally be encountered at imaging. Although symptomatic lesions are uncommon and giant lesions are rare, they should be excised when encountered because there is potential for malignant transformation.

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CONFLICT OF INTEREST STATEMENT

The authors declare that there is no conflict of interest regarding the publication of this article.

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