Extraneural metastasis of meningeal haemangiopericytoma to the liver

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Dear Editor,

Hemangiopericytoma (HPC) is a rare mesenchymal tumor that can occur in any part of the body; however, it has mainly been reported in the extremities, head, neck, retroperitoneum, and pelvic organs (1,2). Meningeal HPC accounts for less than 1% of all intracranial tumors. Although it is curable by complete surgical resection, it behaves aggressively and is accompanied by a high proportion of local recurrence and distant metastasis (1). Here we report two cases of extracranial or extraneural metastasis of meningeal HPC to the liver after 7 years of primary resection and highlight their diagnostic challenges. For the purpose of publication of this report, a separate written informed consent was obtained from each patient.

A 68-year-old woman with a painless abdominal mass and liver lesion revealed upon abdominal computed tomography (CT) performed in 2014 was referred to our institute. In 2007, she underwent excision of parasagittal HPC via craniotomy and completed adjuvant brain radiotherapy.

Apart from a palpable RHC mass, other aspects of physical examination were unremarkable. Laboratory investigations revealed the following results: Serum bilirubin level, 8 µmol/L; alanine transaminase level, 19 U/L; aspartate transaminase, 34 U/L level; alpha-fetoprotein (AFP) level, 2.8 ng/mL, and carcinoembryonic antigen level, 1.4 ng/mL. Hepatitis B surface antigen (HBsAg) and anti-Hepatitis C antibody (anti-HCV) were non-reactive. CT revealed a large, encapsulated, arterially enhancing right hepatic tumor and washout on the portovenous phase typical of hepatocellular carcinoma (HCC) (Figure 1). The patient underwent right hemihepatectomy and recov-

ered uneventfully. No evidence of tumor recurrence was revealed via CT during follow-up 2 years later.

Histopathological examination of the tumor revealed a well-defined mass sized 14×12×7 cm in the liver with areas of necrosis and hemorrhage. On microscopy, the tumor architecture was patternless and comprised thin-walled, staghorn-like branching vascular channels (Figure 2). The tumor was highly cellular with ovoid or spindle-shaped cells, pale eosinophilic cytoplasm, and atypical vesicular nuclei (Figure 3). Mitoses were seen in 6/10 high-power field (HPF). Immunostaining revealed multifocal positivity for CD34 (Figure 4). Finally, metastatic HPC was diagnosed upon detection of diffuse nuclear positivity for STAT6 on immunohistochemistry (IHC).

Our second case was a 30-year-old woman who had been diagnosed with meningeal HPC after cranial surgery in 2007. Seven years later, she was referred for an asymptomatic liver lesion. Prior to the referral, she had experienced two intracranial recurrences in 2008 and 2013, respectively. On both occasions, the tumors were excised and irradiated. Her preoperative liver function test, AFP, HBsAg, and anti-HCV results were normal. Abdominal and pelvic CT revealed a large right lobe liver tumor with an enhancement pattern similar to that of HCC. The tumor was extirpated via right hemihepatectomy, and no recurrence was detected during follow-up.

A well-circumscribed tumor sized 6.5×5.5×5.2 cm was defined on histopathological examination. Microscopically, it was composed of sheets of hypercellular-uniformed cells surrounded by a variably thick pseudocapsule. The cells were homogenous with minimal cytoplasm, whereas the nuclei were oval with small nucleoli and mild nucle-

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Figure 1. CT scan abdomen of case 1 showing huge peripherally enhancing tumour in the right lobe of liver during late arterial phase with washout on portovenous phase.

Figure 2. Microscopic examination showing patternless architecture of the tumour with thin-walled, staghorn-like branching vascular channels.

ar atypia. Mitoses were seen in 6-8/10 HPF. Few staghorn-like branching vascular channels were similarly observed. Immunostaining results were strongly positive for Vimentin and CD99 but mildly positive for CD34 and BCL-2. Considering the patient's background illness and IHC features, metastatic HPC was diagnosed.

HPC is a rare tumor that originates from the pericytes of Zimmermann, which are contractile spindle cells surrounding capillaries and postcapillary venules (1). HPC and solitary fibrous tumor (SFT) have a similar histological appearance and clinical behavior in the peripheral soft

Figure 3. Microscopic examination demonstrated highly cellular cytoplasm consisting of ovoid or spindle-shaped cells with mitoses.

tissue; hence, in 2013, the World Health Organization grouped both these entities as SFT under the category of fibroblastic/myofibroblastic tumors (3). Although most peripheral SFTs are benign, approximately 60% of meningeal SFTs showed local or distant recurrence (2).

Solitary fibrous tumor is smooth, encapsulated, and hypervascular and contains numerous neovascularized vessels in a highly concentrated area of cells. It frequently shows staghorn-like branching vascular channels and mostly spindle-shaped cells (2,3). Its nuclear:cytoplasmic ratio is increased, and its cytoplasm is pale-stained (2). The role of IHC is indispensable to distinguish SFT from other histologically similar neoplasms. CD34, a trans-

Figure 4. Immunohistochemistry staining showing positivity towards CD34.

membrane cell surface glycoprotein originally described on fibroblast, endothelial, and hematopoietic stem cells, was detected in both of our cases. CD34 immunoreactivity is in fact a sensitive marker for SFT (4). Recently, molecular analyses have discovered that almost all HPCs and SFTs harbor an NAB2-STAT6 fusion gene (5). Several studies have suggested that STAT6 IHC staining is a reliable surrogate for detecting this fusion gene; therefore, its immunoreactivity is a highly sensitive and specific marker for SFT (5).

In conclusion, extraneural metastasis of meningeal HPC or SFT to the liver is a rare occurrence particularly after years of successfully treated primary tumor. The HCC-like features of these metastatic lesions on CT render the diagnosis of metastatic meningeal HPC inconceivable at first. Its diagnosis therefore warrants a meticulous review of a patient's clinical history and histopathology as well as an adjunctive immunohistochemistry study of CD34 and STAT6.

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