A 34-year-old male who had a hemangiopericytoma (HPC) within the trigone of right lateral ventricle presented with headache, disorientated consciousness and blurred vision. Computed tomography and magnetic resonance images revealed with homogenous contrast enhancement a 5 cm × 4.5 cm × 4.2 cm lesion within the trigone of right lateral ventricle. Angiographic study revealed a highly vascularized lesion fed from right anterior choroid artery anteriorly and right medial posterior choroid artery posteriorly. The tumor was grossly totally removed via right temporal gyrus. The immunohistochemical study was consistent with HPC. Differential diagnosis of HPC from meningioma and solitary fibrous tumor is crucial because of the high tendency of local recurrence and metastasis of HPCs. Immunohistochemical study is a very valuable method to confirm the diagnosis of HPCs. Complete surgical resection and/or postoperative radiotherapy and/or radiosurgery were beneficial to long-term tumor control. Due to a prolonged time interval between surgery and local recurrence or metastasis, extensive follow-up to rule out local recurrences and delayed extracranial metastases is warranted.

1. Introduction

Hemangiopericytoma (HPC), a vascular mesenchymal neoplasm, arises from the pericytes of Zimmerman, which are leiomyoblastic cells spiraling around capillaries and postcapillary venules.1,2 The tumors may arise in cranio cervical lesions, retroperitoneal tissues, bones, intraoral lesions, the uterus, and intracranial lesion.3 Intra cranial HPC accounts for less than 1% of all central nervous system tumors.4 The most common location of intracranial HPCs is supratentorial related to the dura, similar to meningioma. HPC has rarely been reported as an intraventricular lesion. Herein is a report of a 34-year-old male with right lateral ventricular HPC. Due to the prolonged time interval between surgery and local recurrence, long-term follow up is suggested.

2. Case report

A 34-year-old male patient suffered from headache, blurred vision and disorientated consciousness for two days before admission. After signing an informed consent form he was sent to the emergency department for help where a brain computed tomography with homogenous contrast enhancement revealed a large round mild heterogeneous hyperdense mass (5 cm × 4.5 cm × 4.2 cm) in trigone of right lateral ventricle (Figure 1). Brain magnetic resonance imaging (MRI) revealed an isointensity lesion on T1-weighted image with strong nearly homogeneous enhancement, and an isointensity lesion on T2-weighted image within the trigone of right lateral ventricle (Figure 2). Brain angiography revealed a hypervascular mass in the right lateral ventricle with feeders from right anterior choroid artery and medial posterior choroidal artery. Delayed venous phase showed sunburst appearance (Figure 3). The patient received a temporal craniotomy with grossly total removal of tumor via right temporal gyrus and had an uneventful recovery. During the operation, a very hypervascular tumor arising from choroid plexus of right lateral ventricle was noted. The pathology revealed hemangiopericytoma presenting with staghorn-like blood vessels (Figure 4A). The results showed positive immunostain for vimentin (Figure 4B) and CD34 (Figure 4C) but negative results for GFAP, S-100 protein, and epithelial membrane antigen (EMA). MIB-1 index was approximately between 3% and 5% (Figure 4D). Other immunohistochemical results include positive for hemosiderin, calcification, vascular proliferation and mild focal pleomorphism; there was no evidence of microcyst formation, mitotic activity, presence of gemistocytes, necrosis and eosinophilic bodies. Due to the low MIB-1 index (3–5%), postoperative radiotherapy was not done. The patient underwent postoperative MRI follow up 1 year after surgery and the results revealed no evident tumor recurrence (Figure 5). Long-term follow up is planned.
**Figure 1** (A) Preoperative brain computed tomography without; and (B) with enhancement. The tumor (arrow) approximately 5 cm × 4.5 cm × 4.2 cm located within the trigone of right lateral ventricle.

**Figure 2** (A,B) Brain magnetic resonance imaging revealed isointensity lesion with strongly gadolinium enhancement noted by T1-weighted image. (C) On T2-weighted magnetic resonance imaging image, the tumor looks isointensity when compared with gray matter. The arrow indicates the tumor.
3. Discussion

Intraventricular HPCs probably originate from the pericytes found within the tela choroidea or the stroma of choroid plexus.5 Because there is a high tendency of local recurrence and metastasis, it is important to differentiate HPCs from similar tumors such as meningioma and solitary fibrous tumor. On MRI studies, HPCs are typically isointense to grey matter on T1- and T2-weighted sequences. After injection of gadolinium contrast, heterogeneous strong enhancement was common. The presence of internal serpentine signal voids, although a feature of some meningiomas, seems to be a more frequent finding in HPCs.6 In terms of angiographic features, the characteristics favoring HPCs were6: (1) dual supply from the internal carotid or vertebral and external carotid arteries, with dominant supply from the internal carotid artery branches rather than a primarily external carotid supply seen with meningiomas; (2) a myriad of corkscrew vessels arising from a main feeder within the tumor; (3) a dense fluffy long-lasting tumor stain rather than the sunburst pattern of meningiomas; and (4) a lack of early draining veins.

Preoperative embolization of feeders could be used before surgical removal of intracranial HPC.7,8 This procedure will reduce intraoperative bleeding,9 although the dominant internal carotid arterial blood supply of these tumors often deems them unsuitable for this treatment.10 HPC is difficult to differentiate from meningioma by neuroimaging methods. Immunohistochemistry can differentiate HPCs from meningiomas by showing positive staining for CD34 and vimentin but negative reaction to EMA and cytokeratin.11 The meningiomas give positive immunostain for both vimentin and EMA. In addition, immunohistochemical examinations of CD34, Bcl-2, and reticulin stains are key for the differential diagnosis between HPC and solitary fibrous tumor.12

HPCs are among the very few primary intracranial tumors to metastasize outside of the central nervous system. The most common sites of metastasis are bone, followed by the lung and the liver, and rarely the pancreas and the spleen.13 The rate of distant metastasis at 5, 10, and 15 years is approximately 13%, 33% and 64%, respectively. The median recurrence-free interval is approximately 50 months. The 5-, 10-, and 15-year recurrence rates are 65%, 76%, and 87%, respectively.13 Therefore, irrespective of the extent of surgical resection, the addition of adjuvant external beam radiotherapy is suggested to maximally extend overall survival compared to surgical resection alone. Accordingly, when safe and feasible, surgical resection of recurrent HPC with adjuvant external beam radiotherapy should be the first step in management.14 Guthrie et al. have also recommended total surgical excision of intracranial HPCs and postoperative irradiation before the first recurrence.13 In addition, radiosurgery is a reasonable treatment option for recurrent HPCs. Long-term close clinical and imaging follow-up are necessary because of the high probability of local recurrence and distant metastases. Repeated radiosurgery may be used to treat new or recurrent HPCs over a long follow-up course.15

Figure 3 (A) The tumor was fed by anterior choroid artery anteriorly; and (B) posterior choroid artery posteriorly; (C,D) sunburst appearance was noted during venous phase.
In addition to the rarity of intraventricular HPC, the most important issue is the prolonged time interval between surgery and local recurrence, ranging from 63 months to 104 months. Intra-ventricular HPCs management requires aggressive surgical resection, postoperative radiation treatment, and extensive follow-up to rule out local recurrences and delayed extracranial metastases.

References


