Editorial

Intracranial Hemangiopericytoma: Diagnosis, Treatment and Outcome

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Hemangiopericytomas (HpCs) are rare mesenchymal tumors that may arise in the head and neck, trunk, skin, retroperitoneum and oral cavity. In the central nervous system, HPC is a highly cellular and vascular tumor which is indistinguishable histologically from similar lesions occurring in the peripheral soft tissues. In this issue, Dr. Hsi-Kai Tsou and his co-authors report 2 of 3 (66.7%) and 1 of 3 (33.3%) intracranial HPC patients had local recurrence and intracranial metastasis, respectively, in a relatively short period after surgery. Aggressive growth, tendency to local recurrence and relatively frequent metastases are the clinical features of the tumors.

Neuroimaging study

Plain x-ray films and angiography may distinguish HPC from meningioma. A well-defined, lytic destruction of the adjacent skull favors meningeal HPC, whereas hyperostosis supports meningioma and is exclusive of HPC. Angiography demonstrates a typical blood supply from both meningeal and cortical arteries to form many small cork-screw-like vessels with a marked tumor stain. CT scan and MRI show well-defined tumor with dural attachment and strong enhancement as meningioma, but unlike meningiomas, meningeal HPcs appear to lack calcification.

Histopathological study

HPCs are highly cellular neoplasms composed of plump cells with scant cytoplasm. A dense reticulin network typically investing individual tumor cells is one of the most characteristic features of the tumor. The appearance of stag horn-like vessels separates the tumor cells into small lobules. The characteristic features of meningotheelial cells in part of the tumor differentiate the diagnosis of meningioma from HPC.

HPCs have a particular immunohistochemical profile, which aids in the diagnosis. In Dr. Tsou’s study, one patient (case 2) received X-knife therapy for recurrent tumor, measuring about 2 × 2.5 cm and the tumor decreased in size 15 months later. Stereotactic radiosurgery is indicated for recurrent intracranial HPCs measuring less than 25 mm in greatest diameter.

Predictive factors and outcomes

Prediction of patient outcome is difficult based on current knowledge about the tumors and histological parameters only. However, on the basis of an analysis of central nervous system HPcs, rapid progression is correlated with increased mitotic rate (≥ 5 mitotic figure).
ures per 10 high-power fields), high cellularity, nuclear pleomorphism, hemorrhage and necrosis, in accordance with that of peripheral HPCs. There was some, but not complete, correlation between proliferative labeling in dexamethasone and tumor aggressiveness. The median Ki-67 (MIB-1) labeling index of intracranial HPCs was 10% (0.6-36%), which is at the level of anaplastic meningioma. Lower Ki-67 labeling index is associated with longer recurrence-free interval, lower recurrence rate, lower metastatic rate and higher survival rate, however not at the p < 0.05 level of significance.

Local recurrence rates of intracranial HPCs are almost inevitable and up to 91% of cases, while extracranial metastases occur in 68% of cases at 15 years. In this issue, although no extracranial metastasis was found at the present time, long-term follow-up and high clinical suspicion are mandatory regarding later metastatic potential.

References