Case Reports

Supratentorial hemangioblastoma

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ABSTRACT

يعد ظهور الورم الأرومي الوعائي في منطقة فوق الخيمة بالدماغ من الأمور النادرة جدا، وحتى وقتنا هذا كشف الأدب الطبي عن 128 حالة مصابة بهذه الأورام في منطقة فوق الخيمة. نستعرض في هذا المقال حالة مريضة مصابة بالورم الأرومي الوعائي في منطقة فوق الخيمة، ولم يصاحب ذلك داء فون هيبل–ليندو. ولقد قمنا بوصف الصفات السريرية، وصفات الأشعة العصبية، والمظاهر العصبية المصاحبة للمرض، وقمنا أيضاً بتفسير إجراءات التدخل الجراحي، وطرق المتابعة التي يجب اتخاذها، وأخيراً قمنا بمراجعة الأدب الطبي الذي تطرق لهذا الموضوع سابقاً.

Hemangioblastomas are extremely rare in supratentorial locations, and to date, approximately 128 cases of supratentorial hemangioblastoma have been reported in the literature. Here, we report a female case of supratentorial hemangioblastoma, not associated with von Hippel-Lindau disease. We describe its clinical, neuropathological, and neuroradiological characteristics, elaborate the surgical protocols, and follow-up methods, and review the pertinent literature.

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Hemangioblastomas are benign, highly vascularized neoplasms of uncertain histogenesis, and accounting for approximately 0.99-4.7% of all primary central nervous system tumors.[1] They are the most frequent manifestations of von Hippel-Lindau disease (VHL); an autosomal-dominant inherited cancer syndrome, but they also occur as sporadic non-hereditary tumors. Hemangioblastomas predominantly affect adults, especially of the fifth decade of life, with a male predominance.[2] They arise preferentially in the posterior fossa, becoming the main tumors of the cerebellum, brain stem, and spinal cord. Characteristically, 60% of all hemangioblastomas appear as cystic lesions with an enhancing mural nodule.[3] Supratentorial hemangioblastomas are extremely rare. Currently, approximately 128 cases have been reported.[2,4,5] Here, we report a case of supratentorial hemangioblastoma, not associated with VHL disease, and we review the pertinent literature. Our objective in presenting this particular case is to better delineate its clinical and imaging features and therapeutic strategies.

Case Report. A 19-year-old, right-handed female patient presented with a one-month history of headache. Her medical and family history was uninformative for cancer, or inherited diseases. Neurological examination was unremarkable. Simultaneously, VHL disease was ruled out by abdominal ultrasonography, funduscopic examination, and work-up for mutations of the VHL gene. A skull CT scan revealed a 4.4×3.7cm iso-dense solid-cystic lesion located on the right temporal-occipital lobe with obviously perifocal edema zone (**Figure 1a**). Head MRI indicated that its solid part was hypo-intense on T1-weighted images and hyper-intense on T2-weighted images, with significant enhancement, and non-enhancing multiple cystic compartments (**Figures 1b, c, & d**). At the surgery, the lesion was totally resected by right temporal-occipital craniotomy, and it was observed that the solid part looked like artery-venous malformation, and the cystic fluid was viscous, and xanthochromic. Postoperative skull CT scan illustrated that the lesion was totally removed. Hematoxylin-eosin stain of the specimen demonstrated characteristic features of hemangioblastomas (**Figure 2a**). Immunohistochemical results revealed: S100 (+), CD34 (+), MIB-1 index (MIB-1) 1% (**Figure 2b**). Finally, she recovered uneventfully, and regular MRI follow-up was recommended.

Discussion. The population-based incidence of supratentorial hemangioblastoma is unknown due to its rarity. It is reported that its average age of occurrence is around 36 years.[5] The youngest recorded patient is a 3-week-old male,[6] while the oldest is a 72-year-old

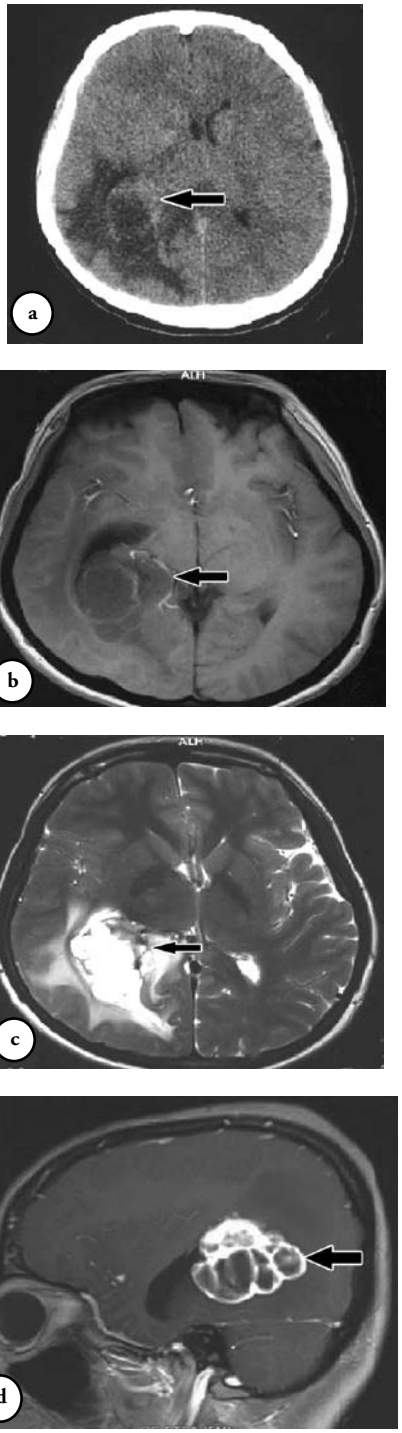

Figure 1 - Pre-operative CT and MRI examinations showing **a)** non-contrast axial plain CT scan shows an isodense cystic-solid lesion in the right temporal lobe (arrow), which involves the right lateral ventricle (**figures b, c, and d**). The solid part of the lesion is hypointense on axial T1 imaging **b)**, and hyperintense on T2 imaging **c)** (arrow). The solid part of the lesion is significantly enhanced (arrow), while the cystic compartments are not on the sagittal gadolinium-enhanced T1-weighted image **d)**.

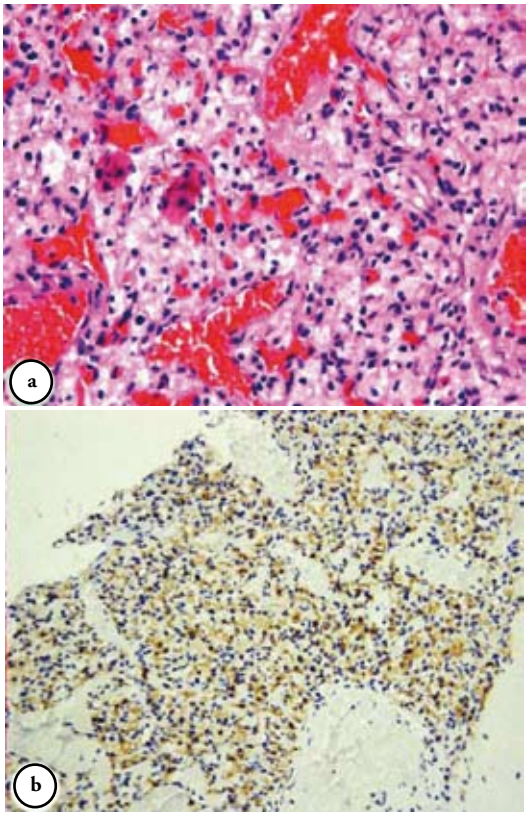

Figure 2 - Histopathological findings. **a)** The lesion is composed of numerous small vessels, and vacuole stromal cells (Hematoxylin and Eosin × 400). **b)** The stromal cells react positively to inhibin α staining (Immunoperoxidase × 200).

male.[5] This entity shows a male predilection (male:female ratio: 2:1).[4] Frank et al[7] concluded that the stromal cell of hemangioblastomas was neither endothelial, neural, epithelial, pericytic, nor neuroendocrine in origin. The failure to specify the cytologic origin of the stromal cells has precluded the development of nonsurgical therapies, and limited the understanding of its basic biology. Clinical presentation of supratentorial hemangioblastoma generally lacks distinguishable specialties from other cerebral tumors. Its clinical manifestations depend mainly on the location and the size of the tumor. A progressive intracranial pressure increase, and systemic polycythemia are usually seen,[5] while weakness of extremities, speech disturbance, memory difficulties, expanding head circumference of infancy, and spontaneous intracranial hemorrhage are less common. If associated with VHL disease, retinal hemangioblastomas, renal carcinomas or cysts, and pancreatic tumors or cysts could be noted.[1] Histologically, hemangioblastomas are composed of a rich network of blood vessels with a single layer of plump, uniform

endothelial cells, and polygonal stromal cells with a swollen, and foamy cytoplasm. The blood space may extend into the large sinuses.[8] Immunohistochemically, the stromal cells of hemangioblastomas react positively to inhibin alpha and CD34.[9] It is important to make a differential diagnosis between hemangioblastoma and angiomatous meningioma. Radiologically, most supratentorial hemangioblastomas arise from the cerebral parenchyma, and 8 reported cases had meningeal involvement.[9] They can present as either isolated or multiple, either cystic or solid lesions. Cystic hemangioblastomas can manifest as large cysts with small nodules. The nodules are contrast-enhancing, while the cystic compartments are not found on the CT and MRI imaging.[3] Morphologically, epidermoid cysts, arachnoid cysts, and cystic glioma should be taken into account in the differential diagnosis. Solid hemangioblastomas are round or round-like with a well-defined border, and they can be iso-intense, or hypo-intense on T1-weighted images with significant enhancement, or hyper-intense on T2-weighted images with earthworm-like flow-void shadows.[5] In addition, meningioma, and ependymoma should be included in the differential diagnosis. The prior treatment protocol of supratentorial hemangioblastoma is surgical removal.[5] During the resection, it is important to remove the nodules of the cystic lesions. For the solid lesions, procedures similar to the management of arteriovenous malformation are implemented. Digital Substraction Angiography is helpful in its diagnosis and treatment. Although the prognosis of sporadic supratentorial hemangioblastoma is more favorable, MRI is recommended as the regular follow-up method. If associated with VHL disease, it is suggested that patients should have lifetime follow-up examinations to monitor recurrence of the disease in the CNS, or discover new lesions in other organs.

In conclusion, a supratentorial hemangioblastoma is an uncommon and benign neoplasm with a favorable prognosis. Its clinical manifestations mainly rely on the location and the size of the tumor. Most appear on the MRI as cystic lesions with an enhancing mural nodule, and they are confirmed by the pathological results showing the characteristics of a rich network of blood vessels with endothelial, and stromal cells. Generally, surgical removal is the rule, and MRI follow-up is recommended.

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