

Intracranial hemangiopericytoma: A case report and review of the literature

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ABSTRACT

Intracranial hemangiopericytomas (HPCs) are rare central nervous system tumors arising from Zimmermann pericytes or mesenchymal cells. It is often difficult to distinguish these tumors from meningiomas based on clinical features and radiological findings. HPCs have been clinically detected in many of the intracranial compartments; here, we present a case with presumed meningioma that was adjacent to the right lateral ventricle and was confirmed by histopathology analysis. A 22-year-old man was referred to our hospital with a slowly expanding, painless mass adjacent to the right ventricle that he had first noticed by accident seven months before. Magnetic resonance imaging demonstrated the presence of homogeneously enhancing mass adjacent to the right lateral ventricle, which was suggestive of a meningioma. The tumor was definitively diagnosed as an intracranial HPC by pathological examination. It is imperative that the surgeon consider this rare diagnosis when evaluating patients with lesions regarded as extracerebral tumors.

KEY WORDS: Hemangiopericytoma (HPC); Magnetic Resonance Imaging (MRI); Stereotactic Radiotherapy (SRT). Meningioma.

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INTRODUCTION

Intracranial hemangiopericytomas (HPCs) are rare central nervous system (CNS) tumors arising from Zimmermann pericytes or mesenchymal cells.¹ It is often difficult to distinguish these tumors from meningiomas based on clinical features and radiological findings. In fact, HPCs were initially regarded as a variant of meningioma. However, detailed research has indicated that they are more similar to the peripheral soft tissue tumors.²

The first documented case of primary intracranial HPC was reported by Begg and Garret in 1954.³ Since then, case studies have revealed that HPCs are more aggressive than meningiomas and have high rates of recurrence and distant metastasis, with local recurrence rates approaching 91% and a 15-year risk of distant metastasis of 70%.⁴ Although HPCs were thought to originate from pericytes in the intracranial compartment, they were usually found as dural-based masses, with non-dural based intracranial HPCs being extremely rare.⁵

Here, we describe a 22-year-old male who had a large HPC adjacent to the right lateral ventricle. Isolated involvement adjacent to the lateral ventricle, as described by our case, is extremely rare.⁶

CASE REPORT

A 22-year-old man was referred to our hospital with a slowly expanding, painless mass adjacent to the right ventricle that he had first noticed by accident seven months before. The clinical examination did not show any positive findings. No regional lymph node enlargement was found. Neurologic examination findings were unremarkable.

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Magnetic resonance imaging (MRI) studies revealed a large dumbbell-shaped space-occupying lesion near the right side of the ventricle, which measured 62mm×40mm×48mm. This lesion appeared to have originated from the right middle cranial fossa, growing upward, rather than from the right lateral ventricle. In addition, it was observed that the third ventricle was markedly compressed and the midline was shifted to the left (Fig. 1A-D). The tumor appeared as isointense on T₁-weighted images, and there was obvious enhancement after gadolinium injection.

A preoperative diagnosis of meningioma was considered. A standard retrosigmoid craniotomy was performed to access the right middle cranial fossa. The tumor appeared to be a well-encapsulated, grey, fibrous lesion adjacent to the right lateral ventricle. Somatosensory evoked potentials and intra-operative facial nerve monitoring were used to ensure the safe separation of the tumor from the right middle cranial fossa and of the vessels from around the mass. The tumor was completely resected.

The patient developed transient right facial paresis and worsening dysarthria after the operation, both of which resolved after three or four days. The patient was discharged from the hospital without any residual deficit. At one month after surgery, the patient had completed adjuvant regional stereotactic radiotherapy (SRT) and gamma knife therapy. Follow-up examinations at one year after surgery, including CT of the brain, chest, abdomen, and pelvis, did not reveal any evidence of local recurrence or extracranial metastasis.

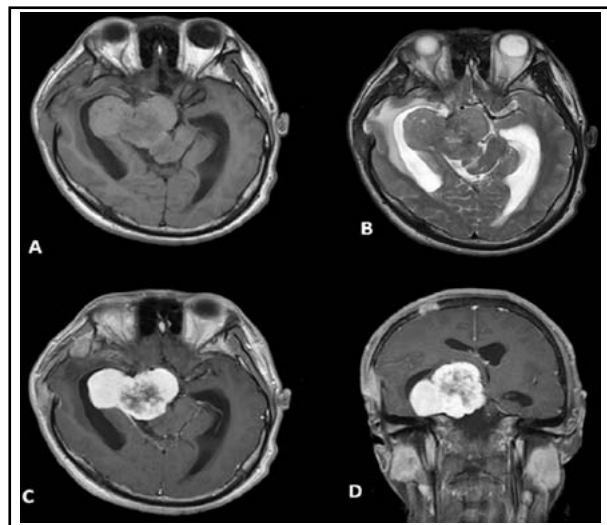


Fig.1: MRI images depicting a heterogeneous mass adjacent to the right lateral ventricle. T1 weighted (A), T2 weighted (B) and post-gadolinium axial (C) and coronal (D) images are shown. Significant perilesional edema is apparent around the lesion.

Histopathological features: Pathological examination was performed after surgical removal of the tumor (Fig. 2A-D). Macroscopically, most of the lesion was solid, with gray-brown hemorrhagic necrotic areas and a jelly-like consistency. Microscopically, the tumor was composed of capillaries and blood vessels lined by a single layer of endothelium. Tightly packed, regular ovoid cells with plump nuclei and scant cytoplasm were found to be arranged in short fascicles. A reticulin stain showed abundant pericytic and vascular collagen. Immunohistochemistry was performed by using antibodies to CD34, epithelial membrane antigen (EMA), and vimentin. The tumor showed immunoreactivity with antibodies to CD34 and vimentin. No EMA expression was detected.

Although some areas of hyalinization were present, suggesting a tumor of mesenchymal origin, the cytology and cellularity of the mass were more typical of HPC. Furthermore, the positive CD34 and vimentin immunoreactivities were more consistent with HPC. A diagnosis of HPC was made.

DISCUSSION

Hemangiopericytomas are rare intracranial tumors, accounting for less than 1% of all intracranial neoplasms.⁷ It has been reported that approximately 2%-4% of all meningeal tumors were HPCs.⁸ HPC was used to be considered as one of the variants of meningioma and was often referred to as 'angioblastic meningioma'. However recent studies suggested that the histomorphology

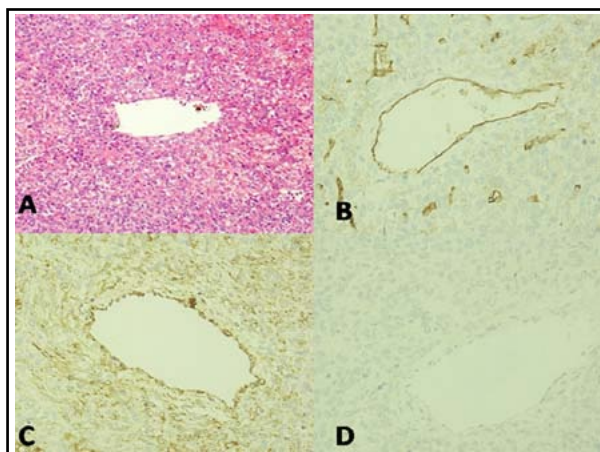


Fig.2: (A) Microscopic examination revealed sheets of homogeneous, randomly-oriented polygonal cells with abundant cytoplasm, oval nuclei, small nucleoli, and moderate pleomorphism. The tumor is seen in close apposition, surrounding mitotically-active staghorn-type vessels lined by flat endothelial cells (hematoxylin and eosin staining; original magnification ×200). (B) Immunoreactivity was observed with antibody to CD34. Blood vessels were CD34-positive, while tumor cells were CD34-negative (original magnification ×400). (C) Extensive and diffuse immunoreactivity with vimentin was observed (original magnification ×400). (D) Immunostaining with EMA was negative (original magnification ×400).

and immunophenotype of HPCs are different from meningioma and hence the World Health Organization classified this tumor type as a distinct entity. In contrast to meningiomas, which are more common in females, HPCs occur more often in males; the HPC male to female ratio is approaching 2:1. The average age of HPC onset is between 38-42 years, which is 10 years earlier than meningiomas.^{9,10}

Radiographically, it is hard to differentiate HPCs from meningiomas, schwannomas, and fibrous tumors. HPCs often arise from dural sinuses, skull base, tentorium, and the falx cerebri. Adjacent to the lateral ventricle involvement, as seen in our case, is extremely rare in the medical literature. HPCs are typically isointense, or mildly hypointense with cortical grey matter, on T1WI. On T2WI, HPCs appear as iso-intense, or slightly hyper-intense. Heterogeneous intense contrast enhancement is a common finding of patients with HPC.

Histomorphologically, HPCs are composed of oval-shaped and elongated tumor cells with ill-defined cytoplasmic borders. The pericytes are located around the vessels and oriented outside the reticular sheath of vessels, but characteristically enclosed by the reticulum. Immunohistochemically, most HPCs express CD34. However, negative CD34 findings have also been reported for some patients with histopathologically proven HPC.¹¹ Nuclear pseudoinclusions and psammoma bodies, which are characteristic histologic features of meningiomas, are not observed in HPCs.¹² Both meningiomas and HPCs are vimentin positive, suggesting a mesenchymal origin. However, only meningiomas are positive for EMA.

Suspected HPCs should be differentiated from other tumors of the CNS. It is important to distinguish HPCs from meningiomas. The immunohistochemical profile of HPCs differs from that of meningiomas. Principally, these two lesions are easily identified by their distinct architectural and cytologic features. Although meningiomas exhibit great variation in nuclear morphology, the nuclei of HPCs are more plump and hyperchromatic, and lack the nuclear pseudoinclusions that typify meningothelial cells.¹² In addition; mesenchymal chondrosarcoma should be differentiated in diagnosis of HPC. Mesenchymal chondrosarcoma has an HPC-like appearance, including the presence of a dense cellular small cell component. The cartilaginous islands of this aggressive chondrosarcoma variant are distinguishing features.

The optimal treatment for intracranial HPCs is complete surgical excision of the tumor. Nevertheless, complete resection is hard to achieve for cer-

tain tumors. Therefore, radiotherapy has been used to treat surgically inaccessible or recurrent tumors. Guthrie et al¹³ proposed that it is important to completely remove an HPC tumor during the first operation and then provide subsequent radiotherapy to prolong the survival period. HPCs are likely to recur at distant sites within the nervous system after resection. Unlike benign meningiomas, extra-neural metastases are much more common. Local recurrences are not rare in patients with HPC. The reported recurrence rates have varied greatly in the literature and are associated with the time of follow-up.¹⁴ Up-front use of radiotherapy has been demonstrated to improve survival. The gamma knife technique may also play a key role in the management of HPCs since it is associated with improved disease-free survival. A recent study suggested that the combination of surgical resection and adjuvant postoperative SRT may prolong survival in patients with newly-diagnosed HPC.¹⁵ Unfortunately, SRT or gamma knife do not appear to provide any significant protection against the development of distant metastases.¹⁵ In our case, the patient was treated with SRT and gamma knife therapy. Although no clinical or radiologic signs of intracranial recurrence or extracranial metastasis have appeared to date, long-term follow-up is necessary.

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