Acta Scientific Clinical Case Reports

Volume 3 Issue 9 September 2022

Case Report

# Intracranial Hemangiopericytoma Presenting as Ophthalmoplegia

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**Received:** August 01, 2022

**Published:** August 25, 2022



## Abstract

**Background:** Intracranial hemangiopericytoma is a rare condition caused by neoplastic transformation of the Zimmerman's pericytes of the meninges.

**Case Report:** A case of a 27-year-old previously healthy female whose chief complaint was headache and painful blurring of vision. Neurologic examination on admission revealed left ophthalmoplegia. MRI revealed a hyperintense middle cranial fossa mass with vasogenic edema with preliminary impression of meningioma. She was initially given steroids then underwent tumor resection. Histopathologic studies revealed findings of hemangiopericytoma. She was discharged stable with improvement of initially observed deficits, but she eventually lost to follow up.

**Conclusion:** Hemangiopericytoma are rare lesions that require a thorough histopathologic investigation since these tumors are difficult to distinguish from meningiomas in preoperative imaging. Our case report was initially diagnosed as a case of meningioma. Our approach to management was complete total resection of the tumor with a plan of post operative radiation therapy and monitoring for tumor recurrence.

**Keywords:** Intracranial Hemangiopericytoma; Ophthalmoplegia; MRI

## Introduction

Intracranial hemangiopericytomas are exceedingly rare lesions with only few cases reported in literature. Known to arise from the Zimmerman pericytes of the meninges, it accounts for 0.5% of all primary central nervous system tumors [1]. The tumor is locally aggressive with a tendency to recur and even metastasize extracranially if not adequately treated [2]. Common presenting symptoms include headache, seizures and focal deficit [3]. We report a case of a patient diagnosed with middle cranial fossa hemangiopericytoma presenting with left eye ophthalmoplegia.

## Case

A 27-year-old right-handed female, with no significant comorbidities, was seen at the ophthalmology clinic complaining of progressive doubling of vision. Eye examination revealed a grade 2 papilledema on fundoscopy and complete left eye ophthalmoplegia. A possibility of an intracranial mass was considered hence cranial MRI was requested and patient was referred to neurosurgery OPD where she was advised admission.

On admission patient was seen by the neurology service. Pertinent neurological findings include MMSE score of 30, ophthalmoplegia and grade 2 papilledema on the left eye and abnormal tandem walk with tendency to fall towards the left. Results of the cranial MRI requested prior to admission showed a middle cranial fossa mass with vasogenic edema. She was started on dexamethasone 4 mg/tab TID for the edema and scheduled for tumor resection. Intraoperatively, there was note of a 7x6x6 cm yellowish, vascular extra-axial mass. The specimen was sent to pathology for examination which revealed a hypercellular stroma composed of spindle cells with monomorphic round to ovoid

nuclei and indistinct cytoplasmic borders scattered with numerous irregularly shaped, thin-walled blood vessels with sharp edges suggestive of hemangiopericytoma (Figure 1). She was eventually discharged on the 7th postoperative day with improvement of left ophthalmoplegia and papilledema.

**Figure 1:** A-B. IA T2 image showed isointense extraxial mass measuring 5.7x5.8x5.5 cm well circumscribed lobulated mass broad dural base at the left middle cranial fossa with vasogenic edema; 1B Tl sagittal showing hyperintense well circumscribed lobulated mass encroaching the left orbital apex and the optic nerve bit without orbit extension.

Figure 1 histopathology of hemangiopericytoma showing (A) haphazard arrangement of hypercellular tumor with areas of necrosis as indicated by the red arrow (B) individual cells exhibit round to ovoid nuclei, dense chromatin, inconspicuous nucleoli and scant eosinophilic cytoplasm. Seen are several thin-walled blood vessels with jagged edges (red arrow) (C) irregularly shaped dilated blood vessels (D) increased mitotic activity shown by the red arrows.

## Discussion

Hemangiopericytomas are highly vascularized tumors of mesenchymal, nonmeningothelial lineage [4]. The diagnosis of this entity is by exclusion. On imaging, its distinction from a meningioma can be difficult due to similar characteristics without depending on the location [5].

Hemangiopericytomas are distinguished from meningiomas by their increase cellularity, higher mitotic index, and microscopic tendency to bulge into unexploded vascular lumens through the endothelium [6]. The immunohistochemical study supports the definitive diagnosis of this tumor, showing positivity for CD34 and vimentin, with negativity for CD31 and S-100 [7].

No standard treatment is available for this tumor due to its rarity [7]. To date, the available published data on hemangiopericytoma are scarce with majority being case reports and retrospective analyses of treatment and tumor biological behavior [8]. For localized disease, like in our case, the primary consensus is complete surgical resection. Among all the available treatment modality, only complete tumor resection increased the overall survival [6] reaching a 100% five-year overall survival and up to an 89% ten-year overall survival [9]. However, rates of local and distal recurrences remain high for intracranial hemangiopericytomas [10].

There are few studies regarding different treatment options for hemangiopericytomas including monoclonal antibodies, mTOR inhibitors, tyrosine kinase inhibitors, target therapies and chemotherapy. However, most published data from these studies are derived from case series, phase 1 and phase 2 studies, making it difficult for oncologists to establish a standard therapy. In addition, part of the studies included patients with various types of soft tissue sarcomas and not solely hemangiopericytomas. In this context, the progression-free survival and overall survival rates reported in these studies are low, such as the use of temozolamide and bevacizumab with a median survival of 10.8 and 24.3 months respectively [11].

## Conclusion

The rarity and unpredictable behavior of hemangiopericytomas pose a challenge in the management of this disease. In the absence of large randomized clinical trials, the only source for its natural history and management are the few published case reports. Based on what is currently published, complete surgical resection is valued to achieve better therapeutic efficacy while the use of conventional chemotherapy is justified with its clinical benefit of reducing symptoms and stabilizing the disease.

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