

## Case Report

# Atypical epidural hemangiopericytoma presenting with visual disturbance

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## Abstract

**Background:** Hemangiopericytomas are a rare entity commonly presenting as subdural tumors.

**Case Description:** A 57-year-old man presented with a progressive visual disturbance over a period of 3 weeks. Cranial computed tomography scans revealed an isodense mass at the tip of the left middle fossa, extending into the orbital apex, and accompanying bony erosions in the sphenoid ridge. On magnetic resonance imaging, the lesion appeared isointense both on T1- and T2-weighted sequences, and intensely enhanced on contrast examinations. A frontotemporal craniotomy revealed a dura-based, capsulized tumor located entirely in the epidural space. A gross total resection was achieved for the tumor and histologically verified as hemangiopericytoma.

**Conclusion:** Hemangiopericytoma should be assumed in a differential diagnosis when encountering epidural tumors, and total resection should be attempted when possible.

**Key Words:** Epidural, meningeal hemangiopericytoma, treatment, visual disturbance

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## INTRODUCTION

Hemangiopericytomas (HPs) are a rare entity that account for only 1% of all intracranial tumors.<sup>[1,5]</sup> Commonly, HP is acknowledged as a dura-based, subdural tumor originating from the Zimmermann's pericytes.<sup>[15]</sup> Although infrequent, HP presents as intraparenchymal<sup>[7,14]</sup> and intraventricular<sup>[9,16]</sup> tumors. Currently, HP and solitary fibrous tumor arising in the central nervous system are managed as a unified entity.<sup>[17]</sup> As HPs have a tendency to cause local recurrence and distant metastases, gross total resection is thought to be the rule for maximizing overall survival.<sup>[2,3,6,10,13]</sup> However, efficacy of adjuvant radiotherapy for HPs has not been determined.<sup>[18]</sup>

HPs rarely present as an epidural tumor in the spinal axis.<sup>[4,12]</sup> However, to our knowledge, there has not been an epidural HP in the cranial cavity.

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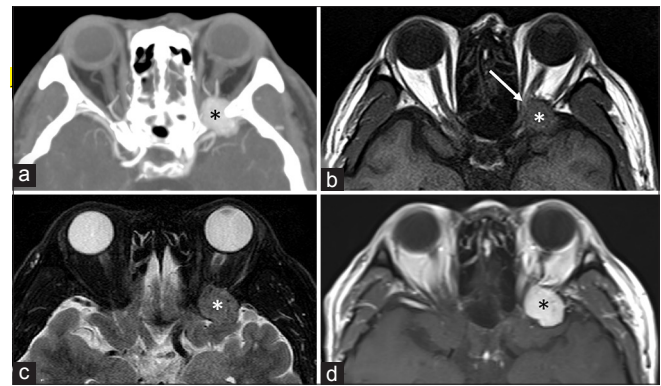
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## CASE PRESENTATION

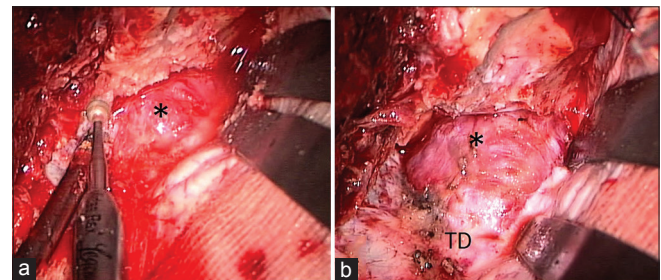
A 57-year-old man presented with a progressive visual disturbance over a period of 3 weeks. His medical history was unremarkable. At presentation, the patient exhibited depressed visual acuity on the left and defects of the temporal visual field. Ophthalmological examination found intact extraocular movements. Other neurological deficits were not noted. Cranial computed tomography scans revealed an isodense mass at the tip of the left middle fossa, 2 × 2 cm in maximal dimension, extending into the orbital apex, and accompanying bony erosions in the medial sphenoid ridge [Figure 1a]. On magnetic resonance imaging, the tumor appeared isointense both on T1- and T2-weighted sequences and intensely enhanced on contrast examination [Figure 1b-d]. The left optic nerve was considerably compressed by the tumor at the orbital apex [Figure 1b]. The patient underwent a microsurgical tumor resection. A frontotemporal craniotomy followed by drilling of the sphenoid ridge revealed a dura-based, capsulized tumor [Figure 2a]. It was elastic, hard, entirely located epidurally, and moderately vascular [Figure 2b]. A meningo-orbital artery was found to pass through the orbitomeningeal foramina and supply the tumor from the upper surface. With the internal debulking maneuver using cavitron ultrasonic surgical aspirator (CUSA), eventually, a gross total resection was achieved. Part of the dura mater at tumor attachment appeared intact and was only given electrical coagulation. Microscopically, the tumor comprised spindle-shaped cells, lacking findings of atypia or necrosis [Figure 3a]. Immunohistochemical stains were positive for bcl2, CD34, CD99, and STAT6 [Figure 3b-e], whereas negative for S100 and epithelial membrane antigen. The MIB-1 index was 10% [Figure 3f]. These were consistent with HP of World Health Organization grade II. Postoperatively, the patient's visual disturbance improved. Systemic <sup>11</sup>C-methionine positron emission tomography/CT scans did not reveal any abnormal accumulation. Immediate adjuvant radiotherapy was not administered. The patient has been under close observation without local recurrence or distant metastasis for 14 months.

## DISCUSSION

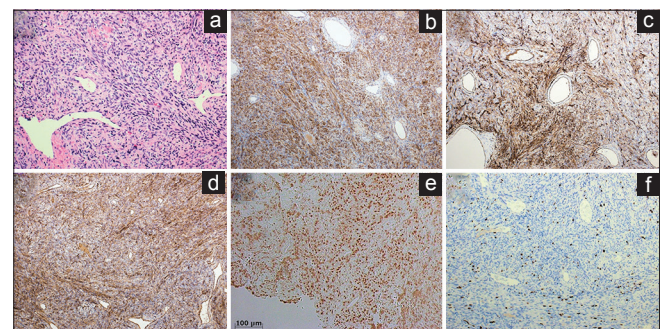
Based on the clinicopathological findings, the present case was thought to be a primary HP that originated from the middle fossa dura and grew as an epidural tumor. To our knowledge, this is the first report of a primary epidural HP originating in the cranial cavity. Intraoperatively, the meningo-orbital artery, a meningeal branch of the lacrimal or supraorbital arteries, connecting the orbit with the cranial cavity and passing through the orbitomeningeal foramina,<sup>[8]</sup> was found to supply



**Figure 1:** Axial computed tomography scan (a) and magnetic resonance images (b-d) showing a well-demarcated mass in the tip of the left temporal fossa extending into the orbital apex, and accompanying bony erosions in the medial sphenoid ridge (a). The tumor appears isointense on both T1- (b) and T2-weighted (c) sequences, and intensely enhanced on contrast examinations (a and d). The left optic nerve is considerably compressed by the tumour at the orbital apex (b, arrow)



**Figure 2:** (a and b) Intraoperative photos showing drilling of the medial sphenoid ridge overhanging the tumour (a) and exposure of a dura-based, capsulized tumour (a and b asterisk). TD: Temporal dura



**Figure 3:** Microscopic appearance of the tumor comprised by spindle-shaped cells, lacking findings of atypia or necrosis (a). Immunohistochemical stains showing positive staining for bcl2 (b), CD34 (c), CD99 (d), and STAT6 (e), with the MIB-1 index of 10% (f). A: Hematoxylin and eosin stain, ×200; b: bcl2; c: CD34; d: CD99; e: STAT6; f: MIB-1, ×100

the tumor. This supports that the present tumor grew extracranially.

The present HP considerably compressed the optic nerve predisposing to radiation injury. For the tumor, total resection was achieved. Meanwhile, the effectiveness of radiotherapy for HPs is not determined.<sup>[18]</sup> Therefore,

prophylactic radiotherapy was not administered immediately after the resection. The timing of adjuvant radiotherapy and modality of administration should be carefully determined on a case-by-case manner according to the tumor location and extent of resection. In our case, stereotaxic radiosurgery may be an alternative option at a local recurrence.<sup>[11]</sup>

HPs tend to cause local recurrence and distant metastases regardless of the histological grades.<sup>[2]</sup> Furthermore, radiological appearance does not necessarily anticipate HP. Therefore, in addition to assuming HP as the differential diagnosis, a total resection should be attempted as possible during surgery of an intracranial epidural tumor.

HP may present as an epidural tumor in the cranial cavity. HP should be assumed in the differential diagnosis during surgery of an intracranial epidural tumor. A total resection should be attempted when possible for such tumors.

### Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

### Conflicts of interest

There are no conflicts of interest.

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