

## CASE REPORT

# An Unexpected Lesion in Cerebellopontine Angle : Hemangiopericytoma

Teoh Jian Woei, MD, MRCS\*, Goh Bee See, MS\*, Shahizon Azura Mohamed Mukari, Mmed Rad\*\*, Siti Aishah Md Ali, DCP\*\*\*, Nor Hafliza Mohd Salleh, MBBS\*\*\*

\* Department of Otorhinolaryngology-Head & Neck Surgery Universiti Kebangsaan Malaysia Medical Centre,

\*\* Department of Radiology Universiti Kebangsaan Malaysia Medical Centre, Kuala Lumpur, \*\*\* Department of Pathology Universiti Kebangsaan Malaysia Medical Centre, Kuala Lumpur

### SUMMARY

**Hemangiopericytoma (HPC) is a rare tumor by definition and intracranial HPC makes up to less than one percent of all the intracranial tumors. It is a dural base tumor and its clinical features and radiological findings are similar to meningiomas. However, cerebellopontine angle hemangiopericytoma had only been reported twice and would almost always be misdiagnosed. Definite diagnosis is important, as the treatment of HPC is different from meningiomas and acoustic neuromas. We report a case of a young female who presented with atypical symptoms of left cerebellopontine angle mass. A literature review of the nature of the disease, radiological findings, immunohistochemical features and treatment options of the tumor are described.**

**KEY WORDS:** *Hemangiopericytoma, cerebellopontine angle tumor, pericyte*

### INTRODUCTION

Hemangiopericytoma (HPC) is an aggressive and rare mesenchymal tumor which originates from the Zimmermann pericyte<sup>1</sup>. The term "pericyte" is also known as vascular smooth muscle cells or mural cells. These spindle-shaped cells surround capillaries and post-capillary venules. Pericytes are formed from the differentiation of mesenchymal cells and also myofibroblast<sup>2</sup>.

This tumor can occur anywhere in the body as it originates from pericytes around the capillaries<sup>3</sup>. The commonest site of this tumor is the lower extremity, followed by the pelvis or retroperitoneum, and head and neck<sup>4</sup>.

The clinical features and radiological findings of this tumor are very similar to meningiomas intracranially. Although classical histologic features of these two tumors are easily distinguishable, occasionally there might be overlap features and immunohistochemistry testing is necessary to differentiate the tumors<sup>1</sup>.

### CASE PRESENTATION:

A 24 year-old female presented with slow progressive left facial weakness for 3 months, which was associated with ipsilateral facial numbness, hearing impairment, tinnitus and intermittent headache for 1 year.

Clinically, there was left lower motor neuron lesion of facial nerve palsy with House-Brackman grade 5. Hypoesthesia of left trigeminal distribution (frontal, maxillary and mandibular

distributions) was also present. Sensorineural hearing loss was noted over the left ear on tuning fork test. Other neurological examinations were normal.

She was treated as Bell's palsy at other hospital. However, due to the progressive worsening of her symptoms, further investigations were done. Pure tone audiometry showed mild to moderate sensorineural hearing loss over left ear. MRI of the brain and internal auditory meatus was done.

MRI findings showed a homogeneously enhancing mass at the left cerebellopontine angle (CPA) with widening of the ipsilateral internal auditory canal. It is isointense on T1WI and hyperintense on T2WI to the grey matter. There were neither serpentine flow void nor dural tail sign seen. A diagnosis of left acoustic neuroma was concluded.

A standard craniotomy was done via a retrosigmoid approach and intraoperative findings noted a well encapsulated grayish tumor at the left CPA extending into the left internal acoustic meatus which appeared to be adherent to the facial nerve. The tumour was excised with facial nerve monitoring and meticulous hemostatic excision was performed as the tumour vascularity is higher compared to schwannomas. The tumour was almost completely removed except the small intracanalicular portion, which is adhered to facial nerve. Post operatively, there was noted cerebrospinal fluid leakage, which resolved with middle ear packing. She had 27 fraction of radiotherapy to her tumour remnant post operatively. There was no sign of worsening cranial nerve deficit during subsequent follow ups and MRI scans which were done twice (6 months and 1 year post operation) showed non-progressive tumor remnant measuring 1.5cm x 0.6cm x 1.0cm at the intracanalicular portion.

Histopathology findings showed that the tissue was diffusely infiltrated by neoplastic cells (Fig. 2A). A large amount of tumor necrosis is present and mitotic figures are 5-8 per 10HPF (Fig. 2A-inset). There is no calcification or psammoma body noted. These neoplastic cells are negative for pan-CK, synaptophysin, CD56, desmin, HMB45, CD31 and GFAP. Histological diagnosis was consistent with anaplastic HPC.

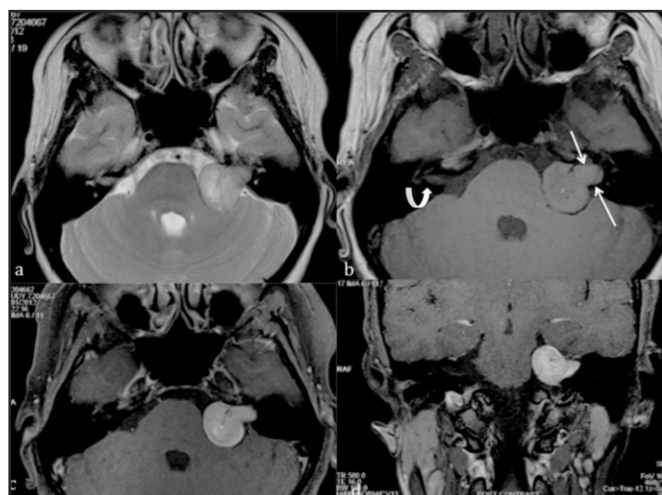
### DISCUSSION

HPCs occur in less than one percent of the entire intracranial tumor and two to four percent of all meningeal tumours<sup>1</sup>. HPCs are always a solitary tumor and mainly arising from the supratentorial region. Intracranial HPC have a male preponderance with a male to female ratio of 2:1<sup>1</sup>. Men are diagnosed slightly earlier at 40 to 50 years old compared to women<sup>2</sup>.

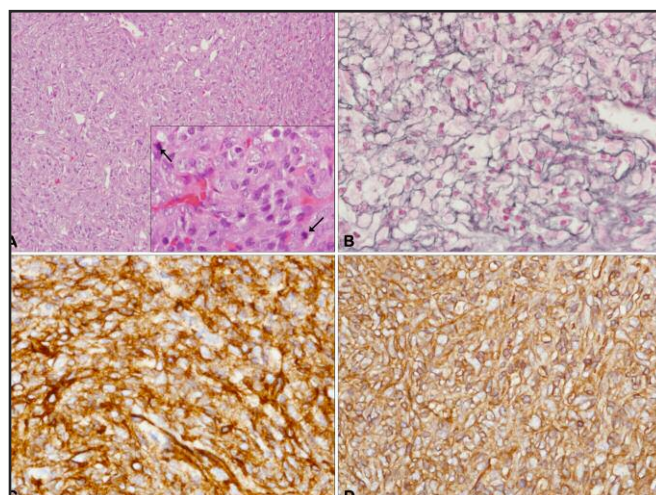
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Corresponding Author: Teoh Jian Woei, Department of Otorhinolaryngology-Head & Neck Surgery Universiti Kebangsaan Malaysia Medical Centre, Jalan Yaacob Latif, Bandar Tun Razak, 56000, Kuala Lumpur, Malaysia

Email: Tjwoei@yahoo.com



**Fig. 1:** MRI of the cerebellopontine angle (CPA). (a) axial T2WI, (b) axial T1WI, (c) axial and (d) coronal T1 WI post gadolinium images. A lobulated extraaxial lesion occupying the left CPA is present, widening the left porus acusticus and internal auditory canal (white arrow). The curved arrow shows the normal right internal auditory canal. Homogeneous enhancement is seen following gadolinium administration.



**Fig. 2:** Histopathological findings of the tumour. (A) Histology of the tumor composed of neoplastic cells with oval to elongated nuclei and vesicular chromatin pattern (H&E stain, original magnification, x200). The mitotic figures (→) are seen (H&E, x600, inset). (B) Numerous slit-like vascular channels are present with a network of reticulin fibres investing the tumor cells (Reticulin stain, x600). The tumor cells are immunoreactive for CD34 (C, x600) and CD99 (D, x400). Using immunohistochemistry (IHC), the neoplastic cells are positive for vimentin, CD34 (Fig. 2C) and CD99 (Fig. 2D).

Any other tumors arising from the CPA might give rise to similar clinical findings but the presenting symptoms might differ slightly in accordance to the location and size of the lesion. The commonest presentations are hearing loss, tinnitus, vertigo, headache, facial hypesthesia and facial weakness. In acoustic neuroma, the most common and earliest presentation would be disturbances of acoustics with 95 percent would have hearing loss. Meanwhile, only 9 and 6 percent of acoustic neuroma patients suffer from trigeminal palsy and facial paresis.

HPCs of the CPA are indistinguishable radiographically from other common CPA lesions like acoustic neuromas or meningiomas. Most lesions present as a broad base lesion, with dural tail sign and heterogeneous enhancement. Subtle radiographic findings allow the differentiation between HPCs and meningiomas such as the absence of calcification and bony hyperostosis, which favor HPCs. Multicentric lesions point toward meningioma, rather than HPC. Acoustic neuromas are typically centered on porus acusticus causing widening of the internal auditory canal, without evidence of dural attachment.

Histopathologically, it is a solid vascular tumor with extensive reticulin network and prominent staghorn sinusoids. Meanwhile psammoma body and nuclear pseudoinclusion are the features that can be found in meningioma. Using immunochemistry, HPCs are positive to vimentin, factor XIIIa, CD-34, CD-99 and Leu-7<sup>1</sup>. Meningiomas are positive to vimentin, epithelial membrane antigen (EMA), protein S-100, neuron-specific enolase (NSE) and Leu-7 antibodies.

Treatment would involve complete gross surgical resection with post operative radiotherapy has the highest overall survival rate. Stereostatic radiosurgery might be recommended in recurring tumor less than 3cm<sup>1</sup>. Intracranial recurrence after excision was noted in the mean period of 12 months to 104 months<sup>5</sup>. Recurrence mainly occurred at primary site but some might showed diffused leptomeningeal spread. Meanwhile 12.9 to 36.4 percent of the intracranial lesions might have extracranial spread. The most common sites of metastasis are lung, bone and liver<sup>5</sup>. Survival rate was five years, ranging from 65%-81.8%<sup>5</sup>.

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