directed cranially to enable the injected blood to track upwards and reach the cervical CSF fistula. However, this argument is deemed less likely as the laterality of symptoms and delayed presentation cannot be explained by this mechanism.

New onset neurological deficit after epidural blood patch may cause much distress to both the patient and the surgeon. The priority is to rule out compressive [9] or ischaemic [12] lesions by an urgent spinal MRI and brain CT scan. These lesions may be treated by surgery, systemic steroid, inotropes or anti-thrombotics. T1-, T2- and diffusion-weighted MRI should provide adequate information in the urgent setting. Electrophysiological investigations may be of interest but these take time to arrange and perform. It is highly likely that new neurological symptoms without imaging abnormalities will improve with time. This complication after epidural blood patch is fortunately rare and runs a benign course.

## Conflicts of Interest/Disclosures

The authors declare that they have no financial or other conflicts of interest in relation to this research and its publication.

http://dx.doi.org/10.1016/j.jocn.2015.03.014

## References

[1] Miyazawa K, Shiga Y, Hasegawa T, et al. CSF hypovolemia vs intracranial hypotension in "spontaneous intracranial hypotension syndrome". Neurology 2003;60:941–7.
[2] Schievink WI, Meyer FB, Atkinson JL, et al. Spontaneous spinal cerebrospinal fluid leaks and intracranial hypotension. J Neurosurg 1996;84:598–605.
[3] Wilkinson HA. Lumbosacral meningismus complicating subdural injection of "blood patch": case report. J Neurosurg 1980;52:849–51.
[4] Olsen KS. Epidural blood patch in the treatment of post-lumbar puncture headache. Pain 1987;30:293–301.
[5] Waguri N, Tomita M, Hayatsu K, et al. Epidural blood patch for treatment of spontaneous intracranial hypotension. Acta Anaesthesiol Scand 2002;46:747–50.
[6] Horlocker TT, Wedel DJ. Neurologic complications of spinal and epidural anesthesia. Reg Anesth Pain Med 2000;25:83–98.
[7] Kane RE. Neurologic deficits following epidural or spinal anesthesia. Anesth Analg 1981;60:150–61.
[8] Epstein NE. The risks of epidural and transforaminal steroid injections in the Spine: Commentary and a comprehensive review of the literature. Surg Neurol Int 2013;4:S74–93.
[9] Tekkök IH, Carter DA, Brinker R. Spinal subdural haematoma as a complication of immediate epidural blood patch. Can J Anaesth 1996;43:306–9.
[10] Kalina P, Craigo P, Weingarten T. Intrathecal injection of epidural blood patch: a case report and review of the literature. Emerg Radiol 2004;11:56–9.
[11] Weir B, Grace M, Hansen J, et al. Time course of vasospasm in man. J Neurosurg 1978;48:173–8.
[12] Huntoon MA, Martin DP. Paralysis after transforaminal epidural injection and previous spinal surgery. Reg Anesth Pain Med 2004;29:494–5.

# Multimodality management of a giant skull base hemangioendothelioma of the sphenopetroclival region

Amol Raheja [a], Ashish Suri [a,*], Shuchita Singh [b], Rajeev Kumar [b], Rakesh Kumar [b], Aruna Nambirajan [c], Meher C. Sharma [c]

[a] Department of Neurosurgery and Gamma Knife, All India Institute of Medical Sciences, Room 712, Ansari Nagar, New Delhi 110029, India
[b] Department of Otorhinolaryngology, All India Institute of Medical Sciences, New Delhi, India
[c] Department of Pathology, All India Institute of Medical Sciences, New Delhi, India

ARTICLE INFO

*Article history:*
Received 22 November 2014
Accepted 3 March 2015

*Keywords:*
Epithelioid
Hemangioendothelioma
Multimodality management
Skull base

ABSTRACT

A 20-year-old man presented with proptosis, nasal obstruction, vision loss and cavernous sinus syndrome, ongoing for 6 years. Imaging and biopsy confirmed a middle skull base epithelioid hemangioendothelioma arising from the left sphenopetroclival region with infratemporal fossa and intracranial-intradural extension into the left temporal lobe. Preoperative embolization of the left internal maxillary artery followed by a combined neurosurgical (front-temporal orbito-zygomatic craniotomy) and otorhinolaryngology (maxillary swing) approach was performed for tumor debulking. Postoperative radiotherapy and maintenance interferon chemotherapy was given to achieve a favorable outcome at 6 months follow-up. We describe the pertinent clinical, genetic, radiological and histopathological features, along with the available therapeutic modalities for a primary giant skull base hemangioendothelioma.



## 1. Introduction

Epithelioid hemangioendothelioma (EH), coined by Weiss and Enzinger [1] in 1982, is a rare vascular tumor (<1% of all vascular tumors) with histopathological and biological behavior between a benign hemangioma and malignant angiosarcoma [2]. EH is primarily a disease involving the viscera, including the lungs and liver, and involvement of the skin, bone, calvarium, brain, spinal cord and cavernous sinus is unusual [3]. As a site of origin of primary EH, the skull base is exceedingly rare with only 11 patients reported to date [4–14]. The continuous spectrum of differentiation and paucity of literature are a hindrance to delineating initial therapeutic protocols and adjuvant therapies thereafter. Recently, Tanas et al. [15] identified the characteristic t(1;3)(p36;q25) chromosomal translocation in EH and the disease defining fusion of the *WWTR1*/*CAMTA1*genes by integrating transcriptomic sequencing with conventional cytogenetics. Herein, we describe a young man who presented with isolated, solitary, primary and giant EH of the middle skull base

* Corresponding author. Tel.: +91 98 68398240; fax: +91 11 26588663.
*E-mail address:* surineuro@gmail.com (A. Suri).

involving large areas of the sphenopetroclival bones with infratemporal fossa and intracranial-intradural extension to the temporal lobe which was managed using a multimodal approach to achieve a favorable outcome at 6 months follow-up.

## 2. Case report

A 20-year-old man presented with progressive headache and non-axial proptosis for 6 years along with a recent onset of left sided nasal obstruction, vision loss, facial numbness and diplopia in the left lateral gaze in the previous 4 months. Examination confirmed the absence of light perception, pan-ophthalmoparesis and fifth nerve distribution hypoesthesia (75% loss to all modalities) on the left side. Radiological imaging revealed a large middle skull base lesion with intracranial extension (Fig. 1a–d). A diagnostic nasal biopsy confirmed that the lesion as EH (Fig. 2). Preoperative embolization of feeders from the left internal maxillary artery was performed to reduce tumor vascularity. Subsequently, a combined neurosurgical (front-temporal orbito-zygomatic craniotomy, anterior clinoidectomy and antero-lateral trans-cavernous approach) and otorhinolaryngology (maxillary swing procedure) approach was performed simultaneously to achieve approximately 80% tumor resection, as confirmed by postoperative MRI (Fig. 1e, f; Fig. 3). The man's neurological recovery was uneventful with acceptable cosmesis and no added deficits postsurgery. He was maintained on nasogastric tube feeding for 2 weeks in view of the partial dehiscence of the palatal mucosa and to prevent palatal fistula formation. Radiotherapy (60 Gy) and maintenance interferon chemotherapy was implemented postoperatively to achieve a stable tumor volume and a favorable outcome was recorded at 6 months follow-up. He resumed his college education shortly after being discharged with no limitations to his activities of daily living.

## 3. Discussion

Predominantly a disease of infants and young adults, intracranial EH has a higher incidence in boys during infancy (3.5:1), but no sex predilection in adults [14]. Reported incidences of local invasion, local recurrence, distant metastasis and mortality rates in intracranial EH are 32%, 24%, 15% and 15%, respectively [14]. The main reported contributing factors for mortality are massive

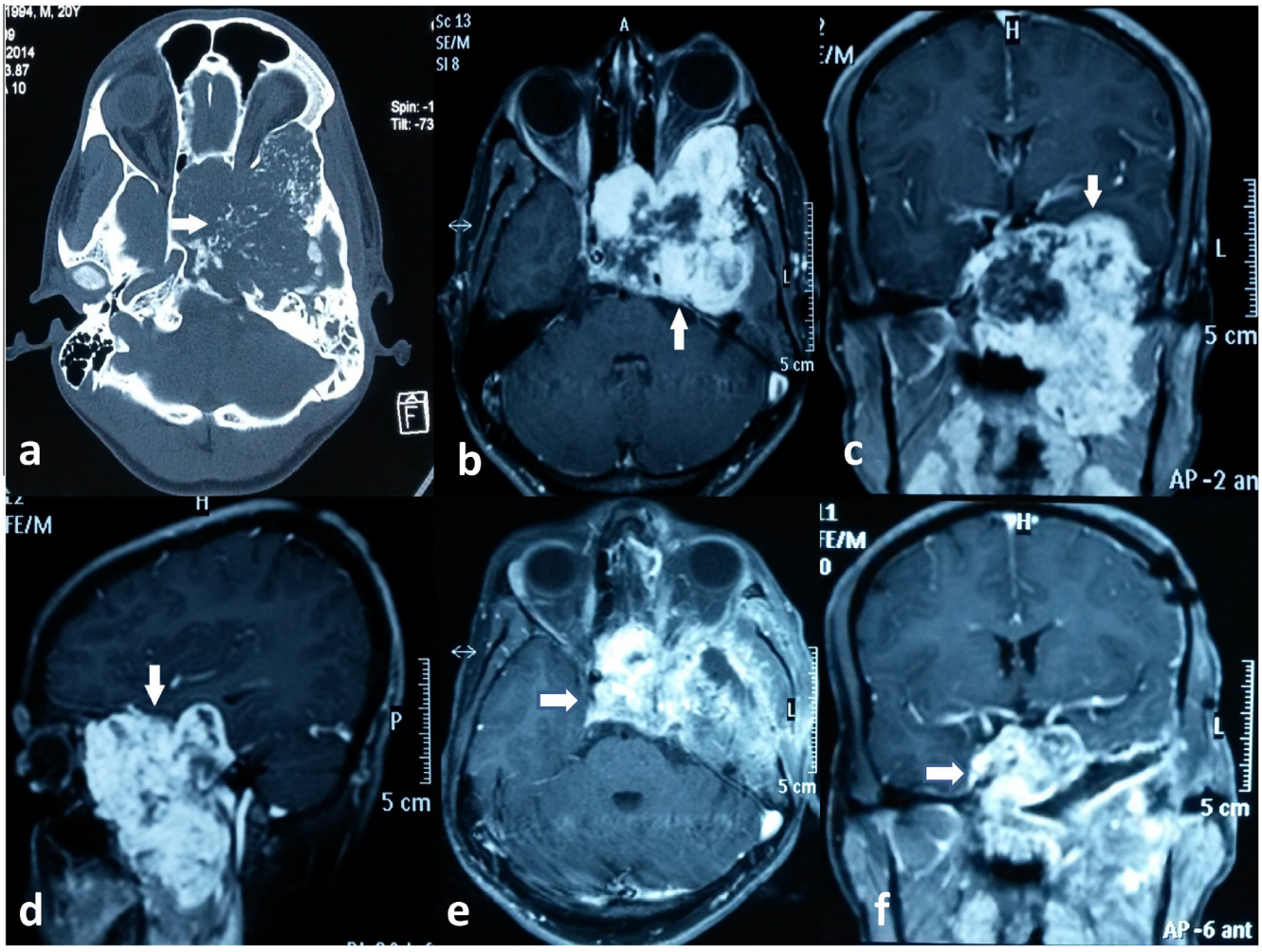


**Fig. 1.** High resolution axial CT scan (a: bony window) showing an expansile osteolytic lesion involving the sphenopetroclival (SPC) region with sclerotic edges and specks of calcification giving rise to a honeycomb appearance with trabeculations (arrow). Preoperative T1-weighted contrast enhanced brain MRI, (b) axial, (c) coronal and (d) sagittal, revealed a giant (8.2 × 6.2 × 6.2 cm) middle skull base lesion arising from the left SPC region with extension into the posterior nasopharynx, pterygo-maxillary fissure, inferior orbital fissure and infratemporal fossa and intracranial-intradural extension into the left temporal lobe. The lesion (arrow) demonstrated intense heterogenous contrast enhancement along with partial encasement of the ipsilateral internal carotid artery. Postoperative T1-weighted contrast enhanced brain MRI, (e) axial and (f) coronal, demonstrated residual tumor (arrow) along the cavernous sinus and petrous bone adjacent to the internal carotid artery.