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Case report

Staged multi-modality treatment approaches for giant cerebellopontine angle hemangioblastomas

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ABSTRACT

Giant hemangioblastomas (HBs) located in the cerebellopontine angle (CPA) present rare, high risk neurosurgical challenges. En bloc resection has been traditionally recommended for HBs, however this approach may pose unacceptable risk with giant tumors. Alternative treatment strategies have not been well described and the relevant literature is scant. This case review includes an illustrative patient with a giant, symptomatic CPA HB. It was felt that the neurovascular and tumor attributes were favorable for a multi-modality treatment strategy rather than circumferential dissection to remove this formidable tumor. A staged approach consisting of preoperative HB devascularization, debulking and piecemeal resection followed by radiosurgery for a small residuum produced an excellent clinical outcome. Variations of this unconventional multi-modality strategy may reduce the perioperative morbidity of carefully selected patients with giant CPA HBs. A thorough literature review is provided.

1. Introduction

Hemangioblastomas (HBs) are uncommon central nervous system (CNS) tumors that account for approximately 2% and 10% of all intracranial and posterior fossa neoplasms in adults, respectively [1,2]. These slow growing lesions have a grade 1 designation by the World Health Organization (WHO) and occur sporadically or in association with von Hippel-Lindau (VHL) syndrome [3]. The histogenesis aligns with a vascular lineage, as HBs are composed of a dense capillary network containing vacuolated stromal (neoplastic) cells [3]. HBs are typically intra-axial lesions located within the cerebellum, brainstem or spinal cord. Exophytic variants within the cerebellopontine angle (CPA) are exceptionally rare and easily mistaken radiologically for common regional pathologies, such as vestibular schwannoma or meningioma [4].

The structure of HBs often contains a highly vascularized tumor nodule associated with a non-neoplastic cyst. In many cases, cyst decompression creates a working corridor adequate to circumferentially dissect and remove the solid tumor en bloc. Safe excision is facilitated by sequentially cauterizing and dividing feeding arteries, leaving large draining veins intact until the arterial supply is extinguished [5]. This strategy, akin to that used for arteriovenous malformations (AVMs), is generally recommended over piecemeal resection in order to avoid profuse intraoperative hemorrhage [5–11]. Less common HB variants do not possess an extramural cyst. The feasibility of safe circumferential dissection of these solid or heterogeneous HBs depends upon tumor size, location and extent of surgical exposure. Large or giant (i.e., >4 cm) HBs within the CPA present rare, extraordinary neurosurgical challenges owing to the intense vascularity and adherence to eloquent structures. The standard retrosigmoid craniotomy used for piecemeal resection of common CPA pathologies provides limited exposure for circumferential dissection of giant HBs, mandating the use of more technically-demanding and higher risk skull base procedures [7,10,12–18].

There are no guidelines to direct HB treatment and non-traditional surgical strategies have not been well described. The current report summarizes the case of a patient with a giant, symptomatic CPA HB. A simplified operative strategy was sought in order to avoid the perioperative risk associated with an expanded surgical exposure and circumferential tumor dissection. This clinical account and case review contribute to the scant literature and support the use of staged, multi-modality treatment approaches in carefully selected patients with these challenging tumors.

2. Illustrative case

2.1. Clinical history

A 45-year-old, married mother of two presented with progressive headaches and three months of gait instability, diplopia and vertigo. She described a sensation of right aural fullness, without

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tinnitus or hearing loss. Her preoperative audiogram was normal and a complete neurological exam was notable only for gait ataxia and nystagmus. The patient's past medical history was non-contributory and there was no family history or personal stigmata of VHL. She was a non-smoker and did not consume alcohol.

Magnetic resonance imaging (MRI) showed an avidly enhancing, solid and cystic mass of approximately 4.2 cm diameter in the right CPA. The tumor produced marked compression of regional structures with associated cerebellar edema, tonsillar herniation and hydrocephalus (Fig. 1). There was no dural tail or extension into the internal auditory canal. The prominent tumor vasculature seen on MRI prompted digital subtraction angiography (DSA) which showed robust, dysplastic tumor feeders from the superior and inferior branches of the right superior cerebellar artery (SCA), with arteriovenous shunting to a large vein that drained into the right transverse and sigmoid sinuses. There was also avid vascularity to the superior aspect of the tumor from a prominent inferolateral trunk arising from the right cavernous internal carotid artery (ICA; Fig. 2).

2.2. Staged tumor embolization and resection

Polyvinyl alcohol (PVA) embolization was initially performed to occlude the superior and inferior SCA feeders to the tumor. Cannulation of the ICA branch that irrigated the superior pole of the HB was deemed unsafe and this vessel was left patent (Fig. 2). Embolization was well tolerated and a standard retrosigmoid craniotomy was performed two days later. As expected, mass effect from the tumor prohibited safe circumferential dissection, but the posterior HB capsule was effectively exposed following mannitol administration, mild hyperventilation and cerebrospinal fluid drainage through a ventricular catheter. There was no readily accessible cyst that could be fenestrated to reduce tumor volume. The capsule was interrogated for the facial nerve, then cauterized and incised in a safe region. Despite embolization, there remained substantial blood supply from the ICA. Piecemeal debulking was performed using sharp dissection, ultrasonic aspiration and electrocautery. The mass was progressively reduced, the brain-tumor interface developed and local cranial nerves protected. The lesion arose from the cerebellar hemisphere and extended superiorly through the tentorial incisura where it was densely adherent to the trochlear nerve and parent SCA. A near total resection was

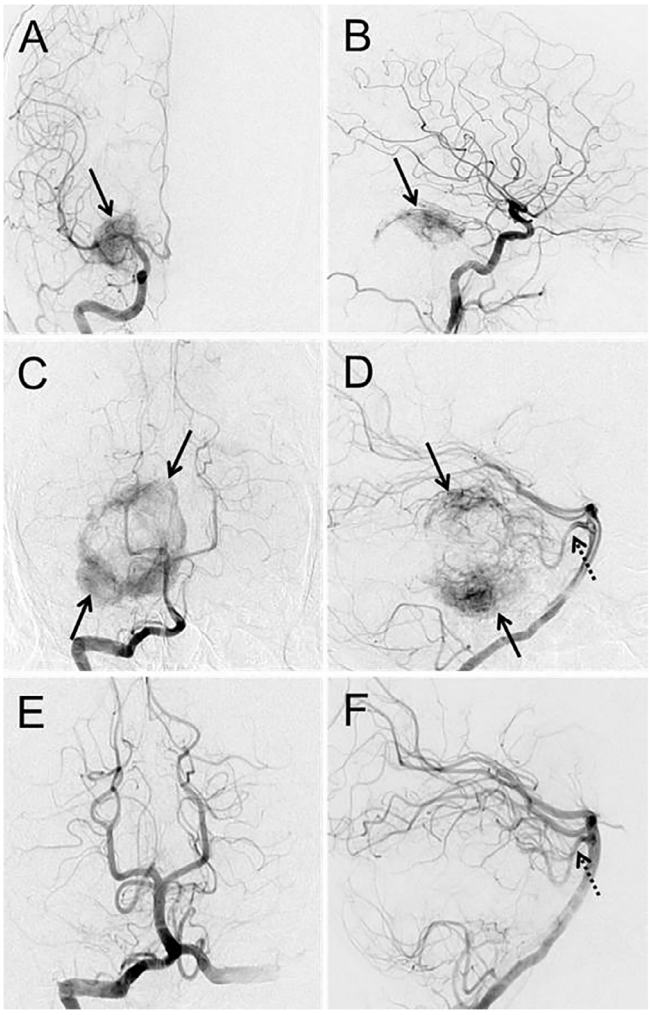

Fig. 2. Preoperative angiography and HB embolization. Anteroposterior (A, C, E) and lateral (B, D, F) angiographic views showing the robust tumor blood supply from the cavernous ICA (A, B) and SCA (C, D). The ICA feeder was not embolized, however the large branches from the SCA to the superior and inferior aspects of the tumor were effectively occluded using PVA particles 2 days prior to resection (E, F). The parent SCA remained patent (dashed arrows). Solid arrows mark the estimated borders of the giant HB.

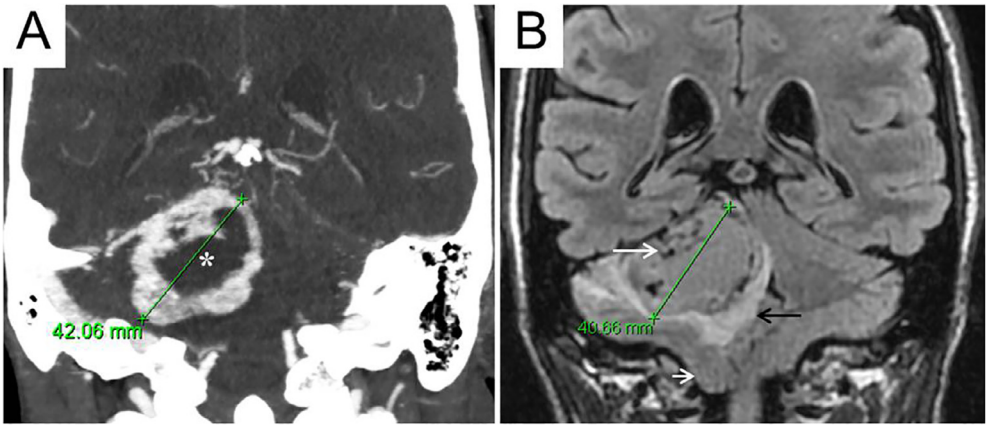

Fig. 1. Radiological imaging of a giant CPA HB in the current patient. (A) Coronal enhanced CT scan of the right CPA tumor showing intense contrast uptake. There was a central cystic region (asterisk) surrounded by thick hypervascular parenchyma. (B) Coronal MRI using a fluid-attenuated inversion recovery sequence. Note the prominent flow voids (white arrow) that, together with the intense enhancement, raised suspicion of a HB diagnosis. The lesion exerted marked mass effect and edema on the surrounding cerebellum (black arrow) and produced tonsillar herniation (short arrow).

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