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Case Report

Hypervascular vestibular schwannoma: A case report and review of the literature ☆,☆☆

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ABSTRACT

Vestibular schwannomas, also known as acoustic neuromas, are benign tumors that arise from Schwann cells near the transition from glial cells to Schwann cells. While most vestibular schwannomas are hypovascular tumors, a small percentage constitute the hemorrhagic and/or hypervascular vestibular schwannomas (HVS) subtype. We describe a case of a 36-year-old female who presented with nausea, vomiting, and an acute decrease in vision in her right eye. Computed tomography of the head demonstrated a hemorrhagic lesion in the right hemisphere with evidence of ventricular effacement. Follow-up magnetic resonance imaging revealed a mass in the right cerebellopontine angle that was hypointense on T1-weighted imaging and mild hyperintense heterogeneous signal on T2-weighted imaging, suggestive of a hemorrhagic vestibular schwannoma. It is important for radiologists to recognize the unique clinical and radiological features of HVS in the initial diagnostic assessment of cerebellopontine angle tumors and to distinguish it from common (hypovascular) vestibular schwannomas and other related pathologies. A preoperative diagnosis of HVS allows clinicians to become familiar with the unique characteristics of the tumor and to devise a feasible operative strategy prior to surgical resection.

Introduction

Vestibular schwannomas (VS) are benign tumors of the vestibular portion of cranial nerve (CN) VIII and have an estimated incidence of 11-13 cases per million every year [1–3]. Hemorrhagic vestibular schwannomas (HVS) are a much rarer phenomenon with an average incidence of approximately 2 cases per year [4]. The clinical manifestation of a unilateral VS at the cerebellopontine angle (CPA) is most commonly ipsilateral sensorineural hearing loss secondary to CN VIII impairment, but varies significantly based on mass effect, location and size of the tumor, and which cranial nerves are compressed. VSs are hypovascular and therefore have a higher tendency to be completely resected. Contrary to this, HVSs are associated with abnormal tumor blood vessels and this additional vascularity may result in excessive bleeding during surgical resection. We present the unique clinical and radiological findings of a case of a patient with an HVS.

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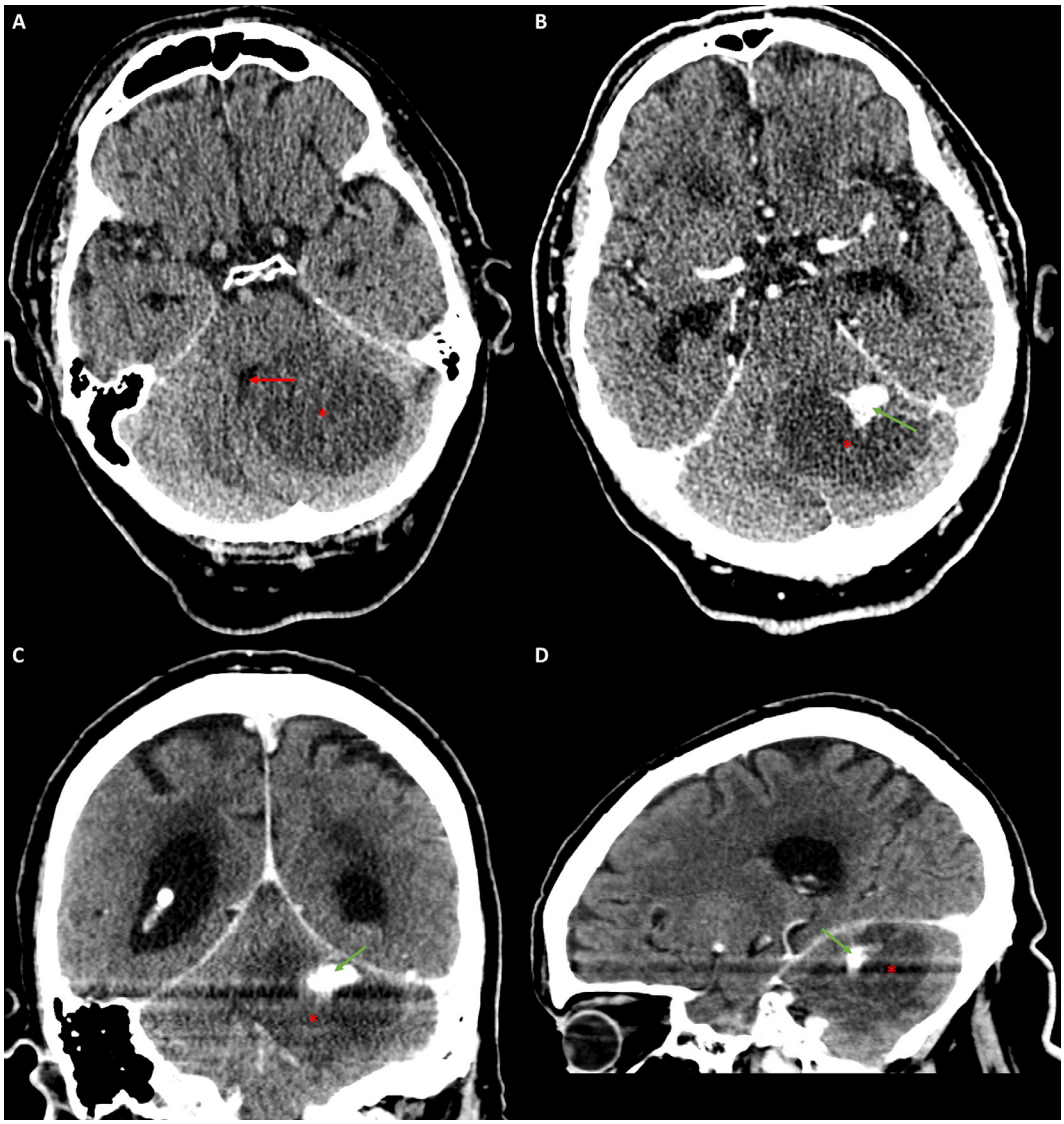

Fig. 1 – CT of the head at presentation demonstrating a large hypodense lesion in the left posterior fossa (red asterisks), with an associated mural nodule along the superolateral margin (red arrow; A). The mural nodule demonstrated homogeneous postcontrast enhancement (green arrow; B-D) (Color version of the figure is available online.).

Case presentation

A 36-year-old female presented to the emergency department with decreased vision in her left and right eye upon waking. For the past 3 days, she had intermittent visual field changes, nausea, and vomiting. Neurologic exam revealed House Brackmann (HB) Grade II right facial dysfunction, ptosis of the right eye, right horizontal nystagmus, and bilateral dysmetria. The patient denied gait difficulties, tinnitus, hearing loss, diplopia, and poor coordination. She had no cutaneous signs of neurofibromatosis and no pertinent past medical, family, or surgical history. The patient was not taking antiplatelet or anticoagulant medication. Computed tomography (CT) of the head with and without intravenous contrast (Fig. 1) revealed a large, nonenhancing hypodense and/or cystic lesion based in the left hemisphere of the cerebellum measuring 3.8 × 5.1 × 3.2 cm. Along the superolateral margin of the hypoattenuating lesion, there was a 0.7 × 1.0 × 0.9 cm mural nodule with avid homogeneous postcontrast enhancement. There was mass effect on the surrounding structures and partial effacement and rightward displacement of the fourth ventricle. The third ventricle and bilateral lateral ventricles were dilated (right greater than the left), consistent with hydrocephalus. The basal cisterns were patent.

Follow-up magnetic resonance (MR) imaging of the brain with and without intravenous contrast (Fig. 2) demonstrated an extra-axial tumor in the right CPA measuring 3.4 × 3.7 × 4.0 cm with acute intratumoral hemorrhage. The mass demonstrated hypointensity on T1-weighted images and mild hyperintense heterogenous signal on T2-weighted sequences. There was diffuse heterogeneous enhancement of this lesion with punctate foci of artifact on the susceptibility-weighted images (SWI), consistent with intratumoral hemorrhage.

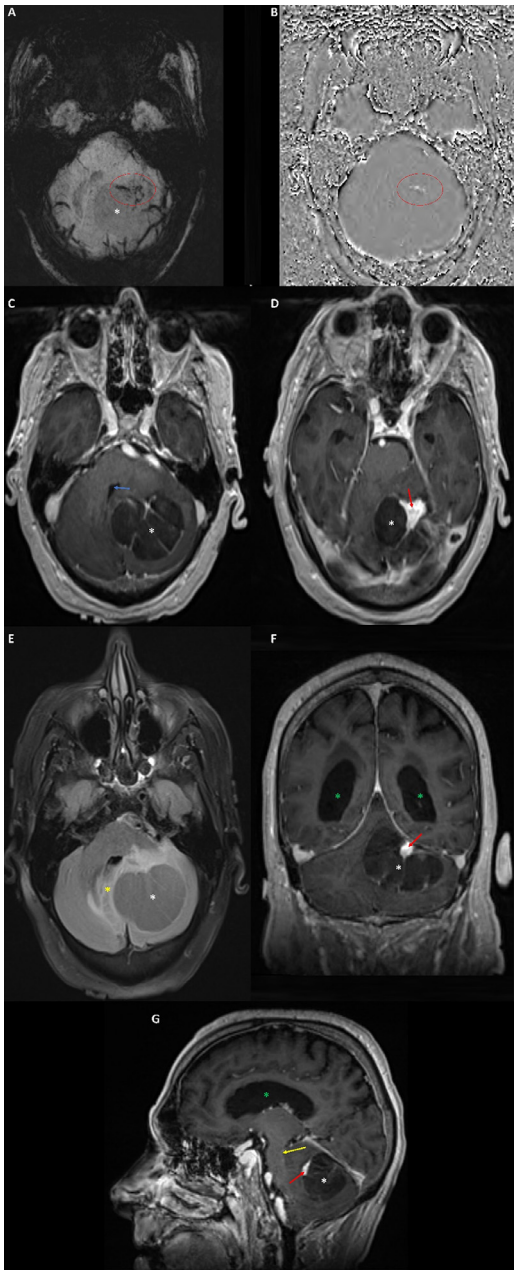

Fig. 2 – Multiplanar MR imaging of the brain redemonstrating a large cystic lesion with thin septations in the left aspect of the cerebellum (white asterisk; A-G). SWI and phase imaging sequences demonstrate increased signal in the anterior aspect of the lesion, suggesting hypervascularity (red circle; A, B). T1-weighted imaging with contrast demonstrates effacement of the fourth ventricle (blue arrow; C) and redemonstration of the mural nodule (red arrow; D). The cystic lesion was associated with surrounding edema (yellow asterisk; E) on the T2/FLAIR imaging sequence. Coronal (F) and sagittal (G) planes using T1-weighted imaging with contrast demonstrate a mural nodule (red arrow; F, G), bilateral lateral ventricle hydrocephalus (green asterisks) and mass effect on the left aspect of the brainstem, including the inferior midbrain and left posterolateral pons (yellow arrow; G) (Color version of the figure is available online.).

Furthermore, SWI demonstrated microcalcifications within the mass as there was decreased signal in the high phase filtered images. The mass was seen extending into the right internal auditory canal as demonstrated by widening of the porus acusticus. There was significant mass effect and effacement of the right inferior midbrain, pons, and the superior margin of the medulla. Mass effect and effacement of the fourth ventricle with upstream obstructive hydrocephalus and partial effacement of the right quadrigenminal and perimesencephalic cisterns was seen. There was increased T2-weighted and fluid-attenuated inversion recovery (FLAIR) hyperintensity along the periventricular white matter, concerning for transependymal flow of the cerebrospinal fluid (CSF). Crowding at the level of the foramen magnum secondary to downward displacement of the cerebellar tonsils was seen. A heterogeneously enhancing mass in the right CPA was most suggestive of a hemorrhagic vestibular schwannoma. A meningioma was determined to be lower on the differential diagnosis primarily due to the lack of intratumoral calcifications on the CT.

To relieve the obstructive hydrocephalus, a ventriculoperitoneal shunt was placed to drain excess CSF from the right lateral ventricle, near the foramen of Monro. Following the shunt placement, the patient denied nystagmus and dysmetria and had subjective improvement in her left visual field. An audiogram reported severe to profound right-sided hearing loss and predominantly mild left hearing loss with a mild 8000 Hz increment. Tympanograms were consistent with normal middle ear functionality bilaterally.

The patient underwent transradial diagnostic cerebral angiogram to evaluate the vascularity of the tumor and to identify potential targets for embolization to reduce her surgical bleeding risk. Selective right external carotid artery angiography demonstrated hypervascular blush of the right CPA mass with brisk rapid contrast filling and venous drainage primarily filling the ascending pharyngeal artery. Additionally, selective right vertebral artery angiography demonstrated brisk filling of the right CPA mass with hypervascular contrast blush and brisk venous drainage into the deep venous system primarily from the right anterior cerebellar artery system. Therefore, it was determined that the hypervascular right CPA tumor had vascular feeders from the right ascending pharyngeal and right anterior inferior cerebellar artery systems.

Two weeks later, the patient underwent surgical resection of the hypervascular vestibular schwannoma via a right retrosigmoid (suboccipital) approach. She recovered well and was subsequently discharged. Post-operative CT without contrast showed expected postoperative changes including small extra axial hematoma and pneumocephalus along the craniectomy site. MR imaging with contrast demonstrated postoperative changes within the right posterior fossa with interval improvement of the extra-axial hematoma and mass effect on the fourth ventricle. The crowding and degree of cerebellar tonsillar descent through the foramen magnum was also significantly decreased. There was redemonstration of the right residual tumor extending into the right internal auditory canal and right CPA adjacent to the right brachium pontis now measuring 3.1 × 2.6 cm. The patient presented 3 days later to an outside hospital with early postoperative complications of headache, potential cerebrospinal fluid (CSF) leak,

pneumocephalus in the subarachnoid and subdural space, and compressive optic neuropathy. The latter of which was likely due to increased intracranial pressure from the chronic obstructive hydrocephalus. The otorhinolaryngology service was consulted, and the right mastoid region was determined to be the source of the CSF leak and pneumocephalus. This finding was supported by MR imaging which showed that the mastoid air cells allowed an egress of CSF into the middle ear cavity and mastoid effusion. This was thought to be secondary to drilling of the internal auditory meatus. This complication was repaired with a mastoidectomy (status post right transpetrosal repair).

To date, the patient's neurologic examination reveals HB Grade IV facial dysfunction, complete right-sided hearing loss, and right eye visual deficits. Recent surveillance imaging demonstrates a gradual decrease in residual tumor with complete resolution of the hematoma within the tumor.

Discussion

Tumors located at the CPA account for 5%-10% of all intracranial tumors. Among these, VS is the most common (75%-85%) CPA tumor[5]. VS may be further classified as hypovascular or hypervascular and/or hemorrhagic tumors. The majority of vestibular schwannomas are hypovascular and can be fully resected, with an estimated hemorrhage rate of less than 1% [6,7]. Most VS cases are characterized as slow growing tumors with low recurrence rates and therefore carry a favorable prognosis. In contrast, HVS represents a rare subtype that is characterized by a VS with a high concentration of abnormal vessels that are branches of both the external carotid artery system and vertebral basilar system [8]. This hypervascularity produces distinct clinical features and operative challenges due to excessive tumor bleeding during surgery [9].

Niknafs *et al.* presented a literature review of 39 cases of intratumoral hemorrhage in VS which found that the occurrence of CN VII palsy was higher among patients with HVS (33.3%) compared to those with non–HVS (6%) [10]. Mortality was more frequently seen in cases of HVS (10%) compared to their non–HVS cohort (0.2%) and tended to occur more commonly in younger patients [8,10]. Large hypervascular tumors are associated with higher rates of surgical morbidity and mortality due to the difficulty in distinguishing the plane between the pia mater and tumor as large tumors can displace the brainstem and cerebellum [11,12]. These data reveal that patients with HVS are more likely to present with neurologic deficits and ultimately may experience worse outcomes than patients with non–HVS lesions [10].

Given the distinct approach and challenges for VS compared to HVS, it is imperative that radiologists provide an accurate pre-operative diagnosis. Initially, patients will typically receive either CT or MR based on their presenting symptoms. Due to the superior contrast resolution, MR imaging is the gold standard in evaluating suspected VS lesions [11,13]. Radiologically, MR imaging of a VS demonstrates a T1-weighted isointense mass relative to the cerebellar parenchyma and demonstrates avid enhancement on postcontrast T1-weighted imaging [11,13]. Intralesional hemorrhage may be represented by T1-weighted hyperintense signal and susceptibility artifacts on T2-weighted gradient-echo sequences [11,13]. Larger VS lesions may often have inhomogeneous enhancement secondary to intralesional hemorrhage and/or cysts. Contrast enhancement may extend into the modiolus secondary to infiltration into the cochlea [13]. While HVS may present with intratumoral hemorrhage, it may be distinguished from VS by tumor hypervascularity, which is characterized by multiple, multi-directional flow voids. Such literature insights corroborate with the radiological findings of our case.

Digital subtraction angiography (DSA) is the gold standard for detecting and evaluating HVS [9,14] HVS will exhibit strong contrast uptake due to the significant vascularity. While less invasive methods, such as with CT or MR angiography or MR venography, are suitable for obtaining accurate static images of the cerebrovasculature, they have not been found to provide an accurate assessment of the tumor vascularity. However, positron-emission tomography, single-photon emission CT, CT perfusion, and MR perfusion studies are able to accurately assess tumor vascularity [15]. In particular, arterial spin imaging (ASL), a technique used in MR perfusion, has been found to accurately evaluate the degree of blood flow in several primary tumors [15–19]. ASL is a noninvasive technique that does not require intravenous contrast or tracers as it relies on magnetically labelling intravascular protons as endogenous tracers [15].

In a study of 103 patients with VS that underwent ASL imaging and DSA prior to surgical resection, it was found that the mean relative tumor blood flow (rTBF) was significantly higher compared to non–HVS tumors. The authors concluded that rTBF as calculated by ASL imaging correlated with VS tumor vascularity [15]. Furthermore, since VS growth rate correlates with the expression level of angiogenic mediators, which in turn correlates with vessel density (as defined by CD31 staining of endothelial cells), ASL may predict tumor growth rate and recurrence rate in VS [20–23].

Due to the distinct differences between HVS compared to VS, several adjunctive treatment strategies for HVS have been developed. Prior to resection, patients may receive preoperative irradiation or preoperative embolization to reduce vascularity and risk of intraoperative hemorrhage. While preoperative irradiation reduces tumor vascularity angiographically and pathologically, the subsequent resection is reported to be more technically difficult, tedious, and aggressive secondary to adhesions. The resection is further complicated by a change in tumor color and an increased risk of cranial nerve ischemia [24–27]. Preoperative embolization to reduce vascularity is increasingly studied. However, embolization is technically difficult given that the tumor blood flow is often from small, thin, tortuous feeders from the pial artery derived from the vertebral basilar system. When successful, the residual tumor tissue is often small and softened secondary to necrosis [17,28,29].

Staged resection, in which there is a deliberate partial resection of the tumor, may be useful in reducing surgical complications. Staged resection results in the residual tumor becoming soft, necrotic, and avascular, therefore allowing for a potentially safer secondary resection [30]. This technique is controversial despite studies showing it to be clinically and pathologically effective. Piecemeal resection may result in in-

traoperative cerebral edema, further obstructing the already small operative window [8,12,28,31]. Furthermore, this surgery is very invasive and requires that the patient undergo at least 2 craniotomies under general anesthesia in a short period of time [8,14,15].

Conclusion

HVS represents a very rare phenomenon and is clinically significant given its association with an increased risk of neurologic deficits, including facial nerve dysfunction. HVS may carry a worse prognosis compared to VS. It is important for radiologists to provide an accurate preoperative diagnosis of HVS as it has important clinical implications and alters surgical management. While MR imaging is vital for initially characterizing VS lesions, further evaluation using DSA or ASL is needed to assess for tumor vascularity.

Patient consent

No consent obtained for this case report as this is a retrospective study with no patient identifiers.

"Formal consents are not required for the use of entirely anonymized images from which the individual cannot be identified - for example, x-rays, ultrasound images, pathology slides or laparoscopic images, provided that these do not contain any identifying marks and are not accompanied by text that might identify the individual concerned."

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