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Hemorrhagic vestibular schwannoma: a case example of vestibular apoplexy syndrome. Illustrative case

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BACKGROUND Acute intratumoral hemorrhage within a vestibular schwannoma, or vestibular apoplexy, is a rare condition. Unlike the typical insidious vestibulopathy typically caused by vestibular schwannoma growth, patients with vestibular apoplexy have an acute and severe presentation with nausea and emesis in addition to severe vertigo and hearing loss. Here, the authors present an illustrative case demonstrating this rare clinical condition and an operative video detailing the surgical management.

OBSERVATIONS A 76-year-old man presented to the emergency department with acute-onset dizziness, left-ear fullness, double vision, gait ataxia, emesis, and facial numbness. Imaging revealed a 2.8-cm hemorrhagic left cerebellopontine angle lesion extending into the left internal auditory canal, consistent with hemorrhagic vestibular schwannoma. The patient subsequently underwent a retrosigmoid craniotomy for resection of the hemorrhagic mass, and by 1 month after surgery, all his presenting symptoms had resolved, allowing his return to daily activities.

LESSONS Vestibular schwannomas typically present with decreased hearing and chronic vestibulopathy. Acute presentation should raise the suspicion for an apoplectic event, and surgical debulking may lead to improvement in most vestibular symptoms.

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KEYWORDS vestibular apoplexy; vestibular schwannoma; hemorrhagic

Vestibular schwannomas are benign lesions that lead to a myriad of progressive symptoms, including hearing loss, dizziness, tinnitus, and headaches.[1–3] An apoplectic event (i.e., intratumoral hemorrhage) is rare but can lead to sudden onset of vestibular symptoms, hearing loss, and facial numbness.[4,5]

Management typically involves resection of the tumor with the goal of functional preservation of facial nerve function. Here, we discuss the presentation and management of a patient with vestibular schwannoma apoplexy with intraoperative demonstration of surgical management (Video 1). When presented with a patient experiencing acute or subacute onset of severe vestibulopathy and lateralized pain, a hemorrhagic vestibular schwannoma should remain on the differential diagnosis, and surgical debulking should be considered to optimize patient recovery.

VIDEO 1. Clip showing the removal of the hemorrhagic tumor. Click here to view.

Illustrative Case

A 76-year-old man with a history of well-controlled hypertension and hyperlipidemia presented to an outside hospital emergency department with acute onset of dizziness, double vision, headache, left-ear fullness, left facial paresthesia, gait ataxia, and emesis. His examination was notable for decreased left facial sensation and decreased left-sided hearing. Magnetic resonance imaging (MRI) showed a peripheral enhancing 2.7 × 2.3 × 2.5–cm mass in the left cerebellopontine angle cistern with intrinsic T1 hyperintensity concerning for a left vestibular schwannoma with intratumoral hemorrhage (Fig. 1A and B). The mass extended into the internal auditory canal (IAC) and was associated with displacement of the ipsilateral trigeminal nerve and brainstem compression with partial effacement of the fourth ventricle without evidence of hydrocephalus. A preoperative audiogram showed profound sensorineural hearing loss of the left ear. Speech reception/detection threshold testing for the left ear revealed 55 decibels hearing

ABBREVIATIONS dBHL = decibels hearing level; IAC = internal auditory canal; MRI = magnetic resonance imaging.
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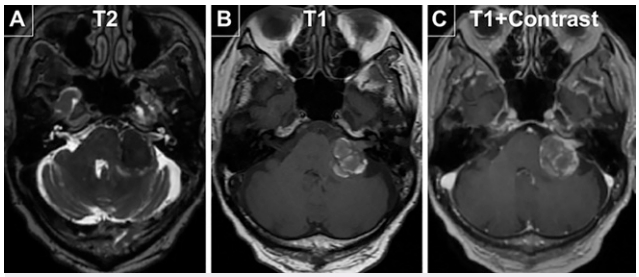

FIG. 1. Preoperative imaging. **A:** T2-weighted MRI shows a hypointense solitary mass compressing the left side of the cerebellum. **B:** T1-weighted imaging shows intrinsic signal concerning for intratumoral hemorrhage. **C:** T1-weighted imaging with contrast shows extension of the tumor into the IAC.

level (dBHL) and 0% speech recognition at 105 dBHL, with no right-ear functional deficits. Overall, the patient's presenting symptoms and imaging findings were concerning for vestibular apoplexy, and he was admitted for management.

Resection was recommended with the goal of maximal tumor removal with facial nerve functional preservation. A retrosigmoid craniotomy was performed to resect the lesion and evacuate the intratumoral hematoma. After exposing the cerebellopontine angle tumor, adjacent subarachnoid hemorrhage and intratumoral hemorrhage were evident (Fig. 2A) in line with the prior apoplectic event. After stimulating the surface of the mass with monopolar electrical current to ensure a safe entry window, the lateral surface of the tumor was entered, revealing a consolidated clot. With the use of an ultrasonic aspirator, the tumor and hematoma were debulked

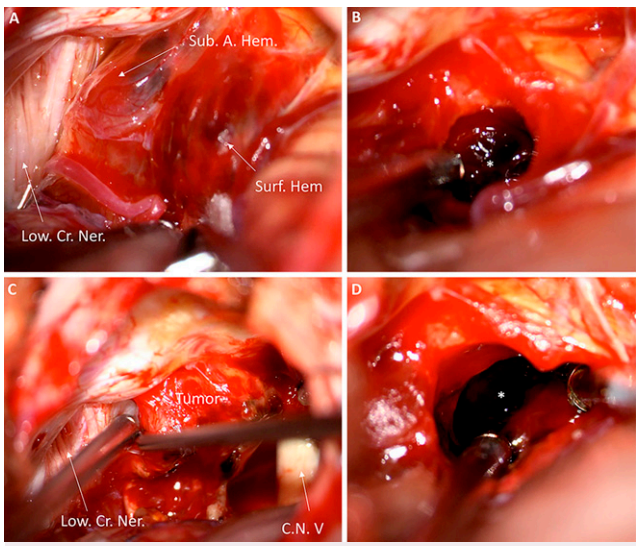

FIG. 2. Intraoperative imaging. **A:** After the tumor was exposed, both subarachnoid and intratumoral hemorrhage was evident. **B:** Debulking of the intratumoral hematoma and tumor proceeded. **C:** The tumor was gradually dissected away from the trigeminal, facial, and lower cranial nerves (Low. Cr. Ner.). C.N. V, cranial nerve V. **D:** Tumor resection of the cisternal component of the tumor was complete, relieving the prior mass effect on the brainstem with preservation of facial nerve stimulation threshold. *Asterisks* refer to the resection cavity (B and D).

with intermittent use of monopolar stimulation to ensure protection of the facial nerve fibers (Fig. 2B and C). Debulking of the cisternal component of the tumor was completed with a residual tumor capsule and no residual mass effect on the brainstem. An adherent tumor capsule to the cisternal facial nerve was left, and there was no change in the stimulation threshold of the facial nerve by the end of the procedure.

The patient's postoperative course was uncomplicated, and his facial nerve function was intact (House-Brackmann grade I). Before discharge, he was ambulating normally and tolerating a regular diet without emesis, with improvement of his vestibular symptoms. A postoperative MRI demonstrated residual tumor within the IAC (Fig. 3A and B), and, at the time of most recent follow-up, the patient had resolution of his vestibular symptoms with preserved facial nerve function (House-Brackmann grade I).

Discussion

Observations

Intratumoral hemorrhage within a vestibular schwannoma leading to acute clinical decline is a rare phenomenon, accounting for $<$1% of all schwannomas.[4–7] Hemorrhage results in acoustic apoplexy syndrome presenting with acute onset of headache, vertigo, gait imbalance, vomiting, and potential cranial nerve deficits. Although many of the same symptoms are similar between hemorrhagic and nonhemorrhagic vestibular schwannomas, the occurrence of sudden headache, acute hearing loss, facial hemiparesis, ataxia, diplopia, dizziness, nausea, and vomiting have been reported to be higher in the setting of hemorrhage.[8] Many of these studies report ataxia, aural fullness, headache, and facial hypesthesia as occurring $>$40% of the time in patients in the setting of hemorrhage. In contrast, nonhemorrhagic vestibular schwannoma cases report these same symptoms $<$10% of the time. In addition, there is a reported 10% risk of mortality in hemorrhagic cases, an important complication when compared with patients with the estimated 0.2% risk of death with nonhemorrhagic vestibular schwannomas.[9–13]

Fewer than 70 cases of hemorrhagic vestibular schwannoma have been reported in the literature, with the incidence estimated to be approximately 2.15 cases per year worldwide.[13] Tumor and patient features that predispose a vestibular schwannoma to hemorrhage are incompletely understood. Hong et al.[14] conducted a retrospective case review series of 53 patients with hemorrhagic vestibular schwannomas that included histopathological analysis, and they found hemorrhage to be associated with highly vascular tissue architecture and larger tumor size. Average tumor size in this series was approximately 3 cm, which is larger than the commonly reported vestibular schwannomas sizes ($<$2.0 cm).[8,13–18] This correlates with the size of the tumor in the present report (2.8 cm). In

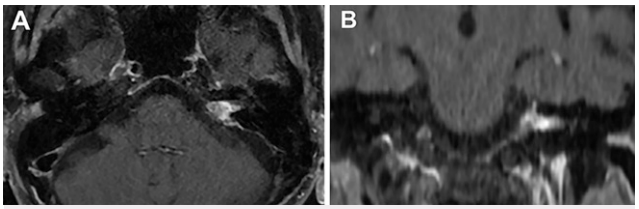

FIG. 3. Postoperative imaging. Axial (**A**) and coronal (**B**) T1 MRI with contrast showing an expected subtotal resection with residual enhancing tumor within the IAC and resolution of brainstem compression.

addition, there has been inconsistent evidence that shows a relationship between various comorbidities, such as chronic hypertension, diabetes, neurofibromatosis, pregnancy, and head traumas, as well as implementation of anticoagulation therapy on the occurrence of hemorrhage incidence.[10,14,19–22] Last, intratumoral hemorrhage is the most common type of bleeding pattern and subarachnoid hemorrhage less so, both of which occurred in our patient.[23]

The management of intratumoral vestibular schwannoma hemorrhage is complex, but our group favors surgical intervention to alleviate the mass effect and accompanying vestibular symptoms that often develop. This is in line with several other groups that have reported on this disease entity.[24–27] Although an adequate comparison of treatment outcomes for medically managed versus surgically operated hemorrhagic cases does not exist, almost all reports proceed with surgical management. Many of the authors have highlighted the decision to operate based on the severe presentation of these patients as well as noted mortality risk outweighing surgical risks and delaying treatment.[4,6,14,18,22,28] If left untreated, there is a potential for worsening edema, mass effect, and rehemorrhage in these patients, and thus observation is not favored.[23,29–32] To the best of our knowledge, there have not been published cases in which observation or radiotherapy was employed as the sole treatment strategy. It is also important to note that the rehemorrhage risk in this population has not been clearly defined.

Resection can proceed with an emphasis on functional preservation of the facial nerve as is typical for nonhemorrhagic vestibular schwannomas. Although intratumoral and subarachnoid hemorrhage from the apoplectic event can provide additional challenges during surgery, our experience has been that aggressive resection and facial nerve monitoring are still possible. Although the estimated preoperative facial nerve deficits that occur in all patients with vestibular schwannomas is approximately 5%, the rate in patients with hemorrhagic occurrences is considerably higher—often 30%–50%.[8,13,33,34] In 2021, Hong et al.[14] published their operative experience with a series of hemorrhagic vestibular schwannomas as well as a systematic review focusing on extent of resection and postoperative outcomes in patients undergoing surgical treatment. Their group observed significantly improved rates of facial weakness (64.1% versus 38.5%; $p = 0.041$) and numbness (46.2% versus 7.7%; $p < 0.001$) in 39 patients treated with resection when compared with their preoperative deficits. Interestingly, an association between extent of resection and symptom improvement was not seen. Surgical outcomes of nonhemorrhagic vestibular schwannomas are similar, with approximately 30%–40% of patients experiencing good facial nerve function postoperatively (House-Brackmann grades I and II), regardless of preoperative deficits.[35,36] Rates of vestibular symptom improvement postoperatively in these patients is unknown, and more work is needed to clearly define expected vestibular outcomes postoperatively.

Lessons

Vestibular apoplexy is a rare complication of vestibular schwannoma presenting with acute and severe symptoms. To the best of our knowledge, Video 1 is the first published intraoperative video outlining the management of a vestibular schwannoma with associated acoustic apoplexy syndrome. The patient had uncharacteristic features of acute intratumoral hemorrhage on a computed tomographic scan for a vestibular schwannoma, with additional subarachnoid hemorrhages discovered intraoperatively. Given the patient's age, he underwent a successful partial resection with, and will receive radiosurgery for, the residual tumor left within the IAC. At last follow-up, the patient's acutely presenting symptoms had resolved.

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Disclosures

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

Author Contributions

Conception and design: all authors. Acquisition of data: all authors. Analysis and interpretation of data: Avalos, Goldschmidt. Drafting the article: all authors. Critically revising the article: all authors. Reviewed submitted version of manuscript: all authors. Approved the final version of the manuscript on behalf of all authors: Avalos. Study supervision: Goldschmidt.

Supplemental Information

Video

Video 1. https://vimeo.com/677735811.

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