Case Report

ANNALS OF MEDICINE & SURGERY

OPEN

Surgical strategies for hemorrhagic vestibular schwannoma with acute neurological decline: a case report with a literature review

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Introduction and importance: Vestibular schwannomas (VSs) are benign tumors of cranial nerve VIII, comprising 8% of primary intracranial neoplasms. Hemorrhagic VSs, though rare, present unique clinical challenges due to their potential for rapid neurological decline.

Case presentation: We present a case of a 68-year-old male with sudden severe headache, nausea, vomiting, and balance issues, initially diagnosed with a hematoma at the right cerebellopontine angle until an magnetic resonance imaging (MRI) scan verified a bleeding schwannoma. Due to deteriorating neurological status and signs of obstructive hydrocephalus on computed tomography (CT), urgent neurosurgical neuromonitoring, successfully managed the hemorrhagic VS, with postoperative recovery intervention required. Neurosurgical treatment, guided by intraoperative outcomes, restoring normal life. Urgent transfer to a medical center with a neurosurgical showing favorable outcomes.

Clinical discussion: Cerebellopontine hematoma verified by CT mandates MRI to exclude tumorous bleeding, which is crucial in cases lacking prior MRI.

Conclusion: Though rare, hemorrhagic VS must be considered in neurologically decompensated patients, especially with risk factors. Timely surgical intervention, despite precipitating a comatose state, can yield satisfactory results.

Keywords: hemorrhagic vestibular schwannoma, hydrocephalus, intratumoral hemorrhage, neurosurgical intervention

Introduction

Vestibular schwannomas (VSs), benign tumors of cranial nerve VIII, constitute 8% of primary intracranial neoplasms, with over 75% located in the cerebellopontine angle (CPA)[1,2]. Typically originating from the superior portion of nerve VIII at the Obersteiner-Redlich transition zone, 5% of cases are associated with type 2 neurofibromatosis[1]. Symptoms, such as tinnitus, asymmetric hearing loss, gait ataxia, vertigo, facial numbness, and late-onset facial weakness due to facial nerve infiltration, commonly manifest when VSs surpass a diameter of 1 cm[1,2]. While intratumoral hemorrhage (ITH) is rare, theories propose vascular wall invasion by tumor cells[3]. Despite slow growth, hemorrhagic lesions can cause abrupt neurological decline, leading to substantial disability and mortality[3].

Case history

A 68-year-old male presented with sudden severe headache, nausea, vomiting, balance issues, tinnitus, and hearing impairment was admitted to the Neurology department of a county hospital. Previously diagnosed with a hematoma at the right CPA, initial management involved vigilant observation. Urgent cranial computed tomography (CT) confirmed the presence of hematoma within the right posterior cranial fossa, exerting pressure on the fourth ventricle, without apparent signs of hydrocephalus (Fig. 1a).

Following our advice via telemedicine consultation, a brain magnetic resonance imaging (MRI) scan delineated a suspected bleeding schwannoma (Fig. 1b). The patient presented with a Glasgow Coma Scale (GCS) score of 15, ataxia with nystagmus towards the gaze direction, mild right-sided facial paresis, and subtle right-sided hearing impairment.

Subsequently, the patient was scheduled for urgent surgery at our department. However, prior to surgery, the patient experienced sudden decompensation to a state of deep somnolence, with CT findings indicative of hydrocephalus (Fig. 2). Upon urgent arrival at the department, the patient exhibited a GCS score of 6 with photoreactive and isocoric pupils.

Urgent surgical intervention ensued, including trepanation at the Kocher point on the right, durotomy incision, and insertion of a ventricular catheter, yielding clear cerebrospinal fluid (CSF) under pressure. Due to the late-night admission, tumor removal surgery was rescheduled for the following day to allow for the accessibility of evoked potentials. The subsequent day, surgery was performed with the patient in a supine position, the right shoulder

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Sponsorships or competing interests that may be relevant to content are disclosed at the end of this article.

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Annals of Medicine & Surgery (2025) 87:355–359

Received 2 September 2024; Accepted 13 November 2024

Published online 9 January 2025

http://dx.doi.org/10.1097/MS9.0000000000002772

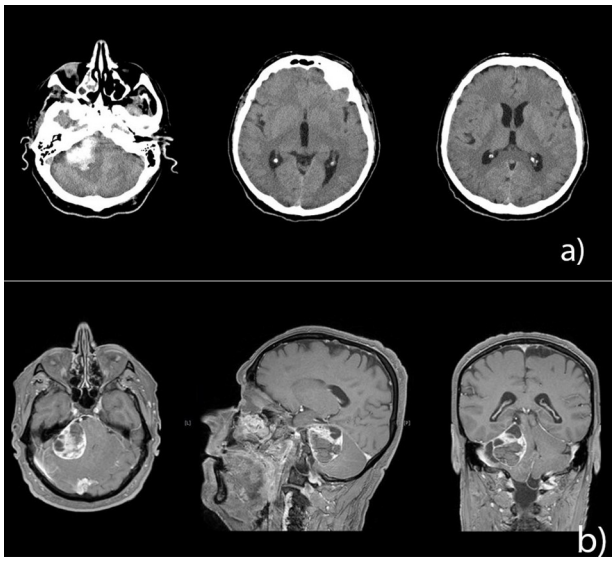

Figure 1. (A) CT scan conducted in a county hospital reveals a hematoma in the right CPA angle without signs of hydrocephalus. (b) MRI with T1 MPRAGE sequence verified the presence of hemorrhage of tumorous etiology (vestibular schwannoma) in the cerebellopontine angle (CPA).

was elevated with silicone under the shoulder and the head turned to the left (semi lateral position), with head immobilization using Mayfield head clamp and a retroauricular incision on the right.

A right retrosigmoid craniotomy was executed, with the evacuation of the intratumoral hematoma and meticulous excision of the tumor abutting the brainstem, facial nerve, and vestibulocochlear nerve complex, guided by evoked potentials. A portion of the tumor adjacent to the internal acoustic meatus was intentionally spared to preserve cranial nerve function. Precise hemostasis was achieved, followed by dural repair and cranioplasty.

Postoperative admission to the intensive care unit revealed a favorable postoperative status on a follow-up CT scan, with no indications of hemorrhage or ischemia. The patient remained verbally responsive with a GCS score of 15 and exhibited no discernible neurological deficits. The external ventricular drain (EVD) was closed on the seventh day and the patient was stable so we removed the EVD on the eighth postoperative day. Subsequent brain MRI confirmed residual tumor remnants adjacent to the right internal acoustic meatus (Fig. 3a and b).

Histopathological analysis confirmed a grade 1 schwannoma according to the WHO classification, with evidence of hemorrhage and abscess formations (Fig. 4). The patient was discharged with a prescribed regimen of dexamethasone and gastroprotection. At the three-month follow-up examination, the patient displayed favorable clinical outcomes with preserved motor and sensory function, House-Brackmann Grade 1, though with persistent hearing impairment.

The follow-up MRI revealed a well-preserved postoperative cavity with stable residual tumor remnants (Fig. 3b). The patient is scheduled for further follow-up assessment, with considerations for gamma knife therapy to address the residual tumor. The work has been reported in line with the SCARE 2023 criteria[4].

Discussion

Hemorrhagic brain tumors, constituting 1% to 11% of intracranial hemorrhages, are linked to higher WHO grades, notably glioblastoma[5,6]. VSs show low bleeding rates globally, about 2.15 cases per year[5]. Yet, ITH within VS may be underestimated. Histological analysis of 274 VS specimens revealed intratumoral microhemorrhage in nearly all cases[7]. In our case, microscopic examination post-tumor resection identified minor hemorrhagic lesions accompanied by scattered siderophages and expanded vascular channels.

The pathophysiology of ITH in VSs remains elusive. It's theorized that multiple hemorrhagic events may lead to cystic VS, seen in 5%–10% of cases with rapid growth and poor surgical outcomes[8,9]. Despite rapid clinical deterioration in our patient, postoperative assessments revealed no major deficits or cystic lesions. Inflammation and angiogenesis likely sustain non-cystic VS growth, even without classic malignancy markers[10]. Sporadic VSs may incite immune responses, fostering chronic inflammation

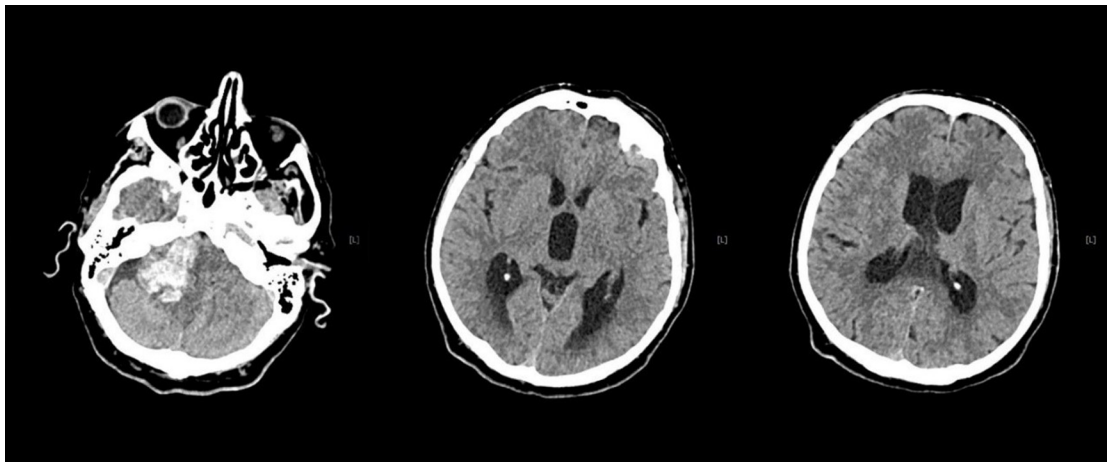

Figure 2. CT scan performed at the county hospital due to the sudden patient's neurological decompensation reveals bleeding in the right cerebellopontine angle (CPA) with enlargement of the lateral ventricles, suggestive of obstructive hydrocephalus.

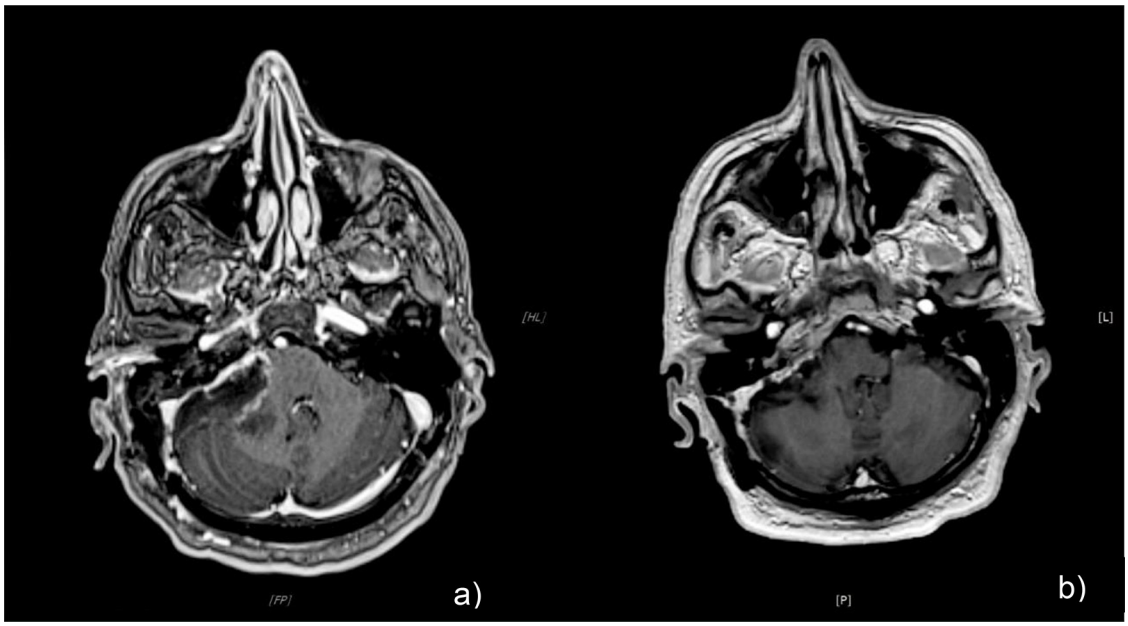

Figure 3. (A) The follow-up brain MRI utilizing T1 MPRAGE sequence immediately post-operation showing no signs of any bleeding or complications. (b) The follow-up brain MRI at 3 months postoperatively, employing the T1 MPRAGE sequence demonstrates the absence of tumor recurrence.

and increased vascular permeability, supported by macrophage infiltration, particularly in Antoni B areas[11,12]. Risk factors for clinically significant ITH include tumor size (>2 cm), accelerated growth, and Antoni A/B area heterogeneity[1-3]. Our case aligns with reported hemorrhagic VS dimensions (3 cm), exhibiting mixed Antoni A/B areas with vascular dilation and abscess formation. Reported predisposing factors for VS hemorrhage include anticoagulant therapy, pregnancy, heavy lifting, strenuous exercise, and hypertension[5,13].

Hemorrhagic VSs pose significant clinical challenges, with symptoms reported in over 40% of cases compared to 10% in non-hemorrhagic VSs, carrying a 10% mortality risk versus 0.2%[8,10,14-17]. This includes severe headache, acute hearing loss, and facial and trigeminal nerve deficits, termed the acoustic apoplexy syndrome[13,18]. The rapid expansion of the tumor due to ITH may contribute to an elevated incidence of deficits evident upon presentation[19]. Our patient presented with acute headache, nausea, vomiting, and balance impairment, deteriorating rapidly to profound somnolence due to obstructive hydrocephalus. Hemorrhagic VSs, while infrequent, may necessitate emergent neurosurgical resection in 5% of cases. In cases like ours, where acute obstructive hydrocephalus is identified as the primary cause of neurological deterioration, expedited EVD may be considered before definitive resective surgery[20]. The average symptom duration for hemorrhagic VSs is 27 ± 12 days, reflecting the rapid development of a more severe clinical course. In contrast, for 89% of patients with typical VS, the interval between symptom onset and diagnosis exceeds 6 months[21].

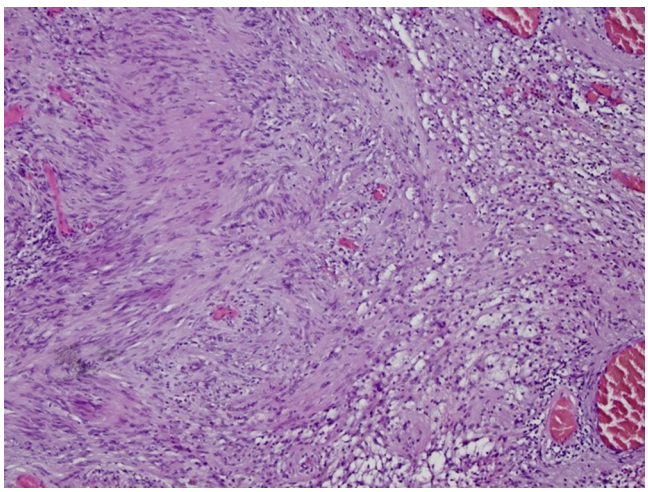

Figure 4. Histopathological exam (HE x100) shows the tumor tissue, which is mostly more cellular (Antoni A areas) and to a lesser extent looser (Antoni B) with hyalinized blood vessel walls.

Distinguishing hematoma from ITH in VS is critical for tailored treatment. Utilizing MRI, we confidently suspected a tumor. CT scans detect recent hemorrhages, while contrast-enhanced MRI distinguishes between hematoma and tumor, showcasing heterogeneous images[21]. Therefore, early MRI is pivotal for etiological investigation[14].

Three standard surgical approaches for VSs include the retromastoid suboccipital (retrosigmoid), translabyrinthine, and middle fossa approaches. Intraoperative neuromonitoring with evoked potentials minimizes nerve injury[8]. The management of hemorrhagic VS primarily involves surgical intervention, although radiation therapy, stereotactic surgery, and observation may be considered in specific cases[5].

In our patient, the initial diagnosis of a CPA hematoma led to a watchful waiting approach until confirmation of hemorrhagic VS and subsequent neurological deterioration. Ventricular catheter placement was necessary due to developed obstructive hydrocephalus. The day after, a right retrosigmoid craniotomy

was performed to remove the hematoma and tumor. Tumor removal was guided by evoked potentials, and a small portion was intentionally left due to its proximity to the facial nerve. The patient was recommended to initiate residual tumor treatment through Gamma Knife therapy.

Despite initial deficits, postoperatively, our patient experienced solely right-sided sensorineural hearing loss, consistent with preoperative status. Preserving hearing poses challenges, with retrosigmoid technique success rates of 20%–40% and middle fossa approach rates of 30%–60%, attributed to cochlear nerve vulnerability. Active treatment, primarily surgical, is vital for improved hearing outcomes[15]. Post-surgery, complications include sensorineural hearing loss (>60%), facial nerve palsy (<10%), surgical site hemorrhage (<3%), and meningitis (<2%). Patients with ITH in VSs postoperatively exhibit sensorineural hearing loss in about 43% and facial nerve palsy in 50% of cases[8]. Long-term follow-ups are crucial for assessing complications accurately. A significant proportion of patients (18%) with fatal hemorrhagic VS experience rebleeding from residual hypervascular tumors, leading to a considerably higher mortality rate compared to non-hemorrhagic VS operations, typically reported at less than 2%[19]. A retrospective study by Gerganov *et al*, which included 400 patients with VS, demonstrated that the development of postoperative hydrocephalus was associated with larger tumor size, irregular tumor surfaces (especially in cases of hemorrhagic VS), and the severity of the preoperative VS. Additionally, patients with a longer duration of symptoms prior to surgery, polycyclic tumors, or inhomogeneous VS were at increased risk for developing CSF leaks[17]. Despite the hemorrhagic nature and severity of the VS in our case, no postoperative hydrocephalus or CSF leakage was observed. Apart from the mentioned hearing impairment, our patient, during regular follow-ups did not exhibit any new deficits or complications.

Conclusion

Patients with hemorrhagic or cystic VS on MRI should undergo intensified surveillance. Confirming CPA hematoma by CT mandates MRI to exclude tumorous bleeding, crucial in cases lacking prior MRI. Though rare, hemorrhagic VS must be considered in neurologically decompensated patients, especially with risk factors. Timely surgical intervention, despite precipitating a comatose state, can yield satisfactory outcomes, restoring normal life. Urgent transfer to a medical center with a neurosurgical department ensures optimal management and potential surgical intervention, crucial for patient recovery.

Ethical approval

Not applicable.

Consent

Written informed consent was taken from the patient prior to the initiation of this project.

Sources of funding

All the authors declare to have received no financial support or sponsorship for this study.

Author contribution

All authors have contributed equally in formation of manuscript.

Conflicts of interest disclosure

All the authors declare to have no conflicts of interest relevant to this study.

Research registration unique identifying number (UIN)

Not applicable.

Guarantor

Bipin Chaurasia.

Data availability statement

Not applicable.

Provenance and peer review

Not commissioned, externally peer-reviewed.

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