

Benign Vestibular Schwannoma: a Rare Cause of Nonaneurysmal Subarachnoid Hemorrhage

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Abstract

Background: Subarachnoid hemorrhage most commonly occurs secondary to trauma, aneurysms, or aggressive tumors. Benign tumors such as vestibular schwannomas are an uncommon cause of subarachnoid hemorrhage. Here we present a case of acute neurologic decline secondary to subarachnoid hemorrhage arising from a benign vestibular schwannoma.

Methods: The methods in this study include case presentation with operative findings and literature review.

Results: A 39 year old male presented with acute hearing loss, severe immediate headache, dizziness, facial numbness, and meningismus. Non-contrast head Computed Tomography (CT) demonstrated subarachnoid hemorrhage. CT angiogram and magnetic resonance imaging revealed a vascular tumor of the cerebellopontine angle with widening of the internal auditory canal, consistent with a vestibular schwannoma. The tumor was approached by retrosigmoid transmeatal craniotomy with complete surgical resection. The pathology revealed vestibular schwannoma.

Conclusion: Although extremely uncommon, benign tumors such as vestibular schwannomas may have an unusual presentation such as spontaneous subarachnoid hemorrhage.

Introduction

Subarachnoid Hemorrhage (SAH) is most commonly caused by trauma, aneurysms, other vascular lesions, and infrequently malignant tumors such as gliomas or metastasis; however, benign intracranial tumors such as meningiomas or schwannomas are an uncommon cause. Vestibular schwannomas, accounting for approximately 8% of all intracranial tumors and 80% of Cerebellopontine Angle (CPA) neoplasms are typified by slow growth and slowly progressive symptomatic presentation such as unilateral hearing loss, or imbalance. Acute presentation of vestibular schwannoma is extremely uncommon and may be associated with Intratumoral Hemorrhage (ITH) or SAH. Here we present a case of a previously asymptomatic gentleman presenting in a manner consistent with SAH, ultimately caused by a benign vestibular schwannoma.

Case Material

This is a 39-year-old male who presented with 1 week of decreased hearing on the left side. He noted intermittent, increasing nausea over that time. Prior to his emergency room presentation, his nausea acutely worsened with episodes of emesis, severe onset headache, left facial tingling and numbness, and left hemibody paresthesias. He took no medications prior to presentation. He did not smoke, did not use drugs, and had no documented hypertension.

Exam: Exam demonstrated a gentleman with an intact sensorium, glasgow coma scale of 15, and his four score was intact. His extraocular movements, visual fields, and visual acuity were intact. He had left V1 thru V3 numbness. Muscles of facial expression were intact bilaterally, giving him a House-Brachmann score of 1. He did however, have dense left sided hearing loss. His tongue was midline, and swallowing was symmetric. He had objective reduction of pin prick on the left hemibody, while strength and reflexes were symmetric. He had demonstrable meningismus.

Radiographic studies

Head CT: Noncontrast CT demonstrated Fischer grade 2 subarachnoid hemorrhages along the left petrous ridge as well as the ambient cisterns (Figure 1). This was in proximity to an extra axial mass in the left Cerebellopontine Angle (CPA) which measured approximately 3.9 x 3.1 x 3.5 cm with widening of the Internal Auditory Meatus (IAC).

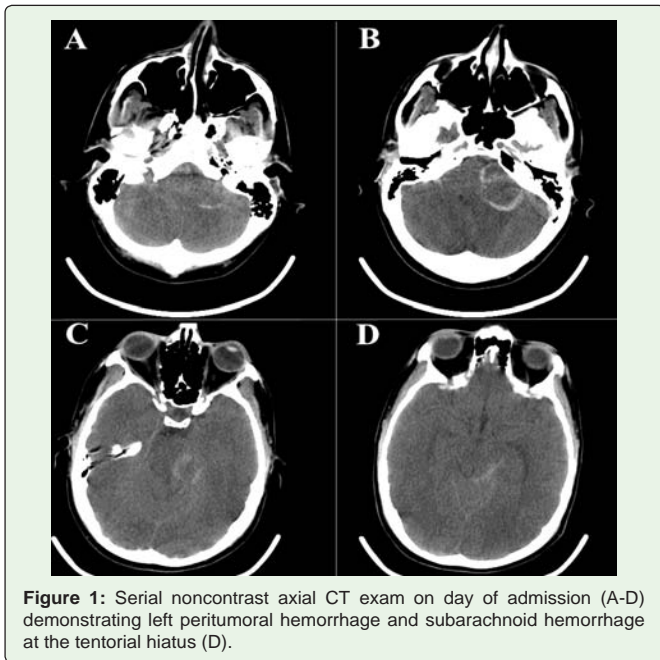


Figure 1: Serial noncontrast axial CT exam on day of admission (A-D) demonstrating left peritumoral hemorrhage and subarachnoid hemorrhage at the tentorial hiatus (D).

CT Angiogram (CTA): The noted left CPA angle tumor was markedly hypervascular with numerous dilated vessels noted within it (Figure 2). The vascular supply was from branches of the left posterior inferior cerebellar artery, superior cerebellar artery and possibly the left posterior cerebral artery. There were no intracranial aneurysms visualized.

MRI/A: Again was confirmed a hypervascular left 3.9 x 3.1 x 3.5 cm cerebello-pontine angle mass with IAC widening consistent with a vestibular schwannoma, there were no intracranial aneurysms visualized (Figure 3).

Surgery: The patient was taken to the operating room within 24 hours and a left sided retrosigmoid transmeatal approach was performed. The tumor was extremely vascular and gross total resection was achieved. Postoperative pathology confirmed

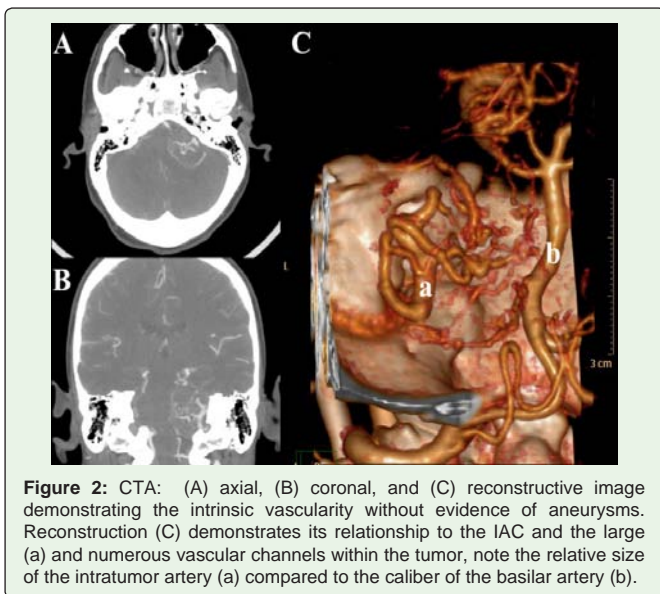


Figure 2: CTA: (A) axial, (B) coronal, and (C) reconstructive image demonstrating the intrinsic vascularity without evidence of aneurysms. Reconstruction (C) demonstrates its relationship to the IAC and the large (a) and numerous vascular channels within the tumor, note the relative size of the intratumor artery (a) compared to the caliber of the basilar artery (b).

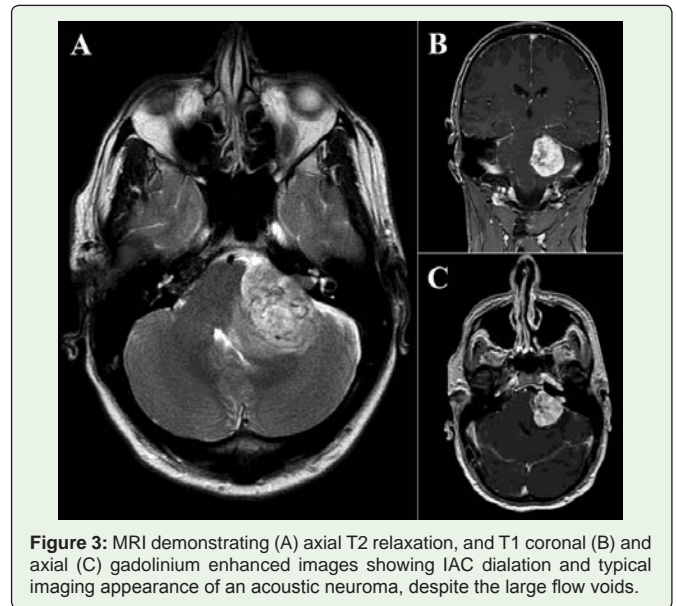


Figure 3: MRI demonstrating (A) axial T2 relaxation, and T1 coronal (B) and axial (C) gadolinium enhanced images showing IAC dilation and typical imaging appearance of an acoustic neuroma, despite the large flow voids.

vestibular schwannoma. Postoperative MRI confirmed a complete resection. During the first postoperative 72 hours, he progressed from immediate postoperative House-Brackmann grade 4 facial weaknesses to grade 6. He was discharge home on postoperative day 3 and had an uneventful recovery.

Discussion

Subarachnoid hemorrhage secondary to an intracranial tumor is rare. Hemorrhage from intracranial tumors accounts for 1-11% of hemorrhage, and is mostly accounted for by aggressive tumors [7,8]. There have been only a few reports regarding subarachnoid hemorrhage associated with vestibular schwannoma as shown in table 1 [1-10]. The first case was discovered at autopsy in 1974 by McCoyd, et al [5].

A literature review of vestibular schwannomas demonstrated 44 cases of documented intratumoral hemorrhages; only 11 of these cases presented with acute SAH (table 1) [1]. Interestingly, there was only one reported death of the 11 cases with documented SAH; the one mortality was from the first reported case by McCoyd [6] in which the tumor was not treated with operative intervention. Most of the 11 cases underwent operative intervention during the hospitalization between post hemorrhage day 4 and 20. Sheppard [7] presented a case in which the patient declined operation and returned 2 years later for operation without complication. Sheppard’s case suggests a relatively more benign course of tumor SAH. Given the extremely rare incidence of SAH with vestibular schwannomas and reviewing the initial plain head CT-scan lead to the intuitive suspicion of aneurysmal SAH due to ruptured large/giant vertebrobasilar aneurysm. However, an aneurysmal source needs to be excluded with CT angiogram or digital subtraction angiogram in such cases. Although the present case was inherently more difficult as all vascular vestibular schwannomas are, the authors did not believe the pre-existent SAH negatively impacted surgery. Additionally, there were no reports of vasospasm or use of calcium channel blockers during hospitalization in the review of the 11 cases. In this case report, there were no documented signs or symptoms suggestive of vasospasm.

Table 1: Summary of VS Patients Presenting with SAH.

Series	Age/Sex	Size (cm)	Presentation	Time to OR	DSA	Outcome
Gavra ³	18F	NR	HA, N/V, MNG	4 days	No	Good
Chu ²	45F	NR	HA, N/V, FN, tinnitus, mild HL	10 days	No	Good
McCoyd ⁶	64F	3.5	AMS, N/V, L hemiparesis	No OR	No	Death
Shephard ⁷	37M	NR	AMS, comatose	10 mos	Yes	Good
Shephard ⁷	66M	3	Ataxia, FN/FW	NR	No	Good
Yonemitsu ^{8,9}	49M	5	HA, N/V	20 days	Yes	Good
Goetting ⁵	19M	3	HA, N/V, FW, otalgia	NR	No	Good
Gleeson ⁴	54F	4	HA, FN, NSG	NR	Yes	Good
Arienta ¹¹	47F	NR	HA, MNG, R FN/FW NSG	NR	NR	NA
Carlson ¹	66M	2	HA, N/V, L FW	NR	No	Good
Present	39M	3.9	HA, N/V, L FN, MNG	1 day	No	Good

Abbreviations: F: Female; M: Male; cm: Centimeters; HA: Headache; N/V: Nausea/Vomiting; AMS: Altered Mental Status; HL: Hearing Loss; L: left; R: right; FN: Facial Numbness; FW: Facial Weakness; MNG: Meningism; NSG: Nystagmus; DSA: Digital Subtraction Angiography.

The mechanisms of hemorrhage of a vestibular schwannoma are not completely understood. There are risk factors that seem to contribute to its occurrence, such as: large size (>2 cm), mixed Antoni type, dilated thin-walled vessels, high vascularity, and the rapid growth of the tumor [2,11]. Some proposed mechanisms to explain hemorrhage in tumors involve vascular endothelial proliferation with subsequent lumen obliteration and distal vessel necrosis, vessel distention and distortion by tumor growth and displacement, and tumor erosion of the vessels [3]. Tumors that are more likely to bleed are rich in endothelial fenestra. Additionally, fenestrated vessels are present in vestibular schwannomas [3,9]. This finding suggests that the key risk factor in determining the risk of hemorrhage is hypervascular vestibular schwannomas, which is the case in our report. Our preoperative imaging suggested an extremely vascular tumor along with the intraoperative resection proved this to be true.

Conclusion

In conclusion, SAH with acute neurological decline is a rare presentation of vestibular schwannoma. Available literature suggests vestibular schwannomas presenting in this manner are hypervascular, as this case demonstrates. Tumor SAH tend to have more benign course than aneurysmal SAH. There have been other proposed risk factor; however, the most recurrent risk factors seem to be the vascularity of the tumor. In such cases, microsurgical resection is the preferred management.

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