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Hemorrhagic vestibular schwannoma: an unusual clinical entity

Case report

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Hemorrhagic vestibular schwannomas are rare entities, with only a few case reports in the literature during the last 25 years. The authors review the literature on vestibular schwannoma hemorrhage and the presenting symptoms of this entity, which include headache, nausea, vomiting, sudden cranial nerve dysfunction, and ataxia. A very unusual case is presented of a 36-year-old man, who unlike most of the patients reported in the literature, had clinically silent vestibular schwannoma hemorrhage. The authors also discuss the management issues involved in more than 1000 vestibular schwannomas treated at their institution during a 25-year period.

Key Words * acoustic tumor * schwannoma * neuroma * hemorrhage * hemorrhagic vestibular tumor

Vestibular schwannomas are the most common tumors found in the cerebellopontine angle (CPA), comprising approximately 80% of all tumors arising in this region. They are usually slow-growing, benign tumors that manifest clinically by directly compressing the neural elements traversing the CPA, including the pons, cerebellum, and the lower cranial nerves. Hemorrhage into a vestibular schwannoma is rare and has been shown to present with acute neurological changes and deterioration. We review the literature on the presenting symptoms of hemorrhagic vestibular schwannomas and describe the case of a man with a large hemorrhagic vestibular schwannoma who presented with signs and symptoms of a slow-growing, insidious compressive lesion. In doing so, we hope to shed light on this unusual clinical entity and discuss management issues.

CASE REPORT

This 36-year-old man had a 6-month history of gradual hearing loss in his left ear. He noted intermittent tinnitus in his left ear and mild intermittent left-sided headaches. His physical examination was otherwise normal. His medical history was significant only for his taking aspirin for pain. Audiography was performed that demonstrated decreased hearing on his left side. A gadolinium-enhanced magnetic resonance (MR) image demonstrated a 2 X 2 X 2-cm peripherally enhancing mass in the left CPA (Fig.

1) with a fluid-filled central cavity consistent with a hematoma. The mass extended into the petrous apex, the pons, and the cerebellar peduncle; it slightly indented the brainstem, but caused no hydrocephalus.

Fig. 1. Magnetic resonance images, axial and coronal views, after administration of contrast material, showing a vestibular schwannoma with intratumoral hemorrhage.

The patient underwent a left suboccipital craniectomy for resection of tumor performed by the combined services of neurosurgery and otolaryngology. Intraoperatively, the tumor was well encapsulated; it grossly compressed the lower cranial nerves and extended from the porus acusticus. After the tumor capsule was entered, the hematoma that had been visualized on MR imaging was encountered. It had components of organized and liquid clot. It was easily removed and a gross-total resection was achieved. The pathological composition of the lesion was that of a schwannoma with no increased vascularity. The patient did well postoperatively.

DISCUSSION

Vestibular schwannomas (acoustic neuromas or acoustic schwannomas) are the most common primary tumors arising from the CPA in adults. Patients usually present with hearing loss, tinnitus, dysequilibrium, headache, and facial numbness. Other more rare symptoms may also manifest such as facial weakness, nausea and vomiting, otalgia, and taste abnormalities. The progression of these symptoms is usually slow and insidious, and often patients will not present to medical attention until their tumors have become quite large.[7]

The tumors' appearance on MR studies is slightly hypo-to-isointense to brain on T_1 -weighted imaging and the signal intensity is decreased on T_2 -weighted imaging. Almost all schwannomas will enhance after gadolinium administration, and the enhancement is homogeneous in 67% of them. There is noted cystic degeneration, but hemorrhage into the lesion is not common.[4,10,12]

Most series have shown that hemorrhagic vestibular schwannomas are rare. Wakai, et al.,[16] demonstrated in their series of 1861 brain tumors that the average rate of all brain tumor hemorrhages

was 5.1%. Their study included 193 vestibular schwannomas, none of which bled. Curati, et al.,[4] also published a series of 35 patients with vestibular schwannomas, and there was no specific mention of hemorrhage in any of them. It appears that the rate of bleeding is low and that most hemorrhagic vestibular schwannomas are presented as case reports.

There is a wide range of presenting symptoms in patients with hemorrhagic vestibular schwannoma, varying from subarachnoid hemorrhage (SAH) symptoms to acute cranial nerve deficits (Table 1).

TABLE 1 PRESENTING SYMPTOMS OF PATIENTS WITH HEMORRHAGIC ACOUSTIC SCHWANNOMAS					
Authors & Year	Age (yrs), Sex	Presenting Symptoms	Acute Onset	SAH*	Prior Insidious Onset
McCoyd, et al., 1974	64, F	nausea, vomiting, lt weakness, mental status changes	Yes	Yes	No
Shephard & Cheeks, 1981	37, M	comatose	Yes	No	No
	66, M	deafness, ataxia, sensory changes, lt facial weakness	Yes	Yes	Yes
Castillo, et al., 1982	61, M	severe rt ear pain, vomiting, dizziness	Yes	Yes	Yes
Yonemitsu, et al., 1983	49, M	sudden headache, nausea, vomiting	Yes	Yes	Yes
Goetting & Swanson, 1987	19, M	lt otalgia, vertigo, vomiting, sweating, facial weakness	Yes	Yes	Yes
Lee & Wang, 1989	65, F	severe headache, vertigo, vomiting	Yes	No	Yes
Brady & Stack, 1994	70, M	diplopia, nausea, headache, facial nerve palsy	Yes	—	Yes
present case	36, M	gradual hearing loss	No	No	Yes
*— = unknown.					

McCoyd, et al.,[11] reported the case of a woman who had tonsillar herniation secondary to hemorrhagic acoustic neuroma. Castillo, et al.,[3] described a man who had "severe pain in the right ear accompanied by dizziness and several episodes of vomiting" and who also had suffered SAH. Shephard and Cheeks[14] reported on two patients: one presented in a coma with SAH and the other with sudden deafness, ataxia, and sensory changes. Yonemitsu, et al.,[18] Brady and Stack,[2] and Lee and Wang[9] all reported cases of patients who presented with severe headache, nausea, and vomiting. Goetting and Swanson[6] treated a patient who presented with acute left-sided otalgia and facial weakness after engaging in weight lifting. Asari, et al.,[1] presented four patients with hemorrhagic acoustic schwannomas: two had developed acute onset of headache and vomiting. Almost all of the case reports of patients with hemorrhagic vestibular schwannomas describe sudden, acute onset of symptoms, neurological deterioration, or frank SAH.

Although most case reports of patients with hemorrhagic vestibular schwannomas cite sudden onset of symptoms and neurological changes, we present a case that was neither symptomatic nor devastating. Our patient's slow, insidious clinical course was very typical of most vestibular schwannomas, and the hemorrhage was silent. The patient does have a history of taking aspirin, which may have predisposed him to bleed into the tumor. Magnetic resonance imaging indicated that the bleeding may have contributed to the expansion of the tumor. The intraoperative exposure demonstrated that there was liquefied and organized clot, suggesting that there may have been multiple bleeds into the tumor at different times.

Of more than 1000 patients treated at our institution over a 25-year period, there have been only a handful with hemorrhagic vestibular schwannomas. In contemporary management of vestibular schwannomas, treatment options consist of observation by means of serial imaging, radiosurgery, or microsurgical resection. The observation option may risk rehemorrhage with acute neurological deterioration as described by other authors (Table 1). The role radiosurgery may play in treating hemorrhagic vestibular schwannomas is difficult to predict. Although radiosurgery appears to be effective in treating highly vascular lesions such as hemangioblastomas[13] or arteriovenous malformations,[15,17] its role in the treatment of many other lesions still needs to be defined.[5,8] We advocate surgical resection of these hemorrhagic tumors by means of either a suboccipital or a translabyrinthine approach. It is our belief that microsurgical resection provides the easiest, most direct route to achieve evacuation of the hemorrhagic component of the tumor, which minimizes risks to critical surrounding nerves and dramatically reduces the mass effect.

CONCLUSIONS

Vestibular schwannomas are the most common primary tumors of the CPA, and they usually present with insidious onset, but without hemorrhage. Almost all cases of patients with hemorrhagic vestibular schwannoma reported in the literature describe sudden, dramatic changes in clinical signs and symptoms. We present a case of a man who appeared to have had multiple bleeds, yet still presented with a slow progressive course. This clinical entity and its management should be of interest to neurosurgeons and neurootologists who may see this unusual presentation.

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