Hemorrhagic Vestibular Schwannoma Presenting with Acute Facial Nerve Palsy: An Unusual Clinical Entity

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Abstract

Sixty six years old female presented with acute onset of Occipital headache, deviation of angle of mouth to left side, giddiness and ataxic gait of one day duration. This was preceded by 2 months history of unsteadiness of gait and mild reduced hearing from right ear. Neurological exam revealed right-sided lower motor neuron facial nerve palsy. Both CT and MRI demonstrated a large cerebellopontine angle tumor with fluid level and intracavitary blood consistent with hemorrhage in acoustic schwannoma. The tumor was excised and histological examination revealed vestibular schwannoma. This is a rare case of isolated hemorrhage into a vestibular schwannoma with atypical acute presentation with ipsilateral facial nerve palsy on the background of hearing loss of short duration. Other differential diagnosis of acute facial nerve palsy may have to be included in such lesions.

Keywords: Vestibular schwannoma; Intratumoral hemorrhage

Introduction

Cerebellopontine Angle (CPA) Vestibular Schwannoma (VS) presenting as sudden onset facial nerve weakness is a rare entity. VS are the most common tumors found in the cerebellopontine angle, comprising approximately 80% of all tumors arising in this region. They are usually slow-growing, benign tumors that manifest clinically by features of chronic compression of the adjacent neural elements traversing the CPA, including the pons, cerebellum, and the lower cranial nerves. They are also known to present acutely in the events of ischemia arising from direct compression of the labyrinthine artery or the Anterior Inferior Cerebellar Artery (AICA). Hemorrhage into VS is known, yet it is a rare form of presentation with acute neurological deterioration.

Case Presentation

A 66 years old elderly female presented with history of vomiting, severe occipital headache, facial deviation of angle of mouth to left, ataxic gait, weakness of right upper and lower limbs of one day duration. Patient also had mild hearing loss in right ear of 2 months duration without associated tinnitus. She was not a known case of hypertension, or on any anti-platelet agents or anticoagulants. On neurologic examination, patient was alert, had right LMN facial paresis (House-Brackman Grade III), with power in right upper limb 4+/5, right lower limb 4+/5 and profound right side cerebellar signs. There were no features of trigeminal nerve or lower cranial nerve compression. A clinical diagnosis of a vascular phenomenon such as AICA aneurysmal bleed, CPA tumor bleed, was suspected. On evaluation, CT brain revealed hyper dense space occupying lesion in right cerebellopontine angle suggestive of hematoma which was extending to ipsilateral quadrigeminal and ambient cisterns (Figure 1). MRI brain showed contrast enhancing, heterogenous lesion in the right cerebellopontine angle with an extension towards internal acoustic meat us with a hemorrhagic component s/o right vestibular schwannoma with hemorrhage (Figure 2). After informed consent, patient underwent Right retro mastoid sub occipital craniectomy and excision of tumor. Intraoperatively, the tumor was well encapsulated; it extended from the porus acousticus and grossly compressed the lower cranial nerves. 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. Clots were easily removed and a gross-total resection of the tumor was achieved. Post operatively patient remained conscious as before and with residual facial nerve paresis (House-Brackman Grade II). She continued to have Sensory Neural Hearing Loss at the time of discharge with right side cerebellar symptoms. Post op CT brain done revealed no residual lesion (Figure 3). She was ambulant with support, taking orally. She was advised facial and limb physiotherapy along with care for right eye from exposure keratitis.
Discussion

Vestibular schwannomas are the most common primary tumors of CPA in adults. Patients usually present with a history of insidious onset, gradually progressive sensory neural hearing loss, tinnitus, headache, disequilibrium, and facial weakness/numbness. Rarely do they present with features of raised intracranial pressure due to mass effect and hydrocephalus. Presentation with lower cranial nerve palsy alone is seldom seen. Most series have shown that hemorrhagic vestibular schwannoma are rare. Wakai, et al. [1], demonstrated in their series of 1861 brain tumors that the average rate of all brain tumor hemorrhages was 5.1% [1]. Other anecdotal case reports of hemorrhagic VS represents patients with simple presentations like headache, nausea, and vomiting to acute onset facial weakness alone, facial with abducens nerve palsy, and with associated trigeminal compression [2-7]. Kim et al. [8], has described an association of large or rapidly growing tumor, mixed Antoni type tumor and tumors with unusually increased vascularity with higher risk of bleed and its acute presentations [8]. A preceding history of hearing loss before an acute onset of facial paresis puts, Bell’s palsy and facial nerve neurona as rare possibility [9]. We describe here a rare case of hemorrhage into a cystic vestibular schwannoma with atypical acute presentation on the background of hearing loss of short duration. The patient presented with sudden onset of occipital headache suggestive of a vascular event. Moreover, there was acute facial nerve palsy on the side of the cyst, which was not viewed as typical signs of a vestibular schwannoma. In our case, the seventh nerve palsy was localized on the ipsilateral side of the tumor. It was suspected that a preexisting non-symptomatic compression of this nerve by the relatively soft cystic tumor was exacerbated by the acute intratumoral hemorrhage causing increase of local pressure in the posterior fossa.

Conclusion

In conclusion, this report describes a rare case of hemorrhage within a cystic vestibular schwannoma appearing on CT and MRI as cyst with fluid level. Although the presented case seems to be a rare occurrence, it should be nevertheless born in mind when investigating acute facial nerve palsy.

References