Hearing Improvement in a Growing Vestibular Schwannoma

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ABSTRACT

Vestibular schwannomas are benign, slow-growing tumors that originate from Schwann cells lining the vestibular nerves, most commonly the superior vestibular nerve. They arise at the neurilemmal/neuroglial junction which is situated within the internal auditory canal. They have an incidence of 1 per 100,000 per year and a prevalence of around 700 per million. A case of a patient undergoing a period of observation for a vestibular schwannoma whose hearing improved despite growth of the tumor is described. This raises interesting questions regarding the pathophysiology of hearing loss in patients with vestibular schwannomas. Possible hypotheses are discussed.

KEYWORDS: Vestibular schwannoma, acoustic neuroma, hearing, follow-up

Vestibular schwannomas are benign, slow-growing tumors that originate from Schwann cells lining the vestibular nerves, most commonly the superior vestibular nerve. They arise at the neurilemmal/neuroglial junction which is situated within the internal auditory canal (IAC). They have an incidence of 1 per 100,000 per year and a prevalence of around 700 per million.¹

We describe a case of a patient undergoing a period of observation for a vestibular schwannoma whose hearing improved despite growth of the tumor. This raises interesting questions regarding

the pathophysiology of hearing loss in patients with vestibular schwannomas.

CASE REPORT

A 76-year-old man presented to a tertiary referral neuro-otology center with a 2½-year history of a progressive right-sided hearing loss. On pure tone audiometry, there was a severe sensorineural hearing loss with a pure tone average of 72.5 dB HL

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Figure 1 (A) Pure tone audiogram at initial presentation. (B) Pure tone audiogram at 1-year follow-up. A marked improvement in masked bone conduction hearing thresholds is shown.

(calculated according to the guidelines of the American Academy of Otolaryngologists; Fig. 1A). There was no measurable speech perception on speech audiometry. He did not have any other neuro-otological symptoms or signs. A magnetic resonance imaging (MRI) scan showed a tumor within the IAC. It measured 10 mm along the long axis of the IAC, 5.4 mm in an anteroposterior direction (measured at right angles to the longitudinal axis of the IAC), and 7.7 mm craniocaudally. The radiological features were consistent with a vestibular schwannoma (Fig. 2A).

A detailed discussion of the management options available for treating vestibular schwannomas was undertaken and the risks and benefits of surgery, radiotherapy, and a period of observation were outlined. The patient elected to follow

a watch, wait, and rescan policy which initially involved annual MRI scanning.

The first follow-up MRI scan, a year after the patient's initial visit, showed that the tumor had grown from 10 mm to 13 mm in a direction along the axis of the IAC (Fig. 2B). In addition, the tumor had extended a little into the cerebellopontine angle and reached the fundus of the IAC. The basal turn of the cochlea also enhanced on T1 imaging following gadolinium injection suggesting enlargement of the tumor into the cochlea. Paradoxically, the patient's hearing had significantly improved with a pure tone average of 61.25 dB HL (Fig. 1B). His speech discrimination had also improved to 30% at 95 dB. As the speech has improved and the tumor is still 4 mm from the cerebellopontine angle, the patient

Figure 2 (A) Initial T2-weighted magnetic resonance imaging (MRI) scan showing the tumor in the right internal auditory canal. (B) Follow-up T1 MRI scan with gadolinium diethylene triamine pentaacetic acid enhancement after 1 year showing the increase tumor size.

remains in the watch/wait/rescan management protocol.

DISCUSSION

Unilateral sensorineural hearing loss is the primary presenting symptom in around two thirds of patients with vestibular schwannomas.² The natural history of this hearing loss is of a gradual deterioration in thresholds, faster than that seen by aging alone. This occurs irrespective of whether the tumor is growing^{3–5} although, in the presence of tumor growth, the hearing probably deteriorates more quickly.^{4,6} There is no evidence that tumor size per se or location of the tumor within the IAC is related to progression of hearing loss.^{4,7} In the few patients who have tumor shrinkage, most series suggest that any pre-existing hearing loss is not reversed. It should also be noted that 10% of patients may develop a sudden-onset acute sensorineural hearing loss.

Improvement in hearing is very unusual in the presence of a vestibular schwannoma but has been documented with tumors that have been stable in terms of their size.⁴ However, hearing improvement in the presence of tumor growth has not been described previously.

The etiology of hearing loss in vestibular schwannoma is not completely understood. It is a reasonable assumption that pressure by the tumor on the cochlear nerve within the IAC is responsible,

at least in part, for the hearing loss. Badie et al have confirmed that there is a correlation between intracanalicular tumor volume and IAC pressure, although they were not able to demonstrate a significant relationship between pressure and hearing.⁸ Similarly, Nadol and colleagues showed no relationship between lateral extent of the tumor and hearing loss.⁵ Lapsiwala and associates have, however, demonstrated that increasing pressure within the IAC intraoperatively produces increases in the latency of wave V observed during measurement of auditory evoked potentials.⁹

The mechanism by which the tumor influences cochlea nerve function is unclear. The neural injury may be due to direct mechanical injury to the nerve or may result from neurotoxic factors produced by the tumor. Equally, injury may result from compromise of the internal auditory artery as it passes laterally within the IAC. Indeed this is the rationale for dissecting the tumor from medial to lateral to increase the likelihood of hearing preservation in the retrosigmoid approach to vestibular schwannoma excision.

Several publications have suggested that hearing loss in patients with vestibular schwannoma is not just neural in origin but may be, at least in part, the result of secondary cochlea damage^{10–12} or ischemia. Degeneration of the inner and outer hair cells, striae vascularis, and the spiral ligament have been documented in cases of vestibular schwannoma.^{13,14} Similarly, reduced distortion product otoacoustic emissions have been documented

in patients with early hearing loss secondary to vestibular schwannoma, presumably reflecting poor outer hair cell function.¹² The most likely explanation for these changes is a reduction in cochlear blood flow resulting from vascular compression by the tumor.

The mechanism by which the hearing improved in this case is unclear. It serves to illustrate that changes in hearing function are not necessarily the result of anatomical changes in the tumor but may result from pathophysiological mechanisms as yet not fully understood. Moreover, it highlights that, while broad conclusions regarding outcomes can be drawn from the literature, on an individual basis eventual outcomes are difficult to predict.

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