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A small vestibular schwannoma arising from the inferior vestibular nerve

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Abstract
The investigation of a complaint of unilateral right sensorineural hearing loss led to the identification of a right 3 mm vestibular schwannoma arising from the inferior division of the right vestibular nerve. On investigation the patient was found to have normal caloric function, this being mediated by the superior vestibular nerve. Both transient and distortion product otoacoustic emissions were absent in the right ear, and ABR was abnormal on this side. These findings are of interest as they indicate that this small lesion produced a hearing loss that was both cochlear and retrocochlear. The anatomical finding that the medial auditory efferents run within the inferior vestibular nerve is considered. This patient did not experience tinnitus or hyperacusis despite the site of the lesion arising from the inferior vestibular nerve.

Key words: Neuroma, Acoustic; Hearing Loss, Sensorineural

Case report

A 53-year-old male presented initially to the Department of Otolaryngology at his local hospital with a complaint of reduced hearing in his right ear. He described the loss as progressive, and believed that it had started four years previously. He had no significant history of noise exposure in the past. He reported only hearing loss without distortion of sound with no tinnitus, hyperacusis nor balance disturbance. The only feature of note in his past medical history was essential hypertension.

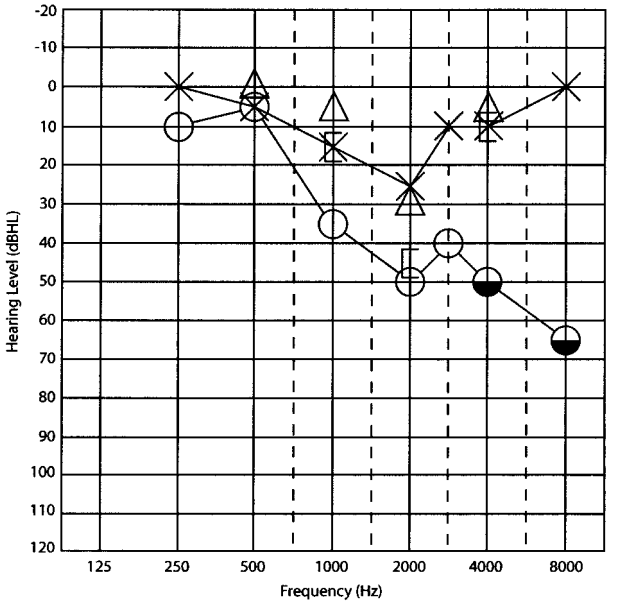

Fig. 1
Pure tone audiogram.

Otological examination was normal, as was cranial nerve examination. The corneal reflex was present bilaterally. On pure tone audiometry a right mild to moderate mid/high frequency sensorineural hearing loss (SNHL) was revealed (Figure 1). Speech audiometry was normal in the left ear, and demonstrated normal reduced maximum speech discrimination (80 per cent) with roll-over in the right ear (Figure 2).

A magnetic resonance imaging (MRI) scan (Figures 3 and 4) demonstrated a 3 mm vestibular schwannoma arising solely from the inferior division of the vestibular nerve. Management of the patient is currently watch and wait with a planned re-scan.

Detailed vestibular investigations were undertaken in order to determine the effect of this lesion upon the clinical vestibular status of the patient. The Romberg test was normal, as was Unterberger's stepping test. Gait was normal as was walking in a line eyes open (WALEO) and closed (WALEC). No indications of cerebellar ataxia were identified. No spontaneous nor gaze evoked nystagmus was evident on clinical observation nor on electronystagmography (ENG). The modified clinical test for sensory

Fig. 2
Speech audiogram.

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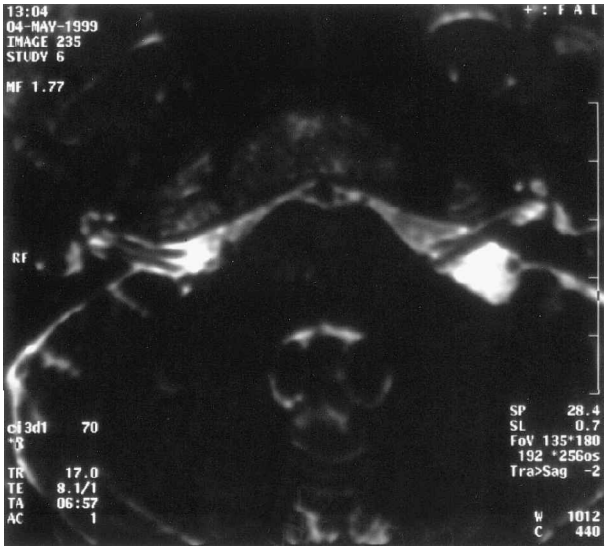

FIG. 3
T2-weighted MRI scan demonstrating inferior vestibular nerve VS.

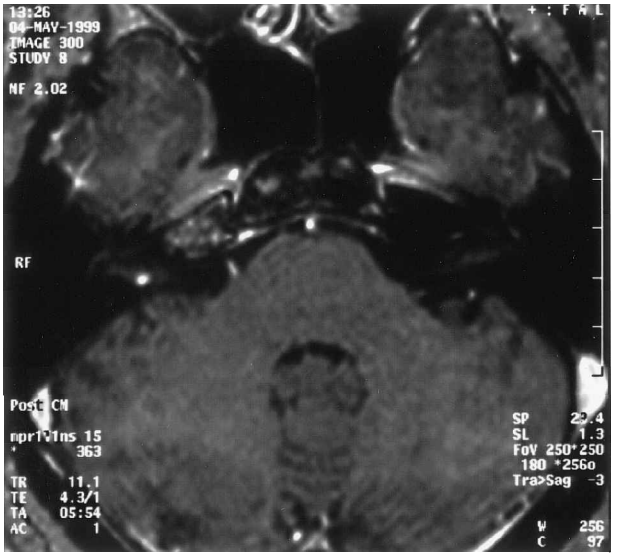

FIG. 4
Gadolinium enhanced T1-weighted MRI scan demonstrating inferior vestibular nerve VS.

interaction of balance (CTSIB)[1] was normal. No abnormalities of conjugate and individual saccades were found on ENG, and smooth pursuit testing was normal. Bithermal caloric tests with water stimuli (30° and 44°C, 20 second flow duration) demonstrated a right cranial paresis of 13 per cent and a directional preponderance of seven per cent, both below the level of clinical significance and hence normal.

Transient (TEOAE) and distortion product otoacoustic emissions (DPOAE) were present in the left ear, but were absent on the right indicative of a cochlear component of the hearing loss.

Auditory brainstem response (ABR) was normal on the left. On the right there was prolonged JIII and JV together with poor morphology of the waveform.

Discussion

This case contains a number of features of clinical interest. In this case a vestibular schwannoma arising from the inferior vestibular division of the vestibular nerve has been identified and investigated. A recent article has demonstrated the origin of a vestibular schwannoma on the superior vestibular nerve.[2] It was previously thought that this was the more common site of origin but it is now suggested that the incidence of schwannomas on the inferior and superior divisions of the vestibular nerve is roughly equal,[3] or greater on the inferior vestibular nerve.[4] Some authors[5] have suggested that the origin of vestibular schwannomas is the vestibular ganglion. The findings in the present case do not support this.

The finding that this patient had normal caloric function is of interest. Caloric testing examines the function of the horizontal semicircular canal which is innervated via the superior vestibular nerve. The results of the caloric tests were normal in this patient demonstrating that this lesion had not impinged upon the superior vestibular nerve sufficiently to reduce caloric function, nor to induce any symptoms of imbalance.

It is also noteworthy that this patient did not experience tinnitus or hyperacusis. The inferior vestibular nerve contains medial and lateral auditory efferent fibres, these having been identified first by Rasmussen.[6] They have been implicated in the modulation of the gain of the auditory system which, in the case of the medial efferent fibres, is by an inhibitory influence upon the outer hair cells (OHCs) (Sahley *et al*).[7] Dysfunction of the medial efferent system has been hypothesized to be a factor in the perception of tinnitus[7–12] and of hyperacusis.[13] Therefore, in the present case, a lesion arising from the inferior vestibular nerve might be expected to cause dysfunction of the medial efferent system, and be associated with tinnitus or hyperacusis. It should be noted that in the presence of a cochlear hearing loss the ability of the efferent system to influence cochlear function would be limited by OHC dysfunction (demonstrated here by absent TEOAE).

Finally there are indications from the audiological investigations that this patient had not only a retrocochlear hearing loss (deduced from the abnormal ABR) but also a cochlear component, demonstrated by the absence of TEOAE and DPOAE together with rollover on speech audiometry. The finding that a proportion of patients with vestibular schwannoma have an associated cochlear hearing loss has been reported previously.[14–16] Moffat *et al*.[17] noted audiological findings that were indicative of a cochlear or mixed cochlear and retrocochlear lesion in 73 per cent of a series of 49 patients with sporadic unilateral vestibular schwannoma. Prasher *et al*.[14] reported absent TEOAE in 19 of 26 patients with vestibular schwannoma (73 per cent): in all these patients in whom TEOAE was absent, a hearing loss of 40 dB or greater was present and was assumed to be cochlear in origin. Telischi *et al*.[18] undertook DPOAE measurements in 44 patients with unilateral vestibular schwannoma. On the basis of the presence or absence of the DPOAE, 26 (59 per cent) tumour ears were classified as having a cochlear loss, 13 (30 per cent) were classified as retrocochlear (DPOAE recorded in the presence of a hearing loss >40 dB) and five (11 per cent) as mixed. Ferber-Viart *et al*.[19] attempted TEOAE recordings in 168 ears with vestibular schwannoma, and in 79 per cent were not able to demonstrate good cochlear function – thus indicating cochlear dysfunction in addition to the hearing loss associated with the tumour. Ferguson *et al*.[15] were unable to evoke TEOAE in 78 patients of a series of 100 with unilateral vestibular schwannoma. Thus a controversial speculation by Bonfils and Uzial[20] that 'acoustic tumours usually produce a cochlear hearing loss' is supported.

Schucknecht[21] considered potential mechanisms for cochlear hearing loss in vestibular schwannoma. Two potential mechanisms were described. The first of these

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was ischaemia causing atrophy of the cochlea and the vestibular labyrinth by compromising blood flow in the labyrinthine artery that runs through the internal auditory canal (IAC). The second was biochemical degradation of the cochlea (which might also affect the vestibular labyrinth). Alterations in the biochemical status of the cochlea in cases of vestibular schwannoma have been reported, specifically increasing protein concentration within the perilymph.[22]

- **Previous cases of vestibular schwannoma have been shown to arise; by MRI, from the superior vestibular nerve**
- **The present case demonstrates a schwannoma from the inferior vestibular nerve. Caloric testing produced normal results indicating that the superior vestibular nerve was not involved**
- **This tiny lesion was associated with indications of both cochlear and retrocochlear hearing loss. The cochlear involvement is noteworthy in a lesion of this size, and the possibility of ischaemia or of biochemical degradation of the cochlea is considered**

Evidence for both ischaemic and biochemical vestibular labyrinth injury in vestibular schwannoma has also been reported by Jahnke and Neuman[23] who studied specimens taken from nine patients during translabyrinthine surgery. Examination with electronmicroscopy demonstrated significant degenerative changes that were thought to be the result of prolonged protein intoxication of the labyrinth (via increased perilymph protein concentrations) and by compression of labyrinthine blood vessels by the tumour. Similar mechanisms were suggested for cochlear dysfunction in these cases.

In addition Prasher *et al.*[14] noted that the influence of a vestibular schwannoma upon the function of the auditory efferent system, and in particular the medial efferent influence upon outer hair cells might cause OHC dysfunction with reduced frequency discrimination, and eventually threshold elevation. Abnormalities have been noted on frequency discrimination tasks in patients with vestibular schwannoma in whom a cochlear component to the hearing loss had been identified.[16] That this is due to medial efferent dysfunction is debatable when one considers the demonstration[24] of normal psychoacoustical hearing abilities on patients following vestibular nerve section and in whom the medial efferent pathway running within the inferior vestibular nerve is hypothesized to have been ablated.

The presence of a mild bilateral notch in the audiogram at 2 kHz is of potential interest. This raises the possibility of an additional aetiology in the hearing status of this patient, perhaps ischaemic due to hypertension or some genetic hearing impairment.

The cochlear hearing loss in the right ear of the present case was associated with a minute tumour (3 mm), and the potential involvement of such mechanisms is worthy of note.

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