THE NEW ZEALAND MEDICAL JOURNAL
Vol 120 No 1265 ISSN 1175 8716

Management of unilateral vestibular schwannoma/acoustic neuroma

Philip A Bird, Martin R MacFarlane

Vestibular schwannomas (VS) or acoustic neuromas (AN) are benign slowly-growing tumours of the vestibular nerve in the internal auditory canal (IAC) and cerebellopontine angle (CPA). The tumours are usually unilateral but can be bilateral in neurofibromatosis type 2 (NF 2) which we will not specifically discuss. These tumours grow from a Schwann cell of one of the vestibular nerves (usually the superior) within the IAC and are correctly classified as a neurilemmoma or schwannoma of a vestibular nerve, although they are commonly called an acoustic neuroma.

The growth of these tumours in the IAC and out into the CPA means that the order of involvement or neural and intracranial structures (and therefore the clinical course) is reasonably predictable—viz: an initial period of silent growth → loss of function of the vestibular nerve of origin and then the associated ipsilateral vestibular nerve, with subtle affects on balance; compression of the ipsilateral cochlear nerve in the IAC → gradual hearing loss (sensorineural) and variable tinnitus; pressure on the trigeminal nerve → ipsilateral facial numbness (hypoaesthesia and hypoalgesia); gradual pressure on the cerebellum and pons → increasing ataxia and ipsilateral cerebellar signs; significant pressure on and stretching of the facial nerve → progressive facial paresis/palsy; significant pressure on and stretching of the glossopharyngeal and vagal nerves → hoarse voice and progressive difficulties with swallowing → aspiration; obstruction to CSF outflow from the 4th ventricle → hydrocephalus, and finally coma and death from a combination of aspiration pneumonia and raised intracranial pressure with coning.

Of course the goal in managing these tumours is to prevent this outcome whilst maintaining quality of life and function of neural structures, especially the nearby cranial nerves.

With increasing awareness of the possibility of unilateral ear symptoms being caused by these tumours, and with the increased availability and accuracy of MRI scanning, more smaller tumours are being diagnosed. Even so, people with unilateral ear symptoms have a <2% chance of actually having a tumour. There is almost certainly an apparent (rather than real) increase in the incidence of vestibular schwannoma from approximately one in 100,000 to one in 50,000.[1–3]

Prior to (and even with) MRI scanning it is likely that many people died/may die with their VS, rather than because of it.

The increase in diagnosed small and medium-sized VS and a better understanding of their natural history has influenced management over the last decade. Management may entail either observation with serial MRI scanning, focused/stereotactic radiotherapy (SR) using X-rays (linear accelerator) or gamma rays (Gamma knife, GN), or surgery incorporating microsurgical techniques. The particular management

modality/treatment selected will depend on the size of the tumour, the age and general health of the patient (and therefore an estimate of their longevity and the risks of surgery), the patient's hearing, other neurological signs, and patient preference. For the purposes of this discussion we will divide the cases according to the size of the largest intracranial (CPA) diameter: small tumours <10mm, medium tumours 10–25mm, and large tumours >25mm.

Small tumours should usually be observed initially, as approximately 80% of these will demonstrate little or no growth over the medium term.[4] Surgical removal when tumours are small (rather than when they are larger) does give the best chance of hearing preservation and excellent facial nerve results, but even the most experienced surgical teams are only able to preserve hearing to preoperative levels in about 60% of cases.[5]

Overall hearing handicap is more strongly related to hearing in the better ear (which is usually very good in these patients) and therefore hearing needs are probably best met by not operating. SR/GN treatment of small tumours carries a very low morbidity and yields excellent results in terms of preventing tumour enlargement, but given the natural history of these tumours almost certainly represents over-treatment in the majority of cases. This treatment does also have an associated morbidity with regard to later deterioration in hearing and facial nerve function.

Medium-sized tumours usually warrant some form of active management in all but the elderly and infirm. A decision between surgery and SR/GN depends on patient age, general health and patient preference. Some patients prefer to have the tumour removed and thus avoid the ongoing monitoring (clinical and MRI) which is necessary after SR/GN.

Others prefer avoiding surgery and are content with ongoing surveillance. SR/GN has an advantage in that it is administered on an outpatient basis and may involve a single treatment or several (fractionated) treatments. In the short-term, this may be a more attractive option than a 4–6 hour operative procedure, a week long stay in hospital and 4–8 weeks off work.

SR/GN fails to halt tumour growth in about 5% of cases over the medium term; it is as yet unknown what the long term (>20 years) results are and there is a very small chance of malignant transformation after SR, but to put this risk in context, it is probably similar to the risk of peri-operative death in a medium-sized tumour.

If surgery is required for SR/GN failures, it is more difficult with worse facial nerve outcomes.[6] If conservative management is undertaken it is important to understand that postoperative functional facial nerve results start to deteriorate when the tumour gets larger than 25mm.

Large tumours should usually be managed surgically to prevent the inevitable compression effects on the cerebellum, brainstem, and lower cranial nerves. SR/GN to lesions >30mm carries the risk of neurological deterioration over the first few months due to associated oedema.

Surgery is generally performed by two surgeons, one an otolaryngologist trained in skull base surgery and the other a neurosurgeon also experienced in such surgery. The operative approach can be translabyrinthine (an incision behind the ear and drilling through the temporal bone with loss of any residual hearing), posterior

fossa/retrosigmoid (an incision a little more posterior behind the ear with the chance of preserving hearing), or by a superior approach (via a small craniotomy through the squamous temporal bone above the ear, also with a chance of preserving hearing).

In the elderly or infirm, elective partial/subtotal removal of tumour rather than complete removal may be indicated, such surgery removing the tumour compressing the brainstem, cerebellum, and lower cranial nerves, but leaving tumour in the internal auditory canal and attached to the facial nerve thus preserving facial nerve function.

In younger patients, complete removal or near complete tumour removal is generally indicated, although if the tumour is particularly adherent to the facial nerve the surgeons may well elect to leave such a small tumour fragment adherent to the nerve rather than risk irreparably damaging or dividing the facial nerve.

Currently in New Zealand SR via a linear accelerator is provided by the Stereotactic Radiosurgery Unit at Dunedin Hospital while Surgery is undertaken in Auckland, Wellington, and Christchurch. An important feature in the assessment and best practice management of patients with VS is that it be undertaken by a team involving both neurosurgeon and otolaryngologist both trained and proficient in the surgical techniques necessary in operating on these tumours.

There has been some publicity in the media over the last few years regarding overseas treatment of these lesions with gamma rays ("Gamma Knife"/GN) but current evidence has not shown this modality to be any more effective than SR using X-rays provided by a linear accelerator (as used in Dunedin).

We do not believe there is significant benefit in New Zealanders seeking overseas treatment, either surgical or SR/GN, for these tumours. In addition to this, there is of course much information on the Internet. Unfortunately, and as all doctors are aware, a lot of the information also relates heavily to advertising and thus the Internet can be an extremely confusing source of information for patients.

Decisions about management of these uncommon benign tumours should be undertaken in centres where they are actively managed. We always endeavour to give patients a full and complete range of information about their tumour including the natural history, expected progression and neurological sequelae and the range of treatment options—i.e. observation/serial imaging, stereotactic radiotherapy and surgery, and this includes information based on our own personal results.

In summary, the treatment of unilateral vestibular schwannoma involves either observation, stereotactic radiotherapy, or surgery. With the larger number of smaller tumours being increasingly diagnosed, more observation is now undertaken.

There is a role for stereotactic radiotherapy, which appears to be effective in the short and medium term, however there are still some unanswered questions about long-term results. For medium-sized tumours in younger people and for large tumours, surgery remains the mainstay of treatment.

Although quite a large number of people with unilateral ear symptoms may be screened to exclude this benign tumour, the majority of scans in these people will be normal.

Competing interests: None.

Author information: Philip A Bird, Otolaryngologist, Martin R MacFarlane, Neurosurgeon, Christchurch Hospital, Christchurch

Correspondence: Mr Philip A Bird, Otolaryngologist, PO Box 4345, Christchurch Hospital, Christchurch. Email: Philip.Bird@cdhb.govt.nz

References:

1. Shin YJ, Fraysse B, Cognard C, et al. Effectiveness of conservative management of acoustic neuromas. Am J Otol. 2000;21:857–62.
2. Tos M, Stangerup SE, Caye-Thomasen P, et al. What is the real incidence of vestibular schwannoma? Arch Otolaryngol Head Neck Surg. 2004;130:216–20.
3. Stangerup SE, Tos M, Caye-Thomasen P, et al. Increasing annual incidence of vestibular schwannoma and age at diagnosis. J Laryngol Otol. 2004;118:622–7.
4. Stangerup SE, Caye-Thomasen P, Tos M, Thomsen J. The Natural History of Vestibular Schwannoma. Otology & Neurotology Inc. 2006;27:547–52.
5. Friedmann RA, Kesser B, Brackmann DE, et al. Long-term hearing preservation after middle fossa removal of vestibular schwannoma. Otolaryngology Head and Neck Surgery. 2003;129:660–5.
6. Slattery WH, Brackmann DE. Results of surgery following stereotactic irradiation for acoustic neuroma. Am J Otol. 1995;16:315–9.

