Acoustic neuroma
an important cause of unilateral hearing loss

Acoustic neuromas are uncommon and their presentations subtle, so a high degree of awareness is needed when evaluating patients with unilateral hearing loss or asymmetrical auditory symptoms. Referral to a centre specialising in the management of acoustic neuroma is required for all patients.

MELVILLE J. DA CRUZ
FRACS, MSc

Dr da Cruz is ENT Surgeon, Westmead Hospital, and Senior Lecturer in Surgery, University of Sydney, NSW.

Acoustic neuromas (also known as vestibular schwannomas) are benign tumours arising from the schwann cell covering of the vestibular nerves. They produce symptoms by compressing the sensitive neural and vascular structures of the inner ear, brainstem and posterior cranial fossa.

The sporadic form of the disease results from an acquired mutation in the gene coding for schwannomas, and has an annual incidence of 1 in 100,000 people. The hereditary form, neurofibromatosis type 2, occurs when the gene mutation is carried in the germline rather than a single cell – bilateral vestibular schwannomas and many other tumours of the central nervous system characterise this form. Neurofibromatosis type 2 is one-tenth as common as the sporadic form of the disease but the disability can be extremely high, including a profound bilateral deafness.

Natural history

Acoustic neuromas generally arise from the vestibular portion of the eighth cranial nerve, which is located within the bony internal auditory canal adjacent to the inner ear; occasionally, they may arise from the cochlear nerve itself. The growing tumour fills the internal auditory canal

IN SUMMARY

- **Acoustic neuromas are benign, slow growing tumours arising from the eighth cranial nerve.**
- **Acoustic neuromas present in the adult years with unilateral hearing loss and tinnitus accompanied by mild balance disturbance. A high degree of awareness is required in order to make an early diagnosis.**
- **MRI scanning is highly sensitive and specific for acoustic tumours. Normal appearances of the inner ear and eighth cranial nerve on a well performed MRI scan can reliably exclude an acoustic neuroma in patients with suspicious symptoms.**
- **For a patient with rapidly progressive symptoms and difficulty in obtaining a specialist referral for MRI scanning, a freely available contrast enhanced CT scan will reasonably exclude the presence of a large tumour with accompanying brainstem compression and hydrocephalus needing urgent treatment.**
- **All patients with acoustic tumours should be referred to a centre specialising in the management of patients with such disorders. The best management option for each patient will depend on symptoms, the size and growth rate of the tumour, his or her age and any comorbidities.**

Downloaded for personal use only. No other uses permitted without permission. © MedicineToday 2001.

Table. Presenting symptoms of acoustic neuroma

- Hearing loss
- Tinnitus
- Balance disturbance
- Headache (hydrocephalus)

and then spills out into the cerebellopontine angle. Further growth results in compression of the brainstem, cerebellum and structures in the posterior cranial fossa, with eventual obstruction of the cerebrospinal fluid (CSF) pathways of the fourth ventricle. This compression of the vital structures of the brainstem is lethal if not relieved by removal of the tumour. Prevention of the inevitable brainstem compression in patients who have acoustic neuromas that continue to increase in size is the therapeutic basis behind treating the disease.

As most schwannomas are extremely slow growing (averaging a 2 mm increase in diameter per year), the natural history is long – spanning years – and the development of auditory and vestibular symptoms is gradual. Some patients may be unaware of gradually progressive hearing loss caused by the tumour until the loss is quite marked. Compensation occurs for the loss of the balance system on one side, and therefore balance disturbance is usually quite mild.

Gradual stretching of the facial nerve and other cranial nerves in the cerebellopontine angle with compression of the cerebellum and brainstem is well tolerated, particularly in young patients. Some tumours may thus be very large before causing any clinically detectable change in facial nerve or brainstem function.

Presentations

Presenting symptoms of acoustic neuroma are listed in the Table. The classic presentation is hearing loss that is gradual and progressive, but occasionally (1% of cases) it can be sudden and dramatic in onset. Some tumours are detected through workplace screening programs when asymmetry in neural hearing thresholds leads to MRI scanning and demonstration of the tumour.

The auditory symptoms are commonly accompanied by tinnitus and by a mild balance disturbance manifested by a tendency to veer to the side of the tumour when walking, particularly in the dark. Rarely, dramatic recurrent episodic vertigo is the first symptom. Very late presentations with massive tumours causing hydrocephalus are usually preceded by a gradual decline in hearing that has been ignored by the patient or doctor.

With the increasing awareness of acoustic tumours and availability of MRI scanners, more patients are being diagnosed early, when their hearing is nearly normal; their only symptom may be a mild hearing distortion when using the telephone or constant unilateral tinnitus. As acoustic neuromas arise at all ages from adolescence onwards, the diagnosis should be considered when evaluating an asymmetrical neural hearing loss or constant unilateral tinnitus in patients in any age group.

Differential diagnosis

Although acoustic neuromas are by far the most common tumour of the internal auditory canal and cerebellopontine angle, other diagnoses (including solid tumours) need to be considered. Of a total of 57 patients with tumours involving the cerebellopontine angle and internal auditory canal who were referred to the skull base unit at Westmead in a 12-month period, the following were diagnosed:

- acoustic neuroma – 41 cases
- meningioma – 8 cases
- glomus tumours – 5 cases
- metastasis – 1 case
- cholesterol granuloma – 1 case
- epidermoid cyst – 1 case.

Normal structures such as blood vessels also enhance on MRI scanning and need to be differentiated from acoustic neuromas. Rarely, lymphoma, metastatic tumours or systemic inflammatory conditions (such as sarcoidosis and tuberculosis) need to be considered.

Sarcomas in children occasionally present as internal auditory canal lesions associated with hearing loss. In many cases, the nature of the lesion can be diagnosed accurately preoperatively with MRI techniques. Serial scanning over nine to 12 months is often used to document the growth rate of the tumour prior to making a decision about surgery.

Investigations

Audiometry

Progressive or persistent unilateral or asymmetrical auditory symptoms are the primary indication for suspicion of an acoustic neuroma, and pure tone and speech audiometry is important to document the hearing levels formally. Note that previously used auditory evoked response audiometry has almost become obsolete in the diagnosis and evaluation of acoustic neuromas.

MRI scanning

After audiometry, follow up with an MRI scan targeted at the internal auditory canal and cerebellopontine angle is mandatory if suspicion is great enough. Gadolinium enhanced T1-weighted MRI scanning has very high sensitivity and specificity for diagnosing acoustic tumours of all sizes, so much so that a normal MRI scan effectively excludes an acoustic neuroma (Figures 1 to 3). The clarity with which the anatomy of the inner ear and adjacent cranial nerves can be seen has made this form of imaging the modality of choice for evaluating patients with suspicious symptoms.

CT scanning

CT scanning has low accuracy for acoustic tumours and is not recommended when this diagnosis needs to be excluded. A

Downloaded for personal use only. No other uses permitted without permission. © MedicineToday 2001.

CT diagnosis depends on asymmetrical appearances of the internal auditory canal bony architecture, which occurs late in the natural history of acoustic neuromas (Figure 4).

A CT scan is useful, however, in the evaluation of patients who have rapidly progressive symptoms, disabling balance disturbance or suspected hydrocephalus, or when there is a delay in obtaining a specialist referral or ordering an MRI scan. A contrast enhanced CT scan will reasonably and quickly exclude large tumours, significant brainstem compression or hydrocephalus. CT scanning still has a place in the evaluation of patients who cannot undergo MRI scanning because of the presence of a pacemaker or other MRI-incompatible implant, but the results need to be interpreted with care because it has low sensitivity for smaller tumours.

Management options

There are several management options for patients with acoustic neuroma, and these need to be considered carefully with regard to the possible morbidity and long term quality of life associated with each option:

- a period of continuing observation with serial MRI scans
- surgery (with or without preservation of hearing)
- radiotherapy.

Factors that will need to be assessed include the tumour size and growth rate (estimated from serial MRI scans), the patient's age and the usefulness of hearing in the nonaffected ear as well as coexisting illnesses. The patient's ability to undergo surgery lasting several hours also needs to be considered.

In general, small tumours producing minimal symptoms can safely be observed for a period, whereas surgery is the best option for patients with fast growing tumours or those that are impinging (or threatening to impinge) on the brainstem. Some tumours show no growth for long periods – observation for an indefinite period may be the preferred option for these cases.

Given the complex and resource demanding nature of surgical treatment for acoustic neuromas, use of radiation has been investigated as an alternative. Stereotactic radiotherapy aims to deliver a high dose of focused radiation to the tumour with the aim of preventing further growth, but a long follow up period with serial MRI scans is required to monitor the rate of tumour growth because the tumour is not actually removed. Control rates of tumour growth approaching 80% have been reported from the most experienced treating centres, with relatively few side effects. The efficacy of radiotherapy has been difficult to judge, however, because 20% of untreated tumours demonstrate no change in size over long periods of time. At this point, the treatment of acoustic neuroma with radiotherapy is still undefined.[1]

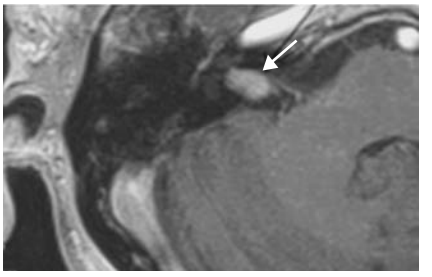


Figure 1. A gadolinium enhanced T1-weighted MRI scan showing a 10 mm acoustic tumour (arrow) confined to the internal auditory canal. This patient presented with a sudden hearing loss on the tumour side.

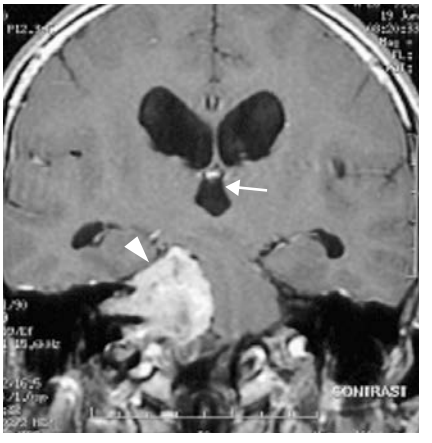


Figure 2. A massive acoustic neuroma (arrowhead) resulting in compression and distortion of the brainstem, obstruction of the CSF pathways causing hydrocephalus (arrow). This 21-year-old patient presented with gross ataxia and was unable to work. The associated progressive hearing loss had been relatively unnoticed.

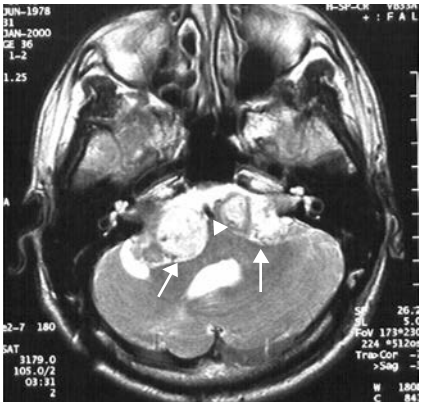


Figure 3. Neurofibromatosis type 2, which is characterised by bilateral acoustic neuromas (arrows) causing extreme brainstem compression. This patient had multiple other intracranial and spinal tumours, including a foramen magnum meningioma (arrowhead) and an extremely poor quality of life. Immediate family members were screened with audiometry and MRI scanning (brain and spine) and found to be clear.

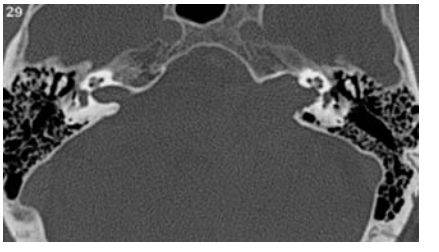
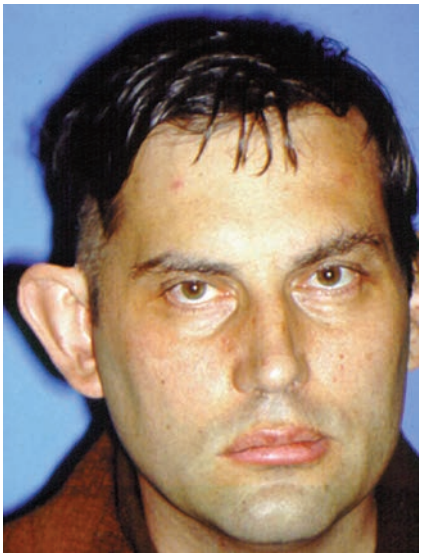
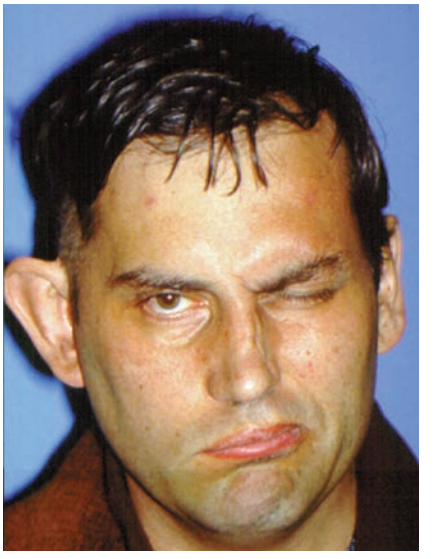


Figure 4. A CT scan of the temporal bones showing normal inner ear anatomy on the right and gross widening of the internal auditory canal on the left due to an acoustic neuroma. This patient's hearing on the left side had been declining for over 10 years.

Downloaded for personal use only. No other uses permitted without permission. © MedicineToday 2001.





Figures 5a and b. Facial nerve function one week after removal of a medium sized (3 cm) acoustic tumour on the right side, shown with facial expression at rest (a, left) and with active eye closure (b, right). Weakness on the side of the tumour is common following surgery but improves gradually. After 12 months, this patient had nearly normal facial nerve movements.

Surgical treatment

The aim of surgery for acoustic neuroma is complete removal of the tumour with preservation of facial nerve function and minimal complications.[2] In specialised units that deal with many acoustic tumours, this is achieved in a high proportion of cases.

Several surgical approaches have been developed, each having advantages and disadvantages. The translabyrinthine approach is most suitable when there is little or no useful hearing because it destroys residual hearing in the operated ear. An attempt to preserve residual hearing in patients with small tumours can be made with an alternative approach through the middle fossa that spares the inner ear, but hearing is preserved in only 60% of these cases (at best), and its contribution to overall hearing is difficult to demonstrate.

Acoustic tumour surgery is a highly specialised and intensive process carried out in only a few major centres. Operating times are long, averaging six hours for medium size tumours and often much longer for giant acoustic tumours.

Long term outcomes

The outcomes of contemporary vestibular schwannoma surgery have improved dramatically since the first successful tumour removal just over a century ago. In specialised units, complete removal is now achieved in 98% of cases. Anatomical preservation of the facial nerve occurs in more that 95% of tumours of small to moderate size, and the rates of hearing preservation for selected tumours are improving.

Nevertheless, recent quality of life studies have shown that, although most patients return to nearly normal activities and full-time employment following removal of the tumour, their quality of life is generally lower than in age and sex matched controls.[3] This is especially true of patients who have minimal symptoms, normal hearing and small tumours – in these patients (about 25% of all cases), a period of observation with serial MRI scanning is justified prior to making a decision to operate.

In general terms, most patients have excellent long term outcomes following modern surgery for acoustic neuroma, and can expect to return to nearly normal social and employment activities with a very high quality of life. However, given the sensitivity of the sensory, neural and vascular structures involved in the surgery, there is a potential for serious complications with devastating consequences, including stroke or death. Overall, serious complications occur in less than 0.5% of operations, mainly in patients with large tumours, significant comorbidity or advanced age. The potential for these poor outcomes strengthens the need for increased awareness about acoustic neuromas and their subtle early presentation with hearing loss or asymmetrical auditory symptoms.

Surgical complications

Following surgery, patients usually take several weeks before being ready to return to their normal working and social lives. It is during this early postoperative period that complications can be most troublesome.

Hearing loss

Hearing loss in the operated ear is the most common complication following surgery, and it is inevitable with the translabyrinthine approach. Patients with minimal residual hearing in the affected ear preoperatively and normal hearing in the opposite ear do not usually notice additional hearing loss following surgery. On the other hand, patients who had nearly normal hearing preoperatively are likely to notice a great change, especially in noisy environments or social situations that depend heavily on binaural hearing.

For patients who have small selected tumours and useful preoperative hearing thresholds, hearing preservation surgery can be attempted. However, the hearing following this form of surgery is only moderate at best, and, at this stage, improvement in this aspect of treatment is still in evolution.

Downloaded for personal use only. No other uses permitted without permission. © **Medicine**Today 2001.

Tinnitus

Tinnitus often persists following removal of the tumour but is rarely a problem. In 2% of cases it worsens following surgery.

Vertigo and imbalance

Vertigo and imbalance occur in the first few days postoperatively. Most patients compensate completely, which allows return to driving, work and other activities about 12 weeks after surgery. A few people feel a little unsteady in the dark or in unfamiliar environments for months – an intensive physiotherapy program can be of help.

Facial weakness

Facial weakness is the most common problem and is usually transient (Figures 5a and b). About 90% of patients with small to medium sized tumours can be expected to achieve nearly normal facial nerve function at 12 months.

In patients who have poor facial nerve function initially or who have a slow or incomplete recovery, special attention needs to be given to eye closure and care of an exposed cornea. Often, occlusive transparent coverings, drops, ointments or artificial tears are all that is required. A temporary tarsorrhaphy or insertion of a gold weight in the upper eyelid may occasionally be required until normal function returns.

If the facial nerve is transected during the surgery or there is no recovery at all, a range of muscle and nerve transfers that may improve facial tone and appearance should be considered. This situation arises in less than 2% of patients who have tumours surgically removed.

CSF leakage

Leakage of CSF from the wound or from the nose (having travelled down the eustachian tube from the site of surgery) occurs in 10 to 15% of operations, generally in the first week after surgery. Most leaks close with temporary CSF diversion via a lumbar catheter for a few days.

Neurological complications

Major neurological complications caused by damage of the lower cranial nerves and strokes are rare, occurring in less than 0.5% of cases, and are generally related to removal of massive tumours. In the specialised units, the death rate is extremely low.

General complications

General complications such as deep venous thrombosis and pulmonary embolism are related to the prolonged nature of the surgery, and occur in about 2% of cases. Special precautions during and immediately after surgery need to be taken to minimise occurrence.

Conclusion

Acoustic neuromas are uncommon, so a high degree of awareness is needed when evaluating patients with unilateral hearing loss or asymmetrical auditory symptoms. More patients are being diagnosed early, but there is a need for further awareness of the subtle early presentation with hearing loss or asymmetrical auditory symptoms. MRI scanning has a high sensitivity for acoustic neuromas and a normal, well performed scan is reliable in excluding the diagnosis. For the best results, patients should be referred to a specialised centre that deals with large numbers of acoustic neuromas. MT

References

1. Pitts LH, Jackler RK. Treatment of acoustic neuromas. N Engl J Med 1998; 339(20): 1471-1473.
2. House WR, Luetje CM, Doyle KJ. Acoustic tumors: diagnosis and management. 2nd ed. San Diego: Singular Publishing, 1997.
3. da Cruz MJ, Moffat DA, Hardy DG. Postoperative quality of life in vestibular schwannoma patients measured by the SF36 health questionnaire. Laryngoscope 2000; 110(1): 151-155.

Downloaded for personal use only. No other uses permitted without permission. © **Medicine**Today 2001.