Acoustic Neuromas

Synonyms: acoustic neurilemoma, acoustic neurinoma, vestibular schwannoma

Acoustic neuromas are tumours of the vestibulocochlear nerve (eighth cranial nerve), arising from the Schwann cells of the nerve sheath. Most arise from the vestibular portion and only a few arise from the cochlear (auditory) division. They are typically benign and slow-growing, but can cause symptoms through mass effect and pressure on local structures, eventually becoming life-threatening. Patterns of growth can vary and a small number may grow rapidly (doubling size in six months). Considering the possibility can enable earlier diagnosis, increase management options and may decrease morbidity.

Within the cerebellopontine angle (CPA), tumours can grow as big as 4 cm in diameter and slow growth allows stretching and accommodation of growth without affecting function. However, tumours within the internal auditory canal produce symptoms much earlier with hearing loss (the most common presenting symptom) or vestibular disturbance.

Epidemiology
Acoustic neuroma represents 8% of all intracranial tumours and 80% of those arising at the CPA. [1]

Risk factors
Confirmed risk factors include:

- Neurofibromatosis. [2]
- High-dose ionising radiation (children who received radiation for benign conditions of their head and neck - for example, to decrease the size of their tonsils and adenoids - were at increased risk of developing an acoustic neuroma much later in life). [3]
- However, the medical use of low-dose ionising radiation, such as used in imaging, has not been established as a risk. [4]

Presentation

Any unilateral sensorineural hearing loss should be considered as caused by an acoustic neuroma until proven otherwise.

Consider the diagnosis of acoustic neuroma in patients with:
- Unilateral or asymmetrical hearing loss or tinnitus, whether progressive or acute in onset.
- Impaired facial sensation.
- Balance problems without other explanation.

Classic presentation of acoustic neuroma confined to the internal auditory canal, involves unilateral progressive hearing loss, vestibular dysfunction and tinnitus.

- A small minority of patients will experience sudden and complete unilateral hearing loss.
- Hearing may also fluctuate.
- A few patients will have normal hearing at presentation.
- Most patients have quite subtle balance disturbance at presentation.
- The severity of tinnitus has been shown to correlate with tumour size. [5]

As the tumour spreads, there is an increase in hearing loss and disequilibrium, and symptoms due to compression of other structures may occur:

- Facial pain or numbness due to involvement of the trigeminal nerve.
- Facial weakness is uncommon despite the tumour pressing on the facial nerve.
- Earache.
- Ataxia due to cerebellar compression.
- Severe brainstem compression can produce hydrocephalus with visual loss and persistent headache and even decreased level of consciousness.

Acoustic neuroma may be diagnosed incidentally and earlier as a result of investigations for unrelated problems.

Associated diseases
Bilateral acoustic neuroma occurs in neurofibromatosis-type 2 (NF2). NF2 is an autosomal dominant disorder (ie has a 50% risk of transmission from a parent) but also shows high levels of mosaicism. 7% of patients with acoustic neuroma also have NF2. Acoustic neuroma due to NF2 tends to present earlier, typically around 30 years old. Genetic screening for NF2 in patients presenting with sporadic, unilateral acoustic neuroma is usually only productive in cases of very early onset (younger than 20 years).[7] NF2 patients are predisposed not only to developing acoustic neuroma but also schwannomas of other cranial nerves.[8]

Increasing symptoms associated with acoustic neuroma increases the likelihood of clinically significant anxiety and depression.[9]

Differential diagnosis
Other CPA tumours include meningiomas, epidermoids, lower cranial nerve schwannomas and arachnoid cysts.

Investigations

Audiology
All patients with unilateral or asymmetric hearing loss should be referred for audiological assessment to quantify and clarify the loss as sensorineural. See reference (below) for examples of audiograms and MRI scans from patients with acoustic neuroma.[10]

Diagnostic imaging
MRI has largely superseded CT scanning as the investigation of choice for suspected acoustic neuroma.[11]

Management

The growth pattern of acoustic neuromas is variable. As many as 75% of tumours have been reported to show no growth. However, there are no reliable predictors of tumour behaviour.[12] There are three treatment options: microsurgery (the technique of choice), stereotactic radiosurgery and observation.[1] When assessing the most appropriate management for each individual patient, consideration needs to be given to future quality of life and symptom relief as well as tumour control, facial nerve function and hearing preservation.

Conservative management
For patients with small neuromas and good preserved hearing, the most appropriate course of action may be to watch and wait with serial scans (usually annually) to monitor growth. Where growth is detected, more active treatment is usually advocated since risk of complications with surgery and ability to preserve hearing are related, in part, to tumour size. There is some evidence that quality of life is reduced in untreated patients.[13] There is no agreed size of tumour triggering active treatment across centres.

Surgical treatment
In the UK, most patients receiving active treatment undergo microsurgery. The surgical approach taken depends on the location of the tumour, its size and the relative importance of hearing preservation. Complete removal is possible in most cases. The risks of surgery include:[14]

- Mortality (risk about 1%)
- CSF leak and meningitis
- Cerebellar injury
- Stroke
- Epilepsy
- Facial paralysis (either partial or complete)
- Hearing loss[15]
- Balance impairment[16]
- Persistent headache[17]

Stereotactic radiosurgery
Stereotactic radiosurgery targets a tumour with a single large dose of radiation using convergent beams of high-energy X-rays, gamma rays (‘gamma knife radiosurgery’) or charged particles and is an alternative, emergent treatment. The aim of treatment is to control (either slow or stop) the growth of the tumour. Stereotactic radiation may be associated with significantly better long-term hearing preservation outcome rates than microsurgery.

Salvage surgery for neuromas post-radiotherapy is technically more difficult than primary surgery. Other potential longer-term risks associated with stereotactic radiosurgery and stereotactic radiotherapy include:

- Radiation-induced brain necrosis.[18]
- Radiation-related cranial nerve injury.
- Malignant change (for example, to a glioblastoma multiforme).[19]

Further reading & references

- Radiosurgery Practice Guideline Initiative - Stereotactic Radiosurgery for Patients with Vestibular Schwannomas; International RadioSurgery Association (IRSA), March 2006
10. Hain TC; Acoustic Neuroma, Dizziness-and-balance.com

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