Asymmetric hearing loss and tinnitus

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A 56 year old man was referred by his general practitioner with a 10 year history of progressive hearing loss and tinnitus, mainly in his right ear. He did not describe otalgia or dizziness. After thinking about it more carefully, however, he admitted that he occasionally felt slightly unsteady but had attributed this to tiredness. His medical history was otherwise unremarkable, and there was no family history of deafness.

Clinical examination showed normal tympanic membranes bilaterally, with unremarkable pharynx and nasal cavity. There were no obvious neurological deficits, and clinical balance tests, including Romberg’s test, Unterberger’s test, and head impulse tests, were normal. Audiological assessment confirmed moderate right sensorineural hearing loss in all frequencies and a mild drop in high frequencies in the left ear.

Because of the asymmetry of his symptoms, magnetic resonance imaging (MRI) of the brain and internal auditory canal was performed (fig 1).

1. What does the MRI scan show?

Short answer

Figure 1 shows a well delineated lesion of the right internal auditory canal extending into the cerebellopontine angle. The lesion is hypointense on T2 weighted imaging (A), isointense on non-contrast T1 weighted imaging (B), and it enhances strongly on T1 weighted imaging after administration of intravenous contrast (C).

Long answer

The axial MRI shows a well defined lesion (arrow) arising from the right internal auditory meatus and extending about 1.5 cm into the cerebellopontine angle (fig 2). The lesion seems to abut the brainstem (white X) and the cerebellum (white star), without compressing them. It is characteristic of a benign lesion or tumour—it is well delineated and does not infiltrate nearby structures.1

2. What is the diagnosis?

3. How does this condition usually present?

4. What are the treatment options for this condition?
On T2 weighted imaging (fig 2A) the lesion is hypointense (dark grey-black) and pushes the cerebrospinal fluid (white hyperintensity) away from the cerebellopontine cistern, whereas it is isointense (light grey) on non-contrast T1 weighted imaging (fig 2B). After administration of intravenous gadolinium (contrast), the lesion enhances (absorbs) strongly and appears hyperintense (bright white) on contrast T1 weighted imaging (fig 2C).

On the left side, the anatomy of the internal auditory canal and the cerebellopontine angle appears normal, with cerebrospinal fluid filling the cerebellopontine cistern and the internal auditory meatus, while the remaining brain structures are normal.

2. What is the diagnosis?

**Short answer**

The diagnosis is a right vestibular schwannoma. Vestibular schwannomas or acoustic neuromas are benign tumours arising from the sheath that surrounds the vestibulocochlear (eighth cranial) nerve.

**Long answer**

MRI shows a lesion in the right internal auditory canal extending 1.5 cm into the cerebellopontine angle, consistent with a vestibular schwannoma (fig 2). Vestibular schwannomas, also known as acoustic neuromas, are benign tumours of the vestibulocochlear (eighth cranial) nerve. They arise from Schwann cells (which form the sheath that surrounds nerves) of mainly the vestibular nerve but also the cochlear nerve. They are found in the internal auditory canal and can extend into the cerebellopontine angle. They may even compress the brainstem, further cranial nerves, and the cerebellum. Most vestibular schwannomas are unilateral. Bilateral vestibular schwannomas are a hallmark of neurofibromatosis type 2.

Vestibular schwannomas are usually slow growing tumours. However, their growth varies widely and is unpredictable. Some tumours remain unchanged for many years, whereas others grow quickly. It has been estimated that around 70% (UK data) of vestibular schwannomas (range in the literature 15-85%) will not grow. These tumours form 6% of all intracranial tumours and about 85% of all tumours of the cerebellopontine angle. The average age of diagnosis is 55 years and both sexes are affected equally. About 10 new cases are diagnosed every year per million people in the United Kingdom and the United States, and the lifetime risk of developing a vestibular schwannoma in England is one in 1000. However, histological studies on human cadavers have shown a much higher prevalence of 0.8-2.7%, suggesting that a large number of cases are asymptomatic (silent) or even misdiagnosed. Interestingly, the prevalence of incidentally diagnosed vestibular schwannomas is about two in 10,000 people in the US (California); this is lower than the histological prevalence but higher than that reported in clinical and epidemiological studies.

Currently, MRI is the gold standard for diagnosing vestibular schwannomas. These tumours typically appear as well delineated hypointense (dark grey-black) lesions on non-contrast T2 weighted scans and isointense (light grey) lesions on non-contrast T1 weighted scans, whereas they enhance strongly after administration of intravenous gadolinium (fig 2).

Because of time considerations and cost effectiveness, non-contrast T2 weighted MRI is usually performed initially. When findings are positive, contrast enhanced T1 weighted and fat suppression imaging are used to differentiate the tumour from other less common lesions of the cerebellopontine angle, such as meningiomas, granulomas, and lipomas.

MRI plays an important role in the differential diagnosis of lesions of the internal auditory canal and the cerebellopontine angle. These lesions are radiologically divided into enhancing and non-enhancing ones. As already reported, schwannomas, mainly arising from the vestibulocochlear nerve but also rarely from the facial nerve, are the most common enhancing tumours in this region. Meningiomas are enhancing lesions that present on MRI with radiological features similar to those of vestibular schwannomas. However, meningiomas typically have a broad base with a “double tail,” giving them a more elliptical (hemispheric) shape, because they arise from the dura and not from the nerves. Metastases in the cerebellopontine angle or the petrous apex are rare, and, although they enhance, they have a more irregular appearance on MRI and patients usually have a history of cancer. Apart from schwannomas, meningiomas, and metastases, other (rare) enhancing lesions of the internal auditory meatus and cerebellopontine angle include sarcoidosis, tuberculosis, and melanoma. Enhancing lesions of the temporal bone and skull base or the cerebellum-brain, such as parangangiomas, endolymphatic sac tumours, lymphomas, gliomas, chordomas, medulloblastomas, and ependymomas, can also invade this area and should be included in the differential diagnosis.

Non-enhancing lesions are also uncommon. Lipomas of the cerebellopontine angle are rare and can easily be differentiated from vestibular schwannomas because they are usually hyperintense (bright white) on non-contrast T1 weighted imaging, they do not enhance after administration of intravenous gadolinium, and they become hypointense (dark grey) on fat suppression MRI. Cholesterol granulomas and epidermoids (hypointense dark grey on T1 weighted imaging) and dermoids (hyperintense-bright white on T1-weighted MRI) are also uncommon and do not enhance on contrast enhanced imaging. Arachnoid cysts show cerebrospinal fluid signal on all MRI sequences (hyperintense white on T2 weighted and hypointense grey on T1 weighted MRI). Recent advances in neuroimaging mean that most lesions of the internal auditory canal and the cerebellopontine angle can be accurately diagnosed on MRI.

In the pre-MRI era, contrast computed tomography and auditory brainstem response audimetry were used to diagnose vestibular schwannomas. Computed tomography with intravenous contrast administration can identify lesions that extend into the cerebellopontine angle but its use is limited in the diagnosis of lesions that lie exclusively in the internal auditory canal.

Auditory brainstem response testing can identify lesions larger than 1 cm but has a low sensitivity for smaller lesions. Currently, contrast computed tomography and auditory brainstem response testing are still used in patients in whom MRI is contraindicated or in people who are claustrophobic.

3. How does this condition usually present?

**Short answer**

Vestibular schwannomas usually present with unilateral or asymmetric sensorineural hearing loss, tinnitus, and dizziness or imbalance. Patients in whom these symptoms are asymmetric or unilateral should be referred to an otolaryngologist.
Long answer

More than 90% of patients with vestibular schwannomas present with unilateral or asymmetric sensorineural hearing loss. The original definition of asymmetric hearing loss was vague and could be interpreted in various ways by clinicians. However, in 1993 the American Academy of Otolaryngology-Head and Neck Surgery set specific criteria for the definition of asymmetric hearing loss. It was defined as an air conduction pure tone audiometry threshold difference of 15 dB or greater between the two ears (threshold measured at 500 Hz, 1000 Hz, 2000 Hz, and 3000 Hz).

Hearing loss is progressive in most cases. However, unilateral sudden sensorineural hearing loss can be the initial symptom and is seen in 7-27% of patients with vestibular schwannomas. Patients with sudden sensorineural hearing loss who undergo MRI have a 2% chance of having a vestibular schwannoma. Hearing is completely normal in the affected ear in 5-12.5% of patients with vestibular schwannomas.

Unilateral tinnitus has been reported as the main presenting symptom in 8-13% of patients with vestibular schwannoma. However, tinnitus can be an accompanying symptom, mainly associated with hearing loss, in 60-83% of such patients. Dizziness or imbalance is the main symptom in 7-26% of patients with a vestibular schwannoma. However, symptoms related to the vestibular nerve are not always described as rotatory vertigo but also as unsteadiness or even “light headedness,” so the precise incidence of vertigo in unclear. In general, balance is affected in a large proportion of patients with vestibular schwannomas.

In clinical settings, apart from taking a detailed medical history, clinicians should also look for gross signs of imbalance, such as unsteadiness in walking (falling to the right or to the left) and nystagmus. They should also perform simple tests such as Romberg’s test, Unterberger’s test, or the head impulse test. In Romberg’s test patients are told to stand erect with feet together, arms outstretched, and eyes closed; the examiner looks for swaying or a tendency to fall to the right or left. In Unterberger’s test patients are told to march on the spot from Romberg’s position; the examiner looks for tendency to fall or abnormal rotation of the body while marching. In the head impulse test patients are told to focus on a stable object while the examiner thrusts the patient’s head; loss of fixation is considered abnormal. Overall, abnormal clinical balance test results raise the suspicion of peripheral vestibulopathy or even central balance disorder and warrant further investigation and imaging studies.

Several studies have looked into the correlation between the severity of vestibulocochlear symptoms (hearing loss, tinnitus, dizziness, and imbalance) and tumour growth or size. Most studies, including rigorous systematic reviews, have found no consistent correlation between tumour growth and hearing loss.

A small proportion of people with vestibular schwannomas also have weakness of the facial nerves or even palsy. Lesions in the cerebellopontine angle that affect the facial nerves have to be differentiated from facial schwannomas or other processes, such as cholesteatomas or invasive growths of the petrous apex. Other cranial nerves, such as the trigeminal nerve and the lower cranial nerves (glossopharyngeal, vagus, accessory, and hypoglossal nerves) are affected only rarely. However, large vestibular schwannomas that extend superiorly can compress the trigeminal nerve. These lesions can present with loss of facial sensation or paraesthesia, loss of corneal sensation, or even facial pain, mimicking trigeminal neuralgia. Large vestibular schwannomas can cause increased intracranial pressure and compression of the cerebellum or the brainstem, and patients may present clinically with dysdiadochokinesis (impaired ability to coordinate and perform fine, rapid movements), ataxia, headaches, and blurred vision.

In conclusion, most vestibular schwannomas present with vestibulocochlear symptoms (hearing loss, tinnitus, and vertigo). Patients with asymmetric and unilateral symptoms warrant referral to an otolaryngologist.

4. What are the treatment options for this condition?

Short answer

The available options are radiological surveillance with clinical review, radiotherapy, or microsurgical excision. Treatment is individualised and depends mainly on tumour size and comorbidities.

Long answer

Currently, there are three options for treating vestibular schwannomas: radiological surveillance with clinical review (conservative management), radiotherapy (γ knife, stereotactic radiosurgery, or microsurgical excision). Treatment depends mainly on the size and growth pattern of the tumour. Small, non-growing, or very slowly growing tumours that do not compress the brain are usually treated conservatively. Large tumours (>3 cm in the cerebellopontine angle) that are compressing the brainstem or the cerebellum are surgically removed, whereas growing tumours that do not threaten the brain and smaller tumours can be treated with radiosurgery. In the case of vestibular schwannomas, radiosurgery usually entails a single high dose of radiation directed solely and accurately at the tumour. This is usually described as stereotactic radiosurgery because its delivery is based on three dimensional evaluation of the patient’s MRI scan; however, the precise radiosurgical protocol may vary.

Management, however, is individualised and depends on factors such as the patient’s age and comorbidities, the severity of the symptoms, and the patient’s wishes. The treatment decision is complex and involves detailed consultation because surgery or even radiotherapy can be associated with serious complications and risks, such as profound deafness, severe balance problems, intolerable tinnitus, and facial nerve weakness or even palsy.

Our understanding of the natural course of vestibular schwannomas, and our attitude towards these lesions, has changed considerably over the years. More patients are managed conservatively or with radiotherapy, with increasing evidence of a better quality of life after such management. Currently, most newly diagnosed vestibular schwannomas will initially be rescanned to identify their growth pattern and to plan further management.

Patient outcome

After detailed consultation, the patient was managed conservatively with clinical reviews and serial MRI. No tumour growth has been noted on follow-up MRI. He remains under radiological surveillance and undergoes annual clinical and audiological assessments. His hearing loss has been treated with a conventional hearing aid.

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