Unusual presentation of more common disease/injury

The middle ear mass: a rare but important diagnosis

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The authors report a rare case of primary intracranial meningioma presenting as a middle ear mass with conductive hearing loss. The authors aim to highlight the importance of diagnosing a middle ear mass, which although rare, may have a substantial impact on ongoing patient management. A discussion of other middle ear pathologies is made in order to demonstrate the subtle differences in presentation.

BACKGROUND

This case describes a rare cause of a traumatic tympanic membrane (TM) perforation. It is the first such case to be reported in the worldwide literature. It highlights the importance of thorough inspection of the middle ear space while performing myringoplasty.

CASE PRESENTATION

A 42-year-old lady presented to the ear, nose and throat (ENT) outpatient clinic with a 25 year history of progressive unilateral hearing loss. There were no other significant otological symptoms. There was no family history of ear disease or history of previous ear surgery. Her medical history was unremarkable apart from carcinoma of the cervix *in situ* 10 years previously.

Clinical examination revealed an intact bulging left TM and a white lesion within the middle ear space (figure 1). The right ear was normal in appearance.

Clinical tuning fork tests indicated a conductive hearing loss on the left side. Examination of the nose, post-nasal space, oral cavity and neck was normal and cranial nerve examination was intact. The audiogram revealed a left-sided moderate conductive hearing loss (figure 2). The working diagnosis was that of a middle ear space mass.

INVESTIGATIONS

A CT scan was performed to evaluate the extent of the lesion, and to identify the degree of bony destruction. There was complete opacification of the left mastoid air cells, middle ear and attic due to soft tissue. The ossicles were intact and there was no bony destruction (figure 3).

In addition, MRI of the head and neck was performed to establish the nature of the soft tissue mass evident on CT. This was coupled with an MR angiogram to establish whether the mass had a vascular origin. A large lesion arising from the left cerebello-pontine angle was confirmed, extending into the postnasal region, with a dural tail. There was distortion of the brainstem and cerebellum. There was infiltration into the left internal acoustic meatus, and jugular foramen. The radiological findings were highly suggestive of a meningioma extending into the middle ear cavity (figure 4).

DIFFERENTIAL DIAGNOSIS

- ▶ Acute otitis media (AOM)
- ▶ Glomus tumour
- ▶ Congenital cholesteatoma
- ▶ Middle ear neoplasm.

TREATMENT

The patient was referred to a tertiary centre for further assessment and treatment planning.

DISCUSSION

This case report has highlighted a rare but serious cause of a bulging TM with associated conductive hearing loss. The differential diagnosis to be considered with this presentation should include AOM, glomus tumours, middle ear tumours and congenital cholesteatoma.

AOM classically presents with an acute history of otalgia, conductive deafness and possibly fever. It is a common diagnosis in children but rare in adults. Clinical examination normally reveals a red and bulging TM. If an acute history, no further investigation is usually required and treatment may consist of oral antibiotics and analgesia.

A typical history for a middle ear mass may include a long-standing or progressive unilateral hearing loss, with or without accompanying otalgia, vertigo, tinnitus and headache. There is often no otorrhoea or history of fever. The TM appears abnormal with increased opacity and less visible middle ear anatomy for example, long process of incus. Depending on the size and specific origin, the middle ear mass may approximate the deep surface of the TM and may cause the TM to bulge if mass volume is sufficiently large. The nature of the underlying mass may alter the colour of the TM. This may be blue or red in vascular lesions and whitish in soft tissue masses.

Meningiomas are the second most common primary cerebello-pontine angle neoplasm and comprise approximately 15% of all intracranial neoplasms. However, less than 1% of meningiomas have extracranial extension or temporal bone involvement.^{1–3} In this particular case, the diagnosis was an intracranial meningioma with middle ear extension. Meningiomas are thought to arise from aberrant

Figure 1 Oto-endoscopic view of left tympanic membrane demonstrating white mass in middle ear space.

Figure 2 Audiogram demonstrating left moderate-severe conductive hearing loss.

Figure 3 Axial and coronal CT demonstrating opacification of the left middle ear space, mastoid air cells and attic. There is also soft tissue density in the sphenoid sinus.

Figure 4 Coronal MRI demonstrating meningioma arising from the left cerebello-pontine angle extending into the middle ear space.

<i>Diagnosis</i>	<i>Symptoms</i>	<i>Signs</i>
AOM	Otalgia Conductive deafness Late otorrhoea	Bulging red/inflamed TM Pyrexia
Meningioma	Conductive deafness Vertigo Tinnitus Headache	Whitish mass in middle ear space
Glomus tumours	Pulsatile tinnitus Conductive/mixed deafness Otalgia Headache Vertigo	Blue/red mass in middle ear space
Congenital cholesteatoma	Conductive deafness	Whitish mass in middle ear space
Middle ear neoplasm	Conductive deafness Otalgia Headache	Whitish if bony, red/pink if soft tissue origin

Figure 5 Table demonstrating the symptoms and signs of middle ear masses (those in bold are more common).

arachnoid cells, and neoplastic advancement occurs along neural tissue such as nerve sheaths.^{2 4 5}

The presentation of middle ear meningiomas is varied in the literature. Otalgia is the most common feature, present in 42% of patients. Conductive hearing loss, facial paralysis and tinnitus are also common, accounting for 32–42%

of symptoms. A lower proportion of patients present with other cranial nerve palsies or vestibular symptoms.⁴ The mean age of presentation is in the fourth decade with a female to male ratio of 9:1. Malignant activity is rarely displayed, and while genuine metastatic activity is not present, malignancy is often regarded as invasion into the brain.^{2 4 5}

Other neoplasms that can present in the middle ear include glomus tumours and rarely schwannomas, carcinomas, melanomas and adenomas.²

Glomus tumours are benign vascular tumours also known as paragangliomas. They are subdivided into glomus jugulare, glomus vagale and glomus tympanicum depending on their location. They are more common in females during the fifth and sixth decades.⁶ A key symptom of glomus tumours is pulsatile tinnitus which may be accompanied by a conductive hearing loss. Otalgia is uncommon but headache and vertigo may be present. Hoarseness and dysphagia may suggest cranial nerve involvement. Glomus tumours often present as an intact TM with a red/blue mass in the middle ear space.

Congenital cholesteatoma is a rare, yet possible, cause of a middle ear mass. The congenital form of cholesteatoma (differentiated from the acquired form by the presence of an intact TM) commonly presents in childhood, sometimes with conductive hearing loss. Clinical examination reveals an intact TM with a whitish middle ear mass normally towards the anterior aspect of the TM. The common symptoms and signs of different causes for middle ear masses are summarised in figure 5.

Middle ear masses require audiological investigation to establish the nature of any hearing loss. In addition, imaging helps identify the origin of the mass and whether or not there is intracranial or extracranial extension. This is crucial in further management and surgical planning if appropriate. MRI with gadolinium enhancement is the most valuable form of imaging in determining disease extent and structural involvement. While CT is not wholly adequate at imaging the neoplasm itself, it is necessary to assess the bony involvement of the disease.^{3 5}

Learning points

- ▶ Middle ear masses are a rare diagnosis and should be considered in systemically well patients with long-standing unilateral hearing loss, tinnitus, vertigo and headache with or without abnormal TM appearance.
- ▶ Early diagnosis is vital in ensuring that subsequent referral, investigation and management is appropriately directed.
- ▶ Such patients should be referred early for assessment in a specialist ENT clinic for audiometric analysis and appropriate radiological investigations to help ascertain the likely origin of a middle ear mass and guide further management.

Competing interests None.

Patient consent Obtained.

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