



Primary meningioma of the middle ear

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Reviewer

Usama Kamel

Extracranial meningioma involving the middle ear is extremely rare.¹ This case report highlights important considerations and pitfalls in the diagnosis and management of this tumour.

Case report

History

A 61-year-old woman presented with a 3-month history of dizziness and right-sided hearing loss which started after an upper respiratory tract infection. She reported no previous ear problems and no symptoms related to the nose or throat.

Examination of the right ear revealed a normal external auditory canal and a congested tympanic membrane. Audiometry showed a 70 db mixed low frequency hearing loss in the right ear (Figure 1) and a flat impedance. An initial diagnosis of right middle ear inflammation with effusion was made.

The patient underwent right-sided myringotomy with grommet insertion. At the time of operation it was noted that the middle ear was full of granulation tissue. At 3-month follow-up, the patient reported mild improvement in symptoms but audiometry revealed a persistent right-sided sensorineural deficit. MRI (unenhanced) showed no significant abnormality at the CP angle, internal acoustic meatus or within the cerebral hemispheres. The appearance of fluid within the right mastoid air cells was noted (Figure 2). The patient was conservatively managed at 12, 18 and 24-month follow-up.

At 36-month follow-up otoscopy revealed a bulging posterior quadrant of the right tympanic membrane. A tympanotomy was performed using endaural incision. A tympanomeatal flap

was raised revealing a middle ear filled with fleshy tumour. This was attached to the incudostapedial joint, filling the hypotympanum. On retrospective evaluation of the MRI, the possible presence of soft tissue in the right mastoid was noted but there was no clear evidence of a lesion consistent with the operative findings.

At follow-up 6 months after surgery, the patient showed no clinical evidence of recurrence. A repeat MRI (gadolinium-enhanced) showed that the soft tissue previously noted had been removed. An area of abnormal signal situated posterior to the middle ear cavity was still present, enhanced and had not changed in size over the interval, most likely representing inflammatory tissue (Figure 3).

Histopathology

Macroscopic examination showed a crescent-shaped brown soft tissue lesion, 11 × 6 × 3 mm. Histological examination showed nests and whorls of uniform small cells with moderate cytoplasm, fine nuclear chromatin and inconspicuous nucleoli (Figure 4). The stroma was densely sclerotic. Psammoma bodies were conspicuous. There was no mitotic activity, nuclear pleomorphism or necrosis. The morphological features were consistent with meningioma (WHO grade 1). There were no atypical features. Immunohistochemical analysis showed positive staining with vimentin and epithelial membrane antigen (EMA). S-100, Cam 5.2 and MNF-116 were negative.

Discussion

Whether it is possible for meningioma to arise primarily from the auditory canal has been debated.²

Figure 1
Initial pure-tone audiogram showing right-sided 70 db mixed hearing loss

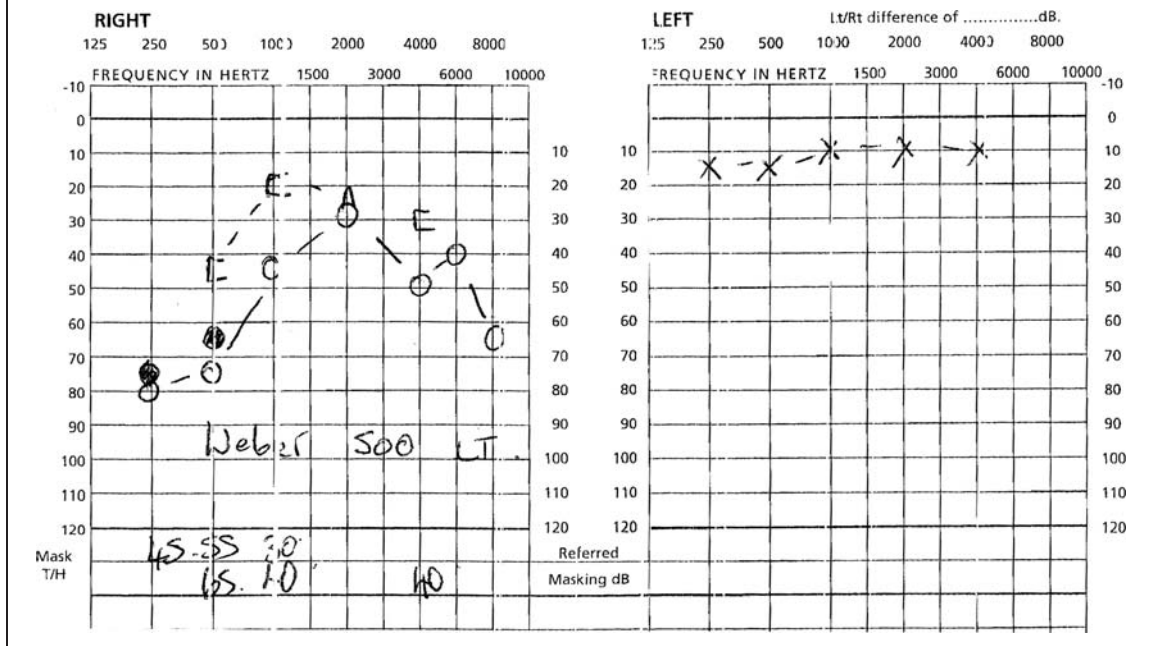


Figure 2
Initial unenhanced MRI showing fluid within the right mastoid air cells

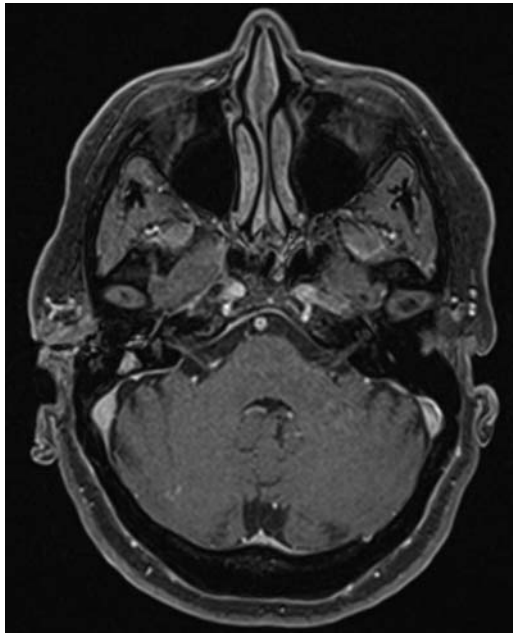


Most of the few reported cases of meningiomas occurring in the temporal bone were prior to the availability of gadolinium-enhanced MRI which is required to exclude an intracranial lesion as the primary origin.¹ Embryologically, it is suggested that ectopic meningiomas could arise from punched-off arachnoid cells outside the neuraxis lying along the line of fusion of primitive nerve and bone sheaths.³

Since enhanced MR imaging has become widely available, very few meningiomas in the ear without intracranial involvement have been reported. These have consisted of extracranial meningioma presenting as an aural polyp with conductive hearing loss,⁴ meningioma arising from the external auditory canal presenting with sagging of the posterior canal wall,³ and one case which, similar to this case, presented with hearing loss and a mass behind the tympanic membrane which was subsequently found to occupy the mastoid antrum and epitympanum.⁵

Meningiomas are relatively benign in nature and so histology and immunohistochemistry become crucial to differentiate the tumour from

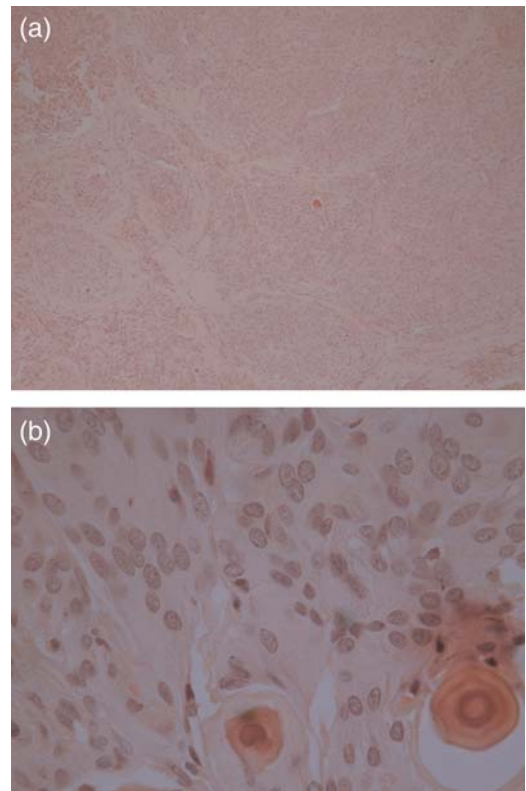
Figure 3
Postoperative MRI with gadolinium enhancement showing removal of soft tissue mass and no intracranial involvement



others including paraganglioma, schwannoma, adenoma, adenocarcinoma, mucoepidermoid carcinoma, glomus tumours and metastatic tumours which are likely to be of more concern.

We report a case where diagnosis was delayed by 3 years from original presentation to the ENT clinic. The rarity of the disease combined with non-specific symptoms contribute to this latency. The common differential diagnosis for a unilateral mixed hearing loss includes otosclerosis, otitis media with effusion, cholesteatoma. Tympanometry is the next investigation to aid diagnosis. A flat tympanogram may commonly suggest a diagnosis of otitis media with effusion,⁶ as happened in this case. Any unexplained sensorineural deafness should lead to obtaining an MRI scan. However in this case, the lesion was not identified by unenhanced MRI. This case highlights the benefit of following up patients with slightly atypical presentation whose symptoms fail to resolve and also raises the question of whether gadolinium-enhanced MRI should be used earlier with unexplained mixed hearing loss.

Figure 4
Sections showing typical meningothelial whorl pattern and psammomma bodies. a. H&E $\times 40$; b. H&E $\times 200$



It is difficult to fully explain the mix of both conductive and sensorineural components. Retrospectively, we would suggest that the conductive element was due to the tumour physically both impeding the movement of the tympanic membrane and reducing the movement of the ossicular chain. The sensorineural component is difficult to explain. It was possibly the result of minimal invasion of the tumour into the cochlea, though this was not seen at surgery.

At initial myringotomy, abnormal granulation tissue was noted behind the tympanic membrane but no biopsy was taken. Biopsy at this stage may have led to an earlier diagnosis and is a useful reminder for surgeons of all levels performing this procedure who do not find serous or mucoid fluid behind the tympanic membrane. It is worth noting that biopsy of middle ear tissue does have risk attached. Any vascular lesions

should be avoided and facial nerve palsy could result from biopsy of a schwannoma.

Management of middle ear meningioma consists of complete surgical excision. This has the potential for complexity owing to anatomy of the middle ear and invasiveness of tumour.

At surgery in this case, a fleshy tumour was seen filling the middle ear space and hypotympanum. Excision of the tumour in the hypotympanum can be carried out in the usual manner. However, tumours fixed to the incudostapedial joint always leave risk of damage to the stapes footplate and subsequent complete sensorineural hearing loss. Such tumours could benefit from excision with the use of laser.

If a tumour is identified prior to surgery then multidisciplinary team involvement may be prudent. Long-term follow-up with serial imaging is necessary to exclude recurrence.³ Estimates of recurrence rate for meningiomas vary widely (7–84%) but prognosis is good and there is not thought to be a reduction in life expectancy.¹

Conclusion

Primary meningioma of the middle ear is rare but needs to be considered by the otolaryngologist in

the differential diagnosis of middle ear pathology. Presenting symptoms are non-specific which may lead to incorrect diagnosis and delayed treatment. Histology and immunochemistry are crucial for diagnosis. MR imaging with enhancement is required to rule out intracranial involvement and periodic follow-up is required to detect recurrence.

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