

Cerebellopontine angle meningioma resulting in middle-ear polyp

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Abstract

Extracranial spread of meningiomas to involve the middle ear is very rare. We present the case of a 43-year-old woman with a known cerebellopontine angle meningioma who subsequently presented with left-sided otalgia and a middle-ear mass extruding through the tympanic membrane due to local invasion of the meningioma. The tumour was excised surgically. A discussion of the relevant literature is also presented.

Key words: Meningioma; Middle Ear; Temporal Bone; Otologic Surgical Procedures

Case report

A 43-year-old woman presented with a long history of left-sided tinnitus, deafness and otalgia of uncertain duration. She had had a dumbbell neurofibroma in the L1/2 region removed 14 years previously but otherwise had no relevant past medical history.

Clinical examination was unremarkable except for left-sided hearing loss, which was confirmed to be sensorineural in nature on audiometry, with a maximal hearing loss of 80 dB at 8 kHz.

Magnetic resonance imaging (MRI) demonstrated an en plaque meningioma of the left cerebellopontine angle. An incidental mucocele of the sphenoid sinus was also present. An MRI scan of the entire spine was unremarkable apart from evidence of the previous surgery.

An initial course of conservative treatment was undertaken. One year later, the patient complained of worsening left otalgia. On examination, a tumour was found to be bulging through the posterior and inferior aspects of the left tympanic membrane, associated with a middle-ear effusion (Figure 1).

High resolution computed tomography (CT) with soft tissue contrast demonstrated an en plaque meningioma of the left cerebellopontine angle extending into the jugular foramen (Figure 2a). High resolution CT using bone windows demonstrated deaeration of the left mastoid air cells, localized hyperostosis and a soft tissue mass within the middle ear (Figure 2b). An MRI scan demonstrated the meningioma en plaque to be extending down to the left jugular canal, through which it extended a short way. There was no evidence of glomus tumour on angiography.

The neurosurgical team undertook a retrosigmoid approach to the lesion, finding a vascular tumour surrounding and anterior to the VIIth and VIIIth nerve complex, extending through the jugular foramen and eroding the petrous bone. The intracranial portion was removed piecemeal leaving the introsseous portion. Histology confirmed the lesion to be an angiomatous meningioma.

The patient made a good post-operative recovery, but progressive symptoms suggestive of lower cranial nerve

compromise (a weak voice and mild swallowing difficulties) prompted a second operation by the otolaryngology team. A radical mastoidectomy was performed, during which a tract of tumour was noted to be passing through the mastoid cavity. The tumour filled the middle ear and extended into the eustachian tube and anterior hypotympanum. The ossicular chain had been destroyed by tumour. Histology confirmed the lesion to be angiomatous meningioma identical to the previous lesion, with no evidence of atypia. The patient made an uneventful post-operative recovery. Post-operative audiometry was unchanged from that pre-operatively. The patient's voice and swallow remained unchanged.

At the time of writing, 20 months after the initial retrosigmoid operation, the patient remained under follow-up care. She had been monitored with MRI scans, which showed some residual meningioma in the lower posterior portion of the petrous temporal bone, extending through the jugular canal, in continuity with a mass of residual tumour lying immediately below the skull base, encasing the internal carotid artery and extending down to the level of the body of C2. This tumour volume has grown 'very slightly' in the nine-month interval between scans. The management plan was to keep the patient under active observation, both clinically and radiologically. There were no plans to intervene surgically provided she remained clinically stable.

Discussion

Meningiomas account for 13 to 18 per cent of all primary intracranial neoplasms; 20 per cent of intracranial meningiomas eventually develop an extracranial extension, most commonly to the orbit.¹ Following invasion of the temporal bone, the most common extension route is through the jugular and lacerate foraminae (as seen in this case) into the nasopharyngeal, retromaxillary, retromandibular and cervical spaces.¹ However, meningiomas involving the middle ear are very uncommon. Meningiomas of the middle ear may be primary (very rare) or



FIG. 1

The left ear demonstrating the tumour mass bulging through the tympanic membrane.

secondary due to extension of an intracranial meningioma.² There has been some debate in the literature as to the origin of primary middle-ear meningioma (also called ectopic meningioma). One theory is that they are the result of differential maturation of pluripotential mesenchymal cells.³ It has also been postulated that they originate from pinched off embryonal arachnoid cell nests which are thought to lie along the lines of fusion of primitive bones and along nerve sheaths.⁴ It should also be noted that most of the reported cases of ectopic meningioma were published before modern imaging techniques were available to exclude an intracranial component, leading to speculation that these were not true primary meningiomas but rather extension of unidentified intracranial lesions.⁵

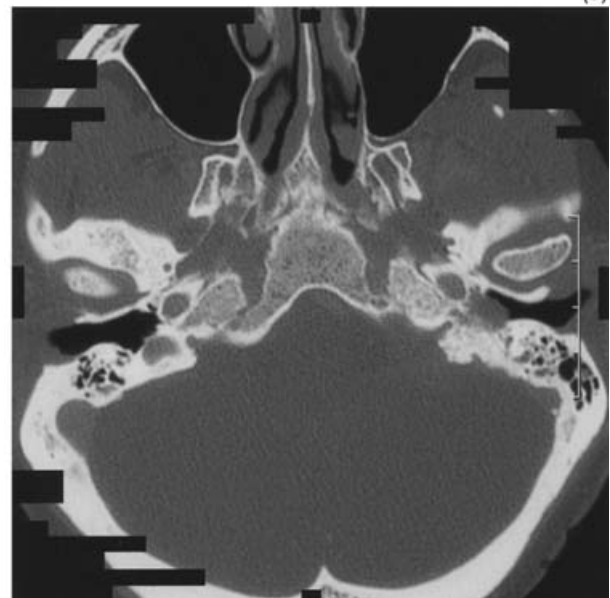
Despite the erosive pattern of growth described in this case and others, such tumours are not necessarily considered to be malignant. Although it is difficult to predict tumour behaviour from histological appearances, it is widely accepted that loss of tissue architecture, necrosis, increased mitotic activity and nuclear pleomorphism are generally associated with more aggressive behaviour; however, it must be emphasized that this is not an absolute.² The presence of brain invasion or distant metastases are accepted as reliable markers of malignancy; neither changes in the tumour suppressor gene p53 nor other immunohistochemical markers have been demonstrated to be reliable in this regard.⁶ Extracranial meningiomas do not differ histologically from those found intracranially. It has been reported that, of the four histological subtypes (meningotheiomatous, transitional, fibrous and angio- blastic), extracranial meningiomas are most commonly transitional or meningotheiomatous.⁷

It is accepted that, in contrast to their intracranial counterparts, meningiomas involving the temporal bone do not demonstrate discrete, well demarcated tumour margins.⁸ This was confirmed in this case by the finding of a rather ill-defined 'tract' of tumour within the mastoid cavity.

The known female preponderance of intracranial meningiomas is mirrored in their counterparts found in the middle ear. In one review of 20 middle-ear meningiomas, 85 per cent of the patients were female (as in this case).⁸



(a)



(b)

FIG. 2

(a) Computed tomography (CT) scan with contrast, demonstrating a soft tissue mass in the left cerebellopontine angle extending into the left jugular foramen. (b) CT scan (bone windows) revealing deaeration of the left mastoid air cells, localized hyperostosis and a soft tissue mass in the middle ear.

There is also an association with obesity.⁹ Approximately 30 per cent of meningiomas have oestrogen receptors and approximately 70 per cent have progesterone receptors; obesity increases the metabolism of androstenedione to oestrone, leading to an increase in circulating levels of oestrogen, which could act via the receptors on the tumour cells to induce proliferation.^{10,11}

Surgery is the preferred treatment for such lesions.^{6,7,9,12,13} The role of radiotherapy for such lesions is not defined but may include control of residual microscopic disease and reduction of the recurrence rate in the case of subtotal tumour excision.¹⁴ In the above case, it could be argued that an infratemporal fossa type A

approach should have been used rather than the jugular foramen approach, as the latter is associated with macroscopic tumour residuum. At the time, it was felt, both from clinical examination and from imaging, that the tumour was confined to the auditory canal and middle ear. At operation, however, it was found to be extending into the upper neck.

- A case is presented of a meningioma invading the middle ear and presenting as a middle-ear tumour with extension as a polyp into the external auditory canal
- Invasion of the middle ear by meningiomas is exceedingly rare
- A review of the literature, including pathogenesis and treatment, is presented

Because of the very slow growth rate of such tumours, most authors have suggested long-term clinical and radiological follow up of such patients; long-term follow up is also recommended even after an apparently extracranial tumour is removed, to exclude intracranial disease.^{7,8,12} There is no consensus as to whether CT or MRI is the imaging modality of choice in this setting.¹⁵

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