

Meningioma Presenting As A Mass In The External Auditory Canal.

V Hemanth, V Sreenivas, J Chaturvedi

Citation

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Abstract

Meningiomas presenting as an intracranial neoplasm are relatively common. (15–18%). However, primary extracranial (ectopic, extracalvarial) meningiomas of the ear and temporal bone are rare, making up for less than 1% of all meningiomas.¹ The known female preponderance of intracranial meningiomas is also seen in the external auditory canal meningioma. They may be primary (very rare) or secondary due to extension of an intracranial meningioma. The possibility of a meningioma should be considered whenever a mass presents in the middle or external ear and a high index of suspicion is needed for diagnosis of such a condition, especially in view of a variety of differentials ranging from benign to malignant conditions. Computed tomography scanning is invaluable, not only for the diagnosis of this condition, but also to see the intracranial extent.

INTRODUCTION

Meningiomas presenting as an intracranial neoplasm are relatively common. (15–18%). However, primary extracranial (ectopic, extracalvarial) meningiomas of the ear and temporal bone are rare, making up for less than 1% of all meningiomas.¹

Meningiomas of the external auditory ear may be primary (very rare) or secondary due to extension of an intracranial meningioma. Among the intracranial meningiomas, cerebello-pontine angle meningiomas have a tendency to involve the external auditory canal in the primary setting.²

We present a case report of a mass arising in the external auditory canal (EAC), presenting as a meningioma in a 45 year old lady, with review of relevant literature.

CASE REPORT

A 45 year old lady presented to the outpatient department of Otorhinolaryngology at a tertiary referral centre in Bangalore, India, with complaints of swelling in the right EAC since the past 2 months. The swelling was insidious in onset and gradually progressive. It was associated with decreased hearing on the same side since one month. There was no history of trauma to the ear or ear discharge, nor was the patient known to be a diabetic.

Examination of the right ear revealed a non tender mass filling the cartilaginous part of the right EAC, extending to

the deep meatus. It was firm on palpation. Examination of the other ear was normal.

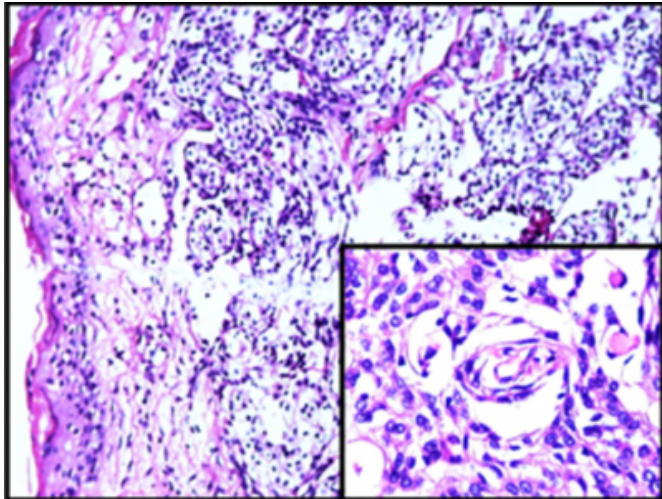
Tuning fork tests and audiometry showed a mild conductive hearing loss in the affected ear.

A presumptive diagnosis of a benign soft tissue lesion of the EAC was made, and we proceeded to take an excision biopsy of the mass in the right EAC under local anaesthesia.

The section of the specimen under microscope showed nests and whorls of cells with vesicular nuclei having few chromatin and pale eosinophilic cytoplasm. Interspersed with these cells were many vascular channels with hyalinised walls suggestive of findings consistent with meningioma. (Figure 1).

Figure 1

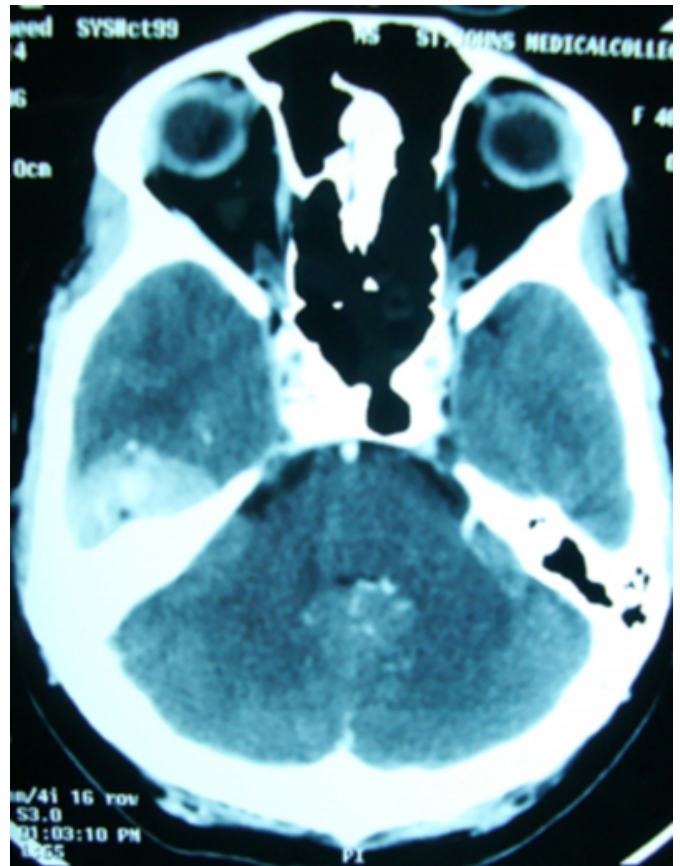
Figure 1: Hematoxylin and eosin stain: original magnification x 200 showing Nests and whorls of cells with vesicular nuclei having few chromatin and pale eosinophilic cytoplasm. Inset – Hematoxylin and eosin stain: original magnification x 400



Since meningioma presenting in the external auditory canal is relatively rare, we proceeded to investigate if the meningioma was purely extracranial or did it have an intracranial extension. A High Resolution Computerised Tomography (HRCT) of the temporal bone with contrast revealed an irregular, heterogeneously enhancing lesion, arising from the Petrous part of the right temporal bone, measuring 2.7 x 1.6 x 1.2 cm, and extending into the Right middle and external ear. (Figure 2).

Figure 2

Figure 2: CECT Temporal Bone: Irregular, heterogeneously enhancing lesion, arising from the Petrous part of the right temporal bone



She underwent neurosurgical intervention for removal of the petrous temporal bone meningioma and is currently maintaining well on regular follow up.

DISCUSSION

Temporal bone meningioma can be considered as a part of the ectopic meningioma group. The known female preponderance of intracranial meningiomas is also seen in external auditory canal meningioma.⁴ Up to 20% of intracranial meningiomas may have extraneuraxial extension,^{5,6,7,8} including the skull, scalp (all cutaneous sites), orbit, upper airway involvement (nasal cavity, paranasal sinuses, nasopharynx), soft tissues, and ear / temporal bone. However, when the scalp, orbit, sinonasal tract, oral cavity, and soft tissues are excluded, the incidence decreases to <1%^{6, 9, 10, 11.}

Ectopic or extracranial meningiomas are classified into 4 subgroups: direct extension from primary intracranial meningioma, extracranial growth from cranial nerve sheath arachnoidcells, extracranial extension from embryonic

arachnoid rests without connection to the skull base or cranial nerves, and distant metastasis from intracranial tumors.³

Meningioma gains access to the temporal bone from 3 principal sites of origin: tegmen tympani and jugular foramen (JF) meningiomas, which spread secondarily into the middle ear cavity and internal auditory canal (IAC) meningiomas, which invade the cochlea and vestibule¹²

Hearing loss is the most common clinical presenting feature in all cases of temporal bone meningioma¹². This is related to ossicular chain encasement by tumor, implying a component of surgically correctable hearing loss with appropriate management. Other symptoms include headaches, dizziness, unsteadiness, vertigo, disequilibrium, tinnitus, otalgia, bleeding, “metallic taste,” and a chronic cough. Facial or other cranial nerves may be involved in late cases.

Histologically, Zulch et al classifies Meningiomas into three types: a.. Meningothelial meningioma (Endotheliomatous) b.. Fibromatous Meningioma c. Angiomatous Meningioma.¹³ However, on histology, ear and temporal bone meningiomas are indistinguishable from their intracranial counterparts.¹

Computed tomographic views of Meningiomas may show presence of hyperostosis and permeative, sclerotic, and/or destructive changes in the skull base.¹² However, Gadolinium -DTPA-enhanced MRI has a much higher sensitivity for meningiomas than does for CT.¹⁴

Differential diagnosis of other lesions in the middle/external ear should be considered. These include schwannomas, paragangliomas, squamous cell carcinomas, adnexal tumors, middle ear adenomas, ceruminous gland tumors, primary and metastatic carcinomas, and malignant melanomas, all of which have a more sinister course than a meningioma.⁴

Left alone, the course of a meningioma appears to be variable. They show a proclivity for local permeation of crevices, suture lines, and foramina of the skull.¹⁴ Bone infiltration in meningioma most commonly occurs by secondary spread, though primary intradiploic tumors are reported. Secondary infiltration of bone may reflect tumor involvement or reactive changes; however, these are not reliably differentiated on imaging.¹²

Although surgery is the treatment of choice, there are a number of challenges because of the invasiveness of the tumors and the complexity of the anatomy of the ear and

temporal bone, especially if cranial nerve function and hearing preservation is intended.¹⁴

Therefore, it probably is best to use a multidisciplinary approach with a combination of intracranial, temporal bone (subtemporal, translabyrinthine, or transcochlear), and skull base (suboccipital) techniques to achieve total resection, possibly including widely exenterative procedures to achieve this end^{15,16,17}.

Radiation therapy has been suggested to yield a possible improvement in survival in cases of meningioma of the central nervous system.¹⁸

CONCLUSION

Meningiomas presenting in the EAC are rare (<1% of meningiomas). Patients most commonly present with complaints of swelling in the EAC and conductive hearing loss. The possibility of a meningioma should be considered whenever a tumor presents in the middle or external ear. A high index of suspicion is needed for diagnosis of such a condition, especially in view of a variety of differentials ranging from benign to malignant conditions.

Computed tomography scanning is invaluable, not only for the diagnosis of this condition, but also to see the intracranial extent. Surgical removal is the main modality of treatment, and should be planned with the active involvement of a neurosurgical team.

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Author Information

V. Hemanth

Resident, Department of Otolaryngology and Head and Neck Surgery, St. Johns Medical College and Hospital

V. Sreenivas

Associate Professor, Department of Otolaryngology and Head and Neck Surgery, St. Johns Medical College and Hospital

Jagdish Chaturvedi

Resident, Department of Otolaryngology and Head and Neck Surgery, St. Johns Medical College and Hospital