



PRIMARY EXTRACRANIAL MENINGIOMA IN MIDDLE EAR: A RARE CASE REPORT

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ABSTRACT

Meningiomas are the most common intracranial extra-axial brain tumor. Rarely, meningiomas can occur extra cranially from ectopically located arachnoid cell rests outside the dural covering of the brain and spinal cord. Extradural meningiomas do not have connections to the dura or any other intracranial structure. We report, case of a 63 years old female and discuss, imaging feature and the histopathologicals data.

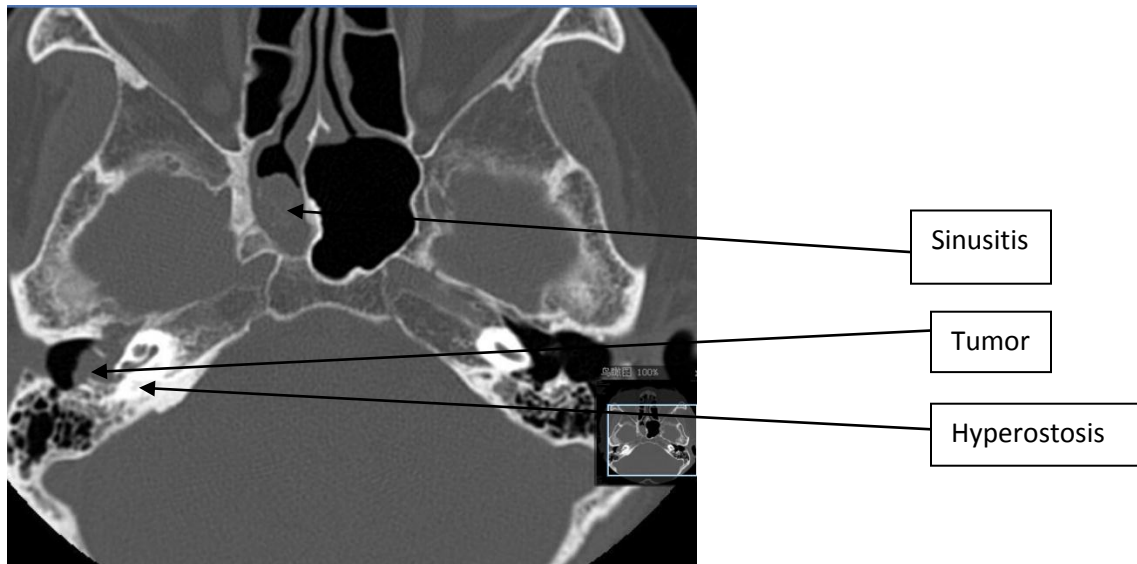
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INTRODUCTION

Meningiomas are non glial cell tumor that originates from the meningocytes or arachnoid cap cell of meninges accounting for 13% to 19% of all primary brain tumors(1). In contrast, extracranial meningioma are rare, and particularly those with primary ectopic meningioma in middle ear(2). It can extend, with diverse clinical findings according to route. Especially, ear and temporal bone meningiomas have been reported to show a variety of clinical findings such as cochlear and vestibular symptoms, moreover cranial nerve palsy and neurological symptoms. We describe a case of primary ectopic meningioma in middle ear.

Case Report:

A 63-year-old female presented at our hospital ENT department with a complaint of right sided hearing loss with intermitted vertigo. There was no significant history of trauma. ENT examination showed congested right tympanic membrane without specific discharge and any sign of infection. Bilateral nasal passage clear, no congestion of pharynx, no swelling of the epiglottis and vocal cord. The patient had no neurological deficit. The laboratory studies were unremarkable. For detail study and doubtful symptom patient was advised for CT scan which revealed a focal tumor with soft tissue density, 15×8 ×13 mm in size and well defined border is shown in tympanic cavity of Middle ear in plain CT scan. The auditory ossicles are wrapped in the tumor with no bone destruction. The structures of inner ear including internal auditory canal are normal (figure 1). Histologically, the tumor was diagnosed as fibrous meningioma because it contained spindle shaped tumor cells, with narrow rod-shaped nuclei. These cells are embedded in abundant collagenous or reticulum background and psammoma bodies are only occasionally encountered. No tumor necrosis is identified. No cerebral tissue is identified (figure 2).



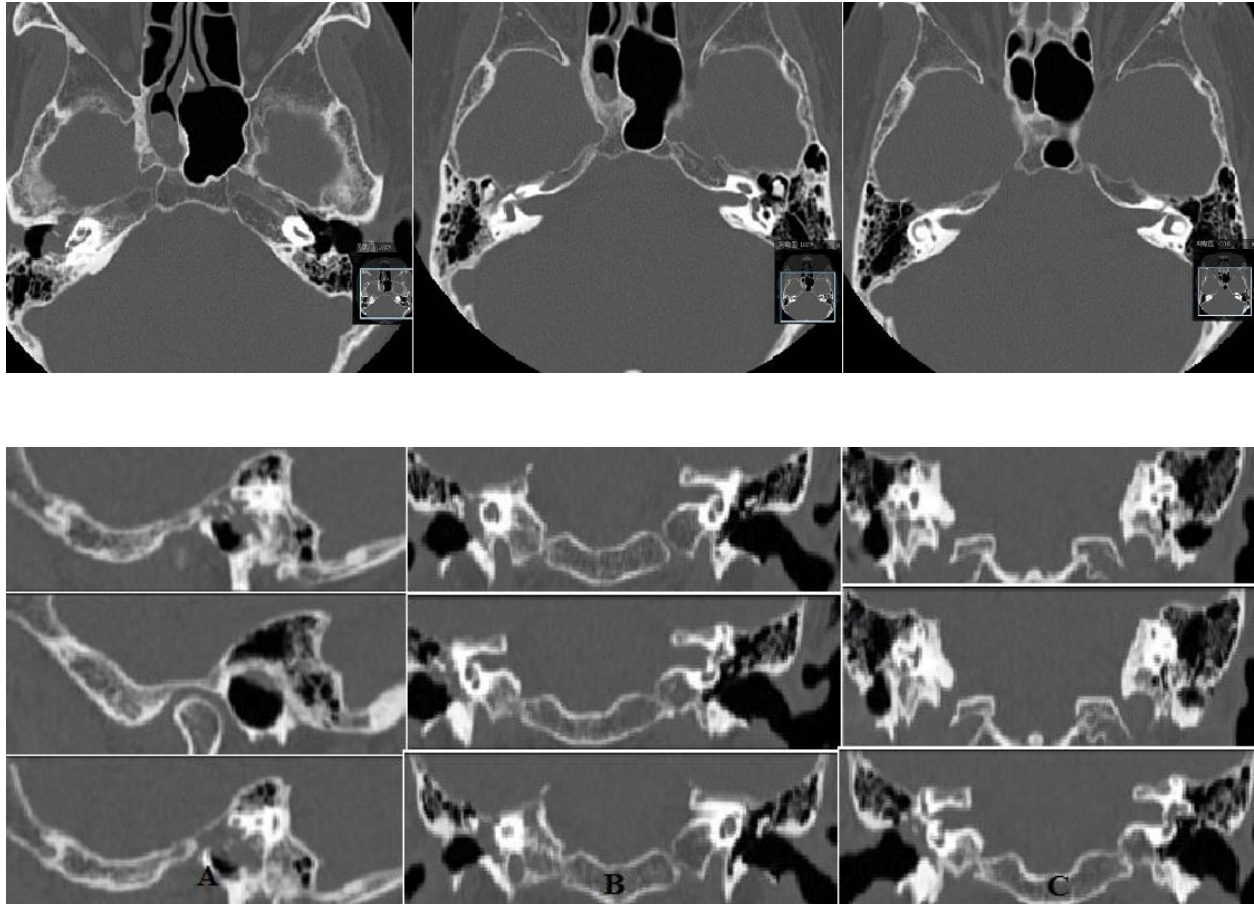


Figure 1: (A , B and C) CT findings: a focal tumor with soft tissue density, 15×8 ×13 mm in size and well defined border is shown in tympanic cavity of Middle ear in plain CT scan. The auditory ossicles are wrapped in the tumor with no bone destruction. The structures of inner ear including internal auditory canal are normal.

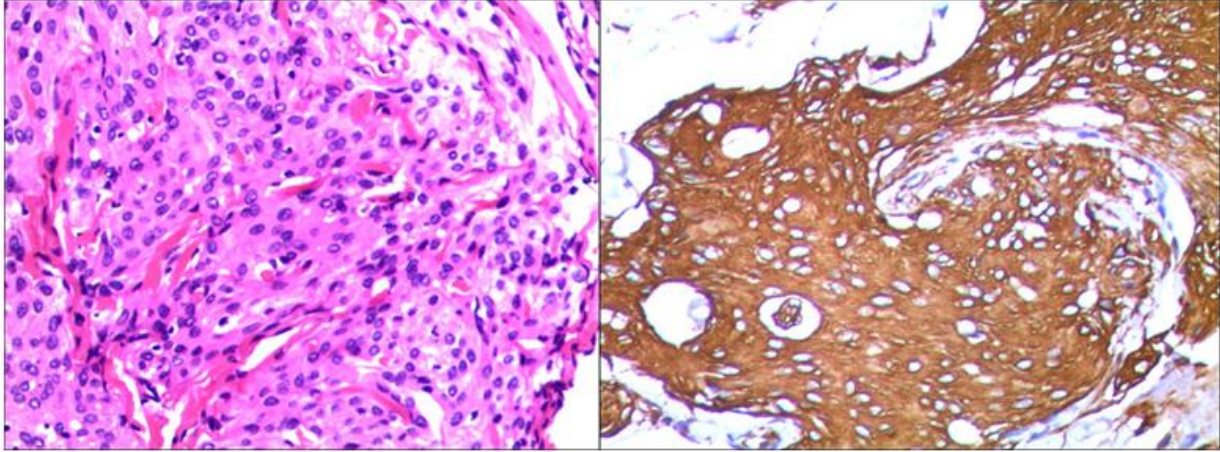


Figure 2: Spindle shaped tumor cells, with narrow rod-shaped nuclei. These cells are embedded in abundant collagenous or reticulum background and psammoma bodies are only occasionally encountered. No tumor necrosis is identified. No cerebral tissue is identified.

DISCUSSION

Extracranial meningiomas are rare; the reported incidence is 1%–2% of all meningiomas (3, 4). The meningiomas arising in locations outside the dural compartment have been called ectopic, Extradural (epidural), calvarial, cutaneous, extracranial, extraneuraxial, or intraosseous meningiomas. To avoid the confusion in nomenclature, Lang et al (5) has proposed a single term, “primary extradural meningioma” for such lesions. This term highlights the origin of these tumors as being separate from the dural coverings of any part of the brain or spinal cord and further differentiates these meningiomas from “primary intradural meningiomas,” which may have secondary extracranial extensions and/or may have metastasized (5).

The most common sites of meningiomas are the sphenoid ridge and the dome of the cerebrum, followed by the falx cerebri and parasagittal sinus. In the arising processes of the extracranial meningioma, Hoyer (6) reported that extracranial meningiomas could be subdivided into the following four main groupings: i) Primarily intracranial tumors with extracranial direct extension. ii) Tumors originating from arachnoid cell rests of cranial nerve sheaths with extracranial growth. iii) Extracranial growth without any apparent connection with foramina or cranial nerves presumably from embryonic rest of arachnoid cells. iv) Benign appearing intracranial meningiomas with extracranial metastases. Pathways for meningioma extension into the middle ear as suggested by Rietz (2) were the tegmen tympani, greater superficial petrosal groove, anterior and posterior surfaces of the petrous pyramid, internal auditory canal, and jugular fossa. Various clinical findings can present according to the route of by which the tumors extend. Rushing *et al.*

(7), Thedinger *et al.*(8), Goel(9), Thompson *et al.*(10) reported that ear and temporal bone meningiomas could show variety of clinical findings as cochlear and vestibular symptoms or cranial nerve palsy. Hamilton *et al.*(11) revealed that hearing loss was the most common clinical presenting feature in temporal bone meningiomas. The complaint of our patient was hearing loss alone, but no tinnitus, vertigo/dizziness and facial nerve palsy. A pure tone audiogram showed a mixed hearing loss in right ear, and almost equals left and right bone conduction. Her imaging findings showed that the right sphenoid ridge meningioma had invaded widely into the right middle cranial fossa and involved the extension along the petrous apex and bilateral optic nerve canals, but the internal auditory canal, facial nerve canal, jugular fossa cochlea, vestibule, semicircular canal, facial nerve canal and ossicles were intact. Therefore, bone conductive loss would be caused by aging change rather than tumor invasion for internal auditory canal, cochlea vestibule and ossicles. In her past history, she previously underwent craniotomy 4 years before for the sphenoid ridge meningioma with right visual impairment. From the above findings, past history and literatures, the symptoms of our case would appear to result from the extension of an extracranial meningioma from the sphenoid ridge to the middle ear via the petrous pyramid.

The patient underwent only partial extirpation with decompression for the optic nerve, rather than total extirpation including middle ear and temporal bone, due to the wide invasion of the middle cranial fossa and cavernous sinus. Some hospitals(12-14) reported that sphenoid wing and temporal meningiomas involved cavernous sinus or optic nerve like as our case. Mirone *et al.*(13) presented that surgical management of patients with sphenoid wing meningiomas could not be uniform. It should be tailored on a case-by-case basis. Therefore, they recommend the optic canal decompression in all patients to ameliorate or preserve visual function.

Differential Diagnosis:

- A. **Metastasis:** Metastatic tumors are very common in the late stages of cancer. Lung, breast, skin, colon, liver, cervix are common site of origin. History, sign and symptoms reveals no any significant near and distant primary tumor.
- B. **Osteosarcoma:** Malignant bone forming tumor occurs in primary and secondary form usually secondary to malignant degeneration of Paget disease, extensive bone infarcts, post-radiotherapy for other conditions, osteochondroma, and osteoblastoma.
- C. **Hemangioma:** Common benign tumors relatively common in head and neck distinct but very rare in temporal bone. Depending on its clinical presentation it can be mistaken for other vascular masses such as glomus tumor or other lesion of the middle ear. Aural bleeding, pulsatile tinnitus, and facial nerve dysfunction are presenting symptoms.
- D. **Giant cell tumor:** Also known as osteoclastoma and are relatively common benign long bone tumor, typically presents with bone pain, soft tissue mass, compression of adjacent structures or acutely

with a pathological fracture.

- E. **Olfactory neuroblastoma:** Rare malignancy of nasal cavity Neuroectoderm often present late with larger tumor which can extend into cranium and usually present with anosmia, facial numbness, nasal congestion, eye pain.
- F. **Paraganglioma:** Highly vascular glomus tumor that arises from the paraganglion cells of the carotid body. It is usually located anterior to the Sternocleidomastoid near the angle of the mandible at the level of the hyoid bone. Contrast enhanced CT is excellent for differentiation from other pathology.
- G. **Melanoma:** Malignant neoplasm that arises from melanocyte a basal layer of the epidermis arises in the skin. However, melanocyte does occur in other unexpected locations. Usually bilateral with multiple lesion.

ABBREVIATIONS

CT: Computed Tomography, MRI: Magnetic Resonance Imaging, ENT: Ear Nose and Throat

CONCLUSION

Identification of typical meningioma area is key for the diagnosis of various types of meningioma. Extracranial meningioma are rare, and particularly those with extension into a middle ear. It can extend, with diverse clinical findings according to route. Especially, ear and temporal bone meningiomas have been reported to show a variety of clinical findings such as cochlear and vestibular symptoms, moreover cranial nerve palsy and neurological symptoms. CT scan together with Histopathology play a role in diagnosis of such type of meningioma.

REFERENCES

1. Kusunoki T, Ikeda K, Miyashita M. A case report of meningioma extending to the middle ear. Clinics and practice. 2012;2(3).
2. Rietz DR, Ford CN, Kurtycz DF, Brandenburg JH, Hafez GR. Significance of apparent intratympanic meningiomas. The Laryngoscope. 1983;93(11):1397-404.
3. Muzumdar D, Vengsarkar U, Bhatjiwale M, Goel A. Diffuse calvarial meningioma: a case report. Journal of postgraduate medicine. 2001;47(2):116.
4. Whicker JH, Devine KD, MacCarty CS. Diagnostic and therapeutic problems in extracranial meningiomas. The American Journal of Surgery. 1973;126(4):452-7.
5. Lang FF, Macdonald OK, Fuller GN, DeMonte F. Primary extradural meningiomas: A report on nine cases and review of the CT-era literature. Journal of neurosurgery. 2000;93(6):940-50.
6. Hoye SJ, Hoar CS, Murray JE. Extracranial meningioma presenting as a tumor of the neck. The American

Journal of Surgery. 1960;100(3):486-9.

7. Rushing EJ, Bouffard J-P, McCall S, Olsen C, Mena H, Sandberg GD, et al. Primary extracranial meningiomas: an analysis of 146 cases. *Head and neck pathology*. 2009;3(2):116-30.
8. Thedinger BA, Glasscock III ME, Cueva RA. Transcochlear transtentorial approach for removal of large cerebellopontine angle meningiomas. *Otology & Neurotology*. 1992;13(5):408-15.
9. Goel A. Extended middle fossa approach for petroclival lesions. *Acta neurochirurgica*. 1995;135(1-2):78-83.
10. Thompson LD, Bouffard J-P, Sandberg GD, Mena H. Primary ear and temporal bone meningiomas: a clinicopathologic study of 36 cases with a review of the literature. *Modern pathology*. 2003;16(3):236-45.
11. Hamilton B, Salzman K, Patel N, Wiggins R, Macdonald A, Shelton C, et al. Imaging and clinical characteristics of temporal bone meningioma. *American journal of neuroradiology*. 2006;27(10):2204-9.
12. Civantos F, Ferguson LR, Hemmati M, Gruber B. Temporal meningiomas presenting as chronic otitis media. *Otology & Neurotology*. 1993;14(4):403-6.
13. Mirone G, Chibbaro S, Schiabello L, Tola S, George B. En plaque sphenoid wing meningiomas: recurrence factors and surgical strategy in a series of 71 patients. *Neurosurgery*. 2009;65(6):ons100-ons9.
14. Stacy RC, Jakobiec FA, Lessell S, Cestari DM. Monocular nasal hemianopia from atypical sphenoid wing meningioma. *Journal of Neuro-Ophthalmology*. 2010;30(2):160-3.