

## **Extracranial Meningioma in Temporal Bone: An Unusual Diagnose**

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**Conflicts of Interest:** Nil

### **Abstract**

Meningiomas are one of the commonest intracranial tumors, but their occurrence extracranially is very rare. Due to rare occurrence, an extracranial meningioma is often misdiagnosed and only after a biopsy and a histopathological examination diagnosis is confirmed. This presentation reports a case of Extracranial Meningioma occurring in temporal region misdiagnosed as soft tissue sarcoma or benign hypertrophy, and only after excision and histopathological examination, the diagnosis was confirmed. This case report also emphasizes on the importance of accurate diagnosis of extracranial meningiomas, in order to determine timely intervention and appropriate therapeutic modality.

**Keywords:** Meningioma, Temporal, Extracranial, Temporal Swelling.

### **Introduction**

Meningiomas are the second most common tumors of central nervous system with an incidence of 10-30% of all

primary intracranial tumors, second only to gliomas<sup>1,2,3,4,5</sup>.

These intracranial tumors very rarely occur at extra cranial sites, with less than 2% cases reported<sup>4</sup>. Most common site for occurrence of meningioma at an extracranial site is in head and neck region, and these tumors have been most commonly identified in temporal region, middle ear, external auditory meatus and paranasal sinuses. Due to rare occurrence, an extracranial meningioma is often misdiagnosed and only after a biopsy and a histopathological examination diagnosis is confirmed.

We report a unique case of extracranial meningioma where an extracranial meningioma was present in relation to temporal bone along with appropriate discussion and management options.

### **Case Report**

A 63 year old female patient reported with a chief complaint of slowly growing painless swelling on right side of the face and head since past 2 years. The swelling was not associated with any relevant symptoms like pain,

change in color of overlying skin, discharge or nerve paralysis. On examination a 6 cm x 7 cm swelling extending from right lateral orbital margin to preauricular region and from lower border of zygomatic arch to mid temporal region was present [Figure 1a and 1b]. The swelling was firm in consistency and non-tender. Patient had CT scan done and the CT report suggested homogenous enlargement of right temporalis muscle extending from calvarium to retro antral region, along with diffuse sclerotic thickening/remodeling of bony calvarium on right lateral wall of orbit, greater wing of sphenoid, frontal and temporal bone. CT report gave a provisional diagnosis of a) soft tissue sarcoma b) Benign hypertrophy of temporalis muscle and reactive thickening of adjoining bones. [Figure 2a and 2b]

An incisive biopsy was planned under local anesthesia, and a 1 cm x 1 cm tissue sample was acquired and send for histopathological examination. The biopsy report suggested presence of normal muscle tissue and after consultation with the pathologist a deeper tissue biopsy was suggested. An excisional biopsy was planned under general anesthesia. A temporal incision was given and layer wise dissection was done [Figure 3]. After elevation of the temporal muscle a white, firm fibrous like mass was encountered adjoining and firmly adherent to the bone. The lesion was carefully detached from the underlying bone and removed in total along with temporalis muscle adherent to the tumor mass. Recontouring of the underlying bone was done using a bone rongeur as an attempt to remove any residual tumor tissue. However due to operative impression of a benign muscle tumor, a conservative approach was agreed upon, and the closure of the wound was done. [Figure 4a and 4b]

Histopathology of the lesion showed spindle cells with mild hyperchromatia, increased cytoplasm, increased mitotic figures and excessive collagenous tissue. There

was presence of psammomma bodies and whorl formation. The tumor was also extending into skeletal muscle fibers. The microscopic features of the tumor were suggestive of meningothelial meningioma extending extracranially in temporalis muscle.

The patient was further referred to neurosurgeon for further management however regular follow up of the patient was done.

### **Discussion**

Neural crest cells are source of origin of arachnoid cells and these arachnoid cells are believed to be the source of origin of a meningioma.

Rushing et al summarized the development of extracranial meningioma as follows:

1. Arachnoid cells present in nerve sheaths or vessels emerge through skull foramina and result in extracranial meningiomas.
2. Pacchionian bodies during embryonic development get entrapped or detached at an extracranial site.
3. Trauma or cerebral hypertension leads to displacement of arachnoid islets.
4. Undifferentiated or multipotential mesenchymal cells, by one mechanism or other consequently give rise to meningiomas at extracranial sites<sup>5</sup>.

Biologically, extracranial meningiomas are benign and slow growing in nature <sup>5, 6</sup>. Extracranial meningiomas can be either secondary i.e. associated with an intracranial mass or primary i.e. not associated with intracranial mass. The tumor may be present in various extracranial sites in head and neck region such as temporal, nasal, paranasal sinus, middle ear, external auditory meatus, Eustachian tube, and jugular fossa<sup>4</sup>. Symptoms of the tumor varies with the location. In the case reported here, facial asymmetry was the main complaint of the patient and the patient only sought treatment for that.

Due to benign nature and slow growth rate the prognosis of extracranial especially temporal bone meningiomas is good following complete surgical resection. However due to difficulty in initial diagnosis, due to incomplete excision, recurrences also fairly common. Recurrent tumors are usually treated with adjuvant radiation therapy, and no role of chemotherapy has yet been suggested. Recurrent rate varies from 6% at 5 years to 20% at 20 years<sup>4</sup>.

### Conclusion

The case report presented here is a rare occurrence of an extracranial meningioma which was misdiagnosed as a) temporalis hyperplasia and b) soft tissue sarcoma. It was only after complete excision of the lesion and histopathological examination the diagnosis was made. Preoperative suspicion of a meningioma, or a contrast-enhanced computer tomography would have led to a more aggressive approach. To summarize, any swelling in temporal region, which is slow growing with no specific signs should be thoroughly investigated clinically and with CECT with a possibility of extracranial meningioma in mind. This would lead to a more specific diagnosis and a planned surgical intervention.

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### Legends Figure



Figure 1a: Preoperative



Figure 1b: Preoperative





Figure 2a : CT View Axial

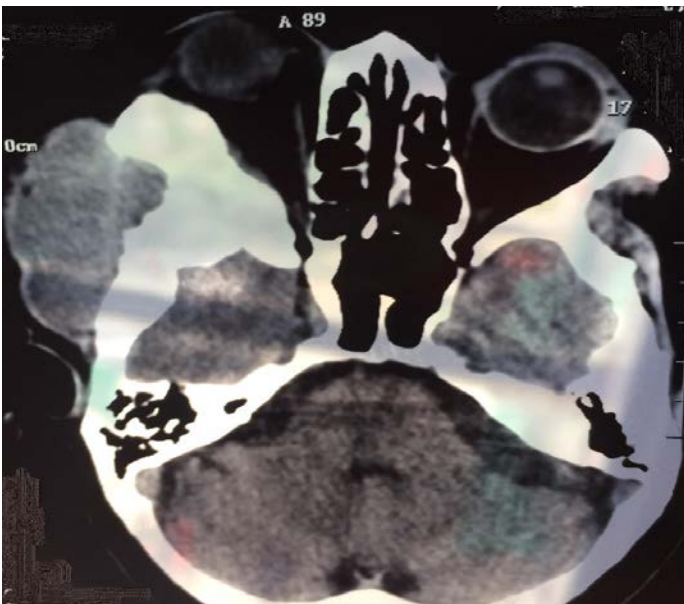


Figure 2b: CT View Coronal



Figure 3: Intraoperative Tumor

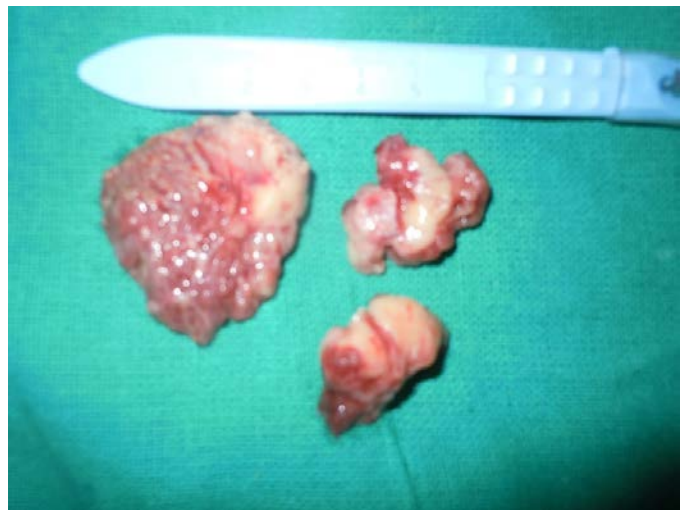


Figure 4: Tumor Tissue Excised



Figure 5: Post Operative



Figure 6: 3 month Post Operative