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Rare Presentation of Meningioma as an External Auditory Canal Mass

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Abstract

This is a case of a 60-year-old male with a history of prior left middle fossa meningioma that was partially resected with an operative report noting diffuse attachment to the middle fossa floor. Gamma knife was recommended but he never completed this management. He then presented about eight years later with a mass from his left external auditory canal. It was reported that two years prior another surgeon operated on the left ear for a cholesteatoma. CT temporal bone showed complete opacification of left EAC, mastoid bowl, and remaining mastoid air cells. In addition, there were irregular bony/hyperostotic changes seen within the left sphenoid and temporal bone. There was dural thickening within the middle fossa adjacent to the previously described hyperostotic bony changes. A mastoidectomy and excision of mass revealed extensive adhesive tissue throughout the middle ear, and mastoid up to the tegmen. Pathology of the portions that were resected confirmed Grade 1 meningioma. Stereotactic gamma knife radiation was completed to the area to prevent further growth. This case highlights extracranial meningioma that did not have definitive management for prior middle fossa floor meningioma. It also highlights the need to think of less common pathology in the middle ear and external auditory canal.

Keywords

Middle Ear, Cranial Base, Otology/Neurotology

1. Introduction

Meningioma is a very common tumor found within the central nervous system and accounts for 10% - 15% of all primary intracranial neoplasms [1] [2]. Extracranial spread usually occurs via transosseous or neural/vascular foramina.

The prevalence of these extracranially spreading meningiomas is around 6% - 20% [3] [4]. The common sites of spread are to the orbit, paranasal sinuses and temporal bone [3] [4]. Due to the slow progression of disease in conjunction with nonspecific findings on computed tomography (CT) temporal bone, extracranial extension of temporal bone meningioma can be confused with more commonly seen middle ear space disease such as chronic otitis media or cholesteatoma. We present the clinical and radiologic case of a 60-year-old male who presented to our clinic and was found to have recurrent grade 1 meningioma within the mastoid, middle ear space (ME) and a large external auditory canal (EAC) mass. There have not been other case reports of a large fungating EAC mass found to be a meningioma from extracranial spread. The patient gave consent for this case report.

2. Case Report

A 60-year-old male with a past medical history of prior meningioma status post excision nine years earlier is now presenting due to left ear tinnitus, otalgia, and hearing loss in November of 2023. He reported that left temporal and anterior craniotomy was performed with an operative report denoting "diffuse attachment to the middle fossa floor." Magnetic resonance imaging (MRI) showed a 4.0 × 3.9 × 2.7 cm extra-axial mass along the left sphenoid wing projecting superolateral and involving the cavernous sinus (**Figure 1**). The meningioma was resected and the dura was repaired duragen. The cavernous sinus was not addressed to avoid injury to critical structures. Follow up MRI showed extra-axial enhancement representative of residual meningioma which was 1 cm in size, located in the left temporal fossa.

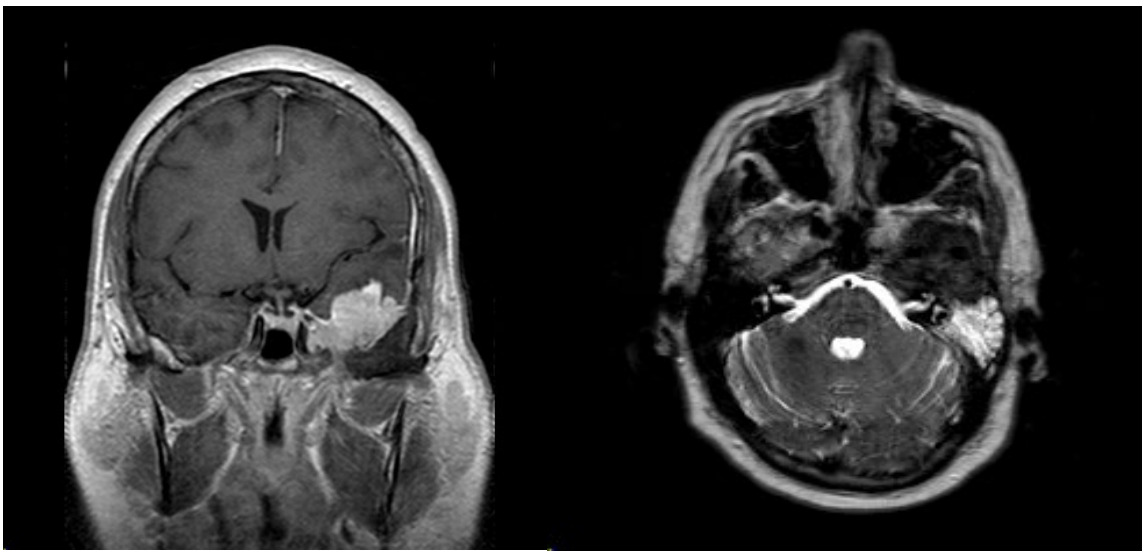


Figure 1. Patient's MRI prior to original craniotomy denoting a coronal view of T1 enhancing left temporal meningioma (Left). Review of the mastoid cavity at this time shows T2 enhancement likely representative of mastoid disease vs meningioma involvement upon initial presentation (Right).

Three years following initial treatment for the meningioma the patient underwent canal wall up mastoidectomy at an outside facility to address suspected cholesteatoma (operative reports unavailable). The patient was then seen in our office 8 years following initial craniotomy with an exam revealing complete EAC occlusion with what appeared to be granulation tissue and purulence in November 2023. CT showed complete opacification of left EAC, mastoid bowl, and remaining mastoid air cells. In addition, there were irregular bony/hyperostotic changes seen within the left sphenoid and temporal bone. There was dural thickening within the middle fossa adjacent to the previously described hyperostotic bony changes (Figure 2). In addition, MRI confirmed residual/recurrent meningioma abutting the left Meckel's cave and along the cisternal segment of the left trigeminal nerve (Figure 3).

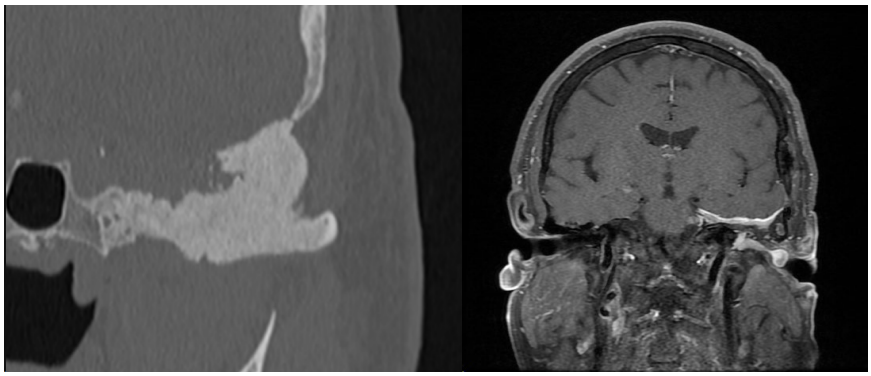


Figure 2. CT temporal bone prior to revision mastoidectomy showing the coronal view of hyperostotic bony changes and erosion along the left middle fossa and tegmen concerning for meningioma (Left). MRI at the same time shows a coronal view of diffusion weighted enhancing mass filling the left EAC in addition to persistent dural enhancement in the area of prior resection (Right).

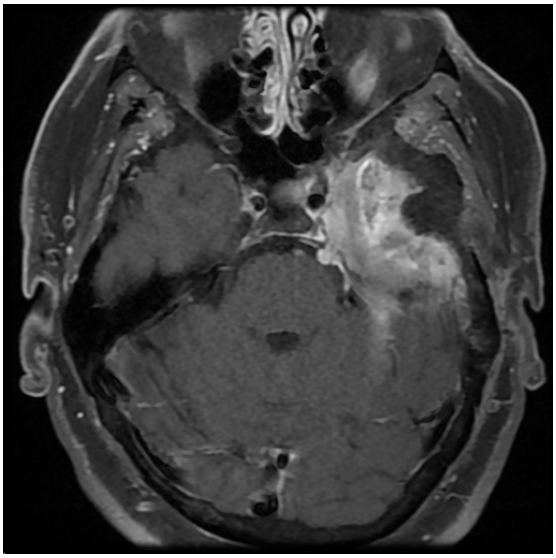
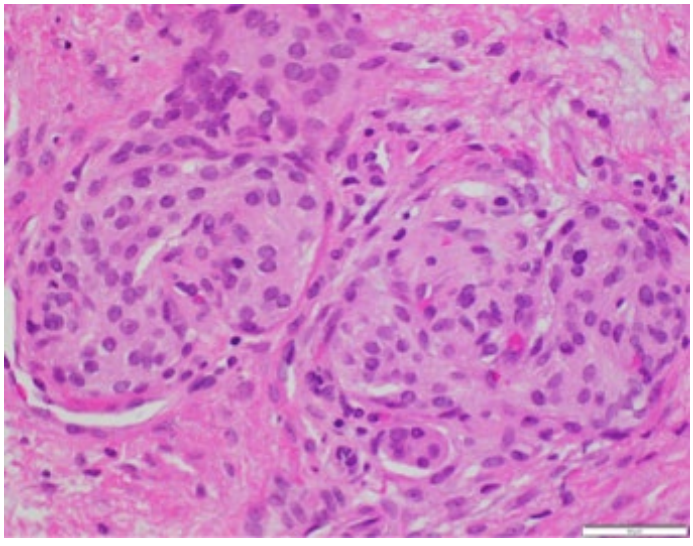


Figure 3. Axial T1 weighted MRI prior to revision mastoidectomy showing residual 6 mm focus within Dorello's canal and adjacent left middle fossa dural enhancement.

The patient returned to the office for biopsy of the EAC soft tissue which showed granulation tissue and epidermal hyperplasia, although clinical history was still concerning for meningioma. Decision was made to proceed with revision mastoidectomy and canalplasty in December 2023. Intraoperatively, the mass was more completely defined as a fibrotic, epithelial appearing lesion which filled the antrum of the mastoid in addition to the mesotympanum and epitympanum. This appeared to originate from the middle ear and external auditory canal. Complete resection of the mass was deferred, as the mass appeared to be highly vascular, densely adherent in all remaining air cells, filling the middle ear with adherence to ossicles, and into the eustachian tube. Complete excision would be difficult due to the extent of disease and increased risk to critical structures. There was also dense, firm bony changes of the EAC surrounding the resected mass. Pathology confirmed WHO grade I meningioma (**Figure 4**). The patient recovered well postoperatively, although on repeat examination and imaging, he was found to have regrowth of the EAC mass with recurrent EAC stenosis. Repeat MRI showed improved caliber of the left EAC, with residual tumor within the middle ear and medial EAC. The patient was referred for evaluation by neurosurgery and radiation oncology. Since the mass was adherent and wide spread and in more an en plaque configuration the decision was made to proceed with sterotactic gamma knife. This is due to the fact that complete gross resection would be almost impossible with this type of spread to areas such surrounding the ossicles, and round and oval windows. Most recent course includes follow up treatment using stereotactic gamma knife radiotherapy in April 2024. Planning included 3.7 cm in the middle ear and mastoid and 6mm around Meckel's cave. 12.5 gray was applied to both the EAC and cavernous sinus. He is scheduled for follow up MRI and audiogram at 6 months post-radiation in October 2024. He has not reported any complications since the gamma knife radiation. It was discussed with the patient that hearing loss, dizziness, tinnitus, and radiation necrosis requiring steroids were all risks of the procedure. Sensorineural hearing loss risk is increased up to two years post radiation.



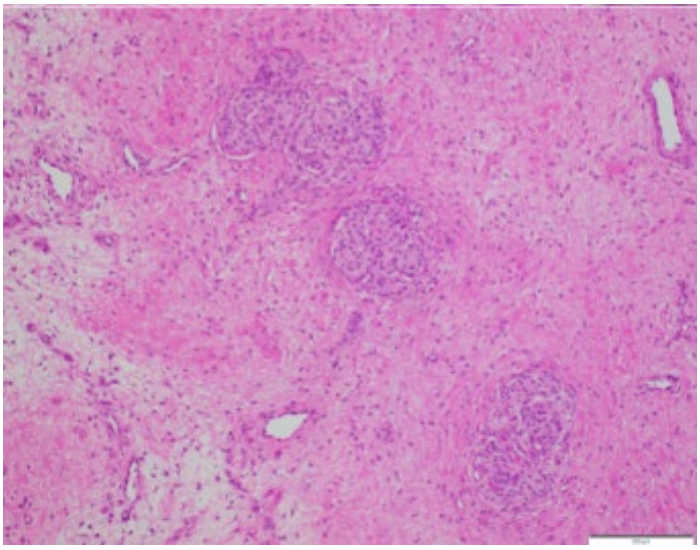


Figure 4. Hematoxylin and eosin stain show the tumors cells grow in a whorled/meningothelial architecture. At higher magnification (Upper), the tumor cells show round to oval nuclei with delicate nuclear chromatin and syncytial pattern (Lower). These features are diagnostic for meningioma.

3. Discussion

Meningiomas are thought to arise from arachnoid cap cells within the outer layer of arachnoid mater with lesions more often affecting women than men. In previously described cases of temporal bone meningioma with extracranial extension, the patient will most commonly present with conductive or mixed hearing loss due to encasement or involvement of the ossicular chain [1]. Previous patient reports have shown a vascular appearing mass, although our patient presented with a fibrotic, polypoid appearing mass with surrounding purulence of the EAC there was a highly vascular mass just medial to this. Meningiomas can further be differentiated into primary and secondary. Primary meningiomas are exceedingly rare and are thought to arise from ectopic arachnoid cells [2]. Secondary meningiomas are felt to extend through the jugular foramen, tegmen tympani, or through the IAC into the bony labyrinth, although there have been reports of direct extension through the mastoid to EAC with no middle ear involvement [3].

Further characterization of these lesions with thin slice temporal bone CT and MRI is an important component of workup. CT can show bony erosion and hyperostotic bony changes, although many times this can remain a relatively nonspecific finding and may mimic other disease processes [3]. A review of 36 temporal bone meningiomas showed that the most common finding on CT scan was "diffuse cloudiness suggestive of severe mastoiditis or otitis media" [4]. MRI will further delineate soft tissue involvement as well as a possible intracranial origin.

Ultimately, soft tissue biopsy is required for diagnosis. Treatment options include observation, surgery, or gamma knife stereotactic radiation therapy. Adjuvant radiotherapy is controversial in low grade tumors and many times be monitored postoperatively. In our patient's case, extensive surgical resection could not

be completed without an elevated risk of injury to facial nerve or other middle ear structures, and he subsequently underwent gamma knife radiotherapy. While meningioma is a commonly found tumor within the central nervous system, extracranial extension of this tumor into the temporal bone is rare. This case is an example of where extracranial extension of meningioma can present with symptoms similar to commonly seen middle ear pathology, but where recurrent meningioma from prior resection should remain within the differential.

This case presented an extracranial extension of a middle fossa meningioma into the mastoid, middle ear and presented as a large mass from the EAC which has a prevalence of only 6% - 20% of meningiomas. There additionally have not been cases reported as a mass protruding from the EAC that was found to be a meningioma. The only situation rarer is of primary ectopic meningioma. These arise independently with no evidence of intracranial disease and are very rare comprising <0.4% of all meningiomas [5]. This patient opted to proceed with stereotactic gamma knife radiation since the meningioma was widespread and adherent to vital structures and could not be resected en block. There might be some instances where and gross total resection could be offered safely as a final treatment.

4. Conclusion

This is a case of an unusual presentation of extracranial spread of meningioma presenting as a large EAC mass that was mistaken as cholesteatoma at an outside facility. This case highlights the importance of taking a good history and keeping a wide differential for ear pathology.

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

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