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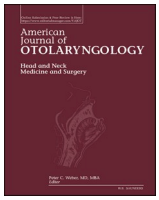

Primary external auditory canal meningioma: Case report and review of the literature

Jeffrey Singh [a,*], Uresh Patel [b], Douglas Backous [a]

[a] Puget Sound Ear, Nose and Throat, 21911 76th Ave W, Edmonds, WA, 98026, United States of America
[b] Swedish Neuroscience Institute, 747 Broadway, Seattle, WA 98122, United States of America

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ABSTRACT

Meningiomas are tumors that arise from arachnoid cells attached to both the pia mater and the inner portion of the arachnoid. They are common intracranial tumors, representing 12-25% of intracranial neoplasms. Intracranial meningiomas can spread extracranially to involve surrounding structures, including the ear and temporal bone. Ectopic meningiomas, described as primary meningiomas with no intracranial involvement, are rare. We describe a case of a primary external auditory canal meningioma with no evidence of intracranial involvement. We present pre-operative imaging findings proving no intracranial involvement prior to surgical intervention. A literature review of this uncommon clinical entity is presented and a discussion regarding its prognosis and treatment is reviewed.

1. Introduction

Meningiomas are tumors that arise from arachnoid cells lining the pia mater and the inner portion of the arachnoid. They are relatively common intracranial tumors and make up approximately 12-25% of all intracranial neoplasms [1,4,5].

It is well-known that meningiomas arising from primary intracranial locations can extend extracranially to involve surrounding structures. The route of spread may be transosseous or through neural/vascular foramina. Previous reports have estimated the prevalence of extracranial extension to be 6-20%, with common sites being the orbit, paranasal sinuses, and the temporal bone [1].

It is also known that primary ectopic meningiomas can arise independently with no evidence of intracranial disease. In general, primary ectopic meningiomas are very rare comprising <0.4% of all meningiomas [1]. Only a limited number of case series and reports have been published describing true ectopic primary temporal bone meningiomas [5,11–13].

It is our goal to review the literature and describe a case of primary ectopic temporal bone meningioma. Imaging studies are detailed, including both a CT and contrast enhanced MRI study of the temporal bone. The patient's case and clinical course is documented and reviewed.

2. Case report

A 42-year-old female presented with an eight-month history of hearing loss in the right ear. Initial examination performed at an outside institution revealed swelling in the external auditory canal (EAC) and she was treated for suspected otitis externa. She completed a three month course of oral and topical antibiotics without any improvement. Given the concern for an ear canal abscess, she was taken to the operating room for incision and drainage. Surgical pathology from this excision demonstrated reactive histiocytic changes. She was then referred to our institution for further care and management.

Upon examination, patient had a significant bulge in the posterior and superior aspect of the right EAC. The swelling was soft and compressible, but did occupy the entirety of the EAC, obscuring visualization of the tympanic membrane and middle ear.

Audiometry showed normal hearing thresholds in the left ear and a very mild conductive hearing loss in the right ear. Her speech recognition scores were 100% bilaterally (Fig. 1).

The patient underwent non-enhanced, high resolution CT scan of the temporal bones and contrast-enhanced MRI scan of the brain and temporal bones. CT scan demonstrated a soft tissue mass, with 'hair-like' dystrophic calcifications in the right EAC, extending into the middle ear cavity (Fig. 2). The EAC walls were intact without permeation or hyperostosis. The mastoid air cells and middle ear cleft were completely

* Corresponding author at: 630-336-0096, Puget Sound ENT, 21911 76th Ave W, Edmonds, WA 98026, United States of America.
E-mail address: jsingh82@midwestern.edu (J. Singh).

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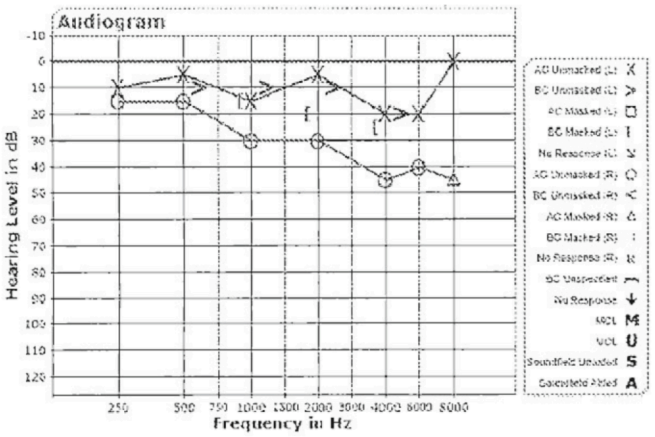

Fig. 1. Audiogram showing normal thresholds at all frequencies in the left ear. The right ear demonstrates a mild mixed hearing loss. Speech discrimination scores were 100% bilaterally.

opacified, likely from obstruction of the eustachian tube. MRI scan demonstrated an enhancing mass (Fig. 3) in the right EAC and middle ear cavity. There was no transosseous or intracranial extension (Fig. 4). The mass demonstrated restricted diffusion (Fig. 5).

After review of these images, a right sided canal-wall intact mastoidectomy and tympanoplasty was performed in effort to obtain tissue for histopathologic diagnosis. The mass in the ear canal (between the posterior ear canal skin and bony ear canal) appeared very thickened and granulomatous. The mucosa of the middle ear was similarly affected. Her ossicular chain was intact and fully mobile. Biopsies of the ear canal lesion as well as the mastoid were taken for permanent histopathological diagnosis.

Final pathology revealed a WHO Grade 1 Meningioma for both the specimens of the ear canal lesion as well as the middle ear. Mitotic activity was 0 per 10 high-powered fields. There was no hypercellularity noted and no cells demonstrating high nuclear to cytoplasmic ratios. There was no necrosis. Acid fast staining and fungal stains were negative.

After reviewing this diagnosis, the decision was made to return to the operating room in effort to excise all remaining tissue and skin of the EAC. An external auditory canalplasty was performed to remove the skin of her ear canal and perform a bony canalplasty. A post-auricular split thickness skin graft was taken to realign the ear canal.

Final pathology from this second surgery demonstrated residual meningioma. Patient has done well following these interventions with no complications and no recurrence to date.

3. Discussion

There have been numerous publications reporting extracranial spread to the temporal bone from an intracranial primary meningioma.

In 2017, Dixon et al. [2], described three cases of EAC meningiomas. All three of these cases presented with otologic complaints including hearing loss, otalgia, and swelling of the external auditory canal. Clinical evaluation and imaging led to a diagnosis of an intracranial meningioma with extracranial spread to the temporal bone in all cases.

In 2007, Marcelissen et al. [3] reported one patient with a history of chronic otitis media of five-years duration. CT showed a soft tissue density in the middle ear. After mastoidectomy and biopsy, permanent pathology yielded a diagnosis of meningioma involving the middle ear. A post-operative MRI was then performed showing evidence of an intracranial meningioma with extension into the middle ear [3].

In 2000 Prayson [4] described six cases of middle ear meningiomas. In this series, five cases were described as extending into the middle ear from an intracranial primary, where one case was thought to be arise from the EAC itself. However, details are not provided regarding this case, nor is imaging reviewed.

In 2003, Thompson et al. [5], published a large case series involving 36 patients with primary ear and temporal bone meningiomas. However, this article mentions an underlying level of uncertainty as to whether or not these represented "true" ectopic primary meningiomas of the temporal bone, as not all patients underwent complete evaluation with an MRI scan to evaluate for intracranial pathology. This highlights the importance of pre-operative MRI in this diagnosis.

Controversy exists in the literature as to the origin of primary ectopic temporal bone meningiomas, and whether or not they can arise as primary lesions without intracranial spread. It is believed that some of the older reports of temporal bone meningiomas that had claimed to be primary ectopic extracranial tumors were indeed extensions from an intracranial component that had not been identified due to lack of MRI evaluation.

Chang et al. [6], reported 21 cases of temporal bone meningiomas arising from the middle ear. However, this report was published in 1998, and it is unknown as to how many of these cases were associated with an intracranial component, as this was before the routine use of MRI [6,14]. Similarly, that same year, O'Reilly et al. [7], described 26 cases of primary extracranial temporal bone meningiomas. Again, only a minority of these cases had imaging, and therefore it is impossible to conclude whether or not these cases truly represented an ectopic primary temporal bone meningioma [7,14].

Other reports have reported on primary extracranial meningiomas not necessarily isolated to the temporal bone. In 2015, Liu et al. [8], presented 19 cases of primary extracranial meningiomas that were isolated to the head. In this review, 19 patients were identified from 1994 to 2012, with evidence of extracranial meningiomas in the head. Of these 19 patients, one patient had evidence of a middle ear meningioma. Per this data, the most common primary extracranial site was the nasal cavity and/or paranasal sinus.

In 2009, Rushing et al. [9] published a literature review with an analysis of 146 cases of primary extracranial meningiomas including all anatomical sites of the body. They identified 13 cases involving the ear and temporal bone. This review does not specify the number of cases evaluated with an MRI scan to fully exclude an intracranial component.

In 2005, Ereno et al. [10], published a case of a middle ear

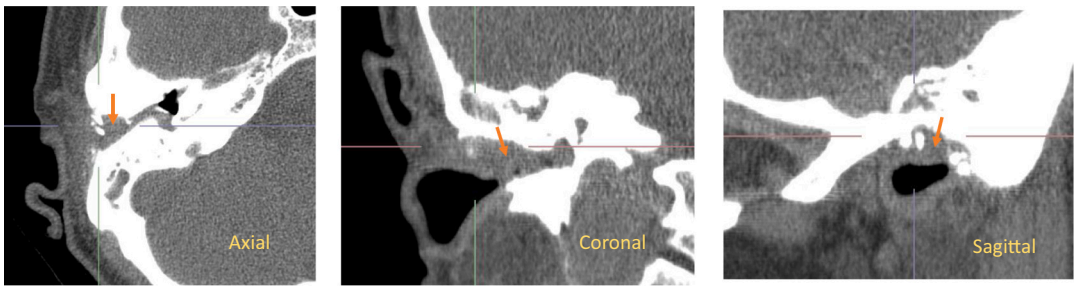

Fig. 2. CT right temporal bone. Soft tissue. Soft tissue mass in the EAC (arrows).

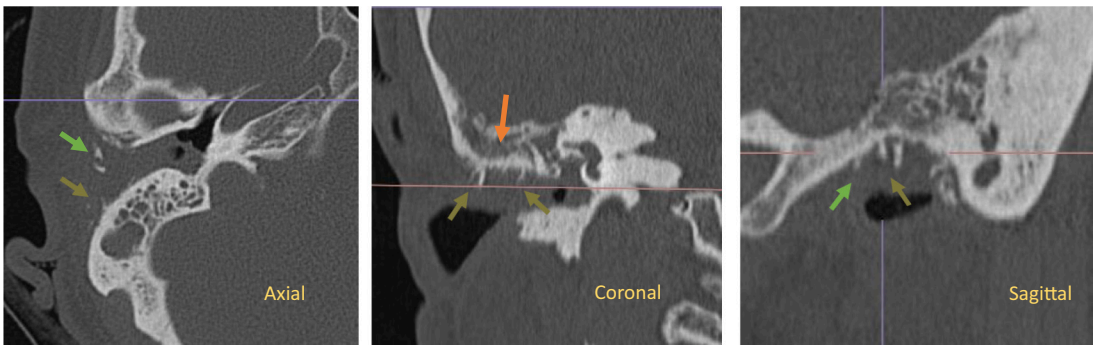

Fig. 3. CT right temporal bone. Hairy dystrophic calcification in EAC mass (green arrows). No EAC bone destruction. Retained mastoid secretions (orange arrow). (For interpretation of the references to colour in this figure legend, the reader is referred to the web version of this article.)

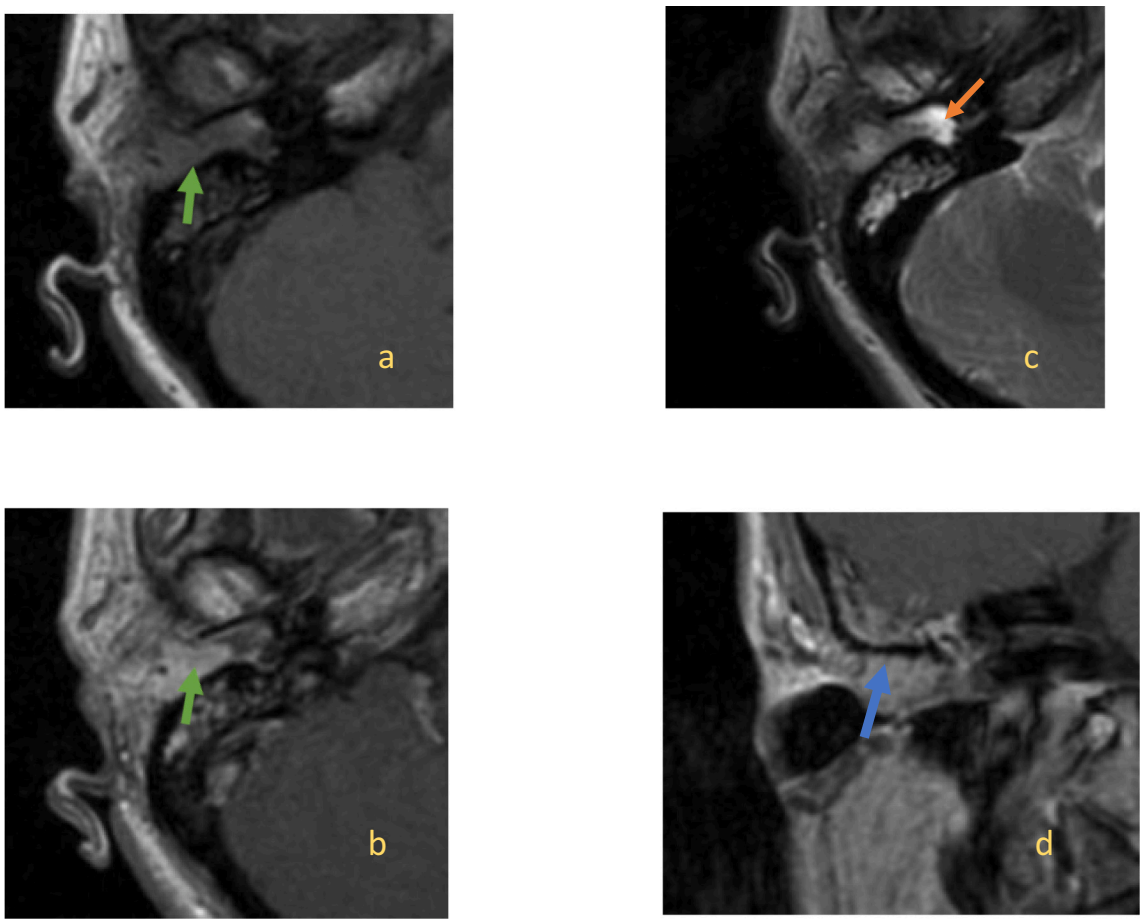

Fig. 4. MRI right temporal bone. a) Axial T1, b) axial T1 post-contrast, c) axial T2, d) coronal T1 post-contrast. Enhancing mass in EAC (a, b green arrows). Retained secretion medial EAC (c orange arrow). Intact superior wall EAC, with retained mastoid secretions above. No intracranial component (d blue arrow). (For interpretation of the references to colour in this figure legend, the reader is referred to the web version of this article.)

meningioma that was removed after obtaining a CT scan showing a mass involving the external auditory canal, middle ear, and mastoid. No MRI scan was obtained in this report to ensure no intracranial involvement.

As can be inferred, these reviews and case reports reflect the importance of both a CT scan as well as a contrast-enhanced MRI scan to completely delineate the extent of the tumor. In order to definitively classify a tumor as ectopic and extracranial, this is a necessity. There are isolated case reports in the literature which seek to identify "true" ectopic primary temporal bone meningiomas [11–13]. It is hypothesized these ectopic meningiomas arise from ectopic arachnoid cells that follow along the line of fusion of primitive nerve or bone sheaths [5,8,9,11,12,14,15].

In 2011, Manjaly [11] described a case detailing a patient who had a mass in the middle ear space that was removed via a tympanotomy with an endaural incision. Final pathology yielded a diagnosis of meningioma. A post-operative MRI showed no evidence of intracranial tumor. Similarly, in 2005, Kumar et al. [12] described a case of presumed primary ectopic middle ear meningioma. Pre-operative CT showed evidence of opacification of the external auditory canal, middle ear and mastoid, with no evidence of erosion. This patient underwent a modified radical mastoidectomy in effort to remove the disease. Post-operative MRI scan showed no evidence of any intracranial pathology. In 2003,

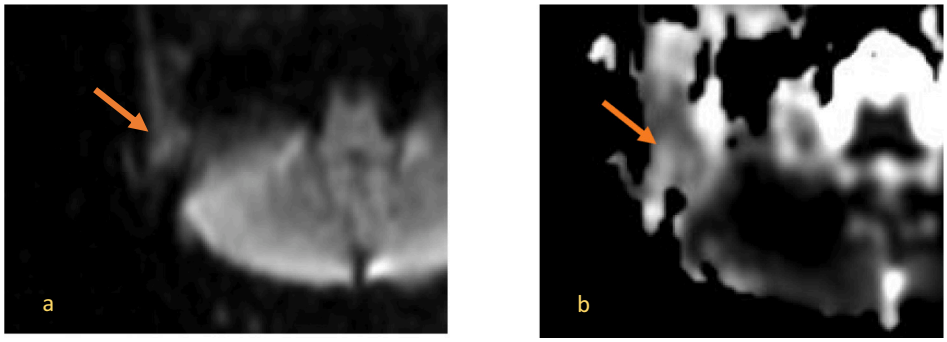

Fig. 5. DWI (a) and ADC map (b). Arrows showing restricted diffusion in EAC mass.

Uppal et al. [13] described the case of a 38 year-old with a diagnosis of primary ectopic meningioma manifested by an aural polyp. Pre-operative CT and MRI demonstrated no evidence of intracranial disease.

It is our goal to add to the literature and describe a case of a true ectopic primary temporal bone meningioma, arising specifically from the external auditory canal. Our patient was thoroughly evaluated with both a pre-operative CT scan, as well as a pre-operative contrast-enhanced MRI scan. MRI scan ruled out any intracranial component, thus defining this as a true primary temporal bone meningioma. Our case report is unique in that we display and report pre-operative imaging of both a CT and MRI scan, proving no intracranial component prior to any surgical intervention. It is also unique in that diffusion-weighted imaging was performed showing restricted diffusion in the tumor. Diffusion-weighted imaging (DWI) plays a central role in the diagnosis of acute ischemic injury in the brain [16]. But it also has a role in evaluating neoplasms. Tumors with a high level of cellularity, such as lymphoma and meningioma can demonstrate restricted diffusion as the cellularity results in reduction of the motion of water molecules in the extracellular space. This reduction in motion is detected at DWI as increased signal with reduction in signal on ADC map (Fig. 5). Restriction of diffusion is not pathagnomic for meningioma, but can be suggestive [17]. DWI should be considered in the routine use of imaging temporal bone tumors.

Surgical removal remains the goal and treatment of choice regarding primary ectopic meningiomas of the temporal bone with a good prognosis. In Thompson's et al. review of 36 patients with ear and temporal bone meningiomas, 5-year raw survival was 80% and 5-year disease-free survival was 77.1%. However, 28% of patients developed recurrence ranging from 5 months to 4.8 years following initial presentation [5]. This highlights the importance of long-term follow up in these patients, and some recommend yearly follow up with CT and/or MRI imaging [8,11,12].

Given the complexity of anatomy in the temporal bone, it becomes a challenge to ensure complete en-bloc removal of the tumor. Extent of surgical removal varies, with some case reports performing modified radical cavities to fully remove the ear canal, while others perform surgical removal of the mass only [10–14]. It may be reasonable to tailor the extent of surgical treatment pending the pathologic characteristics of the disease, with more aggressive removal (modified radical mastoidectomies) reserved for more advanced disease, but no data exists to support this claim. The utility of radiation therapy or chemotherapy has also been questioned, but no data is available in regards to its specific use in the treatment of ectopic primary temporal bone meningiomas [5,12,14].

4. Conclusion

Although rare, primary extracranial meningiomas have been described to involve the ear and temporal bone without evidence of intracranial involvement. The Otolaryngologist should be aware of this entity in the differential diagnosis of an external auditory canal or middle ear mass. We describe a case of primary temporal bone meningioma with pre-operative images ruling out intracranial involvement. Treatment of these lesions should be complete surgical removal. Because of the complexity of the temporal bone, it may be difficult to obtain clear margins. Due to the paucity of cases, the literature does not provide guidance on surgical planning and treatment. Prognosis is good for these patients, but recurrences can be high, so these demand close long-term follow up.

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