Temporal Bone Adenoma: A Comprehensive Analysis of Clinical Aspects and Surgical Outcome on a Very Rare Entity

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Abstract

Objective The aim of this study is to present our experience in dealing with middle ear adenomas (MEAs), very rare tumors of the middle ear.

Methods The medical notes of individuals with MEAs treated in tertiary referral; academic settings were retrospectively reviewed. We recorded the presenting symptoms, imaging findings, and pathology results. We additionally examined our surgical outcomes, follow-up period, recurrence, and morbidity.

Results We identified four patients with MEAs: two males and two females with an average age of 36.25 years (range = 27–51 years). Despite the detailed imaging studies, including computed tomography and magnetic resonance imaging with intravenous contrast administration, a biopsy was essential in setting the diagnosis. Total surgical resection was achieved in all patients without any recurrence over an average of 6 years (range = 3–10 years). Complete ipsilateral deafness was the commonest surgical morbidity due to footplate infiltration by the tumor.

Conclusion Total surgical resection is the treatment of choice in MEAs to minimize the risk for recurrence; this can come with morbidity, mostly sensorineural deafness. Given the very limited literature, long-term follow-up is recommended.

Keywords

- adenoma
- facial nerve
- mastoid
- middle ear tumor

Introduction

Middle ear masses are rarely of adenomatous origin. Adenomatous masses can present as neuroendocrine adenomas, endocrine adenoma, and temporal bone or middle ear adenoma (MEA). The relationship of these tumors to each other remains controversial. They may represent the same tumor in different stages of glandular and neuroendocrine differentiation.¹ Based on this perspective, these tumors are classified under Saliba and Evrard neuroendocrine adenoma of middle ear classification based on immunohistochemistry and metastasis.²

In general, MEA are rare tumors with few cases reported in the literature.^{1–8} They can present with vague symptoms, which usually lead to delayed diagnosis, even years following the initiation of their symptoms. Although their management is believed to be surgical, the outcomes of such intervention are poorly documented because of the rarity of MEA.

This scarcity of scientific reports on MEAs leads to a lack of widely accepted approaches to this condition. Our objective was to present our experience in managing MEAs focusing on their presenting symptoms, management, and long-term outcome and add to the limited literature on MEA.

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Materials and Methods

Basic Settings and Patient Selection

We performed a retrospective case series in a tertiary referral university center. The project was approved by the local audit committee.

Patients with MEAs were identified through a lateral skull base database of nearly 1,000 patients, including patients with vestibular schwannomas (most cases), cerebellopontine angle meningiomas, glomus tumors, petrous apex cholesteatomas, and additional less common conditions. The skull base dataset includes patients that have been referred to our tertiary center from other hospitals, covering a catchment area of 2.2 million.

The medical records and imaging of four patients with MEAs were retrieved from our regional skull base dataset.

Recorded Factors and Analysis

The medical notes of patients with MEAs were retrospectively reviewed. Recorded data included:

- Demographics
- Presenting symptoms
- Radiological findings, including computed tomography (CT) and magnetic resonance imaging (MRI)
- Treatment modality and outcome/morbidity
- Recurrence rates and follow-up period
- Given the number of patients, we performed a qualitative analysis of the data.

Results

We identified four patients with MEAs: two females and two males with an average age at diagnosis 36.25 years (range = 27–51 years). They all presented with hearing loss and aural pressure; additionally, patient 2 had a mild facial nerve weakness at presentation (►Table 1).

The initial assessment included otoscopy, audiometry, and temporal bone imaging with CT and MRI with intravenous contrast (►Figs. 1 and 2). The size and the extension of the tumors varied from restricted within the tympanic cavity (cases 3 and 4), extending toward the eustachian tube (case 1) or even infiltrating the lateral skull base up to the jugular foramen (maximum diameter 3.5 cm; case 2; ►Table 1).

Given the nonspecific radiological findings, a biopsy through a tympanotomy was performed in all patients (►Fig. 3), which confirmed the diagnosis of MEA.

All tumors were resected completely. Sensorineural deafness was the commonest morbidity (in two patients, cases 1 and 4), while one patient experienced maximum conductive hearing loss due to blind sac closure; this was necessary because of intraoperative cerebrospinal fluid leak, which was dealt with repair and blind sac closure (case 2). Of note patient 2, who initially presented with mild facial nerve weakness, experienced no additional weakness following the resection (House-Brackman grade 2 postoperatively).

All patients remained under clinical and radiological surveillance (combination of MRI and CT with intravenous contrast administration). All patients were seen within 6 weeks and 3 months postoperatively when baseline postoperative imaging

Table 1 Analytic data of the four patients with middle ear adenomas

N	Age	G	Symptoms	Location	Bone involvement	Treatment	Follow-up	Recurrence	Morbidity
1	32	F	Aural pressure Mild conductive hearing loss	Middle ear lesion invading the promontory and extending down the ET	Infiltration of the round window niche	Total resection	3 years	No	Deafness
2	27	M	Facial nerve weakness (House-Brackmann grade 2) Aural pressure Mild conductive hearing loss	Mass filling the tympanic cavity and infiltrating the temporal bone up to foramen jugular (not involved)	Erosion of the lateral skull base	Total resection (extended lateral) Temporalis flap	6 years	No	Facial nerve weakness (grade 2) Maximum conductive hearing loss (60dB)
3	35	M	Mild conductive hearing loss Lesion visible behind tympanic membrane	Mass attached to the tympanic membrane not filling the cavity	Ossicles invasion but no inner ear invasion	Transmastoid total resection Ossiculoplasty	10 years	No	No issues
4	51	F	Mild conductive hearing loss Aural pressure	Mass within the tympanic cavity	Invasion of the footplate	Transmastoid total resection	5 years	No	Deafness

Abbreviations: ET, eustachian tube; F, female; G, gender; M, male.

Fig. 1 Right middle ear adenoma (arrows) compromising the eustachian tube opening in the middle ear extending along the promontory as shown in computed tomography scan (A) and postcontrast T1 weighted magnetic resonance imaging (B), where it demonstrates some enhancement; the location and the enhancement is atypical for a paraganglioma, ruling out such differential diagnosis. Biopsy was necessary for setting the diagnosis.

was obtained. Following this, they were kept under annual clinical and annual/biannual imaging review. After an average follow-up of 6 years (range = 3–10 years), no recurrence has been seen.

Discussion

We present our experience in managing MEAs; these tumors present with nonspecific symptoms. Detailed imaging identifies a neoplastic mass that will raise the suspicion, but biopsy is necessary to confirm diagnosis. Surgical management creates morbidity, namely hearing loss while earlier diagnosis reduces the need for radical surgery. Although benign, longer term follow-up is recommended to monitor for recurrence.

Given the rarity of MEAs, their origin remains unknown with some hypothesizing aberrant migration of cells from salivary glands. However, a neoplastic change due to an inflammatory stimulus of the mucosal epithelium of the middle ear has become more of an acceptable explanation for these tumors.⁶ Initially they were thought to be a different entity from carcinoid tumors and neuroendocrine tumors of

the middle ear. However, the current perspective in the literature suggests that they represent the same disease in different stages. This was first suggested by Saliba and Evrard in 2009 with criteria based on immunohistochemistry and metastasis.⁷ As the cell of origin remains unknown, a maldifferentiation of the epithelial stem cells of the middle ear would be a plausible explanation to their origin. It is difficult to tell whether MEAs are always secretory tumors or not since due to their small size, one could hypothesize that they are not big enough to produce a detectable amount of hormones. In our series, we had no tumor with neuroendocrine features.

All our cases presented with conductive hearing loss. Most case reports of middle ear adenomatous masses are first thought to be cholesteatoma based on radiological findings.^{3–5} However, CT with contrast will confirm the enhancing features of the MEA on postcontrast imaging. Apart from cholesteatoma, the differential diagnosis should include glomus tympanicum and facial nerve schwannoma.⁸ Glomus tympanicum tumors are highly vascular soft tissue masses in the middle ear cavity. Facial nerve schwannomas usually arise from the geniculate ganglion but can also arise from the tympanic and mastoid segments of the facial nerve, with radiological

Fig. 2 Preoperative appearances of extensive left middle ear adenoma (arrows) as seen on MRI (A) and CT (B) scan and postoperative appearances (dotted arrows show the extend of the resection with significant bony defect) following radical resection (MRI-c and CT-D), showing no evidence of recurrence in 6-year follow-up; *shows the internal carotid artery and + the jugular bulb. CT, computed tomography; MRI, magnetic resonance imaging.

expansion of the bony facial canal and tumor enhancement following contrast.⁸ Given the nonspecific symptoms and radiological findings, a biopsy is essential to confirm diagnosis.

A systematic review reported 94 cases of middle ear adenomatous masses of which 19 were middle ear adenoma.⁷ The commonest symptoms included hearing loss, aural fullness, tinnitus, and otalgia. The CT scans were used to localize the mass, which occurs in the mastoid in 18% of the cases.⁷ Most reports of middle ear adenoma report no bone invasion or erosion; this has been clearly demonstrated in cases where the ossicles and mastoid bone are removed. However, entrapment and destruction of the ossicles is possible.^{6,7} In our cases, extension through the oval/round windows into the inner ear was identified intraoperatively in two out of four cases. This can be difficult to identify on the preoperative imaging. Literature suggests that to prevent recurrence of MEA, complete surgical excision along with ossicular chain removal is necessary. One report demonstrated that all recurrences occurred in tumors treated with ossicular preserving surgery.⁷

Chemotherapy has never been reported as a mode of treatment, while the use of radiotherapy is not recommended, as these tumors are not radiosensitive.^{1,2} In our series, complete resection was achieved in all patients without any recurrence at the time of the study. However, sensorineural deafness occurred in two cases as the result of the surgical

resection when the tumor had extended into the inner ear. This complication should be anticipated if the tumor is adjacent to either window. When complete excision is planned, patients should be counseled for possible permanent hearing loss.

Although long-term follow-up—even life-long—has been suggested, it is unclear mostly because of the small number of published cases what the ideal follow-up time should be. Given the lack of strong evidence in the literature, we would suggest a minimum 10-year follow-up, while further imaging for larger tumors or young patients can be argued. In case 3, patient with small presenting tumor and good outcome, we have not planned further review following the 10-year mark. The remaining patients will stay under review. Overall, the precise length of follow-up is open for discussion, but it should be long enough to rule out recurrence.

The present study has only four patients and was retrospective, which are the main weaknesses of the study. However, given the rarity of MEAs, this is one of the better-completed case series with MEAs in the literature with the longest follow-up.

Conclusion

MEAs are rare, benign tumors, and a diagnostic challenge not only because they present with vague and nonspecific

Fig. 3 Histology of middle ear adenoma (patient 1) ($\times 200$) showing cribriform islands of tumor (A) and positive staining for synaptophysin (B).

symptoms but also because radiological findings are non-diagnostic. Complete surgical resection, following histological confirmation, is the recommended treatment to prevent recurrence. Long-term follow-up is also recommended. Patients should be warned of the possibility of inner ear invasion and permanent hearing loss.

Conflict of Interest

None declared.

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