Middle ear adenomatous neuroendocrine tumors (MEANTs) are uncommon causes of middle ear masses that account for <2% of the primary ear neoplasms. Up to now, approximately 150 cases have been identified in the literature to date. This tumor was first described by Hyams and Michaels in 1976. Murphy et al. found that these tumors exhibit neuroendocrine differentiation. Histologically, the presence of neuroendocrine and glandular structures has led to the use of a wide variety of terminologies such as adenoma, carcinoid tumor, and neuroendocrine tumor. The patients usually have nonspecific symptoms such as unilateral hearing loss, auditory fullness, tinnitus, and otalgia. There is no specific radiological finding. A definitive diagnosis is based on complete removal of the tumor and combined histopathology and immunohistochemical examination. In this case-report, we describe a patient with MEANT who complained of hearing loss and auditory fullness in her left ear. Otomicroscopy revealed a mass of polypoid tissue filling the left external auditory canal. In the pure tone audiometry test, the pure-tone average was reported as L45/5 R10/0, and the tympanogram was type B on the left ear. In temporal bone computed tomography, the soft-tissue mass located in the middle ear was found to extend to the antrum and the external auditory canal. Biopsy taken under local anesthesia from the visible mass in the external auditory canal was reported as MEANT and the diagnosis was confirmed by histopathology and immunochemistry after surgery.

Keywords: Adenoma, Carcinoid tumor, Middle ear, Neuroendocrine tumor

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Address for correspondence: Ozan Ozdemir, MD. Saglik Bilimleri Universitesi Istanbul Egitim ve Arastirma Hastanesi, Kulak Burun Bogaz-Bas Boyun Cerrahisi Anabilim Dali, Istanbul, Türkiye

Phone: +90 530 569 43 97 E-mail: opdrozanozdemir@gmail.com

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Case Report

A 38-year-old female patient presented to our clinic with complaints of increased hearing loss and aural fullness in the left ear for several months. Otomicroscopic examination of the patient revealed that the left external ear canal was filled with polypoid tissue, so it was not possible to visualize the eardrum and the right tympanic membrane was intact and natural in appearance. The rest of the clinical examination was normal.

In the audiometry test, the pure-tone average was reported as L45/5 R10/0 and the tympanogram was type B on the left ear. In temporal bone CT, the soft-tissue mass located the middle ear was found to extend to the antrum and the external auditory canal (Fig. 1a).

The MRI showed that the soft-tissue mass extending into the ear canal was isointense in T1-weighted images, and it was close to the intensity of the gray matter signal in T2-weighted sequence (Fig. 1b). After imaging, a biopsy was taken from the visible mass in the external ear canal under local anesthesia. Histopathology and immunohistochemistry have proven it to be an MEANT. The patient consulted with the endocrinology department and an octreoscan was carried out. There was no carcinoid syndrome and no distant metastases were described.

Complete surgical excision of the mass was planned with tympanomastoidectomy under general anesthesia. During the surgery, it was found that the tumor surrounded and eroded the ossicles, but there was no bone erosion in the tegmen or scutum. It was not possible to separate the tumor from the malleus and incus. When the tumor was found to have spread to the sinus tympani, the posterior wall of the external auditory canal was removed and the canal wall down mastoidectomy was performed to reduce the risk of recurrence. There was no dehiscence on the bony canal of the facial nerve tympanic segment. The graft prepared from tragal cartilage was laid on the suprastructure of stapes with spongostan supporting it.

Post-operative immunohistochemistry confirmed the diagnosis. There were no complications during the patient’s follow-up period, and no recurrence was detected in the MRI of the 1st year postoperatively.

Discussion

The mucosa of middle ear is derived from the endoderm, and the middle ear lacks enterochromaffin cells and skin cells with neuroendocrine properties. Therefore, the pathophysiology of MEANTs is unclear. Thompson and Torske suggested that this tumor may originate from an undifferentiated stem cell.[9] This theory explains the epithelial components as well as the neuroendocrine markers, such as pancytokeratin, synaptophysin, and chromogranin-A (Fig. 2).

In Saliba and Evrard’s classification in 2009, MEANTs were divided into three types according to their immunohistochemistry findings and metastases.[10] MEANTs with positive immunohistochemical markers and negative metastases are classified as Type 1, negative immunohistochemical markers and negative metastases as Type 2, and carcinoid tumor with positive immunohistochemical markers and
metastases are described as Type 3. Our case was evaluated as Type 1 according to the classification of Saliba and Evrard.

The MEANT typically surrounds the ossicles and may extend into the mastoid antrum and auditory canal. It is often seen in the 5th decade and there is no significant difference among gender. The most common symptoms of these tumors are unilateral conductive hearing loss with auditory fullness and accompanying otalgia, tinnitus, and facial nerve palsy. The coexistence of these symptoms may mimic COM. COM is also characterized by tympanic membrane perforation, hearing loss, and inflammation of the middle ear mucosa. Torske and Thompson reported a case presented with facial nerve palsy. The tumor did not infiltrate the nerves in any of these cases. Facial nerve palsies are thought to be associated with tumor compression or anatomical abnormalities. In addition, regional or systemic symptoms may rarely appear in middle ear carcinoid tumors associated with the carcinoid syndrome. In accordance with this information, our patient had a neuroendocrine tumor mimicking COM characterized by a polyp filling the external ear canal and had no evidence of facial paralysis and no systemic symptoms.

The definitive diagnosis of MEANTs is based on combined histopathology and immunohistochemistry. CT and MRI are used in the pre-operative evaluation, but radiological images are not specific. CT images highlight homogeneous, hypodense, and well-circumscribed lesions that can extend to the mastoid bone and external auditory canal. MEANTs tend to surround the ossicles without erosion. On MRI, it is isointense on T1 images and is close to the intensity of the gray matter signal in T2-weighted sequence.

The recommended treatment for MEANT is complete surgical removal. Although tumors confined to the mesotympanum can be excised transcanally, mastoidectomy is preferred for large tumors extending into the epitympanum and mastoid cavity. As it is expected, the rate of recurrence is higher in the transcanal approach (14%) than in mastoidectomy (9%). In cases of ossicular involvement, the ossicles are removed and reconstructed, although it may be preferable to preserve the ossicular chain with repeated debulking procedures. The recurrence rate showed a better long-term outcome in cases where ossicles were removed and reconstructed. Radiotherapy, chemotherapy, or somatostatin analogs are not recommended for the treatment of MEANTs. Our patient was also successfully treated with canal wall down Type 3 tympanomastoidectomy and there was no evidence of recurrence or metastasis at 1-year follow-up.

Conclusion

MEANTs are rare middle ear masses with benign morphological features and slow local growth. They usually do not cause systemic symptoms and mimic chronic otitis. The etiology of these tumors is unclear, their classification is not well defined, and the treatment is complete tumoral excision. Patients need to be monitored lifelong using CT/MRI scans for early detection of recurrence.

Disclosures

Informed consent: Written informed consent was obtained from the patient for the publication of the case report and the accompanying images.

Peer-review: Externally peer-reviewed.

Conflict of Interest: None declared.


References