Middle Ear Carcinoid Tumor With External Auditory Canal Extension: A Case Report

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Significance Statement
The middle ear cavity is an uncommon location for neuroendocrine tumors. They commonly appear as retrotympanic masses, with intact tympanic membrane, and rarely mimic acquired cholesteatoma, as in our case. The clinical and radiological findings are not specific; therefore, the final diagnosis is often delayed. We present a rare case of a middle ear carcinoid tumor with external auditory canal extension that was treated successfully with radical mastoidectomy with no evidence of recurrence or metastasis at 16 months of follow-up.

Case Description
A 44-year-old man was referred to our ear, nose, and throat (ENT) department with a 2-year history of recurrent right ear otorrhea, accompanied by fullness, tinnitus, and progressive hearing loss. For the past 2 years, he had been treated with antibiotics, as chronic otitis media was the initial diagnosis, with only temporary relief.

Otomicroscopy revealed a vascular mass occupying the right external auditory canal (EAC) with extension to the middle ear via perforation of the tympanic membrane (pars flaccida). His left ear was normal. No swelling or tenderness was noted in the mastoid region. His facial nerve function was normal. Both the Weber and Rinne tests indicated conductive hearing loss on the affected side. A full ENT examination did not reveal any pathological findings. Audiometry confirmed conductive hearing loss at low and high frequencies with sensorineural hearing loss of high frequencies on the right side. Computed tomography (CT) of the temporal bones revealed a mass with soft tissue density in the right middle ear with extension to the EAC. The ossicular chain was intact, the mastoid process was normal, and there was no evidence of bone destruction (Figure 1).

We performed a biopsy under local anesthesia to confirm the diagnosis. Both the tegmen tympani and EAC were found to be intact on CT; therefore, a biopsy was a safe approach (encephalocele and meningocele were excluded from the differential diagnosis). The biopsy showed findings (morphology and immunochemistry) relevant to a well-differentiated neuroendocrine neoplasm (grade 1, carcinoid tumor) (Figure 2). Chest and abdominal CT and magnetic resonance imaging were performed, the findings of which excluded another tumor location. Ten days later, radical mastoidectomy was performed. A polypoid, vascular, gray tumor was identified filling the middle ear cavity (mesotympanum and epitympanum) and extending to the EAC. The handle and head of the malleus and body and short process of the incus were surrounded by the tumor, with no ossicle erosion. There was minor expansion to the antrum and no expansion to the mastoid or Eustachian tube. There was no evidence of facial nerve involvement or bone erosion. The tympanic cavity was cleared of the mass, and the malleus and incus, tympanic membrane, and part of the skin of the EAC were excised with the aim of securing negative margins of resection. Cartilage-perichondrium composite graft was harvested and placed over the intact stapes. The facial nerve was not injured, and hearing remained at the same level. The final pathological examination confirmed the negative margins of resection and neuroendocrine nature of the mass: well-differentiated neuroendocrine neoplasm (grade 1, carcinoid

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Additional radiotherapy or chemotherapy was not performed. Follow-up after 16 months with otomicroscopy and new CT did not reveal any recurrence or metastasis of the tumor.

**Discussion**

The most common locations of neuroendocrine tumors are the lungs, pancreas, and gastrointestinal tract, and very rarely, the middle ear cavity.\(^1\)\(^,\)\(^2\) The main symptoms are otorrhea, fullness, conductive hearing loss, and tinnitus\(^3\)\(^,\)\(^4\) while facial nerve palsy is a rare manifestation\(^5\)\(^,\)\(^5\).

Middle ear carcinoids are mostly retro tympanic masses with intact tympanic membrane. Extension from the middle ear cavity to neighboring areas such as the EAC, Eustachian tube, and mastoid is uncommon\(^3\)\(^,\)\(^5\). Differential diagnosis includes a spectrum of multiple pathologies, such as cholesteatoma (acquired or congenital), glomus tumor, chronic otitis media, mucosal adenoma, ceruminous adenoma, adenocystic carcinoma, pleomorphic adenoma, schwannoma of the facial nerve, and meningioma\(^6\)\(^,\)\(^7\).

The treatment of choice for middle ear neuroendocrine tumors is surgical. Transcanal is a safe and minimally invasive approach for small tumors located in the mesotympanum, with no invasion of the adjacent tissues. If the tumor is embedded in the osseous chain, the ossicles should be removed. Tumors extending to the EAC or mastoid necessitate tympanoplasty and/or mastoidectomy. Sometimes, canal wall down mastoidectomy is unavoidable to prevent the possibility of residual tumor.\(^8\) Cervical node or distant metastasis has also been reported in advanced-stage tumors. In cases of regional metastasis, parotidectomy and/or neck dissection should be performed.\(^6\)

Chemotherapy has not been reported in the literature thus far. The efficacy of adjuvant treatment with radiotherapy has...
not been established, although it is reserved for metastatic cases. Additionally, radiotherapy poses a risk of malignant transformation of tumors.\textsuperscript{2}

Recurrence rates range from 18\% to 22\% and are influenced by several factors.\textsuperscript{2,5} Patients who undergo radical mastoidectomy exhibit lower rates of recurrence than those exhibited by patients who undergo a less extensive surgical approach.\textsuperscript{2} Total removal with negative margins of resection is strongly recommended. Disease-free intervals range from 53 to 158 months, as reported in the literature.\textsuperscript{6}

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