The First Reported Case of Recurrent Carcinoid Tumor of the External Auditory Canal

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Abstract

Objectives: Neuroendocrine tumors (NETs) of the ear canal are exceedingly rare, typically called Merkel cell carcinoma or well-differentiated and carcinoid when indolent. When arising in the temporal bone, NETs usually originate in the middle ear/mastoid. We present the first case of a recurrent carcinoid tumor originating in the external auditory canal (EAC).

Study Design: Case report.

Methods: Examination of medical records; literature review.

Results: A 38-year-old woman presented to an outside otolaryngologist in 2007 with a pedunculated mass originating from the superior aspect of the medial bony ear canal. Physical exam and CT scan found no bone erosion, and the patient underwent uneventful soft tissue excision. Histology demonstrated a well-differentiated NET positive for chromogranin – eventually diagnosed as carcinoid. In 2015, she returned with a recurrent mass now involving the anterior, superior, and posterior aspects of the medial ear canal. The tympanic membrane was invaded, and tumor had entered the middle ear. CT confirmed physical findings, and an MRI (brain, face, neck, abdomen, and pelvis) found a possible nasopharyngeal (NP) lesion and an unrelated lung nodule. Incisional biopsy of the ear lesion revealed an infiltrative NET with a low Ki-67 index, again diagnosed as carcinoid. NP biopsy was negative. The patient consented to lateral temporal bone resection; no adjuvant therapy is planned.

Conclusions: We describe the first case of a recurrent low-grade cutaneous NET (carcinoid) of the EAC. Despite its low proliferative index, gross total resection of tumor and surrounding bone is recommended to prevent recurrence.

Introduction

NETs of the skin were first described in 1972 by Toker, who reported that detailed trabecular growths arising from the dermis layer of the skin had the potential to metastasize through lymphatic pathways.1 These lesions are now considered to be NETs when well-differentiated (carcinoid, atypical carcinoid) or neuroendocrine carcinoma when poorly-differentiated (e.g., small cell NET).2 Merkel cell carcinoma (MCC).3 Well-differentiated neoplasms tend to have a more indolent course, while poorly differentiated neoplasms are more aggressive. Head and neck NETs are extremely rare and especially uncommon within the ear. While several cases of neuroendocrine lesions have been reported in the middle ear, there are less than 30 reported cases that have been isolated to the external ear, with only five cases originating in the EAC.2,4

Case Report

The patient is a 38-year-old female who presented to a community-based otolaryngologist in 2007 with otalgia, aural fullness and decreased hearing in the left ear. Physical examination revealed a palpating mass in the bony ear canal. Temporal bone CT imaging revealed no bone erosion or extension of the mass beyond the ear canal (Figure 1A). A wide local resection was performed, and histologic examination revealed a well-differentiated NET with positive margins. The patient then presented to us in 2015 with identical symptoms from 2007. Physical exam found a recurrent mass in the left ear canal that was partially obstructing the view of the tympanic membrane. A temporal bone CT revealed a lesion measuring approximately 6 mm (Figure 1B). A small component of the mass was projecting anterior to the head of the malleus. MRI noted that the mass extended into the middle ear and was present along the manubrium of the malleus. Further imaging revealed the presence of an NP lesion and lung nodule. Biopsy of the NP lesion revealed normal adenosin tissue and the lung nodule was felt to be benign based on imaging characteristics. Pathology sections of the EAC mass confirmed the presence of cutaneous NET (Figure 2). The patient opted for her most definitive surgical treatment option – a lateral temporal bone resection (Figure 3). Surgical pathology confirmed the presence of a low-grade NET. This patient will be followed with annual MRI and PET scans; she is considering hearing rehabilitation with a bone anchored hearing device if she remains disease free for 2-3 years.

Discussion

There have been approximately 50 reported cases of carcinoid NETs found within the middle ear, however, we could only find five cases of neuroendocrine lesions located in the EAC.2 While there have never been any reports of distant metastasis from carcinoid NETs of the ear, these lesions do have the potential to recur.5 The tumor recurred in this patient, because the extent of initial resection was not adequate. The presentation of EAC lesions can vary, but in the case of our patient, she had the sensation of aural fullness, subjective hearing loss, and otalgia. Unlike carcinoid NETs within the GI or bronchial systems that are known to produce symptoms of flushing and diarrhea, cutaneous carcinoid NETs are considered functionally silent.6 Surgical removal of these lesions is currently the standard of care. Adjunct radiation and chemotherapy can be considered for high grade lesions or when surgical resection is incomplete.

Conclusion

This is the first report of a recurrent carcinoid NET of the EAC, recurring 8 years after primary resection. While the recurrence rate of middle ear carcinoid NETs has been estimated to be 22%, there is little known about the potential for recurrence or distant metastasis amongst lesions originating in the EAC. Due to the low proliferation index and the maximal nature of the surgical resection, observation (serial imaging) without chemo or radiation was recommended in this patient.

References


Figure 1: A) Axial CT image from 2007 demonstrates a small soft tissue lesion (arrow) at the anterior aspect of the junction of the osseous and cartilaginous portions of the left EAC. B) Coronal CT images from 2015 show a new mass (arrow), in the medial aspect of the EAC with extension into Prussak’s space.

Figure 2: Immunohistochemistry demonstrating positive A) CD56 (x600), B) Synaptophysin (x600), and C) MAK-6 pancytokeratin (x600) antibody staining. D) Ki67 proliferative index is 3% (x400).

Figure 3: Clinical image taken during lateral temporal bone resection. Tumor arising from the anterior and superior bony ear canal walls (box) with extension into the epitympanum and involvement of the ossicular chain (arrow).

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