The Journal of International Advanced Otology

J Int Adv Otol 2021; 17(6): 577–580 • DOI: 10.5152/iao.2021.21230

Case Report

A Case of Carcinoid Tumors in the Ear Canal With Long-Term Postoperative Follow-Up

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Cite this article as: Tanahashi S, Oonishi M, Mori K, Umeda M, Wakaoka T, Takahashi H. A case of carcinoid tumors in the ear canal with long-term postoperative follow-up. *J Int Adv Otol.* 2021;17(6):577-580.

Carcinoid tumors in the ear canal are very rare. In this report, we experienced a case of carcinoid tumor of the ear canal that underwent total tumor resection. This study included a 39-year-old man presented with a chief complaint of right-sided hearing loss. Computed tomography scan showed a shadow from the ear canal to the right tympanic chamber. There were no suspicious findings of metastasis in the cervical lymph nodes or other organs. At the time of surgery, the tumor was simply removed because it was small and there was no adhesive invasion. Postoperatively, the patient has been under observation for 11.5 years without any recurrence. Carcinoid tumors in the ear canal can rarely metastasize or recur after more than 10 years. It is important to follow up with the patient for a long time after surgery, using the Ki-67 index of the removed tissue as a prognostic reference.

KEYWORDS: Carcinoid tumor, ear canal, neuroendocrine tumor

INTRODUCTION

Carcinoid tumors are very rare in the ear canal, and only 7 cases have been reported.[1] There have been cases of recurrence 8 years after the initial surgery,[2] so long-term follow-up is necessary.

We experienced a case of carcinoid tumor in the ear canal, performed relatively long-term follow-up after tumor resection, and reported the case with literature review.

CASE PRESENTATION

A 39-year-old male was referred to our department with temporary hearing loss and swelling of the right ear. A swelling was found in the right ear canal just before the tympanic membrane (Figure 1). A biopsy of that area revealed a pathological diagnosis of adenoma or low-grade adenocarcinoma, and Computed tomograpghy (CT) scan showed shadows from the ear canal to the right tympanic cavity, but no bony destruction (Figure 2). Positron emission tomography-CT showed no signs of metastasis in the cervical lymph nodes or other organs. There was no complaint of hearing loss at the time of the patient's visit to our department, and pure tone audiometry showed no significant hearing abnormality. Surgical treatment was chosen, and tumor removal was performed in accordance with right tympanoplasty type I plus mastoidectomy. The operation was started through a posterior incision of the right ear under general anesthesia. White tumors were found in the bone part of the ear canal to the tympanic ventricle in the 2, 9, and 12 o'clock directions. The size of the tumor was less than 1 cm, and the area around the ear ossicles was granulation. The mucosa surrounding the tympanic membrane and the skin of the ear canal where the tumor was located was removed. Postoperative pathology showed polygonal and round epithelial-like cells with unevenly distributed nuclei and eosinophilic sporangia proliferated in an enhanced manner, a tendency to form solid nests and areas of irregularly fused glandular ducts (Figure 3). Immunostaining showed chromogranin A positivity and CD56 positivity, and the Ki-67 index was 3%-4% (Figure 4), so the diagnosis of carcinoid tumor in the ear canal was made. Endoscopic examination of the right ear canal at 8 months postoperatively showed no recurrence (Figure 5).

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Received: February 7, 2021 • **Accepted:** March 13, 2021
Available online at www.advancedotology.org

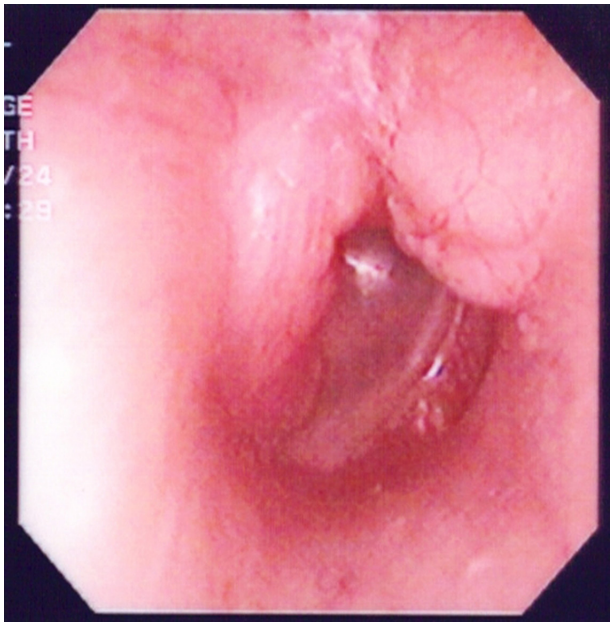

Figure 1. Right external auditory canal findings on initial examination. Swelling was found in the right external auditory canal just before the tympanic membrane.

Since then, the patient has been undergoing follow-up with regular outpatient observation of the ear canal and annual CT scans. There was no recurrence at 11.5 years after the surgery, and the patient is still under follow-up.

DISCUSSION

Carcinoid tumors are considered neuroendocrine tumors because they are derived from neuroendocrine cells[3] and usually occur predominantly in the lungs, trachea, and gastrointestinal tract. Although occurrence in the ear region is rare, there are relatively many reports of carcinoid tumors occurring in the middle ear. Among carcinoid tumors, the incidence in the middle ear is less than 0.7%.[4] And although the incidence is unknown, cases occurring in the ear canal are even rarer.[1]

Neuroendocrine tumors are mainly derived from primary endocrine cells scattered in tissues related to the archenteron, such as the gastrointestinal tract, trachea, and bronchi. They are classified according to their site of origin as tumors of foregut, midgut, or hindgut origin.[5] The mucosa of the tympanic ventricle is derived from the foregut, and carcinoid tumors in the middle ear are thought to be caused by some kind of pluripotent endoderm-derived stem cell that resides mainly in the mucosa of the tympanic ventricle, and that carcinoid tumors develop by a mechanism similar to that of the lung.[6] However, in the normal skin of the ear canal, neuroendocrine cells do not seem to exist except for Merkel cells localized in the basal layer, and the origin of carcinoid tumors when they occur in the ear canal is reported to be poorly understood.[1] It is possible that the mucosa of the middle ear cavity may have strayed into the ear canal during the developmental stage, but the pathogenesis of the tumor, in this case, is unknown, and future studies are expected.

In this case, we performed pathological diagnosis of the removed tumor by immunostaining and found that it had the characteristics

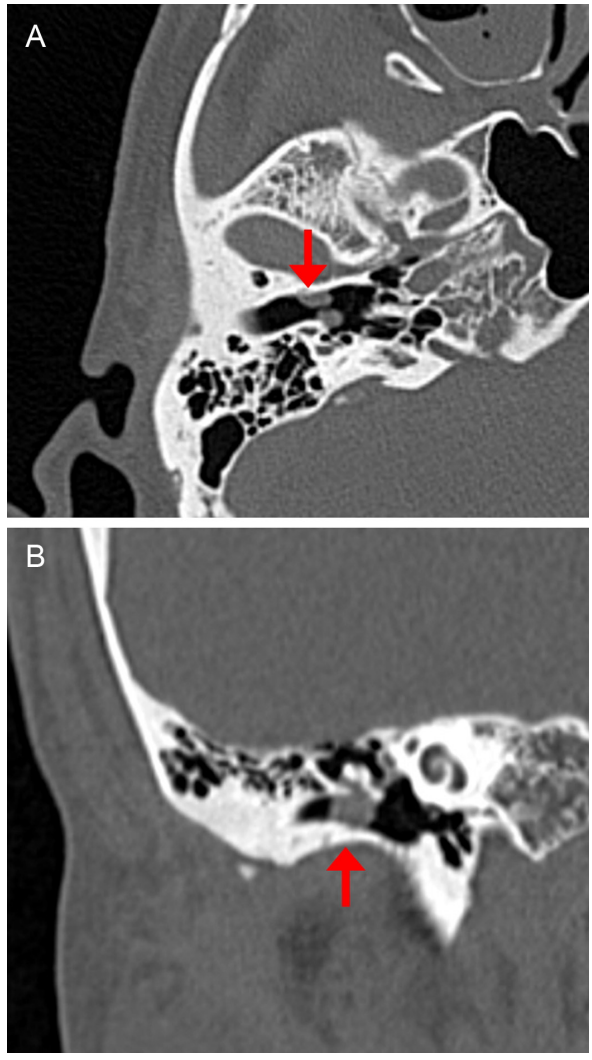

Figure 2. Computed tomography horizontal (2-A) and coronal (2-B) views of the right ear, showing an equidense mass in the right external auditory canal without bony destruction (arrow).

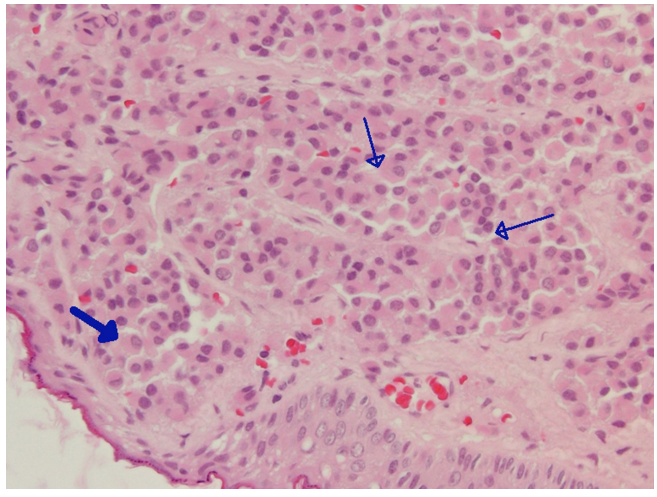

Figure 3. Postoperative pathology. Polygonal and round epithelial-like cells with unevenly distributed nuclei and eosinophilic sporangia proliferated in an enhanced manner (thick arrow). There was a tendency to form solid nests and areas of irregularly fused gland ducts (thin arrows). The nucleus had components of unequal size and irregular nuclear rim.

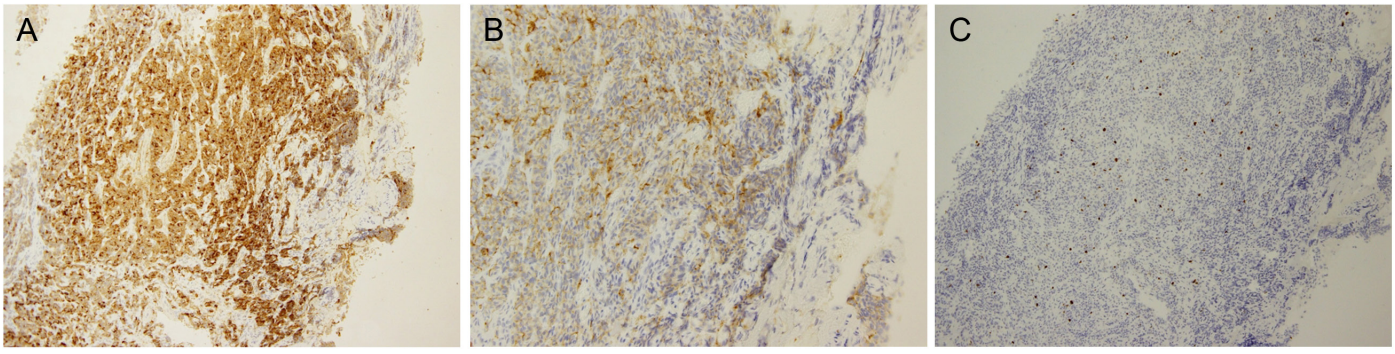

Figure 4. Immunostaining findings. (3-A) Chromogranin A positive, (3-B) CD56 positive, and (3-C) Ki-67 expression level was 3%-4%.

of a neuroendocrine tumor, such as chromogranin A positivity and CD56 positivity, which led to the diagnosis of carcinoid tumor of the ear canal. The Ki-67 index was 3%-4% in this case. In lung carcinoid tumors, it has been reported that cases with low Ki-67 index have a high survival rate.[7] According to this report, the Ki-67 index of normal lung carcinoid tumors is less than 10%, and cases with a high Ki-67 index have a poor prognosis. Although the prognostic cutoff value for carcinoid tumors of the ear canal is not clear, the Ki-67 index was less than 10% in the present case, which suggests a good prognosis.

The best treatment is the surgical removal of the entire tumor. If the tumor is small (<2 cm) without metastasis or bone destruction, a local tumor resection can be performed as for any other organ.[8] If there is bone destruction, extended surgery such as middle ear radical surgery or temporal osteotomy is necessary as in the case of cancer of the ear canal and middle ear. And if there are cervical lymph node metastases, additional neck dissection is necessary. In this case, the tumor was small in size and its margins were clearly separated from the surrounding tissues, so a simple tumor resection could be completed. The efficacy of external beam radiation for carcinoid tumors in the middle ear is unknown, and it is not considered necessary to perform it aggressively.[9,10] Therefore, we did not perform external beam radiation in this case as well. Among radiotherapies, peptide receptor radionucleotide therapy (PRRT) is an internal radiation therapy targeting somatostatin receptors, which are highly expressed in neuroendocrine tumors and is approved and used as a new treatment for neuroendocrine tumors.[11] At this time, the efficacy of PRRT for neuroendocrine tumors in the ear region is unknown, and further study is expected.

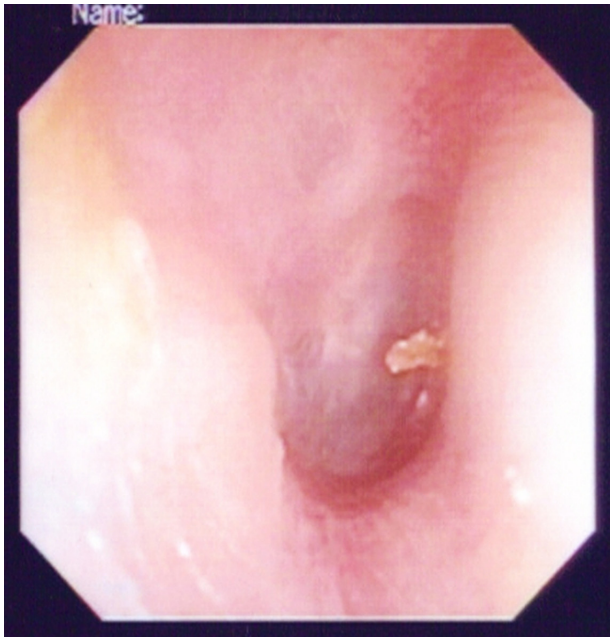

Figure 5. Right external auditory canal findings at 8 months postoperatively.

Postoperative recurrence rates in the middle ear have been reported to range from about 18% to a long-term local recurrence rate of 22%.[9,12] Because of their slow growth, carcinoid tumors in the middle ear develop slowly, and the time from initial surgery to recurrence is long, ranging from 15 to 33 years, with an average of 11 years.[12] Therefore, compared with ordinary malignant tumors, it may take a much longer time before they can be judged as cured. In this case, the lesion was mainly in the ear canal, and no recurrence was observed after 11.5 years of outpatient follow-up with simple tumor resection alone. It is possible that carcinoid tumors in the ear canal follow the same gradual development process as those of the middle ear. We plan to continue long-term follow-up of the patient.

CONCLUSION

In conclusion, carcinoid tumors in the ear canal are a disease with a good prognosis if surgical removal is possible. However, there are rare cases with high malignant potential, such as metastasis. Therefore, it is recommended to treat it as a malignant tumor although the malignancy is low. In addition, it is important to follow up with the patient for a long time after surgery, using the Ki-67 index of the removed tissue as a reference for prognosis.

Informed Consent: Informed Consent: Informed consent was obtained in writing from the patient who participated in this study.

Peer-review: Externally peer-reviewed.

Author Contributions: Concept - M.O.; Design - S.T.; Supervision - M.O.; Resource - S.T., K.M., T.W.; Materials - S.T., M.O.; Data Collection and/or Processing - S.T., K.M., T.W., H.T.; Analysis and/or Interpretation - S.T., M.O., K.M.; Literature Search - S.T.; Writing - S.T.; CriticalReviews - M.O., M.U.

Acknowledgments: We would like to acknowledge the assistance of Dr. Tomoya Hori of Ogaki Municipal Hospital.

Conflict of Interest: The authors have no conflict of interest to declare.

Financial Disclosure: The authors declared that this study has received no financial support.

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