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Carcinoid Tumor of Middle Ear: A Case Report

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Introduction

Primary carcinoid is an unusual neoplasm of middle ear and it is rarely discovered clinically. Carcinoid tumor presents with nonspecific clinical symptoms, and often it has been misdiagnosed. Here we reported a case of an elderly female presented with right ear polyp and was diagnosed as carcinoid tumor of the right middle ear. The tumor cells were positive for chromogranin, synaptophysin, S 100, CD56 and NSE with low Ki-67 proliferative activity (< 1%).

Keywords: Middle ear, carcinoid, ear polyp, immunohistochemistry, case report

Introduction

Carcinoid tumor, also known as neuroendocrine tumor, is a very rare neoplasm in the middle ear. Middle Ear carcinoid tumor was first reported in 1980 by Murphy.[1] Patients usually presents with nonspecific symptoms of conductive hearing loss, aural fullness, tinnitus, otalgia, and headache. The common differential of middle ear carcinoid tumor on histopathology is middle ear adenoma by histopathology because they are benign gland-forming neoplasms with an overt tendency to develop neuroendocrine and mucinous differentiations. The definitive diagnosis is made by immunohistochemistry of biopsy materials. [2,3] They have an indolent course and are usually confined to the tympanum and lacks the capacity for metastasizing. However, several recent reports suggest that the carcinoid should be classified as a low-grade malignancy supported by the evidence of local recurrence, regional cervical lymphatic metastasis and distant metastasis.[4] Here we present a rare case of carcinoid tumor of middle ear.

Case Report

A 72 years old female presented with decreased hearing more on the right side. There was no complaint of pain, bleeding, dizziness and facial palsy. Otoscopic examination revealed a mass in the middle ear. Pure tone audiometry revealed bilateral mixed hearing loss with severe to profound hearing loss on right ear and mild to moderate hearing loss on left ear. No swelling or tenderness in the mastoid region. No cervical lymph nodes were palpated. A computer tomography scan showed an isodense mass without osteolytic invasion in external auditory meatus and middle ear. Provisional diagnosis of ear polyp was made. The mass was excised and was received in the department of pathology. The histopathological findings showed a polypoid tissue covered with stratified squamous epithelium and tumor

cells nest in the subepithelial area. The tumor cells had round and oval with finely-dispersed chromatin, arranged in cords and nests without necrosis and mitotic activity.(Figure 1A) The immunohistochemical staining showed positivity for chromogranin, synaptophysin, S 100, CD56 and NSE with low Ki-67 proliferative activity (< 1%).(Figure 1 B,C,D) A diagnosis of primary middle ear carcinoid tumor was given.

Discussion

The primary neoplasm in the middle ear are rare, out of which carcinoid tumors are the most uncommon tumors.[5] The age of presentation is 16-64 years with male preponderance. It presents with non specific clinical and imaging findings that is why they are easily misdiagnosed. [1] More than 90% of the patients complain of hearing loss, and 20%-30% of patients suffer from ear fullness, tinnitus, ear discharge. Some patients develop facial paralysis.[4] Carcinoid tumors are currently categorized as well-differentiated neuroendocrine carcinomas that belong to a group of neuroectodermal neoplasms with epithelial differentiation. A typical carcinoid is considered as Grade I, according to the latest WHO classification of head and neck tumors.[4] The diagnosis of middle ear carcinoid mainly depends on the pathological diagnosis, and it has the histopathological features of both adenoid and neuroendocrine. Immunohistochemistry can improve the detection rate of neuroendocrine granules and the accuracy of diagnosis.[1] In fact, middle ear carcinoids may be under reported as it is challenging to distinguish it from other middle ear adenomatous tumors by histopathology. Based on histopathology alone, it is difficult to distinguish between middle ear carcinoid tumor and other middle ear adenomatous tumors such as adenoma.[3] Middle ear carcinoid need to be identified with adenomas, paragangliomas, adenocarcinoma, adenoid cystic carcinoma and metastatic carcinoma of the middle ear.[1] With routine light microscopy, both are composed of small round to oval uniform cells with finely stippled chromatin arranged in glandular, trabecular, and solid patterns without pleomorphism, necrosis, and mitosis. However, middle ear carcinoid tumor cells are immunohistochemically reactive to antibodies to chromogranin A, neuron-specific enolase, keratin, and synaptophysin, while middle ear adenoma show negativity for the neuroendocrine markers. Furthermore, the electron microscopy would demonstrate numerous neurosecretory granules in the carcinoid tumor cells.[3] They are negative for S-100 which has been used to differentiate carcinoid tumors from paraganglioma. Ki-67 proliferative index of these tumors are generally less than 10%, whereas most small-cell carcinomas show a value that is substantially higher than 25%. Several investigators believe that Ki-67 serves as a significant predictor for local recurrence, osteolytic enlargement and metastasis. A low Ki-67 index is associated with longer survival in carcinoid tumors. Primary middle-ear carcinoids generally develop slowly, and its local invasion is usually nondestructive.[4] Middle ear carcinoid appears to follows a less aggressive course compared to carcinoid tumors elsewhere in the body, yet studies have shown cases where regional lymph node metastasis indicate.[3] Middle ear carcinoid is mainly treated by surgery. If the tumor is confined to primary site with no distant metastasis, radiotherapy and chemotherapy is not recommended. On the contrary, if there is distant metastasis, radiotherapy and chemotherapy is considered. The recurrence rate of middle ear carcinoid is as high as 25%. Therefore, it is necessary to evaluate the cervical and parotid lymph nodes carefully. If there is a metastasis, lymph node dissection should be performed. [1]

Conclusion

The incidence of middle ear carcinoid is low, with non specific clinical and radiological features. The diagnosis is mainly based on histopathology. Definitive diagnosis can be made using histopathology. Due to its rare occurrence, reporting the clinical presentation and features of each new case would help to delineate the characteristics and therefore, increasing the awareness towards the disease.

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Conflict of Interest: We declare that there are no conflicts of interest amongst the authors.

Ethical Approval : Not Applicable As It Is A Case Report

Legends Figure

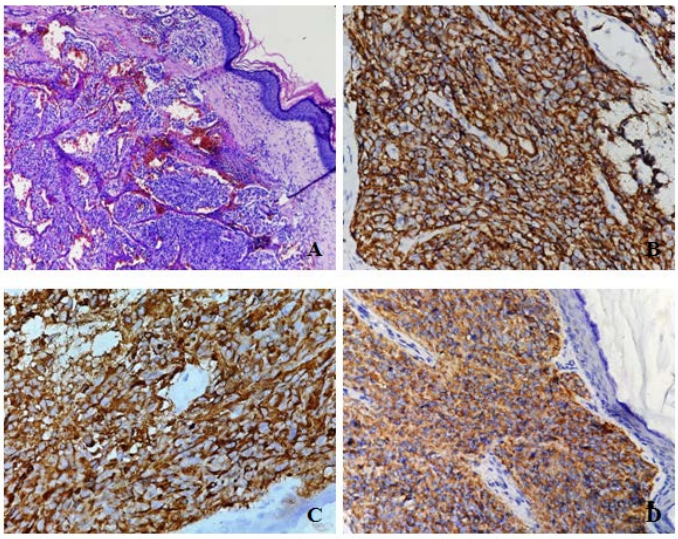

Figure 1 : A : Carcinoid tumor of middle ear (H & E, 100X); **B** CD 56 positivity (IHC, 400X)**, C :** Chromogranin positivity (IHC, 400X)**; D** Synaptophysin positivity (IHC, 100X)

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