Carcinoid Tumor of the Middle Ear: A Case Report

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Abstract

Primary carcinoid of middle ear is a kind of neuroendocrine tumor, and it is rarely discovered clinically. Carcinoid does not have typical clinical symptoms, and it has been misdiagnosed frequently. It is also nonspecific in clinical and radiologic aspect. Its diagnosis depends on pathologic result. This article will report a case which takes “discovering external auditory canal tumor” as chief complaint, and diagnosed as “primary carcinoid of middle ear” eventually, and discuss its clinical and pathologic features treatment and prognosis.

Keywords: Middle ear neuroendocrine tumor carcinoïd pathology

Introduction

Carcinoid tumor, also known as neuroendocrine tumor, is a group of heterogeneous tumors that originated in peptide neurons and neuroendocrine cells. It growth in many organs and tissues, most common in the gastrointestinal tract and pancreas, and rarely occurs in the liver, gallbladder, bronchial, lung, adrenal medulla, paranganglia, thyroid, parathyroid, etc., especially rare in sinuses, throat, eye socket and the middle ear. Middle ear primary neuroendocrine tumors are particularly rare. The first middle ear neuroendocrine tumor was discovered and reported by Murphy in 1980 [1], only 56 cases being reported by the end of 2004 overseas [2], followed by only hundreds of cases reported domestic and overseas [3]. The first case of middle ear carcinoid tumor in China was first reported by Hong Zhu in 2001 [4].

Case Presentation

Male, 27 years old, presented with discovery of "left external auditory canal mass, accompanied with 8 months of hearing loss". This patient suffered from left external auditory canal slowly bleeding with no obvious incentives, as well as hearing loss from 8 months ago, without tinnitus or aural fullness, founding that the left external auditory canal neoplasm growing quickly; fully fill in the external auditory canal, which has no swelling or tenderness. Being treated by methylprednisolone in local hospital, the bleeding symptom was relieved, but the volume of the tumor did not change significantly. The patient had a history of fatty liver disease for 3 years and had no any other diseases. Physical examination: the left external auditory canal is blocked by a mass, which has a rough epithelioid tissue-like surface, and it’s flexible and full of external auditory canal (Figure 1), without abnormal secretions. The right tympanic membrane was integrated and had a clear signs; No swelling or tenderness in the bilateral mastoid region. Facial: bilateral forehead is symmetrical, bilateral eyelid can be closed, the nasolabial groove is normal, cheek blowing does not leak, tooth is in the center, No enlarged lymph nodes can be touched in the neck. Auxiliary examination: pure tone test: right ear is normal, left ear BC: 20-25-20-70dBHL, AC: 65-75-70-100 dBHL. WT test: 256Hz inclined left. Temporal MRI: left tympanic cavity and external auditory canal’s occupying lesion, malignant tumor is not excluded; left middle ear mastoitis (Figure 2). Admitting diagnosis: left external auditory canal and middle ear mass; left middle ear mastoitis. We performed an open radical mastoidectomy, and delivered the external auditory canal neoplasm of the left ear to the pathology department as frozen. The pathological result was “polypoid tissue covered with stratified squamous epithelium, tumor cells nest can be seen infiltrated the epithelium”. The antrum and tympanum were filled with flexible neoplasm, and the mass of the external auditory canal was connected to them. The structures of the stapes were not connected with the footwall. We did an excision of malleus, incus, stapes and chorda tympani nerve, cleaned up the mass in the tympanic cavity, and delivered the mass to the pathologic department. Then we closed the eustachian tube with small piece of muscle, and performed conchoplasty. Postoperative pathology: general: a piece of polypoid tissue, 2.5 x 1cm x 1cm. Microscopic examination: polypoid tissue covered with stratified squamous epithelium, small and circular, relatively uniformed tumor cells nest can be seen infiltrated the epithelium, solid nesting, cords like, trabecular or arrayed like acinus.
Temporal MRI: left tympanic cavity and external auditory canal's occupying lesion, malignant tumor is not excluded; left middle ear mastoitis.

The imaging findings of middle ear carcinoid are not specific, which refers to a medium and lower grade highly differentiated neuroendocrine tumor. Ki-67 positive index ≤2% as level one(lower level),3%-20% as level two(medium level), >20% as level three(higher level) [7,8]. The tumor cells of lower differentiated NEC are organ like, Kikukata like, or diffuse distribution, nucleolus are visible. The division is easy to see, and often accompanied with laminar or atalike necrosis. In this case, the patient's Ki-67 is less than 5%, which refers to a medium and lower grade highly differentiated neuroendocrine tumor.

Differential diagnosis

Middle ear carcinoid need to be identified with adenomas, paragangliomas, adenosaccinoma, adenoid cystic carcinoma and metastatic carcinoma of the middle ear. Middle ear adenomas: both of them are rare occurred in the middle ear, and they have many similar pathological features and clinical behaviors, and have certain difficulty in differential diagnosis. Middle ear paragangliomas: more common than middle ear carcinoid, there are some similarities between them in endocrine aspect. Neuron specific olefin staining can be positive in both of them, but 5-HT, cytokeratin and silver staining present positive in the middle ear carcinoid only. Adenocarcinoma, adenoid cystic carcinoma and metastatic carcinoma: the Grimelius silver staining of adenocarcinoma and adenoid cystic carcinoma were negative. In addition, neuroendocrine granules were also absent under the electron microscope. Temporal metastatic carcinoma are infrequent, up to now, only one case of colonic carcinoid metastasis to the middle ear has been reported [9].

Treatment and prognosis

At present, middle ear carcinoid is mainly treated by surgery.
If the tumor is confined to the primary lesion, with no distant metastasis, radiotherapy and chemotherapy is not recommended [5]. On the contrary, if there is distant metastasis, radiotherapy and chemotherapy was considered. The recurrence rate of middle ear carcinoid is as high as 25% [10]. Therefore, remove the invaded otosteon and the lesion completely is recommended. Subtotal resection of temporal bone can be performed according to the recurrence range for recurrent. There have been reports of middle ear cancoaid metastases to the neck, parotid, basis cranii, posterior cranial fossa, internal organs and skeleton [6,10,11]. Therefore, it is necessary to evaluate the cervical and parotid lymph nodes carefully. If there is a metastasis, lymph node dissection should be performed as soon as possible. Middle ear NET prognosis is closely related to the degree of tumor differentiation, well-differentiated NET is mainly treated by surgery. Mastoidectomy can be performed, to remove the lesion completely. For poorly differentiated NEC, surgery plus radiotherapy or chemotherapy should be adopted.

**Conclusion**

The incidence of middle ear carcinoid is low, and its clinical and imaging features are not specific. The diagnosis is mainly based on pathology. At present, there is a lack of follow-up for middle ear carcinoid in biological behavior and therapeutic effect. It is mainly treated by surgery. It is according to histological classification and grading of tumor and whether distant metastasis occurs to decide whether postoperative supplementary radiotherapy and chemotherapy should be performed.

**References**