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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 pathological featurestreatment and prognosis.

Keywords: Middle ear neuroendocrine tumor carcinoid 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, paraganglia, thyroid, parathyroid, etc., especially rare in sinuses, throateye socket and the middle ear. Middle ear primary neuroendocrine tumors areparticularly rare. The first middle ear neuroendocrine tumor was discovered and reported by Murphy in 1980 [1], only 56 cases being reportedby 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 tinnitusor 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 bymethylprednisolone 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: 65-75-70-100dBHL. WT test: 256Hzinclinedleft. 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 neoplasmof 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 under 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 conchaplasty. 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 infiltratedthe epithelium, solid nesting, cords like, trabecular or arrayed like acinus.Immunohistochemically

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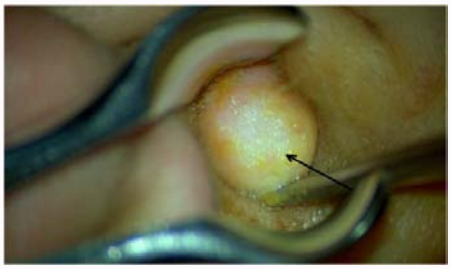

Figure 1: The left external auditory canal is blocked by a mass, which has a rough epithelioid tissue-like surface, and it's flexible and full of external auditory canal.

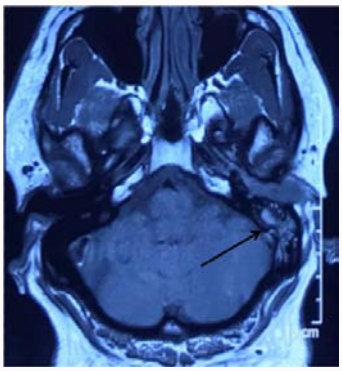

Figure 2: 256Hz inclined left. Temporal MRI: left tympanic cavity and external auditory canal's occupying lesion, malignant tumor is not excluded; left middle ear mastoitis.

staining: CK+, CK7 a little +, CK20-, CD56+, Syn+, CgA a little +, ck5/6 a little +, CEA-, P63 a little +, s-100 -, Actin-, Ki67 is less than 5%. Diagnosis:neuroendocrine neoplasms in squamous epithelium mucosa, combined with Immunohistochemically results considered as middle ear carcinomas (Figure A and Figure B).

Discussion

Clinical manifestation

Middle ear carcinoid tumor is an untypical clinical entity with nonspecific clinical symptoms. The age of onset is 16-64 years old, more common in men. It can only presentgeneral ear symptoms, such as tinnitus (24%), aural fullness (34%), ear discharge (17%), otalgia (10%) and hearing loss (90%), etc. [1,3], facial nerve paralysis can occur in some severe cases, specific carcinoid syndrome (intermittent flushing, diarrhea, and bronchospasm) is not common in most of the cases. In this case, except for normal ear symptoms such as plugging sensation in ear and hearing loss, the patient present with the characteristic symptom of slowly bleeding in the left external auditory canal.

Diagnosis

The imaging findings of middle ear carcinoid are not specific, which is why this kind of diseases are easily overlooked or misdiagnosed. Soft tissue density and osteolytic destruction in the middle ear can be shown in temporal CT scan ordinarily.In this case, soft tissue density can be seen in the left tympanic cavity, but there is no obvious osteolytic destruction. The diagnosis of middle ear carcinoid mainly depends on the pathologic result, and it has the histopathological features of both adenoid and neuroendocrine. Under electron microscope, argentaffin and Dense neuroendocrine granules are the

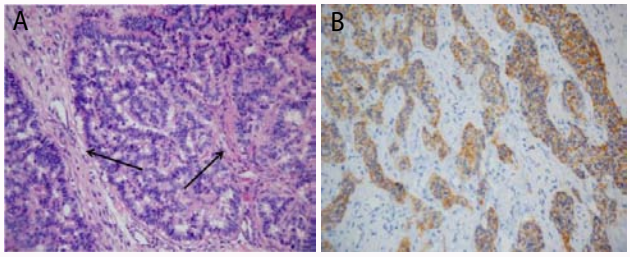

Figure A, B: Neuroendocrine neoplasms in squamous epithelium mucosa, combined with Immunohistochemically results, considered as middle ear carcinomas.

characteristic manifestations of carcinoid. Immunohistochemistry can improve the detection rate of neuroendocrine granules and the accuracy of diagnosis. Markers of neuroendocrine differentiation of typical carcinoid can often expressed as NSE, synaptophysin CsA, CD56, CD57 and epithelial markers (CK7, CK8) [5]. According to malignancy,carcinoid can be classified into two categories, slowly growing low grade and highly differentiated NeuroEndocrine Tumor (NET) and highly metastatic malignant poorly differentiated neuroendocrine carcinoma (NeuroEndocrine Carcinoma, NEC) [6]. Highly differentiated NET's histopathological features can be listed as follows [1]:The morphology of the tumor cells is consistent, solid nesting, cords like, trabecular or arrayed like acinus [2]; Tumor cells are in small or medium size, circular or ellipse, with medium or abundant amount of cytoplasm, eosinophilic or graininess, nuclear chromatin is slightly condensed, and nuclear division can be seen occasionally [3]; There are abundant of small vessels and fibrous interstitial around the tumor nests. According to the proliferation activity of the tumor cells, which can be indicated by the positive index of Ki-67, NET can be classified. 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 accompaniedwith laminar or atatlike 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, adenocarcinoma, 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 stainingand 5-HT 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 cancroid 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

1. Sahan M, Yildirim N, Arslanolu A, Karslioglu Y, Kazikdass K. Carcinoid tumor of the middle ear: report of a case. J American Journal of Otolaryngology. 2008;29(5):352-6.
2. Soga J, Ferlito A, Rinaldo A. Endocrinoearcinomas(carcinoids and their vailants)of the larynx: a comparative consideration with those of other sites. Oral Oncol. 2004;40(7):668-72.
3. Chan KC, Wu CM, Huang SF. Carcinoid tumor of the middie ear: a casereport. Am J Otolaryngol. 2005;26(1):57-9.
4. Zhu H, Xu G, Wen J. A recurrence of a primary carcinoid tumor of the middle ear: a case report and review of literature. Chinese Journal of Clinical Oncology and Rehabilitation. 2001;8(5):60-2.
5. Lu D, Yang H, Chen F. Clinical analysis of typical carcinoid primary carcinoma in the ear, nose and throat [J]. Chinese Journal of Otorhinolaryngology Head and Neck Surgery. 2014;49(10):864-66.
6. Mei J, Li GM. Primary neuroendocrine neoplasm in the middle ear: a report of 2 cases [J]. Chinese Clinical Oncology. 2016;21(6):573-4.
7. Chinese gastroenteric pancreatic neuroendocrine tumor expert group. Chinese pathologic consensus for standard diagnosis of gastrointestinal and pancreatic neuroendocrine neoplasm. Chinese Journal of Pathology. 2011;40(4):257-62.
8. Chinese gastroenteric pancreatic neuroendocrine tumor expert group. Chinese pathologic consensus for standard diagnosis of gastrointestinal and pancreatic neuroendocrine neoplasm [J]. Chinese Journal of Pathology. 2013;42(10):691-4.
9. Bates GJ, Luscombe FE. Carcinoid metastasis in the middle ear: anaesthetic and surgical management. J R Soc Med. 1989;82(3):171-2.
10. Ramsey MJ, Nadol JB, Pilch BZ, McKenna MJ. Carcinoid Tumor of the Middle Ear: Clinical Features, Recurrences, and Metastases. Laryngoscope. 2005;115(9):1660-6.
11. Fundakowski CE, Chapman JR, Thomas G. Middle ear carcinoid with distant osseous metastasis. Laryngocope. 2013;123(3):779-82.