Clinics in Surgery

Case Report
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A Rare Case of the Pilomatrix Carcinoma in Auricle and Literature Review

*Fei Wang, Jingru Zhang and Lian Hui**

Department of Otorhinolaryngology, The First Affiliated Hospital of China Medical University, China

Abstract

Introduction: This case report describes the diagnosis and treatment of a rare auricular pilomatrix carcinoma in a 37- year-old man.

Case Presentation: The patient underwent four local excision operations in 2 years, but the tumor still recurred. CT and MRI showed no characteristic manifestations. We performed wide resection under an otology microscope and repaired his temporal wound with a submental island perforator flap.

Discussion: pilomatrix carcinoma was considered an extremely rare carcinoma, lesions are located most frequently on the head as flesh-colored or bluish, asymptomatic cystic or solid masses. There is an ongoing dispute over the criteria for the histopathologic diagnosis of PC. Surgical wide resection is the recommended treatment such as mohs micrographic surgery.

Conclusion: Preoperative diagnosis was crucial, especially its histopathologic features. Wide surgical resection was the best treatment and prevented the mass from reoccurring.

Introduction

Pilomatrix Carcinoma (PC) is a rare malignancy originating from hair matrix cells with a high local-recurrence rate and a propensity to metastasize [1,2]. PC most commonly occurs in the head and has rarely been reported elsewhere [3]. Here, we report a rare case of PC in the auricle, the outermost part of the ear.

Case Presentation

A 37-year-old man presented with a 2-year history of a rapidly growing, originally hazelnut-sized, mass in the left auricle. He underwent four local-excision operations, but the mass recurred and continued growing rapidly. Examination of the auricle and neck area revealed a 3 cm × 3.5 cm irregular mass with ulceration and the involvement of two 1 cm × 1 cm hard lymph nodes in the left neck area II.

Enhanced temporal-bone Computed Tomography (CT) revealed a lumpy soft-tissue shadow in the left auricle, which was lobulated with uneven density, further intensified by enhancement. Multiple, small lymphoid nodules could be seen in the left parotid gland and neck. Enhanced temporal-bone Magnetic Resonance Imaging (MRI) revealed a diffusely inhomogeneous mass containing mixed low- and high-intensity areas. No metastasis was observed.

Surgery was performed under general anesthesia. The auricle was removed, but the earlobes were preserved for reconstruction. We removed part of the temporal muscle and performed lateral temporal-bone resection, left parotidectomy, and left neck lymphadenectomy with margins >2 cm, and then repaired the temporal wound with a submental island perforator flap. Most procedures were performed under otomicroscopic guidance. Pathological analysis confirmed that the parotid gland and cervical lymph nodes were tumor-free. The tumor in the auricle was malignant, of cutaneous adnexal origin, and the cancer cells were densely arranged in nests with aggressive growth and obvious atypia. Immunohistochemistry revealed a diagnosis of PC *via* the following markers: Cytokeratin (CK) (+), S-100 protein (-), Vimentin (-), P63(+), P40 (weakly+), B-cell lymphoma (Bcl) (-), CD30 (+), Epithelial Membrane Antigen (EMA) (+), CD10 (-), and antigen Ki-67 (70%+).

The patient was discharged and followed-up for 6 months without recurrence. All study protocols were approved by the ethics review committee of the first hospital of China Medical University.

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Correspondence:
Lian Hui, Department of Otorhinolaryngology, The First Affiliated Hospital of China Medical University, Shenyang, China, Tel: +86-024-8328-3126;
E-mail: wang19820126@163.com

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Discussion

PC was first reported in 1980 [4] and is also known as pilomatrical carcinoma, malignant pilomatircoma, malignant pilomatrixoma, matrical carcinoma, and calcifying epitheliocarcinoma of Malherbe. With only 125 to 135 cases reported to date, PC is considered exceedingly rare [1,5].

Clinical characteristics

PC is male-predominant and usually occurs at 40 to 60 years of age. Lesions are most frequently located in the head as flesh-colored or bluish, asymptomatic cystic or solid masses [6]. Most PCs are 0.5 cm to 20 cm in size; however, larger PCs have occasionally been encountered. PC may clinically resemble basal cell carcinoma, epidermal cyst, pilomatrixoma, or lipoma [2]. Here, the PC presented as an auricular mass following repeated simple excision. Although it will rarely be clinically diagnosed, clinicians should be mindful of possible local recurrence.

CT and MRI

Few reports have described PC-associated imaging findings. Niwa et al. [7] reported CT findings of a well-circumscribed, sand-like calcified mass and MRI findings of diffusely inhomogeneous, mixed signal intensities with inhomogeneous enhancements. There were some similar, albeit non-characteristic CT and MRI manifestations in this case.

Histopathologic features

Mutations in the exon-3 encoded, phosphorylated region of CTNNB1 have been reported [8]. There is ongoing dispute over the criteria for the histopathologic diagnosis of PC and aggressive pilomatrixoma and pilomatrixmoma. Tumors comprised pleomorphic basaloid cells with prominent nucleoli and atypical mitoses and central areas with keratotic material, shadow cells, and foci of necrosis. The tumor nests were surrounded by a desmoplastic stroma and infiltrated the adjacent tissues [5,9].

Treatment

PC recurred in >50% of cases with simple excision [3,6]. Surgical wide resection is the recommended treatment, when feasible, particularly for margins ranging from 5 mm to 30 mm [5]. Mohs micrographic surgery has been suggested as an alternative with greater margin control [2,10]. The role of chemotherapy and radiotherapy as an adjunct is unclear. Electron-beam radiation therapy has been successfully used is some cases.

Prognosis

PC tends to recur and has a high systemic metastasis rate (10%) [11]; thus, following definitive diagnosis, patients should be followed-up closely.

Conclusion

We reported an unusual case of auricular PC in a young man. We performed wide resection under otomicroscopic guidance and repaired the temporal wound with a submental island perforator flap. Preoperative diagnosis was crucial, especially regarding the histopathologic features. Wide surgical resection was optimal and prevented recurrence.

Established Facts

. Pilomatrix Carcinoma (PC) is a rare malignancy originating from hair matrix cells.

. Lesions are most frequently located in the head as flesh-colored or bluish, asymptomatic cystic or solid masses.

. There were some similar, albeit non-characteristic CT and MRI manifestations.

. Surgical wide resection is the recommended treatment

Novel Insights

. We reported an unusual case of auricular PC in a young man.

. Wide surgical resection was optimal and prevented recurrence.

Statement of Ethics

Written informed consent have given the patient to publish the case (including publication of images). Study protocols were approved by the ethics review committee of the First Hospital of China Medical University.

Authors Contribution

Lian Hui did the operation and supervised the paper. Fei Wang was involved the treatment of the patient and wrote the clinical details of the case. Jingru Zhang was involved the treatment of the patient and provided the images.

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