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Case Report

Exceptionally rare site:the occurrence of pilomatricoma of the ear lobule

Tapasaya Gund, Rahul A. Telang, Kireet Yathati*, Prakhar Kumar

Department of Otorhinolaryngology, BJGMC and Sassoon Hospital, Pune, Maharashtra, India

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***Correspondence:**
Dr. Kireet Yathati,
E-mail: tapasayagund@gmail.com

ABSTRACT

Pilomatricoma, a rare benign tumor primarily affecting the head and neck region, presents diagnostic challenges and requires histopathological confirmation for accurate diagnosis. A case of pilomatricoma in a 70-year-old female with a chronic swelling on the Left ear lobule is reported. Surgical excision was performed, confirming the diagnosis histopathologically, highlighting the importance of accurate recognition and management. Timely surgical excision of pilomatricoma is crucial, with low recurrence rates reported. Accurate diagnosis aids in appropriate management and optimal patient outcomes.

Keywords: Pilomatricoma, Benign tumor, Histopathological confirmation, Surgical excision, Accurate diagnosis

INTRODUCTION

Pilomatricoma, an infrequent benign tumor originating from hair follicle matrix cells, with a male to female ratio of 2:3.[1] It typically occurs in children and young adults, with the highest incidence before the age of 21, although it can also occur in the elderly. The most common sites of occurrence were the neck (30.2%), cheeks (16.8%), scalp (16.2%), and brow and periorbital area (14.0%).[2] On the face, the most affected region is the cheek accounting from 18.8% to 36% of all cases, followed by the periorbital region with an incidence between 6.6%-14%.[3] Pilomatricoma constitutes a small proportion, approximately 0.12%, of all skin tumors. Initially described in 1880 by Malherbe and Chenantais as calcifying epithelioma, its understanding advanced with Forbis and Helwig's 1961 discovery, which established its origin from hair matrix cells. Despite its distinctive histopathological characteristics, pilomatricoma is often missed in diagnosis and is frequently omitted from differential considerations. It typically presents as a superficial, firm, slow-growing, and usually painless mass, with possible skin discoloration or a bluish tint. Here, we report a case of pilomatricoma in a 70-year-old female located on the Left earlobe, emphasizing its importance in the differential diagnosis of ear lobule masses as an educational example.[4-8]

CASE REPORT

A 70-year-old female resident of Pune presented to the ENT OPD of tertiary care center with a chronic swelling over the left ear lobule. The swelling had persisted for one year, accompanied by recent dried-up pus discharge for the past three days. The patient complained of intermittent, dull-aching pain over the lesion. She denied any history of trauma, fever, chills, weight loss, fatigue, numbness, or tingling sensation associated with the swelling. The patient had not undergone any prior medical or surgical treatment for the swelling. Upon examination, a spherical, firm swelling measuring approximately 2×1×1 cm was noted. The overlying skin appeared tense, and there was mild tenderness upon palpation. However, no local rise in temperature was observed. The clinical presentation suggested an underlying infection, supported by the presence of dried-up pus discharge and tenderness. Given the clinical findings, investigations were ordered to confirm the

diagnosis and assess the extent of the lesion. These included a complete blood count, culture and sensitivity test of pus discharge, fine-needle aspiration cytology, and imaging studies. Surgical excision was performed, and the excised tissue specimen was sent for histopathological examination.

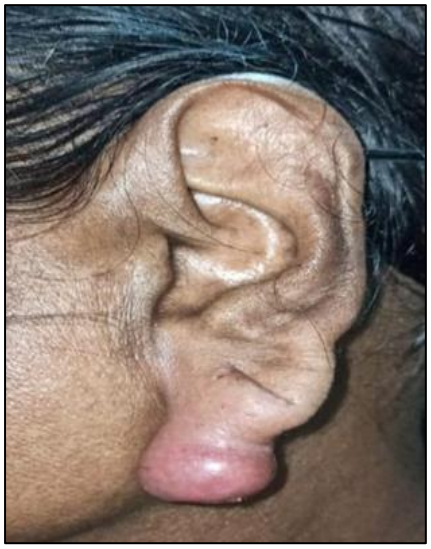

Figure 1: Left ear pilomatricoma.

Figure 2: Left ear pilomatricoma.

Figure 3: Gross image of the benign tumor and pilomatricoma.

The examination revealed a lobulated and circumscribed dermal-based neoplasm. Islands of basaloid cells exhibiting abrupt keratinization without intervening granular layer (trichilemmal keratinization) were observed. Ghost/shadow cells were also present, with basaloid cells showing mitotic activity, although abnormal mitoses were absent.

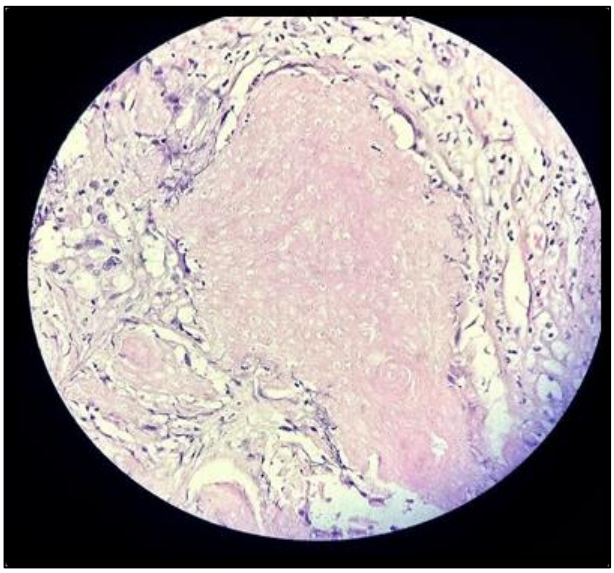

Figure 4: H and E histopathology image of pilomatricoma-showing ghost cells.

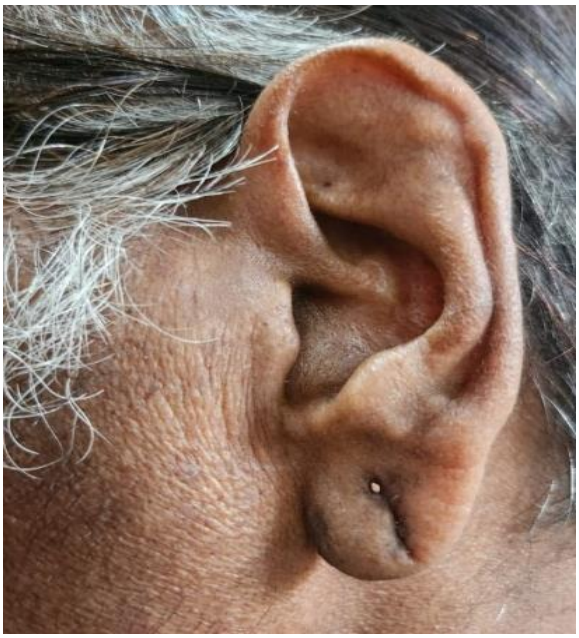

Figure 5: 6 months post operative image of pilomatricoma, no recurrence.

Based on the histological findings, the diagnosis of pilomatricoma was made. Although benign, surgical excision was warranted due to the persistent symptoms and risk of recurrence. The patient underwent successful surgical excision with no immediate complications.

DISCUSSION

Pilomatricoma, commonly known as calcifying epithelioma of Malherbe, is a rare benign tumor primarily occurring in the head and neck region, often diagnosed in women during their first two decades of life.[6,7] While its etiology remains debatable, hypothesis suggest its origin from either a branchial cleft or an ectodermal source, possibly linked to hair matrix cells.[8] Genetic mutations, including APC gene or CTNNB1 gene mutations, and overexpression of the bcl-2-proto-oncogene, are implicated in its pathogenesis.[9] Clinically, pilomatricoma manifests as a firm, subcutaneous mass with characteristic signs such as the tent sign and teeter–totter sign, aiding in its diagnosis.[4] However, definitive diagnosis often requires histopathological examination, which reveals a unique histological pattern with basaloid, polygonal squamous, shadow, and foreign-body giant cells, along with calcified deposits.[10] Despite its distinct features, cytological diagnosis can be challenging due to the variability in cellular composition among patients.[11]

Misdiagnosis is not uncommon, with lesions often mistaken for epidermal inclusion cysts or basal cell carcinomas, underscoring the importance of accurate diagnostic methods.[12,13] Imaging techniques, though of limited value, may aid in the differentiation of pilomatricoma from other lesions.[14] Treatment primarily involves complete surgical excision, with low rates of recurrence reported postoperatively.[15] Enucleation has also been reported as an alternative with no recurrence in certain cases.[16] Malignancy is rare, suspicion should be raised in cases of recurrence.[17] Distinguishing pilomatricoma histopathologically from other skin neoplasms, such as basal cell carcinoma with matricial differentiation, is crucial for appropriate management.[18] Despite challenges, accurate diagnosis and timely intervention remain pivotal in ensuring optimal patient outcomes.

CONCLUSION

Pilomatricoma is a rare but important benign tumor that predominantly affects the head and neck region, with characteristic clinical and histopathological features. Accurate diagnosis and appropriate management, including surgical excision, are crucial for optimal patient outcomes. Recognition of pilomatricoma's clinical presentation and histopathological patterns is essential to prevent misdiagnosis and ensure timely intervention.
The rarity of this tumor, low incidence of cases and diverse clinical presentations have led to a high number of misdiagnoses. However, differential of Pilomatricoma should be kept in mind when dealing with clinical case of swelling of ear lobule.

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