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Case Report

Pilomatrixoma of the Cheek: A Case Report

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ABSTRACT

Pilomatrixoma “Calcifying epithelioma of Malherbe” is a benign calcifying tumor that arises from the hair matrix. It is more common in females and usually presents during the first two decades of life as an asymptomatic, mobile, firm to hard mass. Most of the cases are benign and affect the face. Surprisingly, a preoperative diagnosis of pilomatrixoma is rarely considered by clinicians and often mistaken for other skin-based and subcutaneous lesions. We report a case of pilomatrixoma of the cheek in female Libyan patient, that was clinically diagnosed as a benign salivary gland tumor or reactive lymph node with histopathological findings, and a brief review of the literature.

INTRODUCTION

Pilomatrixomas are benign neoplasms originating from the cells of hair follicles. Malherbe and Chenantais described pilomatrixoma as a benign tumour of sebaceous glands in 1880⁽¹⁾. In 1961, Forbis and Helwig proposed the term pilomatrixoma to avoid a connotation of malignancy⁽²⁾. They typically present as a slowly enlarging, solitary mass^(3,4). Majority of all pilomatrixomas were reported to affect the head-neck region and rarely the upper extremity⁽⁵⁾. It is predominately affecting children⁽⁶⁾. According to histopathological findings, pilomatrixomas are chronologically divided into four phases: early, fully developed, early regressive and late regressive.

In the late regressive period, shadow cells, calcification, ossification and fibrotic stroma develop, and a complete loss of the basaloid epithelial component is observed⁽⁷⁾. It usually presents as an isolated lesion and rarely undergoes malignant transformation⁽⁸⁾. It is not uncommon for these tumors to be misdiagnosed. On clinical examination, the consistency of pilomatrixoma varies considerably, depending on the degree of calcification⁽⁹⁾. Fine needle aspiration (FNA) cytology can be used in diagnosis^(10,11), and ultrasound examination may be helpful in diagnosis^(2,12).

This lesion is usually reported by dermatologists and plastic surgeons, and not commonly encountered in dental practice⁽¹³⁾. Its histological resemblance to the calcifying odontogenic cyst (the Gorlin's cyst) make it relevant, both to the maxillofacial surgeons and the pathologists⁽¹⁴⁾.

The purpose of this report is to describe a case of pilomatrixoma of the cheek in a young Libyan female patient. We believe that its consideration is essential in the preoperative differential diagnosis of firm nodular masses of the oral and paraoral regions.

Case Report

A 22-year-old woman had a one year history of slowly growing, painless mass of the right cheek. On physical examination a palpable firm 0.5x0.5cm mass was detected at the level of the angle of the mouth, the lesion was freely mobile with normal mucosa and overlying skin color. The preoperative differential diagnosis included pleomorphic adenoma and reactive lymph node. The surgical procedure consisted of enucleation of the lesion using intraoral approach. At the time of surgery, the tumor was easily shelled out of the surrounding tissue. The tissue was routinely processed and embedded in paraffin wax and Hematoxylin and Eosin (H and E)-stained sections were studied.

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Figure 1: show area of mass in right buccal mucosa

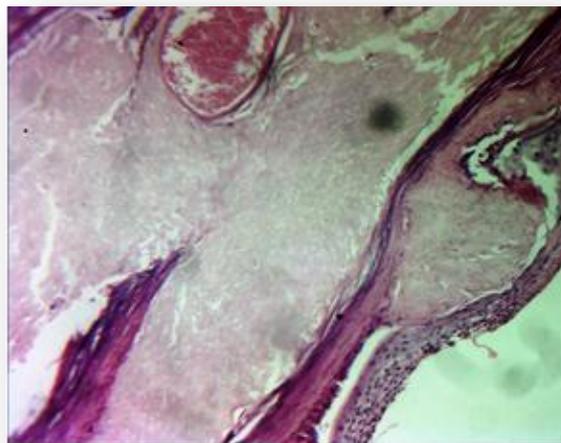


Figure 3: peripheral basaloid cells are darkly stained, round or elongated, with indistinct cell border and minimal cytoplasm and central ghost cells

Histopathologic Findings

Microscopically, the lesion was well encapsulated tumor composed of two distinct cell populations; basophilic basaloid cells located towards the periphery and eosinophilic ghost cells occupying the central portion in fibrovascular connective tissue stroma.

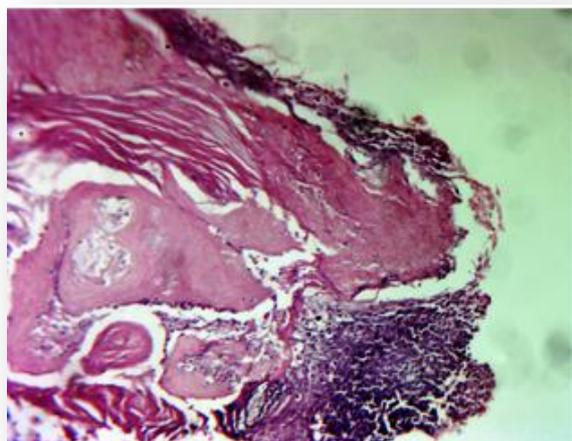


Figure 2: photomicrograph of pilomatrixoma showing the characteristic basaloid and shadow cells (H and E, 10).

DISCUSSION

Pilomatrixoma, or calcifying epithelioma of Malherbe, is a benign skin neoplasm that arises from hair follicle matrix cells. In 1880, Malherbe and Chenantais first described this lesion, referred to as the calcifying epithelioma, though it was thought to derive from sebaceous glands⁽¹⁾. Later, the term pilomatrixoma was coined by Forbis and Helwig in 1961, thus, avoiding the term epithelioma which carries the connotation of malignancy and it was suggested that the cells of origin are the outer root sheath cells of the hair follicle⁽²⁾. These lesions are typically found in the head and neck region, but they have also been described in various upper extremity locations.

The reported incidence of pilomatrixoma is 0.08% in surgical pathology cases and 1.04% when benign dermatological lesions are included. The aetiology is not known; a relationship to trauma, insect bite or surgery was noted in the history of fewer than 5% of cases⁽⁷⁾. These lesions present most commonly in children and young adults, and they are noted more commonly in females. Most of cases of pilomatrixoma occur sporadically without an apparent genetic predisposition. However, few are associated with genetic disorders such as Gardner's syndrome, basal cell nevus syndrome and xeroderma pigmentosa⁽¹⁵⁾.

Histologically the neoplasm is well encapsulated containing irregularly shaped, lobulated islands of cells separated by fine, fibrovascular connective tissue

stroma. The islands demonstrated two distinct cell populations comprising of basaloid cells at the periphery and the ghost or shadow cells occupying the central portion. A rare malignant counterpart, pilomatrix carcinoma, has been described in the literature. Many key features are similar between these benign and malignant counterparts; the primary differentiating characteristics include a high mitotic rate with atypical mitoses, central necrosis, infiltration of the skin and soft tissue, and invasion of blood and lymphatic vessels. Pilomatrixoma show positive for cytokeratin, CD3, CD20, CD68, PCNA, and CD3411⁽¹⁶⁾. Pilomatrixomas are often misdiagnosed on preoperative evaluation, however palpation of superficial firm nodule that is not painful or tender is characteristic. Rarity of this lesion and its histological resemblance to the calcifying odontogenic cyst (the Gorlin's cyst) make it relevant, both to the maxillofacial surgeons and the pathologists⁽¹⁴⁾. The differential diagnosis is primarily with dermoid cyst, however dermoid cysts may contain other teratomatous elements in small areas⁽¹⁷⁾.

For the best of our knowledge, we describe here the first case of pilomatrixoma between Libyans in the cheek region of a young female patient and we believe that its inclusion in the differential diagnosis of subcutaneous and/or submucosal nodular masses of cheek and Paraoral regions is essential.

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