

An Unusual Presentation of Pilomatrixoma

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ABSTRACT

A case of twenty years old female is reported, which was treated inappropriately. She complained of a painless lump located on the right cheek at middle half of the face. An early diagnosis of sebaceous cyst was made. Excision was performed and histopathological findings revealed Pilomatrixoma. Pilomatrixoma is an uncommon, harmless, skin lesion derived from hair matrix cells. It is composed of an epithelial component of the most proximal portion of hair follicle. It is most often diagnosed in young children but may also affect adults. Single skin-coloured or purplish lesions arise on the head and neck, but they may occur at any site. The key to diagnosis is identification of darkly stained 'basophilic cells' and 'shadow cells'. Calcium deposits are found in most of the lesions. Complete surgical excision is the treatment with rare chances of complications.

KEY WORDS: Pilomatrixoma. Epithelioma. Calcium. Treatment.

INTRODUCTION

Pilomatrixoma is the most common type of pilar tumours¹, most frequently appearing in the first or second decade of life. The tumour is usually a deep-seated, solitary, firm nodule with overlying normal epidermis.² This is a rare, benign, circumscribed, calcifying epithelial neoplasm that is derived from hair matrix cells. However, multiple pilomatrixomas are uncommon.³

CASE REPORT

A twenty years old female presented in the Department of Oral and Maxillofacial Surgery, Liaquat College of Medicine and Dentistry, Karachi, with a complaint of a painless lump on her right cheek on middle half of face for the last eight years. The swelling was round, 3-4cm in diameter, initially being superficial but subsequently involved the deeper tissues. It was not associated with any pus or discharge (**Figure I**). A diagnosis of Sebaceous cyst and Acne cyst was established in consultation with two dermatologists respectively. Intraregional corticosteroid injections were administered (twice within six months) but the lesion was not responsive to treatment. Physical examination revealed that the lump was hard, non-tender, 1x1cm (approximately) in size and was attached to the overlying skin. She had no other signs or symptoms and her routine laboratory tests were normal. A preliminary diagnosis of Sebaceous cyst was made and excisional biopsy of the lump was performed under local anaes-

thesia. The wound was sutured with 3-0 silk and dressing was done. Patient was advised for antibiotic therapy and follow up after one week (**Figure II**). Postoperative findings revealed a hard mass with an irregular surface, carrying attached portion of overlying skin. Microscopic description revealed fibrocollagenous tissue showing a nodular lesion exhibiting nests of basophilic cells with few areas showing ghost appearing cells. The nuclei were round to oval. Intervening stroma was fibrocollagenous and in areas showing giant cell reaction. Osseous metaplasia was also identified. Focally skin tissue was identified on the surface. No evidence of any cyst formation or malignancy was seen (**Figures III-IV**).

FIGURE I:
PRE OPERATIVE CLINICAL PICTURE OF PATIENT



FIGURE II: POST OPERATIVE CLINICAL PICTURE AFTER ONE WEEK



FIGURE III: A PHOTOMICROGRAPH OF THE HISTOLOGICAL SECTION OF PILOMATRIXOMA AT LOW POWER (Hx E)

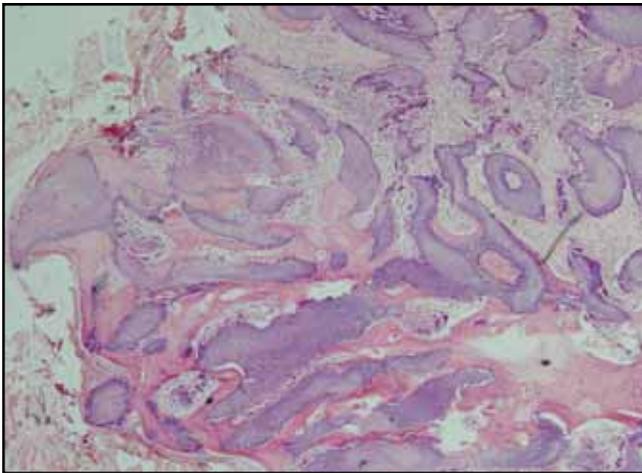
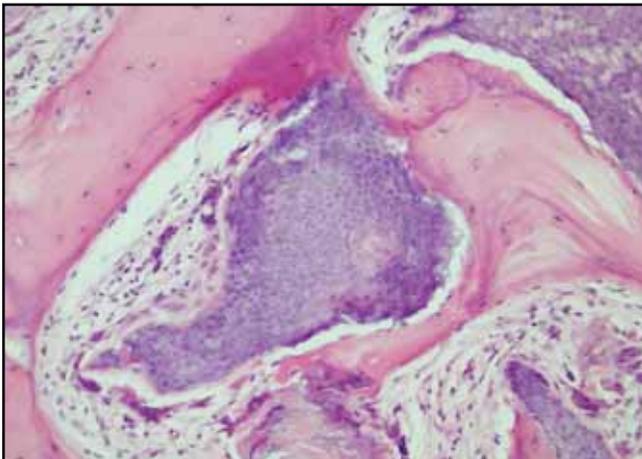


FIGURE IV: A PHOTOMICROGRAPH OF THE HISTOLOGICAL SECTION OF PILOMATRIXOMA AT HIGH POWER (Hx E)



DISCUSSION

Pilomatrixomas are benign skin neoplasms of hair follicle origin. They are one of the most common superficial masses of the head and neck, excised in children. Although the entity has been well studied in the literature, few studies have been undertaken to evaluate the clinical characteristics of head and neck pilomatrixomas specifically in children. In a retrospective chart review ninety-one cases of pilomatrixoma were confirmed in 86 patients. The age range was 5 months to 17 years. The median age at time of excision was 6.0 years. The most common sites of occurrence were the cheek (36%), neck (20%), periorbital region (14%), and scalp (9%). The male to female ratio was 1:1.5. Multiple lesions were found in 8.2% of patients.⁴

The distribution of these lesions did not correlate with the density of the hair follicles, but it was in accord with the distribution of intermediate hair such as those in the hair border. This relationship may have etiologic significance.⁵ Histopathologic examination of the excised tumors revealed variable basophilic hair matrix cells and sheets of non-viable eosinophilic shadow cells. Foci of dystrophic calcification were also seen in the necrotic tumor areas.⁶

Beta-catenin is a downstream effector in the WNT-signalling pathway, acting as a signal for differentiation and proliferation. Immunohistochemical and molecular analysis of beta-catenin reveals that mutations in CTNNB1, the gene encoding beta-catenin, are present in a wide variety of benign and malignant neoplasms and pilomatrix carcinomas may arise from their benign counterparts.⁷ Multiple periorcular and facial pilomatrixomas can occur in children in the clinical absence of myotonic dystrophy, Gardner's syndrome and sarcoidosis.⁶ Other diagnoses include sebaceous and dermoid cysts, foreign body reaction, calcification in lymph gland, and fat necrosis. Factors contributing to misdiagnosis include cystic lesions with varying consistency, punctum-like appearance, atypical location, and absence of clinically recognizable calcification. Despite close excision, the recurrence rate is low.⁸ All pilomatrixomas had been treated for solitary tumors with simple surgical excision and closure. There are no reported adverse outcomes and no tumor recurrences at the surgical sites. These findings support the use of simple surgical excision as the treatment of choice for these tumors.⁹ The importance of three pillars of diagnosis i.e. history, examination and investigation is well established. A clinician shall always correlate the findings with each other before undertaking any treatment. Pilomatrixoma is a cutaneous neoplasm that is one of most common causes of superficial head and neck masses in children. Although the pre-surgical diagnosis may be difficult in some cases, pilomatrixoma must be kept in the differ-

ential of superficial head and neck masses in children.⁴ Pilomatricoma was an unusual finding in the Oral Surgery Department. Awareness regarding the inclusion of dermal appendages diseases is very important while establishing the differential diagnosis for chronic, painless head and neck swellings.

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