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Case Report **Open Access**

Ulcerated Pilomatrixoma: A Frequently Misdiagnosed Tumour

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Case summary

Pilomatrixoma is a rare superficial benign skin tumor arising from the hair follicle matrix cells. Pilomatrixoma can occasionally become malignant. Complete surgical excision is often curative. It is often clinically misdiagnosed, hence the reason to share this case. We report a case of a 19-year-old man who presented with an ulcerated non-painful swelling involving the medial part of the right upper eyelid for two years. It was initially clinically diagnosed as keratoacanthoma and later basal cell carcinoma. However, histopathological examination after complete surgical excision revealed features of pilomatrixoma. This report highlights that pilomatrixoma may be misdiagnosed in clinical practice.

Keywords: *Pilomatrixoma, Eyelid, Epithelioma, Pilomatrix carcinoma.*

Introduction

Pilomatrixoma is a rare benign tumor of the hair follicle matrix. Although it can occur at any age, it most frequently appears in the first or second decade of life with female preponderance (1,2). These tumors occur as solitary lesions however, multiple nodules have been reported (3,4). Pilomatrixoma is most commonly found in the head and neck region with rare involvement of the trunk and extremities (3,4). Pilomatrixoma occurring in the head and neck region commonly involves the cheeks, followed by the neck, periorbital region and scalp. Lesions are characteristically painless, slow-growing, hard subcutaneous nodules. Pilomatrixoma recurrence is due to incomplete excision and rarely undergo malignant transformation to pilomatrix carcinoma (3,4).

There has been an increase in understanding of the morphologic features and clinical presentation of pilomatrixoma. Yet, challenges still persist in making clinical diagnoses (5). This may lead to a delay in definitive treatment and posing chances for malignant transformation. Few cases have been reported worldwide, and to our knowledge, no case had been reported previously in Tanzania. This case write-up aims to share a case of a rare tumor to raise clinicians' awareness of the possibility of pilomatrixoma as a cause of solitary skin nodules, especially those on the head and neck. We report a case of eyelid pilomatrixoma in a young adult with no evidence of local recurrence 2 years after surgical excision.

Case Presentation

A 19-year-old male from the Southern part of Tanzania was referred to the eye clinic from the dermatology unit with a clinical diagnosis of keratoacanthoma of the upper eyelid. He reported to have the swelling on the medial aspect of the right upper eyelid for 2 years, which was

insidious in onset and progressively increasing in size, and later on the swelling **became** ulcerated. It was painless, not bleeding nor discharging. However, it was cosmetically disfiguring. He reported **having** been treated with ointments and antibiotics from various health facilities without remission.

Figure 1. Hard ulcerated mass involving the right upper eyelid

The patient had normal vital signs on general examination. Local examination revealed a stony-hard, painless swelling below the brow margin of the right eye near the medial canthus. The swelling was ulcerated with yellowish crusts at the base of the ulcer. Margins of the lesion were flat, and the lesion was not fixed to the orbital rim or underlying tissues. The swelling measured 3cmx2cmx1cm (Figure 1). There were no palpable pre-auricular or submandibular lymph nodes. The rest of the ocular examination of both eyes was unremarkable.

The initial clinical diagnosis was basal cell carcinoma. A wide surgical excision of the swelling with a 3mm clear margin was done. The surrounding tissues, including the orbital margin were free from the tumor. The defect was reconstructed with **an** advancement skin graft on the same sitting. Macroscopically, the tumor was **a** hard, calcified, dull white encapsulated nodule (Figure 2).

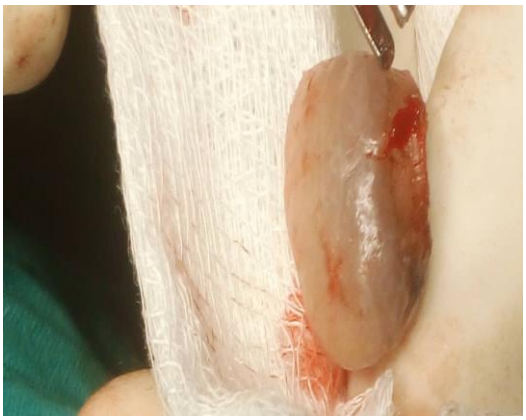

Figure 2. Well-circumscribed, hard, calcified, dull white nodular mass

Histopathological examination revealed dermal solid nests of eosinophilic ghost cells with a trichilemmal type of keratinization in the background of calcified stroma (Figures 3 and 4), which confirmed the diagnosis of eyelid Pilomatrixoma.

Wide local excision is curative in most cases. There were no signs of recurrence during the first 2 years of follow-up. The patient has been kept on long-term follow-up.

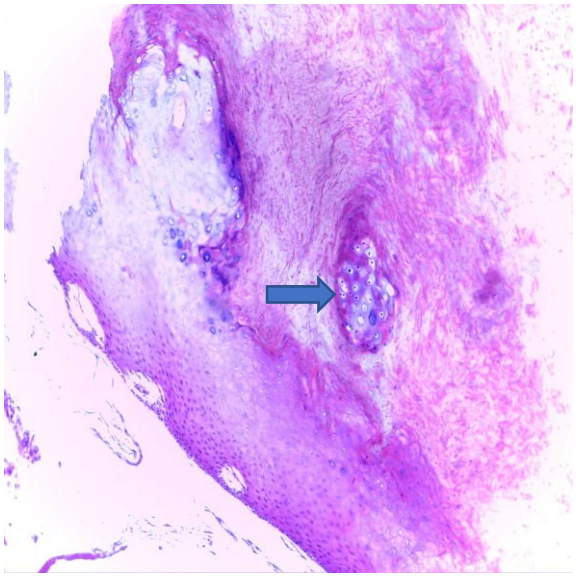

Figure 3. Lesion composed of a dermal solid nest (arrow) with remnants of trichilemmal keratinization in the background of calcified stromal tissue (HE X100)

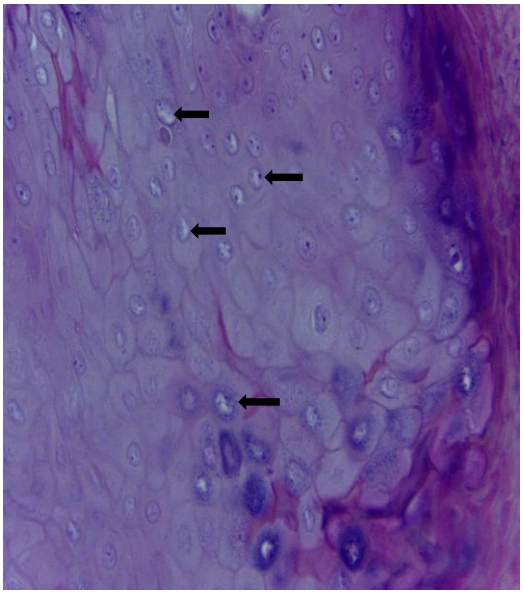

Figure 4. Details of dermal solid nests showing multiple eosinophilic ghost cells (arrows) (HE X400)

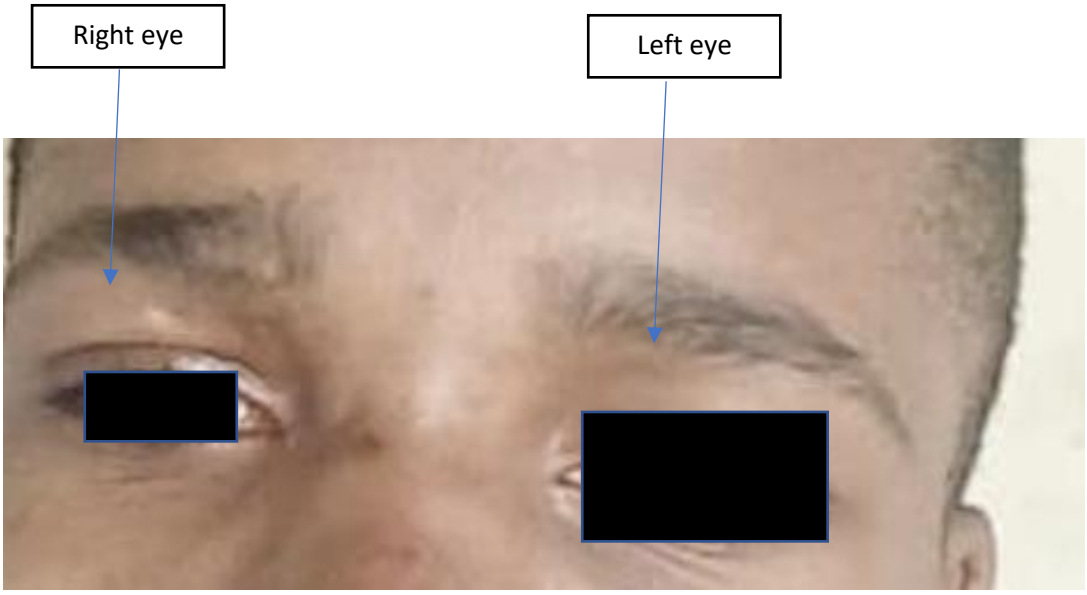

Figure 5. One year post excision and advancement flap skin graft

Discussion

Pilomatrixoma, also known as calcifying epithelioma of Malherbe, is a rare benign tumor of hair follicle matrix cell origin, making up to 1.04% of all benign tumors of the skin (5,6). It is commonly benign in nature with few cases reported to have malignant transformation (5).

There are debates on the origin of pilomatrixoma, some believe pilomatrixoma is epithelial choriostoma, whereas others propose pilomatrixoma to be of ectodermal origin (1,7).

Diagnosis can be made clinically with histopathological confirmation. The “Tent” and “Teeter-totter” signs are pathognomonic and most helpful in making a clinical diagnosis, the former occurs when the skin overlying the lesion is stretched which produces multiple facets and angles while the latter refers to the protrusion on the skin when the pressure is applied on the opposite side of the lesions (8). It is possible that these typical clinical features were initially overlooked in our patient and were thereafter challenging to elicit as the lesion ulcerated.

The clinical presentation of pilomatrixoma may look more or less similar to many benign tumors of the skin. Hence in the list of differential diagnoses of benign skin lesions. Our patient was clinically diagnosed with keratoacanthoma. In a case review of pilomatrixoma, only up to 30% of all pilomatrixoma were correctly diagnosed clinically (5,8).

The presence of ulceration with elimination of material through the lesion, combined with the histological presence of epidermal ulceration with extrusion of eosinophilic tumor cells, may configure a rare, atypical, and infrequent form called perforating pilomatrixoma (9–11). Our patient had an ulcerated lesion but with no histological features of perforating pilomatrixoma.

Another challenge clinicians face is differentiating pilomatrixoma from pilomatrix carcinoma as the histologic features are almost the same. However, a lesion >4 cm, infiltrating border with fascia and skeletal muscle involvement, a predominance of basaloid cells, nuclear pleomorphism, abnormal mitotic figures, vascular, lymphatic and perineural invasion warrants the possibility of pilomatrix carcinoma (6,12). Our patient did not have these features both on clinical and histopathological evaluation.

A missed clinical diagnosis is associated with unnecessary investigations, delay of treatment or getting the wrong treatment. Our patient stayed with a disfiguring facial lesion for two years without a correct diagnosis.

The definitive treatment of pilomatrixoma is complete lesion excision, as in our patient. Local recurrence is reported to occur in case of incomplete surgical excision(3). The present patient had no signs of local recurrence 2 years after surgical excision.

Conclusion

Pilomatrixoma is a rare tumor that continues to cause difficulty in clinical diagnosis. A missed diagnosis leads to unnecessary investigations, wrong treatment procedures, and delays in definitive treatment which adds to the chances of malignant transformation. Careful clinical examination can lead to early definitive diagnosis and treatment.

Ethical consideration

A written informed consent was obtained from the patient for sharing his photos and medical information.

Authors' contributions

Assistant surgeon (SN), drafting manuscript (SN and CM), revision of histopathological evaluation (NS and AM), critical revision of manuscript (CM, SN, NS and AM). All authors reviewed and approved the manuscript.

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