

Case Report

Giant Ulcerating Pilomatrixoma: A Case Report

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Received: 07 July 2024

Accepted: 22 October 2024

Epub Ahead of Print: 28 December 2024

Published:

DOI

10.25259/IJPGD_140_2024

Quick Response Code:



ABSTRACT

Pilomatrixoma is a rare benign adnexal neoplasm originating from hair follicle matrix cells in which giant, bullous, ulcerating, anetodermic and lymphangiectatic variants may be seen. Giant pilomatrixomas (size > 5 cm) rarely ulcerate, and are often a diagnostic challenge as they resemble cutaneous malignancies. We report the case of a 45-year-old male who presented with a slow-growing, ulcerating nodule on the left forearm.

Keywords: Adnexal neoplasm, Hair tumour, Shadow cells

INTRODUCTION

Pilomatrixoma, also known as calcifying epithelioma of Malherbe, is a rare benign adnexal neoplasm originating from hair follicle matrix cells, comprising 0.12% of skin tumours. It predominantly affects children and young adults, with a female preponderance.^[1] Typically slow growing and well circumscribed, it presents as a cystic or firm nodule measuring 0.5–2 cm, often located on the head and neck, upper extremities and trunk.^[2] Rarely, bullous, ulcerating, anetodermic, lymphangiectatic and giant variants occur. Giant pilomatrixomas (size > 5 cm) rarely ulcerate, and are often a diagnostic challenge as they resemble cutaneous malignancies.^[1] This report aims to highlight one such diagnostic challenge.

CASE REPORT

A 45-year-old male presented with a gradually enlarging mass on his left forearm for 2 months. Physical examination revealed a 5 × 5 cm² exophytic, centrally firm, tender ulcerated nodule with yellowish crusting on the flexor aspect [Figure 1]. No fixity or discharge was noted, and surrounding skin was unaffected. There was no history of trauma, infection or prior skin cancers; general examination, routine tests and chest X-ray were normal. Dermatoscopy showed irregular whitish streaks, reddish pink areas, bluish grey regions and multiple dotted and few irregular linear vascular patterns [Figure 2]. Differential diagnoses included squamous cell carcinoma, keratoacanthoma, lymphoma, melanoma, sarcoma and metastasis. A 4-mm punch biopsy showed shadow cells, basaloid cells, inflammatory infiltrate and metaplastic calcification foci, suggestive of pilomatrixoma. In a small biopsy specimen, evaluating the structure and growth patterns of a neoplasm can be challenging. This difficulty extends to distinguishing between pilomatrixoma and pilomatrix carcinoma, as well as discerning between their aggressive and proliferative variants. The lesion was excised under local anaesthesia with histopathological differential diagnoses of aggressive pilomatrixoma, proliferating pilomatrixoma and pilomatrix carcinoma

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in mind. Macroscopically, the lesion was clearly defined and had a grey-tan colour. Microscopically, an unencapsulated



Figure 1: Giant ulcerating pilomatricoma: Erythematous ulcerated nodule with yellowish crusting seen on flexor aspect of the left forearm.

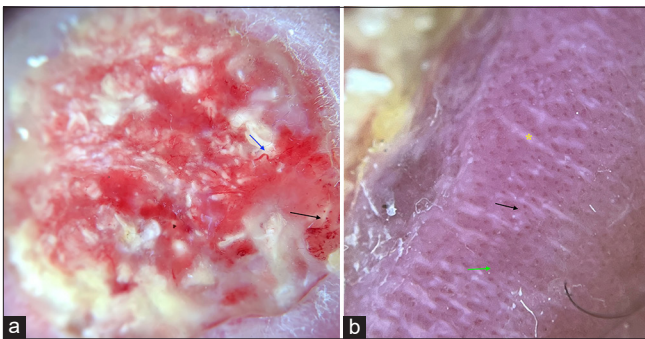


Figure 2: Dermatoscopy (using dermatoscope IDS-1100, under polarised light with $\times 10$ magnification) in giant ulcerating pilomatricoma showing (a) dotted vessels (black arrow) and linear irregular vessels (blue arrow), (b) irregular whitish streaks (asterisk), few reddish pink homogenous areas (green arrow) and dotted vessels (black arrow), IDS: Illuco dermatoscope.

neoplasm involving the whole dermis, with aggregates of basaloid matrical cells peripherally and eosinophilic shadow cells centrally, which favoured a fully developed pilomatricoma, was present. Foci of metaplastic ossification were seen [Figures 3]. No necrosis, haemorrhage or invasion was observed. Few mitoses were noted. Follow-up showed no complications or recurrence.

DISCUSSION

Pilomatricoma, a benign tumour from hair follicle matrix, has been inaccurately diagnosed preoperatively (up to 30% cases) due to its similarity to other tumours.^[3] Although its cause is unclear, B-cell lymphoma-2 gene and beta-catenin pathway have been implicated.^[4,5] While generally not hereditary, multiple lesions can occur with Gardner's syndrome, Steinert disease and Turner's syndrome.^[3] Dermatoscopic features include irregular, whitish streaks indicating calcification and cornification, reddish homogeneous zones, hairpin-like, linear irregular and dotted vessels representing vascular structures and structureless grey-blue areas corresponding to melanin within basaloid cell aggregates or siderophages in inflammatory infiltrate.^[6-8]

Pilomatricoma features 'shadow cells,' a unique microscopic sign of flawed hair differentiation.^[9-12] Shadow cells are named so because although their silhouettes are discernible, the nuclei are not observed. They form a characteristic feature of pilomatricoma and are significant because they represent differentiation towards the hair cortex, seen as structureless eosinophilic cells lacking nuclei.^[4] Pilomatricomas progress through four stages: (a) Early cystic lesions, (b) fully developed large cysts, (c) early regressive with basaloid cells, shadow cells, lymphocytic infiltrate and multinucleated giant cells and (d) late regressive with numerous shadow cells, no basaloid or inflammatory cells and eventual calcification and ossification.^[4,12]

In our case, proliferating pilomatricoma, aggressive pilomatricoma and pilomatricoma carcinoma were shortlisted as

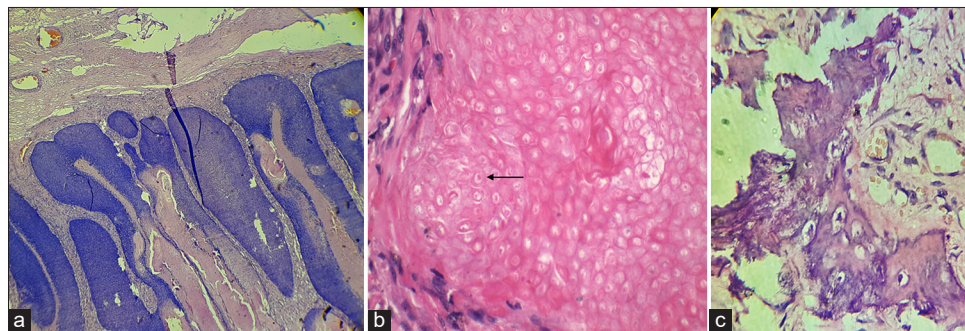


Figure 3: Histopathology of giant ulcerating pilomatricoma showing (a) well-circumscribed unencapsulated neoplasm involving the whole dermis, composed of aggregates of basaloid matrical cells aligned at the periphery and shadow cells centrally (H&E $\times 10$) (b) shadow cells (black arrow) in H&E $\times 40$ (c) Foci of metaplastic ossification (H&E $\times 40$). H&E: haematoxylin and eosin.

histopathological differential diagnoses after examination of limited biopsy specimen and diagnosis was confirmed after examination of surgically excised specimen. Proliferating pilomatricomas typically occur around the seventh decade, while aggressive and benign forms are more common in children and young adults.^[1,4] Proliferating and aggressive pilomatricomas represent intermediate stages of a histological spectrum between benign pilomatricoma and pilomatrix carcinoma.^[4] Our case differed from proliferating pilomatricoma by lacking nuclear atypia and mitotic figures. Aggressive pilomatricomas typically show a higher matrical cell component, increased mitotic activity (7/10 high-power fields [HPF] vs. 1–2/10 HPF), single cell necrosis, prominent nucleoli, invasive growth patterns and nuclear pleomorphism, all absent in our case.^[4,11] Pilomatrix carcinoma was ruled out due to the absence of abnormal mitosis, nuclear crowding, pleomorphism, necrosis and lymphatic or perineural invasion.^[4,5]

Surgical excision is the preferred treatment for pilomatricomas, with a low recurrence rate (more common in proliferating types). Fixed tumours are excised along with the overlying dermis.^[13]

CONCLUSION

Pilomatricomas pose diagnostic challenges due to their histopathological complexity. Giant ulcerating pilomatricomas can mimic malignancies, highlighting the importance of recognising shadow cells, a key histopathological clue essential for accurate pre-operative diagnosis.

Ethical approval

Institutional Review Board approval is not required.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

Use of artificial intelligence (AI)-assisted technology for manuscript preparation

The authors confirm that there was no use of artificial intelligence (AI)-assisted technology for assisting in the writing or editing of the manuscript and no images were manipulated using AI.

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How to cite this article: Sangam VB, Patel NM, Shah HA, Chauhan J. Giant Ulcerating Pilomatricoma: A Case Report. *Indian J Postgrad Dermatol*. doi: 10.25259/IJPGD_140_2024