CASE REPORT

Digital Pilomatrixoma
An Unusual Site of Presentation

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Abstract
Pilomatricoma is an uncommon, harmless, skin lesion derived from hair matrix cells. It is also spelled 'pilomatrixoma' and sometimes known as 'calcifying epithelioma of malherbe'. It is most often diagnosed in young children but may also affect adults. Single skin-coloured or purplish lesions arose on the head and neck, but they may occur on any site. They are characterized by calcification within the lesion, which makes it feel hard and bony. FNAC can help us in the diagnosis but excisional biopsy and histopathological examination is recommended for confirmation and curative treatment. We present a rare case of pilomatricoma of the digit in an adult female.

Key Words
Pilomatricoma, Basaloid cells, Ghost cells

Introduction
First described by Malherbe and Chenantais in 1880 as a benign neoplasm of sebaceous gland origin, our understanding of pilomatricoma has evolved significantly (1). It is now understood that pilomatricomas are calcifying epitheliomas which may differentiate towards the hair matrix, cortex, follicular infundibulum, outer root sheath and hair bulge (2). Etiology has been linked to mutations such as B-catenin and bcl (3,4). Although pilomatricomas represent only 0.12% of all cutaneous tumors, it is relatively common in children, but occurrence in adults is increasingly being recognized (5,6,7) Grossly, it is a nodular, subepidermal benign tumor with areas of calcification. Microscopically, it is composed of solid nests of small basaloid cells that may lead to a mistaken diagnosis of basal cell carcinoma. The main feature is that these basaloid cells undergo abrupt keratinisation leading to the formation of 'ghost' and 'shadow' cells (6). Foreign body reaction, calcification and ossification are common. The present case is reported due to the rare occurrence of a pilomatricoma at an unusual site - the digit of an adult female.

Case Report
We encountered a case of pilomatricoma in a 48 year old female patient. The patient came to the surgical OPD, with the complaint of swelling 2nd digit, left foot. On clinical examination, it was a superficial and firm mass. Pain was elicited on palpation. The overlying skin was ulcerated and exhibited a bluish discoloration (Fig 1). The lesion was surgically excised and submitted for histopathological examination.

Grossly, it was 1 cm in size skin covered lesion. Cut section revealed gritty sensation. Microscopically, it is composed of solid nests of small basaloid cells that may lead to a mistaken diagnosis of basal cell carcinoma. The main feature is that these basaloid cells undergo abrupt keratinisation leading to the formation of 'ghost' and 'shadow' cells (6). Foreign body reaction, calcification and ossification are common. The present case is reported due to the rare occurrence of a pilomatricoma at an unusual site i.e digit (2nd toe left foot) and that too in an adult female.

Discussion
Pilomatricomas represent 70% of all adnexal tumors in the first two decades of life and show a female preponderance (8,11). In a study done by Golpur M (11), the female-male ratio was 0.97:1. Duflo S et.al (9) reported majority of these cases in paediatric population where as Joel Juin Li Chan et.al (10) reported two out of three cases in adult males. Our discussion is regarding a rare presentation of pilomatricoma in a 55 yrs old female.

In a study done by Hernandes - Perez E (8) and Pirouzmanesh A et.al (12), the most common sites of occurrence were the neck (30.2 %), cheeks (16.8 %),
Our case is the rare one because of its unusual presentation in the digits.

Pilomatrixomas are derived from hair matrix cells. The cause is unknown. Recently, some genetic changes have been found in the affected hair cells - an overactive Proto-oncogene called BCL-2 suggests the normal process of cell death is suppressed and mutations in TNNBI suggest loss of regulation of a protein complex called beta-catenin/LEF(4).

Features that can obscure the diagnosis on presentation include telangiectasia, hyperkeratosis, hemosiderin deposition and erosion (2). Our patient presented with the nodule over the dorsum of 2nd toe with ulceration of the overlying skin. The classical histology is said to be defined by the presence of ghost or shadow cells and basophilic cells; however, calcification is also common(4). Differential diagnosis to consider with pilomatrixomas include sebaceous cyst, dermoid cyst, branchial cyst, preauricular cyst, ossifying hematomas, chondroma, lymphadenopathy, foreign body reaction, fibroxanthoma, giant cell tumor and osteoma cuits(12).

Attempts have also been made to use fine needle aspiration (FNA) to diagnose pilomatrixoma. Kumar and Verma (13) found that the presence of basoloid cells, ghost cells and foreign body giant cells are essential for a confident diagnosis of pilomatrixoma. Additional supporting features included calcification, naked nuclei and nucleated squamous cells. In our case FNAC of the lesion was done but it revealed only giant cells and calcification.

Pilomatrixoma can be diagnosed clinically and excisional biopsy is recommended for diagnosis and curative treatment. Because the recurrence rate after excision is low, pilomatrical carcinoma should be considered in the case of local recurrence (14).

Complications of pilomatrixoma are rare. However, occasionally they grow to giant size (several centimeters in diameter) and pilomatrical carcinoma has been very rarely reported (6).

References