CASE REPORT

Digital Pilomatrixoma An Unusual Site of Presentation

Archana Gupta, Sheetal Singh*

Abstract

Pilomatricoma is an uncommon, harmless, skin lesion derived from hair matrix cells. It is also spelled 'pilomatrixoma' and sometimes known as 'calcifing epithelioma of malherbe'. It is most often diagnosed in young children but may also affect adults. Single skin-coloured or purplish lesions arised 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 i.e digit (2nd toe left foot) and that too in 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, the overlying epidermis was keratinized. The underlying dermis showed solid nests of small basaloid cells and ghost cells (*Fig. 2*). Focal areas of calcification were also seen. The lesion was diagnosed as a case of pilomatricoma with its presentation at an unusual site - digits.

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%),

From the Department of Pathology, *Surgery, Acharya Shri Chander College of Medical Sciences & Hospital, Sidhra,Jammu-J&K
Correspondence to : Dr Archana Gupta, Department of Pathology, Acharya Shri Chander College of Medical Sciences & Hospital. Sidhra. Jammu

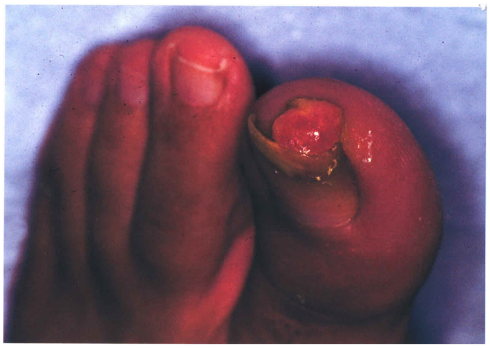

Fig.1 Swelling 2nd digit, left Foot with Overlying Skin ulcerated & Exhibited a Bluish Discoloration

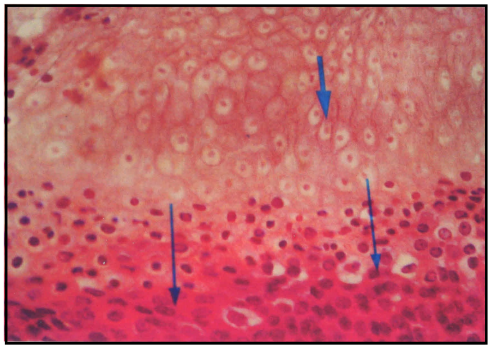

Fig.2 *Dermis Showing Solid Nests of Small Basaloid Cells and Ghost Cells*

scalp (16.2%) and brow and periorbital area (14.0%). Our case is the rare one because of its unusual presentation in the digits.

Pilomatricomas 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 to 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 pilomatricomas include sebaceous cyst, dermoid cyst, branchial cyst, preauricular cyst, ossifying hematoma, 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 pilomatricoma. Kumar and Verma (13) found that the presence of basaloid cells, ghost cells and foreign body giant cells are essential for a confident diagnosis of pilomatricoma. 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.

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

References

1. Malherbe A, Chenantais J. Note sur I' epithelioma calcifie des glands sebaces. *Progres Medical* 1880;8:826-8.
2. Kurokawa I, Yamanaka K-i, Senba Y, *et al*. Pilomatricoma can differentiate not only towards hair matrix and hair cortex, but also follicular infundibulum, outer root sheath and hair bulge. Experimental dermatology. *Exp Dermatol* 2009;18(8):734-7.
3. HassanienA, Glanz S. B-catenin expression in benign and malignant pilomatrix neoplasms. *Br J Dermatol* 2004;150(3):511-6.
4. Farrier S, Morgan M. bcl-2 Expression in pilomatricoma. *Am J Dermatopathol* 1997;19(3):257-7.
5. Julian CG, Bower PW. A clinical review of 209 pilomatricomas. *J Am Acad Dermatol* 1998;39(2):191-5.
6. Lan MY, Lan MC, Ho CY, Li WY, Lin CZ. Pilomatricoma of the head and neck: a retrospective review of 179 cases. *Arch Otolaryngol Head Neck Surg* 2003;129(12):1327-30.
7. Moehlenbeck FW. Pilomatrixoma (calcifying epithelioma): a statistical study. *Arch Dermatol* 1973;108(4):532-4.
8. Hernandez-Perez E, Cestoni-Parducci RF. Pilomatricoma (calcifying epithelioma): A study of 100 cases in El Salvador. *Int J Dermatol* 1981;20(7):491-4.
9. Duflo S, Nicollas R, Roman S, Magalon G, Triglia JM. Pilomatrixoma of the head and neck in children: a study of 38 cases and a review of the literature. *Arch Otalaryngol Head Neck Surg*1998;124(11):1239-42.
10. Joel Juin li Chan. Multiple pilomatricomas : Case presentation and review of the literature. *Dermatology Online Journal* 2010; (16): 4
11. Golpour M. Evaluation of characteristics of patients with pilomatricoma in Mazandaran province, 1996-2006. *Pak J Biol Sci* 2009;12(6):548-50.
12. Pirouzmanesh A, Refinish JF, Gonzalez-Gomez I, *et al*. Pilomatrixoma: a review of 346 cases. *Plast Reconstr Surg* 2003;112(7):1784-9.
13. Kumar N, Verma K. Fine needle aspiration (FNA) cytology of pilomatrixoma. *Cytopathology* 1996;7(2):125-31.
14. Taaffe A, Wyatt EH, Bury HP. Pilomatricoma (Malherbe). A clinical and histopathologic survey of 78 cases. *Int J Dermatol* 1988:27(7):477-80.