



PILOMATRICOMA OF THE THIGH: A RARE SITE OF PRESENTATION

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ABSTRACT

Pilomatricoma is a benign appendageal tumor of hair matrix also known as Malherbes calcified epithelioma. A 20 year old female presented with a solitary, bluish nodule measuring one x one cm in the antero-medial aspect of left thigh since one year. Excision biopsy of the nodule was done which on histopathological examination revealed features of pilomatricoma. The sections were subjected to IHC for beta catenin and bcl2 which were non expressive. Although this tumor is most commonly seen in head, neck and upper extremities, it can rarely arise in lower limbs and thus should be carefully evaluated.

KEYWORDS: Pilomatricoma, shadow cells, thigh, unusual site.

INTRODUCTION

Pilomatricoma is a benign appendageal tumor arising from the matrix cells of the hair follicle.^[1] It is rare and comprises 0.2% of all routine dermatopathological specimens.^[2] It was first ever described by Malherbe and Chenantisback in 1880 and therefore came to be known as Malherbes calcified epithelioma. Though it can occur at any age, it is usually seen in the first 2 decades of life and more frequently in women. The common locations of occurrence include head, neck and upper extremities.^[1,3]

Pilomatricomas present as a cystic or firm nodule that is slow growing and is often misdiagnosed as a sebaceous cyst, foreign body reaction, soft tissue tumor or epidermal inclusion cyst.^[2,4] It is known to be associated with tuberous sclerosis and high levels of beta-catenin caused by either a mutation in the APC gene or beta-catenin gene.^[5,6]

We present a case of pilomatricoma of thigh which is an uncommon location of its presentation.

CASE REPORT

A 20 year old female presented with a solitary nodule in the antero-medial aspect of left thigh since one year. She developed pain in the same over the past one month. There was no history of trauma, bleeding or discharge from the swelling.

Local examination revealed a subcutaneous, tender nodule measuring one x one cm with bluish discoloration of the overlying skin. A clinical diagnosis of sebaceous cyst was made. Excision biopsy of the nodule was done and sent for histopathological examination.

Macroscopic examination showed a nodule measuring one x one cm. The cut surface was homogenous, grey white. Microscopic examination of the lesion revealed fragments of stratified squamous epithelium with hyperkeratosis and dual population of cells comprising predominantly of ghost or shadow cells in the centre along with a very few peripheral basaloid cells. There were areas of mild chronic inflammation and fibrosis. The histological features were consistent with pilomatricoma.

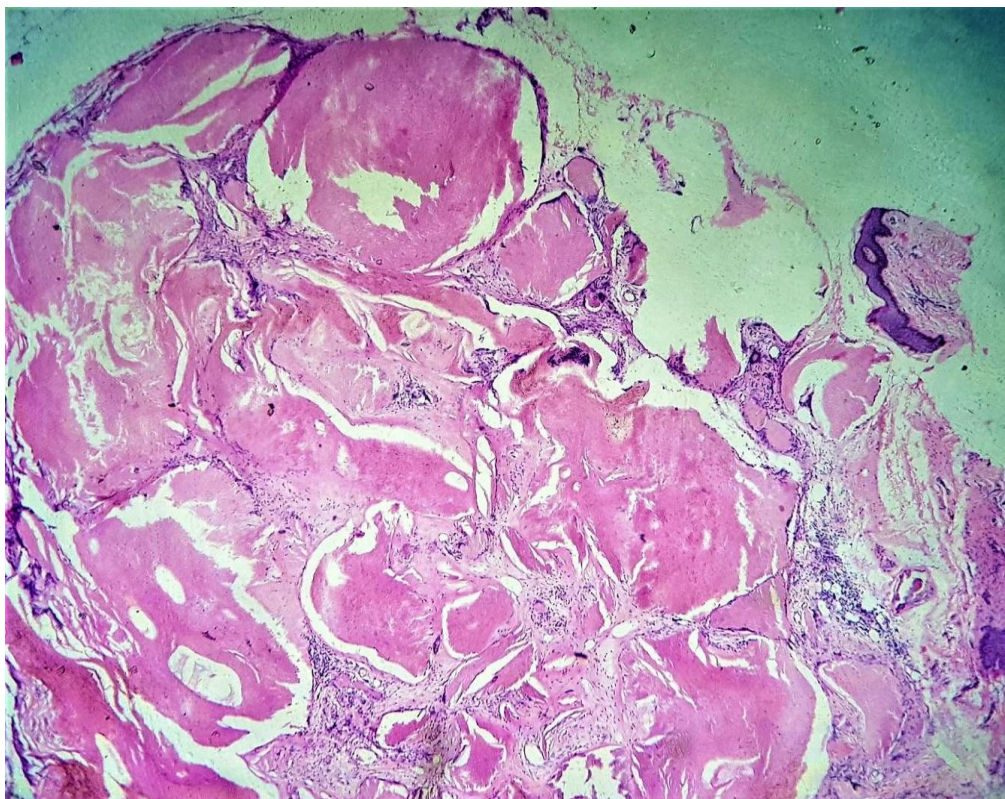


Figure 1: Microsection shows a lobulated lesion in the dermis with characteristic shadow/ghost cells (H &E, 40X).

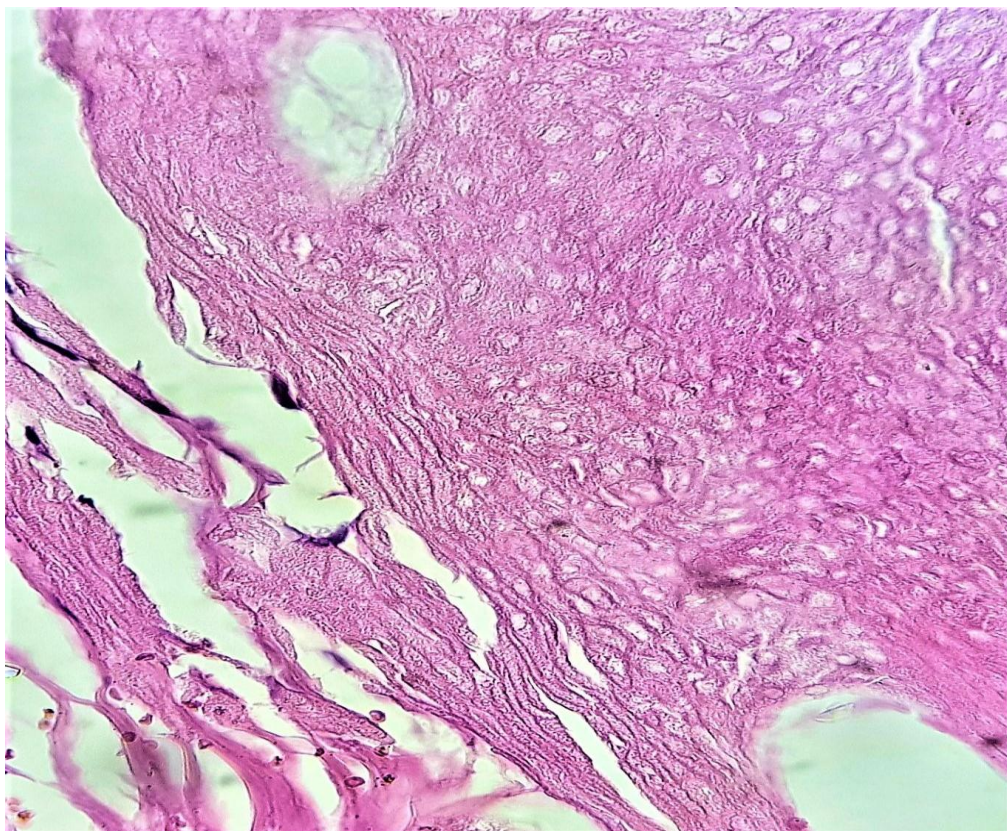


Figure 2: Microsection show ghost cells with keratinization (H & E, 400X).

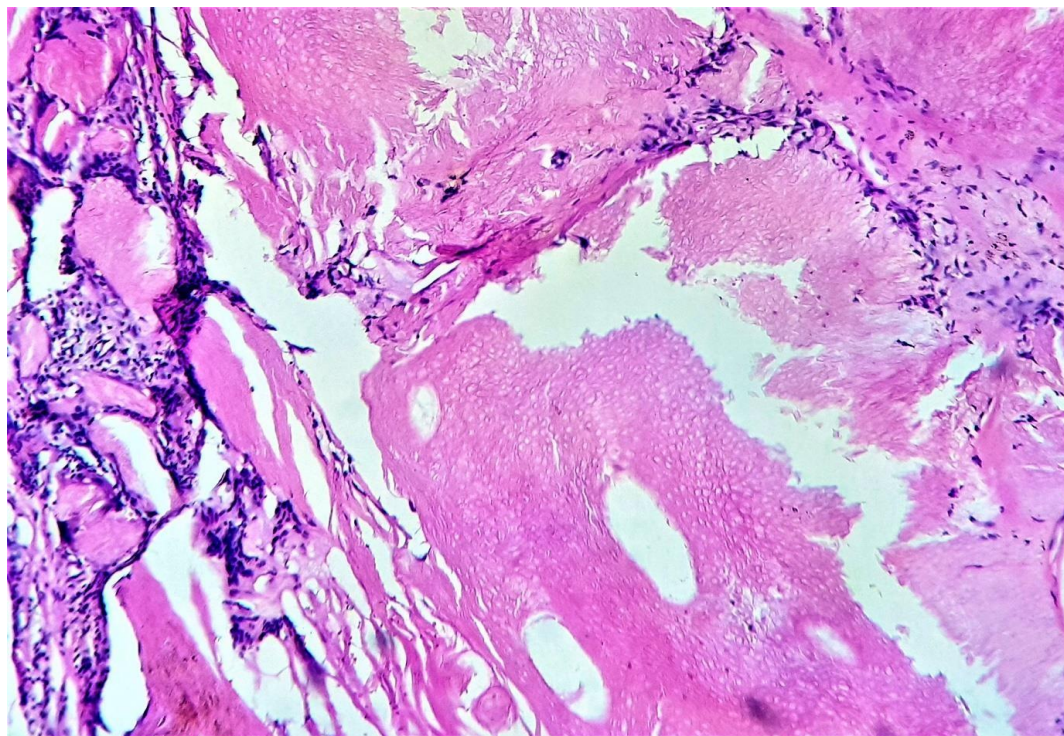


Figure 3: Microsection shows pilomatricoma with eosinophilic, enucleated shadow cells, very few basophilic cells with lymphocytic infiltration (H & E, 100X).

Immunohistochemistry for beta catenin and bcl2 were also performed, which were non- expressive.

DISCUSSION

The Incidence of Pilomatricoma is reported to be 0.1 to 0.3%.^[1] It occurs as a solitary, deep dermal subcutaneous nodule with blue- red overlying skin. They are generally located in the head and neck region(40-77%) followed by trunk and upper extremities. Their occurrence in the lower extremities is very rare. As these tumors are unusual in the lower limb, they need to be differentiated from sebaceous cyst, tuberculoma, hemangioma, keloid, dermoid cyst, foreign body etc. Trauma and infection are thought to be possible triggers of this tumor however the etiology remains unknown.^[1,3]

Microscopically, these lesions typically comprise of basaloid cells, transitional cells, shadow cells, amorphous debris and areas of calcification.^[7] Calcification is reported to be present in 80% of the cases. It is known that basaloid cells transform into shadow cells via transitional cells with keratolytic changes.^[8,9]

Investigative procedures that can be carried out are FNAC, ultrasonography, X-ray, CT and MRI. The final diagnosis requires histopathological confirmation before surgical excision.

The mode of treatment is complete surgical excision along with one to two cm of surrounding tissue, however, recurrences can occur after excision.^[3] The rate of local recurrence is reported as zero to three percent.^[1]

In most of the studies, the tumors are solitary and less than two cm in diameter. Our patient also presented with a solitary tumor of one cm size.

Pigmented and giant forms have also been reported in the literature.^[5,10] Multiple pilomatricoma have been documented in association with genetic disorders like myotonic dystrophy, gardener syndrome, Xeroderma pigmentosa and basal cell nevus syndrome.^[6] Malignant transformation is rarely observed in these tumors.^[3]

Beta-catenin is a subunit of cadherin protein complex that acts as an important regulator of development and differentiation of hair follicle. It is also responsible for adhesion between epithelial layers and among the cells. It has recently been shown to play an important role in the formation of hair follicle derived tumors, including pilomatricoma.^[5,9] IHC studies have demonstrated this association, where basaloid cells exhibited strong nuclear and membranous staining while transitional cells and shadow cells did not express beta- catenin at all.^[9] Our case was negative for the expression of beta catenin because of lack of basaloid cells and predominance of shadow cells.

The anagen, catagen and telogen cycle of normal hair growth are regulated by apoptosis. BCL2 is a proto-oncogene that suppresses apoptosis in benign and malignant lesions. Faulty suppression of apoptosis contributes to pathogenesis of these tumors.^[6,8] In a study of ten cases of pilomatricoma, all were found to be positive for bcl2.^[6] However, our case gave a negative result with bcl2.

CONCLUSION

Our aim of presentation of this case was to emphasize on the unusual location of pilomatricoma which warrants evaluation of such lesions even in the lower extremities and to demonstrate the correlation between expression of bcl2 and beta-catenin in these lesions. Our case did not show overexpression of bcl2 and beta-catenin because of predominance of shadow cells. The pattern and intracellular localization of beta catenin is still controversial and is a topic of interest for research.

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