Pilomatricoma at Uncommon Site: A Case Report
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Case Report
Subject: Pathology

Abstract:
Pilomatricoma is benign skin adnexal tumour arising from the hair follicle matrix. Its most common location is maxillofacial region followed by upper extremities. Lower extremities are a rare site for this tumour. It presents as a skin nodule and rarely exceeds 5 cm in diameter. We are thus presenting an unusual case of pilomatricoma on thigh of a young male which was clinically thought to be a malignancy due to its larger size and rapid growth.

Keywords: Pilomatricoma, Lower Extremity, Benign.

Introduction:
Malherbe and Chenantais first described Pilomatrixoma, or calcifying epithelioma of Malherbe in 1880 [1,2]. It is a benign tumor originating from the matrix cells of the hair follicles [3] and is classified as a benign skin appendage tumor belonging to the group of suborganoid tumors with hair differentiation [4, 5]. The term pilomatrixoma was introduced by Forbis and Helwing in 1961 [6], in reference to its hair cell-derived lesion which was later changed to pilomatricoma to be more correct etymologically [7].
Pilomatricoma may be located in any part of the body except the palms and soles, and shows a predilection for the maxillofacial region [5]. According to most authors head, neck and upper extremities are more commonly involved [1, 3, 8].

A study done by Kwon et al concluded that head and neck tumors predominate with 70% of all pilomatrixomas, followed by the upper extremity with 22% while other locations include the hair-bearing back, chest and lower extremities [9]. Hence it can be made out that lower extremity is a fairly uncommon location for pilomatricoma.

Case Report:
We received a skin nodule in our histopathology laboratory which was excised from thigh of a 25 year old male patient. Clinically the nodule was 6cm in diameter, which had rapidly increased in size in 2 months. There was no significant history like trauma or similar nodule anywhere else or in any family member. Hence a clinical suspicion of malignancy was made. Grossly the nodule was skin covered, firm and well circumscribed. Sections were taken including the margins, keeping in mind the clinical suspicion of malignancy.

Histopathological examination showed a well delineated subepidermal tumor with solid nests of basaloid cells undergoing abrupt trichilemmal-type keratinization (Figures 1 and 2). Ghost cells were seen with calcification. Foreign body type of giant cell reaction was seen at places. There was no evidence of any atypia or abnormal mitotic figures. Hence a final diagnosis of pilomatricoma was made, without any evidence of malignant transformation. The patient was kept in follow up and there was no evidence of any recurrence or any similar swelling anywhere else.
Discussion:

Pilomatricoma is a slow growing benign tumor arising from hair follicles [1, 5]. The median age at presentation is generally less than 30 years [10, 11] but according to few authors it has a bimodal age distribution with first peak in first and second peak in sixth decade [1, 5]. Almost all the previous studies indicate a slight female preponderance [1, 3, 9, 10, 11, 12] however in a study conducted by Guinot-Moya et al there was a slight male preponderance [5]. Our patient was a 25 year old male which is in accordance to other studies although its size had increased quite rapidly over a small period of time which is quite contradictory to common belief.

Pilomatricoma presents as a skin nodule and according to most authors its size ranges from 0.5-5 cm. Usually it is asymptomatic and the overlying skin is unremarkable [10], while some authors also report secondary changes like hyperemia or ulceration [1, 5]. In our case the lesion was slightly larger (6cm) than generally reported in earlier studies.

All the old literature reveals the following decreasing order of site of occurrence of pilomatricoma: head and neck, upper extremities, hair bearing back, trunk and lower extremities [1, 3, 5, 8, 9, 10, 11, 12]. Hence lower extremities are a very uncommon site for pilomatricoma. Moreover, according to Guo et al, lesions with average size 1.22 cm were commonly located in head and neck and upper extremities [11]. Our case was quite unusual as not only was the lesion present on thigh but also its size was quite big at presentation.

Trauma has been implicated as an etiological factor [3, 5, 10] while familial preponderance is also reported [5, 10, 12]. Few also reported incidence of multiple pilomatricomas [5, 12]. Pilomatricoma can be associated with other diseases such as myotonic dystrophy, Gardner syndrome, Steinert’s disease, Turner syndrome and sarcoidosis [1, 9, 10, and 13]. In our patient there was no history of trauma. Nor was there any history of familial predisposition or multiple lesions or any associated syndrome.

Pilomatricomas are quite commonly misdiagnosed clinically with some authors reporting diagnostic accuracy rates of 0-30% while others reporting rates up to 54.4% [1, 3, 10, 11]. The lesion in our case was also clinically misdiagnosed as malignancy, mainly because there was history of rapid increase in size and the lesion was 6 cm at presentation.

Surgical excision is generally curative with negligible relapse rates [1, 5, 9, 14]. The lesion of our patient was surgically removed without any recurrence after 6 months.

Conclusion:

It can be concluded that pilomatricoma may present as an unusually large size nodule even in the lower extremities. Since it is often misdiagnosed clinically and it’s a common benign neoplasm of skin it should always be kept in differential diagnosis while dealing with skin nodule biopsies from relevant sites.
References:


