Pilomatricoma - Calcifying Epithelioma of Malherbe: Case Report and Review of the Literature

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SUMMARY

A case of pilomatricoma is described in a 13-year old girl who presented with a small stony hard mass on her right thigh. Malignant change can occur, therefore such lesion should be removed completely.

INTRODUCTION

Cutaneous calcification and ossification occurs in association with a wide variety of conditions, including inflammatory processes, neoplasms, scarring and metabolic conditions. The most common cutaneous neoplasm with ossification is the pilomatricoma.

CASE REPORT

A 13-year old girl presented to Skin Outpatient Department, Shaikh Zayed Hospital in February 1999 with a small mass on middle of lateral aspect of her right thigh for the last 6 years. It was slowly growing and painful on pressure. There was no history of trauma at the site of the lesion, neither there was any history of bleeding or discharge from the mass. Systemic review was unremarkable. Cutaneous examination revealed a mobile, tender, subcutaneous nodule measuring 1.5 cm x 0.5 cm in size, with stony hard consistency, in the middle of the lateral aspect of her right thigh. The overlying skin was normal. Regional lymph nodes were not palpable. Provisional diagnosis of calcinosis cutis was made. Excision biopsy of nodule was done and submitted for histopathological examination.

Macroscopic examination of the serial sections of 1.5x0.5 cm nodule showed multiple greyish white solid areas.

Microscopic examination of the lesion revealed the keratinized, stratified squamous skin with features of hyper and parakeratosis. There was dense fibrosis with mild chronic inflammatory cells infiltrate in the upper dermis. The deeper dermis and subcutaneous tissue revealed circumscribed nodule (Fig. 1) composed of extensive areas of calcification within cystic spaces (Fig. 2) surrounded by nests of ghost cells (Figs. 2, 3) foamy macrophages, few plasma cells, lymphocytes and multinucleated foreign body type of giant cells (Figs. 1, 3). Keratin flakes were present. Areas of fibrosis and hyalinization were also seen. There was no evidence of granuloma formation or malignant change. These features were consistent with a diagnosis of pilomatricoma.

DISCUSSION

Calcified lesions in the skin have been noted since ancient times. Galen, before 200 A.D, described stones in some tumors, as did Ambrosie Pare in 1585. A more precise description was given by Wilcken's in 1856. However, the first complete work, based on a series of patients, was published by Malherbe and Chenantais in 1880. They described calcifying epitheliomas, initially thought to be tumors of sebaceous glands. Malherbe himself in 1905 corrected this view. The term pilomatrixcoma, to denote origin from hair matrix cells, was suggested by Forbis and Helwig in 1961. This was later corrected to pilomatricoma. These tumors have a wide variety of signs, which often cause misdiagnosis. These are rare but still the
Fig. 1 Circumscribed nodules in dermis. Chronic inflammatory infiltrate and multinucleated foreign body type of giant cells.

Fig. 2 Areas of calcification within cystic spaces, surrounded by nests of ghost cells.

Fig. 3 Chronic inflammatory infiltrate, multinucleated foreign body type of giant cells and ghost cells.

Most common benign tumors of hair matrix. These accounted for one in 500 histologic specimen taken from 209 cases collected during 20 year period. Pilomatrixoma occurs at any age from infancy and is frequently seen in children. The majority of the patients are under 20 years of age and females are affected more often than males. It is not hereditary but a number of familial cases are recorded. It usually presents as a solitary lesion but on occasions multiple tumors are evident as part of an autosomal dominant inherited disorder. Rarely it may represent a dermatological marker of systemic disease e.g. dystrophia myotonica or Gardner's syndrome. The tumor presents as a slowly growing, firm to hard nodule on head, upper limbs, neck, trunk or lower limbs. The skin over the tumor is normal and tumor has a lobular shape on palpation. In adults there may be quite short history and there is usually no evidence of a preceding cyst. It may be subjected to periodic inflammation and on occasions presents as a granulomatous swelling. Calcification is seen in 80% of lesions. Chalky deposits are sometimes evident clinically or may be revealed by radiology. Malignant change is extremely rare. It arises chiefly in large lesions that have been present for many years. It occurs more often in middle aged men in the regions of head and neck, rarely on the eyelid. Pilomatrix carcinoma can metastasize to the lungs, bone and viscera with a subsequent poor outcome.

The anagen, catagen and telogen cycles of normal hair growth are regulated by programmed
cell death (apoptosis). bcl-2 is a proto-oncogene that helps to suppress apoptosis in both benign and malignant tumors. Both apoptosis and bcl-2 are critical factors in normal hair follicle development. In one series of 10 histologically proven cases of pilomatricomas, bcl-2 expression was studied by immunohistochemical methods. All of the cases were strongly decorated by bcl-2 immunostaining and faulty suppression of apoptosis was attributed to the pathogenesis of pilomatricomas.

As regard its pathological aspect, it is situated in the dermis and is composed of well-circumscribed, rounded islands giving a lobulated contour. The outer cells are small and their rounded nuclei crowded together make this region deep basophilic. Mitotic figures can usually be seen. The cytoplasm is scanty and the cell margins indistinct. Towards the center of the mass, the cytoplasm becomes more abundant and eosinophilic. The nuclear outline persists, but the chromatin is sparse and clumped in dark granules, then, all the basophilic material disappears, leaving a "Ghost cell". Melanin may be present. The stroma that encapsulates the masses usually contains inflammatory and foreign body cells and occasionally may ossify. Rarely malignant change can occur.

In one study after histopathological examination of 118 lesions of 116 patients with pilomatricoma, four distinct chronological stages were categorized: early, fully developed, early regressive and late regressive. Early lesions were small cystic structures lined by squamoid and basaloid epithelium containing keratin filaments and faulty hair matrix material composed of shadow cells. Fully developed lesions were large neoplasms lined by basaloid epithelium at their periphery and within, composed of regularly shaped, densely packed zones of cornified masses containing shadow cells. Early regressive lesions had no apparent epithelial lining but did have basaloid cell foci at the periphery; within, they were composed of pink hair matrix material with shadow cells surrounded by granulation tissue with inflammatory infiltrates and multinucleated histiocytic giant cells. Late regressive lesions had no epithelial component and were composed of irregularly shaped, partially confluent masses of faulty hair material and calcified or ossified shadow cells embedded in a desmoplastic stroma with little or no inflammatory infiltrate. Thus the lesion begins as an infundibular matrix cyst and ends up as a calcified and ossified nodule with no visible epithelial component.

In one study two patients with subcutaneous nodules (later histologically proved to be pilomatricoma) on forearms underwent imaging tests. Plain films showed non-specific well circumscribed lesions. Ultrasonography revealed nodular, well circumscribed hyperechoic lesion in one patient. In both cases spin-echo (SE) T1-weighted images (TiWI) showed homogeneous, intermediate signal intensity (SI). On gadolinium-enhanced TiWI (1 patient), no enhancement was observed. Both lesions showed predominant low to intermediate SI on T2WI.

**Treatment**

Local excision is required for benign lesions. Wider excision will be needed if malignancy is suspected. Role of radiotherapy and chemotherapy in the treatment of disseminated disease is unclear. Our case highlights the fact that any cutaneous lesion with similar clinical features should be suspected as pilomatricoma and diagnosed histopathologically after complete removal to avoid subsequent serious consequences.

**REFERENCES**


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