Case Report

Cytomorphological diagnosis of pilomatricoma in an unusual location

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Abstract

Pilomatricomaisa benign skin adnexal tumor that is derived from hair matrix. Pilomatricomas usually present in children and adolescents but can occur at any age. Pilomatricomas are generally asymptomatic and found mainly in the head and neck area. Upper extremities not a common site for this lesion. It israrely identified on the chest, trunk, or lower extremities. They areeasily diagnosed on histology due to their typical histological features but diagnosis in cytology is generally difficult as the features may mimic other skin lesions and leads to misdiagnosis.

Here we describe a case of pilomatricoma of left arm in a 25 year old male, which was diagnosed on cytology. The cytological smears were cellular and consist of aggregates of anucleate squames, basaloid cells and shadow cells. Subsequent histopathologyof the excised lesion confirmed the diagnosis of Pilomatricoma. Through thisreport we highlight the cytomorphological features that helps us in arriving the correct diagnosis of Pilomatricomaon Fine needle aspiration cytology (FNAC) smears.

Keywords: FNAC, Ghost cells, Nodule, Pilomatricoma

Introduction

Pilomatricomaisan uncommon benign tumourof hair follicle that is derived from hair matrix [1]. Most commonly pilomatricoma affects children and adolescents but they can be diagnosed in adults also. They areslightly more common in females than males.

Pilomatricoma most commonly occur in head and neck region, may occur in upper extremity but rare on chest, trunk, and lower extremities [2]. Mostly theyare asymptomatic and usually present as solitary, skin colored to purplish, firm, subcutaneous nodule for which the patient seeks attention [3,4]. Here wedescribe a case of pilomatricoma of left arm in a 25-year-old male, which was diagnosed on Fine needle aspiration cytology and subsequent histopathology of the excised lesion confirmed the diagnosis of Pilomatricoma. Through thisreport we highlight the cytomorphological features that helps us in arriving the correct diagnosis of Pilomatricomaon Fine needle aspiration cytology (FNAC) smears**.**

Case Report

A 25year young adult male came to surgery OPD with the complain of swelling in the left forearm for 3 months. Swelling wasnot associated with pain or fever. There was no history of any associated trauma. On clinical examination the swelling was about 1x1cm, firm to hard in consistency and non-tender. Overlying skin was purplish-red in color. FNAC was performed with a 22G needle and thick grayish material aspirated was smeared on the slide. Both thewet and dry smear was made and subjected to staining. Microscopic examinationof smear revealed richly cellular smears showing clusters and sheets of basaloid cells, ghost cells along with foci of calcification and foreign body giant cells. The basaloid cells show mild pleomorphism, over lapping, scant cytoplasm having regular round to oval nuclei.

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Case Report

Background shows keratinous debris and anucleate squames. These features strongly suggested diagnosis of Pilomatricoma. Later on, excision biopsy specimen was received. Grossly thespecimen was 1.5x1.5x1.2 cm. On cut shows homogenous chalky-white area. Specimen was processed and microscopic examination revealed characteristics features of Pilomatricoma.

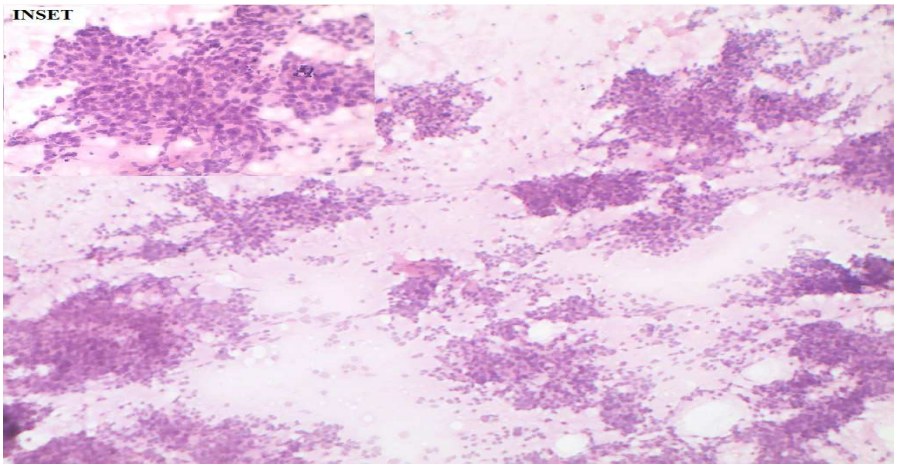

Figure-1: Photograph showing many clusters of basaloid cells with keratinous debris and squames in the background (H&E stain, 4x), INSET : Clusters of basaloid cells (H&E stain,40x)

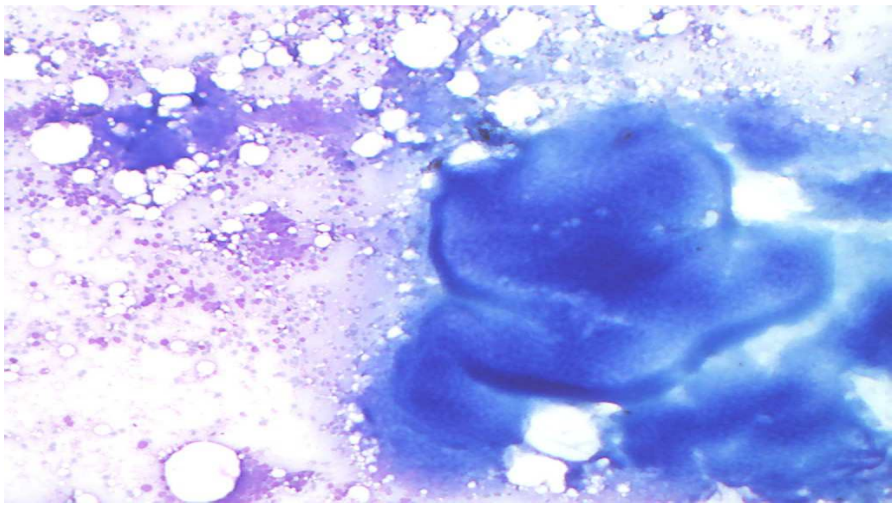

Figure-2: Photograph showing clusters and scattered population of basaloid cells, ghost cells and inflammatory cells in the background. (Giemsa stain,4x)

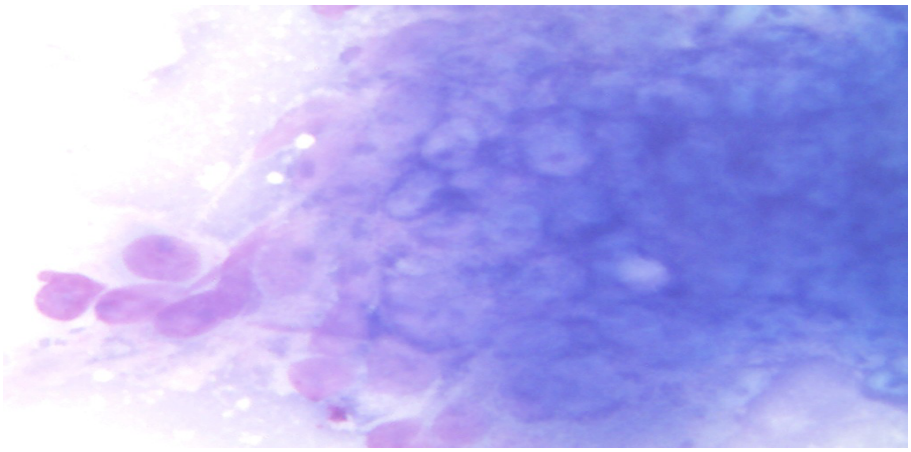

Figure-3: Photograph showing clusters of ghost cells. (Giemsa stain ,40x)

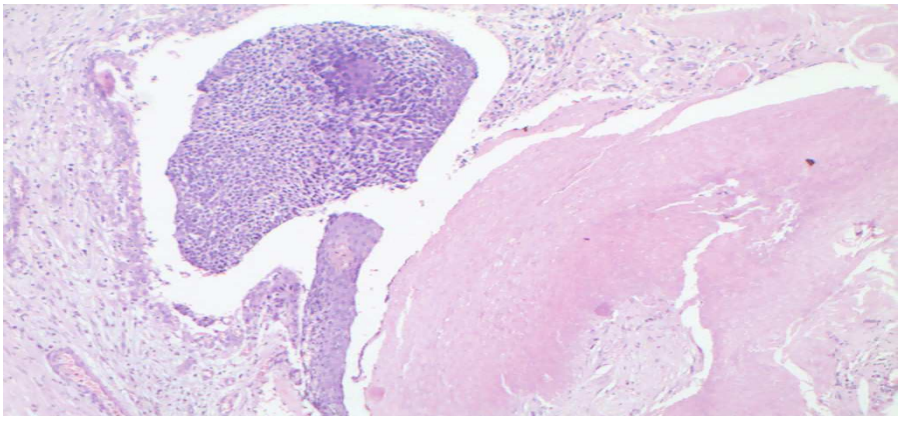

Figure-4: Photograph showing island of basaloid cells and eosinophilic shadow cells (H&E stain -4x)

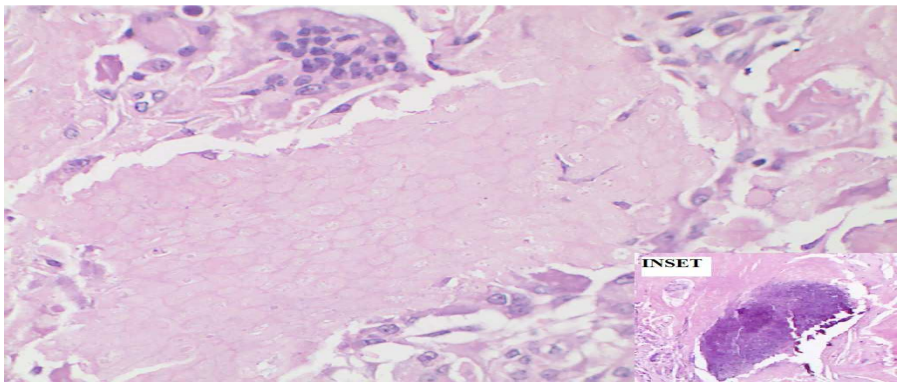

Figure-5: Photograph showing sheets of eosinophilic shadow cells along with foreign body giant cells (H&E stain– 40x) ; INSET : Foci of calcification (H&E stain,10X)

Discussion

Pilomatricomais anuncommon benign skin adnexal condition that is known to be derived from hair follicular cells [1]. It isalso known as Calcifying epithelioma of Malherbe as it was first described by Malherbe and Chenantaisin 1880 as a benign neoplasm of sebaceous gland origin [5]. It was in 1961 when Forbis and Helwigcoined the term pilomatrixoma to better suit its histology as cells of origin were the outer root sheath cells of the hair follicle and thus, avoiding the term epithelioma which leads to intimation of malignancy [1].

The patho physiology behind Pilomatricomaisfaulty suppression of apoptosis that contributes to the pathogenesis of these tumors.This isevidenced by in a study of 10 pilomatrixoma lesions, in which bcl2 immunostaining results were strongly positive [6].

Pilomatricoma formationre presents a disturbance of the hair follicle cycle in which limited cytologic differentiation of pilarkeratinocytes occur but further development into mature hair fails to take place [7]. In ourcase the patient presented as a nodule on the left forearm which was not the common site as seen from the previous literatures that most common location of the Pilomatricomais head and neck region [2]. The lesionwas a solitary firm purplish nodule which are the common presentation of Pilomatricomaas seen from the previous studies [3,4]. Fine needle aspiration cytology of the lesion was done. Cytology smearswere cellular and revealedsheets and aggregates of basaloid cells and shadow cells along with foci of calcification,foreign body giant cells, anucleatesquames. Later excisional biopsy of the lesion was performed. At lowpower the histological pattern usually seen in pilomatricomais of a well-circumscribed nodulo-cystic tumor. While predominantly seen within the lower dermis, extension into the subcutaneous tissue is not uncommon. In our case histological section of the lesion revealed characteristic histological findings of Pilomatricoma. Prominent features were a circum scribed lesion, nests of basaloid cells, eosinophilic shadow cells (ghost cells) along with mixed inflammatory cells, foci of calcification and foreign body giant cells in the intervening connective tissue stroma that confirmed our cytological diagnosis of Pilomatricoma.

As seenin our case most of the component of cytological features of the Pilomatricoma were present so we were able to diagnose it as Pilomatricomawhich was later confirmed on Histology. But itis not easy to diagnose in all the case as predominance of one component over other leads to erroneous diagnosis with multiple differentials. Depending upon thecomponent the lesion may be mistaken for trichilemmal cyst, epidermal inclusion cyst, benign appendageal tumour (like eccrine spiroadenoma, cylindroma, Hidrea-denoma), granulomatous lesions, squamous and basal cell carcinoma, lymphomas, small round blue cell tumor, foreign bodies [8-12]. Differentiation isbased mainly on the type of predominant component. If thereis predominance of squamous cell component with paucity of basaloid cells, then Epidermal inclusion cyst may be mis interpreted as it is also superficially located and smears show sheets and scattered population of anucleate and nucleate squames. If thecyst wall ruptures, then there may be foreign body granulomatous response along with inflammatory cells [13,14].

Similarly, predominance of basaloid cell component may lead to misdiagnosis of skin appendageal tumors, such as cylindroma, eccrine spiradenoma and hidradenoma. As smearsfrom these lesions contain mainly basaloid cells in cohesive, smoothly contoured groups in contrast to the typically irregular, saw-toothed edges of the cohesive to loosely cohesive monolayer sheets of basaloid cells seen in Pilomatricoma.

Shadow cells, mature nucleated squamous cells and multinucleated giant cells are rare to absent. Similarly, highcellular yield of basaloid cells, the presence of small primitive-appearing cells with a high nuclear-cytoplasmic ratio, prominent nucleoli in a background rich in debris and inflammatory cells may be mistaken for malignancy [14].

As Pilomatricoma itself it is a rare condition and chances of conversion to Pilomatricoma cancer is also very rare, so definite management of the lesion is wide local surgical excision. Rarity ofthe lesion provides little evidence of follow up recommendation, however study suggests overall recurrence rate after excision is 2.6% [15,16].

Conclusion

1. Pilomatricomaalthoughvery rare condition, it should be kept in mind by the clinicians and pathologists not only for nodules located in the head and neck region but also for nodules located in the upper extremities as seen in our case and other rarer sites.
2. Adequate sampling of the lesion from the FNAC and complete spectrum of cytological findings helps in accurate diagnosis of the lesion.
3. Excisional biopsyof the lesion further confirms the lesion due to its characteristic morphology.
4. In thisstudy we have highlightened the cytological features of the pilomatricomanodule located on the left forearm and also how to differentiate it from other mimicking lesions.

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