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A Rare Case Report on Pilomatrixoma of the Arm Diagnosed Cytologically

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Introduction - *Pilomatrixoma is a benign neoplasm derived from hair follicle matrix cells, is seen commonly in head and neck. Occurrence of pilomatrixoma in the upper extremities is not common and has been reported irequently in the available literature.*

Case Report - *We present the case of an eighteen year-old girl underwent fine needle aspiration (FNA) of a firm subcutaneous nodule on the lateral aspect of right arm. FNA smears shows clusters as well as dispersed population of round to oval cells along with few multinucleated giant cells. Few ghost cells were seen. A cytologic impression of pilomatrixoma was rendered, which was confirmed on histopathology.*

Conclusion - *Pilomatrixoma should be considered in the cytologic differential diagnoses of subcutaneous masses even in unusual locations like arm.*

Key words – *Pilomatrixoma, benign, FNA*

INTRODUCTION –

Pilomatrixoma, also known as pilomatricoma or calcifying epithelioma of Malherbe, is a benign neoplasm that derives from hair follicle matrix cells. It is most often seen in children and the most common anatomical location is the head and neck region[1]. Occurrence of this lesion in the arm is unusual and has been described in few reports in the available English literature[2-5]. It presents as a solitary, slow growing dermal or subcutaneous nodule and is rarely diagnosed clinically. Though histologic diagnosis of pilomatrixoma, even in

unusual locations, is straight forward, the same is not true for aspiration cytology. One case of pilomatrixoma of the arm was diagnosed as round cell tumour on cytology. The final diagnosis was rendered on histopathology of the resected mass[4].

We present a case of pilomatrixoma in the arm of a young girl.

CASE REPORT

An 18 year old girl presented to the surgical outpatient department with a nodular swelling over right arm. She had noted the mass one and half year before initial presentation which is gradually increasing in size for the last 3-4 months. There was no history of trauma, fever, chills, weight loss, fatigue, numbness, or tingling. Local examination revealed a firm to hard, well circumscribed nodular, tender, mobile subcutaneous swelling, ms 1x2 cm in size on the lateral aspect of the right arm. The overlying skin was normal in appearance. Plain radiographs were unremarkable.

We received requisition for cytological examination and fine needle aspiration (FNA) was performed from the swelling. FNA was performed using 22G needle and 10mL syringe. The smears were air dried and stained with Giemsa stain.

FNA smears were cellular showing presence of dispersed as well as tight cohesive clusters of basaloid cells which are round to oval cells having bland nuclei with scant to moderate amount of pale blue cytoplasm (Fig 1). Clusters of ghost cells were also present (Fig 2). Admixed multinucleated giant cells were also noted (Fig 3). Foci of necrosis and calcium granules were identified in the smears examined. A cytologic diagnosis of pilomatrixoma was rendered and excision biopsy advised.

Following the cytologic impression, an excision of the mass was carried out and specimen was received for histopathological examination. Grossly, grey white soft tissue mass ms 3x2 cm in size was received. Microscopically, section showed presence of ghost cells and clusters of basaloid cells along with foci of calcification and few giant cells (Fig 4&5). Histological diagnosis of pilomatrixoma was made. So, cytological diagnosis was correlated well with histopathological diagnosis.

DISCUSSION

Pilomatricomas are of ectodermal origin and arise from the outer root sheath cell of the hair follicle[6]. Calcifying epithelioma was originally described in 1880 by Malherbe and Chenanlais as a neoplasm of sebaceous glands[7]. The term pilomatrixoma was introduced based on further studies performed in 1961 by Forbis and Helwig who demonstrated that the cells differentiated in the direction of cortical cells of the hair follicle[8]. Although pilomatricoma can develop in patients of any age, it occurs most often in children and young adults and they are noted more commonly in females[9]. Pilomatrixoma is typically found in head and neck region, though it has been reported in upper extremities and other sites. In a large series of 346 pilomatrixomas, about 15.3% were seen in upper extremities[1].

A previously reported case of pilomatrixoma of the arm was diagnosed cytologically as blue round cell tumor due to the presence of round to ovoid cells with occasional rosette-like appearance. Histopathology in this case showed features of pilomatrixoma. The authors suggested that early rapidly growing lesions, composed predominantly of basaloid cells, may lead to over diagnosis of malignancy[4].

A rare malignant counterpart, pilomatrix carcinoma, has been described, and approximately 90 cases have been reported in the literature. It is locally aggressive and can recur. In several cases, it has demonstrated metastases. Many key features are similar between these benign and malignant counterparts; the primary differentiating characteristics include a high mitotic rate with atypical mitoses, central necrosis, infiltration of the skin and soft tissue, and invasion of blood and lymphatic vessels[10,11].

Although pilomatrixomas are usually solitary, multiple lesions have been reported in association with genetic disorders, such as myotonic dystrophy, Gardner syndrome, xeroderma pigmentosum, and basal cell nevus syndrome[12, 13].

Diagnostic tests and Imaging studies are often unnecessary in the work up of a superficial benign lesion like pilomatrixoma however tests are done to exclude malignancy or to determine the depth of lesion. Fink and Berkowitz found ultrasound to be helpful in children[14].

The differential diagnosis of these lesions should include dermoid, epidermoid cysts, metaplastic bone formation, foreign body reaction, sebaceous adenoma or carcinoma, trichoepithelioma and basal cell epithelioma, juvenile xanthogranuloma, capillary hemangioma, and rhabdomyosarcoma[1, 15].

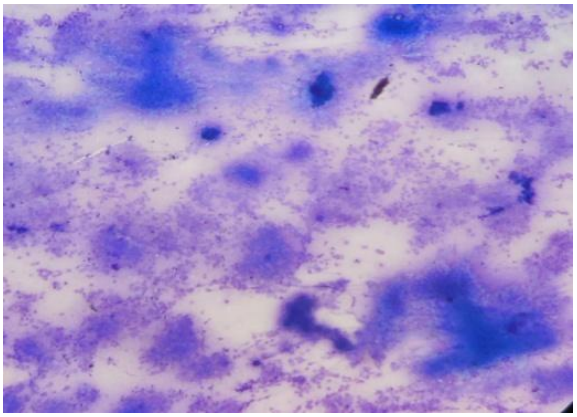

FIG 1: Clusters Of Tight Cohesive Basaloid Cell With Scant Cytoplasm And Presence Of Ghost Cells (Mgg, 10x)

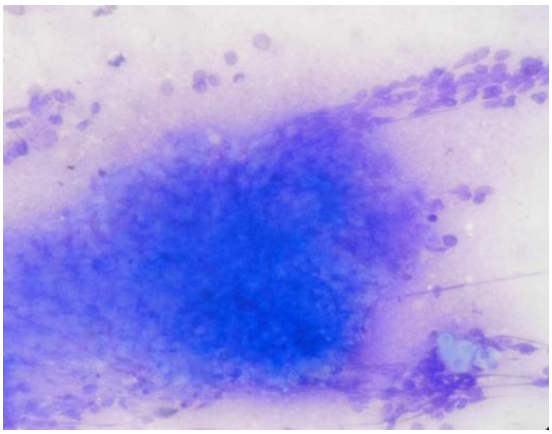

FIG 2: Ghost Cells Cluster And Few Dispersed Basaloid Cells (Mgg, 40x)

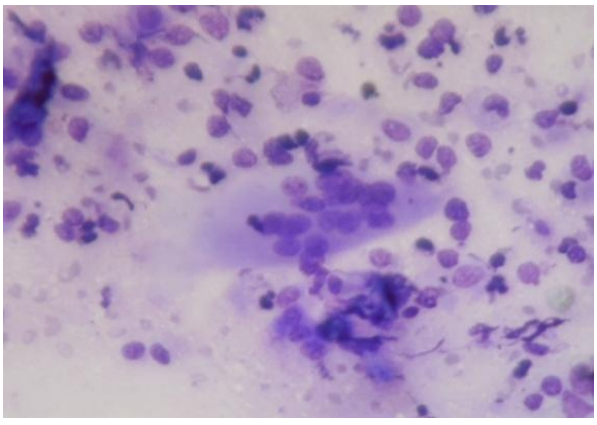

FIG 3: Smear Showing Presence Of Giant Cell (Mgg, 40x)

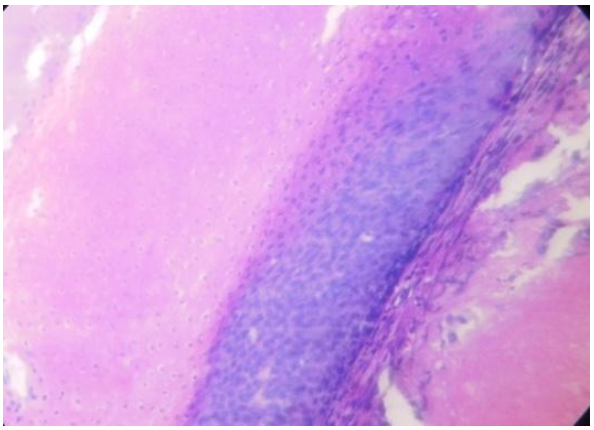

FIG 4: Strip Of Basaloid Cells Along With Ghost Cell (H&E, 40x)

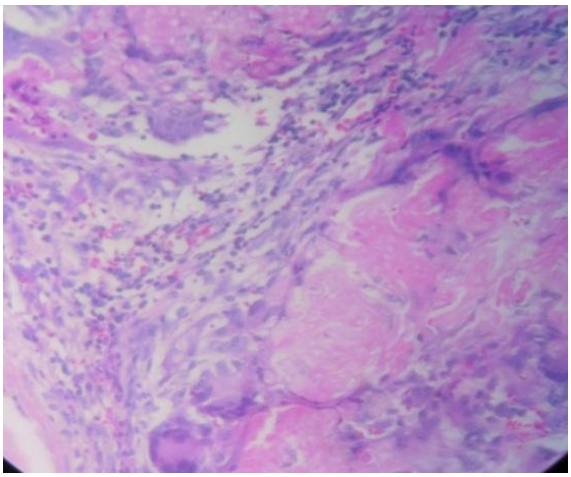

FIG 5: Shadow Cells, Giant Cells Along With Dispersed Basaloid Cells (H&E, 40x)

CONCLUSION

The present case highlights the importance of considering pilomatrixoma in the clinical and pathologic differential diagnosis of dermal or subcutaneous nodule even in locations other than head and neck region. Although a very rare tumour and often misdiagnosed as epidermal or dermoid cyst. Since spontaneous regression never occurs, cosmetic problems and reports of its malignant transformation demand its complete excision.

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