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CYTODIAGNOSIS OF PILOMATRIXOMA- A Case Report

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INTRODUCTION

Pilomatrixoma (Pilomatricoma, PMX, calcifying epithelioma of Malherbe), is a relatively common benign neoplasm arising from hair matrix of skin adnexa. It was first described by Malherbe and Chenantais in 1880[1]. It accounts for almost 20% of pilar tumours with differentiation towards the hair matrix of the hair follicle. It was found particularly on the head and neck and upper extremities. It may arise in the persons of any age, but 40% arise in children younger than 10 years and 60% in the second decade of life. It manifests clinically as a firm to hard dermal or subcutaneous nodule, measuring 0.5-3.0 in diameter, covered with normal skin. [2]

Very few reports are available on the cytological features of pmx in literature.In many cases pmx has been erroneously diagnosed cytologically as an epidermal inclusion cyst, giant cell tumour, squamous cell carcinoma, or malignant adnexal tumour [3] or round cell tumour of soft tissues [4]. We are presenting a case of pilomatrixoma diagnosed by FNAC in a 8- year- old girl.

CASE REPORT:

An 8-year-old-girl presented with a swelling in the left arm that she had noted 6 months earlier. The swelling increased in size gradually to attain the size of 1x1 cm over a period of six months. It was firm, mobile, non tender, skin over the swelling was not pinchable and there was bluish discolouration over the swelling.

FNA of the swelling was performed using a 22G needle fitted to a 10-ml syringe and yielded grayish white material. Two smears were prepared, one was wet fixed in 95% ethanol and stained with H&E and other smear was air dried stained with Leishman's stain.

The smears were composed of many ghost cells forming sheets, honey comb pattern, with thick and deeply stained cytoplasmic membrane and lightly stained central area (Fig 1). Numerous multinucleated giant cells were present along with epithelioid granulomas(Fig 2, 3). Scattered neutrophils and squames were present (Fig 4, 4a). Very few basaloid cells which had high nuclear cytoplasmic ratio and scant basophilic cytoplasm (Fig 5) were seen in the background. Excisional biopsy of the swelling showed the histological features of pilomatrixoma(Fig 6,6a).

DISCUSSION:

Pilomatrixoma is a skin appendageal tumour that manifests itself as a firm to hard dermal or subcutaneous nodule covered by normal skin. Occasionally it may be more superficial causing a blue-red discolouration of skin[1,3]. Although the lesion can appear at any age, it is commonly seen in children and is more common in females. Majority of the tumours are in head and neck region, the other sites being commonly affected are the upper extremities, trunk and lower extremities. The maximum size reported is 3 cms[3].

The history of pilomatrixoma shows two types of cells, basaloid cells and shadow cells or ghost cells. The basaloid cells have round to oval hyperchromatic nuclei and scant cytoplasm. There is abrupt transition to ghost cells, which are eosinophilic with a distinct border and possess a central unstained area of shadow of the lost nucleus. Calcification, foreignbody giant cells and keratinisation are a constant feature[4,5,6]. There are several reports in literature describing its cytologic features and addressing the main differential diagnosis and its pit falls [7]. However a review of literature showed that out of 52 reported cases the correct cytologic diagnosis was reported in only a few cases[3,7]. Most of the cases were reported as carcinoma, mainly squamous cell carcinoma or suggestive of carcinoma, epidermal inclusion cyst, giant cell tumour, malignant adnexal tumour, basal cell carcinoma or round cell tumour of soft tissue[1,3,,4,7].

The cases of erroneous diagnosis include lack of awareness of cytologic features of pilomatrixoma [8]; presence of monomorphic population of mitotically active basaloid cells with squamous differentiation but without anucleated ghost cells, chronic inflammatory cells or foreignbody giant cells;[9] high cellularity; high nuclear/ cytoplasmic ratio and the presence of anucleate squames [10]; predominance of one component over the other two and non-representative FNA smears. Atypia in nucleated squamous cells and misinterpretation of basaloid cells as malignant can lead to a diagnostic dilemma [8]. Similarly, misinterpreting the basaloid cells as intermediate cells will result in a misdiagnosis of mucoepidermoid carcinoma of salivary gland[11.]

Layfield et al [9] and Bansal C etal [3] reviewed the FNA cytology findings in large series of cutaneous masses including pilomatrixoma. Lesions that mimic pilomatrixoma are skin appendageal tumours, such as trichilemmal and epidermal inclusion cyst, cylindroma, malignant neoplasms such as squamous cell and basal cell carcinoma and granulomatous lesions. Salivary gland lesions also enter in the differential diagnosis of pilomatrixoma, especially if they are located in the parotid or submandibular areas[12].

Though diagnostic pitfalls do exist, certain cytologic features have been found useful in many others in making a precise pre-operative diagnosis of pilomatrixoma on FNA cytology smears. These include the presence of ghost cells, primitive- appearing basaloid cells with a high nuclear cytoplasmic ratio with nuclei exhibiting evenly dispersed chromatin and prominent nucleoli, naked nuclei , nucleated squamous cells, calcification, foreign body giant cells and pink fibrillary material enveloping basaloid cells . The presence of ghost cells seem to be the key or pathognomonic feature for recognizing pilomatrixoma[1]. Sanchez et al recommended the importance of examining smears with papinicolaou stain and Diff-quick since the nuclei of basaloid cells may appear unduly enlarged in smears stained with Diff-quick [14].In the case reported by Ma et al, ghost cells were not present in the first FNA smears but were easily recognized after repeat aspiration [9].

CONCLUSION:

Although there are several reports in the literature describing features and addressing the main differential diagnosis and diagnostic pitfalls of pilomatrixoma, this lesion continues to cause difficulty in cytodiagnosis. Atypia in nucleated squamous cells, misinterpretation of cytologic features of pilomatrixoma seem to be the important

factors that lead to the erroneous diagnosis of malignancy. However, the presence of ghost cells, naked nuclei, a dual cell population(both squamous and basaloid) and calcification should alert the cytologist to a case of pilomatrixoma.

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Legends to figures

Fig 1 : Ghost cells forming sheets, honey comb pattern[arrow] [H&E, 100X]

Fig 1a: Ghost cells in sheets [H&E, 100X].

Fig 1a: Ghost cells in sheets [H&E, 400X].

Fig 2: Showing multinucleated giant cell [H&E, 400X]

Fig 2a: : Showing multinucleated giant cells [H&E, 100X]

Fig 2b: Showing epithelioid granulomas [H&E, 400X]

Fig 3: showing scattered neutrophils and squames[H&E , 100X]

Fig 3a: showing scattered neutrophils and squames [H&E , 400X]

Fig 3b: showing scattered neutrophils and squames [H&E , 400X]

Fig 4: Showing basaloid cells [H&E, 400X]

Fig 5: Excisional biopsy showing features of pilomatrixoma[H&E, 40X]

Fig 5a: Excisional biopsy showing features of pilomatrixoma[H&E, 400X].

Fig 5b: Excisional biopsy showing features of pilomatrixoma [H&E, 400X].