Pilomatrixoma of right arm: a rare case diagnosed on fine needle aspiration cytology

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Abstract

Pilomatrixoma, a benign adnexal tumor frequently observed in head and neck. The incidence of pilomatrixoma in upper extremities is not common and sparsely reported in the available literature. Diagnosis by pre-operative aspiration cytology has been limited to few cases of this uncommon tumor. A 4-years-old girl underwent fine needle aspiration cytology (FNAC) of a right arm subcutaneous nodule. FNAC reveled predominantly anucleated shadow (ghost) cells with few basaloid cells. A cytology impression of pilomatrixoma was rendered, which was confirmed on histopathology.

This case report highlight the role of FNAC in diagnosis of uncommon skin adnexal tumor (pilomatrixoma) to avoid misdiagnosis and over diagnosis in view of varied cytomorphology.

Keywords: FNAC, Diagnosis, Pilomatrixoma, Adnexal tumor.

Introduction

Pilomatrixoma (Pilomatricoma) or calcifying epithelioma of Malherbe is an uncommon tumor with differentiation towards hair cells, especially hair cortex cells.(1) It is benign skin appendageal tumor most commonly observed in head and neck region. Presence of this lesion in arm is uncommon and has been reported in few cases in available literature.(2) Histopathological diagnosis of pilomatrixoma even at unusual location is straightforward, the same is not true for aspiration cytology.(3) Pilomatrixoma is misdiagnosed as epidermal cyst, monomorphic adenoma or benign cysts or over diagnosed as malignant condition as carcinoma. An accurate diagnosis of this benign uncommon lesion on cytology is imperative, considering that excision is curative.(4) We highlight the role of cytology in diagnosis of pilomatrixoma in arm of young girl.

Case Report

A four-years-old girl presented to pediatric surgery OPD with history of gradually increasing swelling over right arm since 2 months. No significant past, personal or history of pain/ trauma noted. Local examination revealed firm subcutaneous swelling measuring 1.5x1.2cms on lateral side of right arm. The swelling was nontender, not fixed to overlying skin or underlying structure. Soft tissue lesion was clinically suspected and FNAC was performed from swelling. The procedure was performed under all aseptic precaution with 23 G needle and 10ml syringe.

Cytology: The smear was prepared and stained with pap and toluidine blue stain. FNA smear predominantly showed anucleated shadow (ghost) cells (Fig. 1a, b) admixed with a few aggregates of round to oval basaloid cells with scanty to moderate amount of pale blue cytoplasm and vesicular nuclei (Fig.1c, d). Necrosis or mitotic activity or multinucleated giant cells were not seen in smear examined. Cytological diagnosis of Pilomatrixoma was rendered and excision biopsy was done.

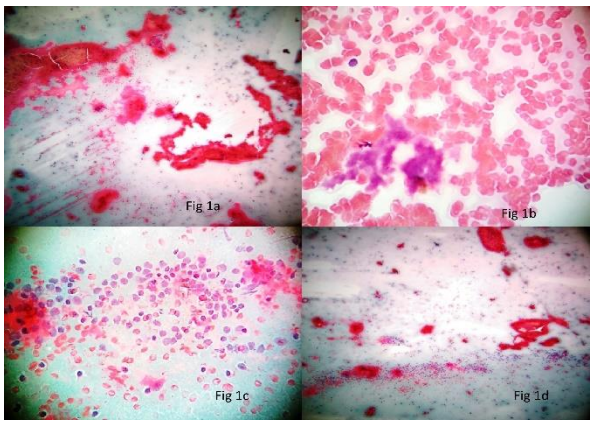

Fig. 1:

a. Cytology showing predominantly anucleated shadow /ghost cells clusters (Pap,x100)
b. Acellular ghost cells on toluidine blue (TBS,x400)
c. Basaloid cells with round to oval nuclei (Pap,x400)
d. Combination of bimodal population of shadow and basaloid cells clusters (Pap,x100)

Histopathology: We received a nodular, well circumscribed soft tissue mass measuring 1.4x1.1cms with attached skin tag. Cut section showed a round circumscribed grayish white lesion measuring 1.3x1.0 cms in diameter with foci of whitish material (calcified) (Fig. 2a). Multiple section showed features of classical pilomatrixoma with bimodal population of foci of

anucleated shadow (ghost) cells, cluster of basaloid cells(Fig. 2b, c) and many multinucleated giant cells (Fig. 2d), thus confirming the diagnosis of pilomatrixoma. On subsequent follow up patient is doing well with no local recurrence.

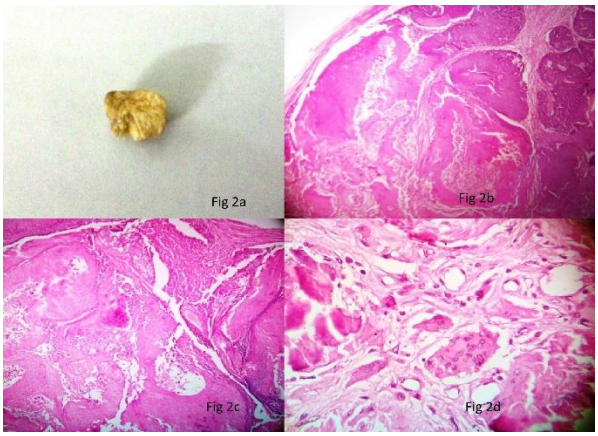

Fig. 2:

a. Cut section of resected arm mass with calcified whitish material

b. Histopathology of dual morphology of circumscribed tumor with shadow and basaloid cells clusters (H&E,x100)

c. Ghost cells clusters with periphery arranged basaloid cells nest (H&E,x100)

d. Multinucleated giant cells with calcified material (H&E,x400)

Discussion

Pilomatrixoma was first described as "epithelioma cutis necrotic calcification Malherbe" by Malherbe and Chenantais in 1880 as a benign, subcutaneous tumor of sebaceous gland.[5] Pilomatrixoma now called as pilomatricoma as proposed by Forbis and Helwigin 1962.[6] Thus avoiding the word epithelioma, which may erroneously described as malignancy by some researchers.[6]

Pilomatrixoma is a benign skin adnexal tumor with hair follicle matrix cells differentiation. It has bimodal peak during first and sixth decades of life. It is usually found in head and neck region and upper extremities with other sites are uncommon. Radiology has limited value in diagnosis of pilomatrixoma, as it shows only variable amount of calcification.[4]

FNAC is useful in preoperative diagnosis and most favored diagnostic modality, usually shows characteristic features of pilomatrixoma.

Presence of ghost cells and shadow cells is must for diagnosis by FNAC. Other features include basaloid cells, clusters, few nucleated squamous cells, and calcification with giant cells response to keratin. Despite these features pilomatrixoma may be mistaken for benign cystic lesions like epidermal inclusion cyst to monomorphic adenoma or may be over diagnosed as carcinoma in view of basaloid cells with anaplasia.[4,7] Histopathological features of pilomatrixoma showed deep sub epidermal tumor consisting of irregular islands of keratinized shadow cells surrounded by peripheral basaloid cells.[3] Calcification and foreign body giant cells reaction to keratin may be seen as seen in other cases. Treatment of choice is wide local excision with 1-2 cms of healthy skin margins. Malignant transformation and recurrence is very rare as per literature in pilomatrixoma.

Conclusion

In the present case, we highlight the role of FNAC in the preoperative diagnosis of pilomatrixoma. The cytological smear should be examined carefully to avoid diagnostic error due to predominance of one component over the others. Cytopathologists should keep in mind the variability of cytologic features depends on cellular composition of the lesion.

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