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Gaint Pilomatrixoma on Chest wall- A Rare Case Report with Review of Literature

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Abstract: *Pilomatrixoma also termed as calcifying epithelioma of Malherbe is derieved from hair follicle matrix cells. It is a very rare benign neoplasm which is often mistaken for malignancy. It can even originate from peri areolar piliform bulbs and can be mistaken for breast malignancy. We present a case 31 year old male with pilomatrixoma on left chest wall and review literature regarding presence of pilomatrixoma on chest wall*

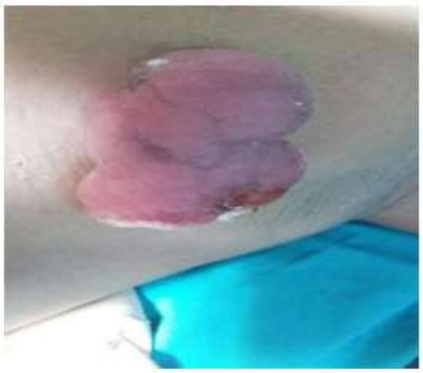
I. Introduction

Pilomatrixoma Or Pilomatricoma is a benign neoplasm derived from hair follicle matrix cells. These are mostly found on head and neck region and rarely on other sites(chest/breast 2.6%). Due to such rare presentation on chest, discussion is mostly confined to literature of pathology and dermatology.

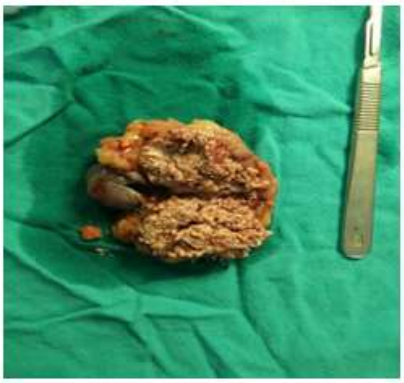
Case report

A 30 year old male presented with a lump on chest wall which gradually increased in size from size of pea to that of a golf ball over a period of 6 months. The size of lump increased rapidly in the last one month , there is no history of discharge from lump , fever or weight loss.

Examination showed an exophytic lesion of size 5*4*2.5cm, non tender , firm in consistency over left chest wall. Axillary lymph nodes were not palpable bilaterally, there were no engorged veins and dimpling of skin on chest wall .



Pre Op.lesion on chest



Post Op lesion

X ray chest was done and was found to have no abnormality, followed by FNAC which was suggestive of Round cell Neoplasm. After proper work up excisional biopsy of the lump was done under general anesthesia . Histology revealed Pilomatrixoma

II. Discussion

Pilomatrixoma or calcific epithilioma of Malherbe is a benign neoplasm of skin which originates from piliferous follicles[1]. Dr. Chenantals Malherbe in 1880 first described it and thought it to be calcified epithelioma of sebaceous gland[2]. Forbis and Helewig in 1961 introduced the term Pilomatrixoma which better conveys the histological source[3].

Pilomatrixomas are mostly seen in children usually on head and neck area . it is very rare presentation to see pilomatrixoma in a male patient and that too on chest wall.

In 1980 Lapansri et al described about malignant pilomatrixoma which is rare and only a few articles are available[4]. It is an aggressive neoplasm which has high chances of recurrence . few cases have also been reported to have malignancy. To differentiate malignant from benign lesions the salient featurtes to be noted are high mitotic rate with atypical mitosis , infiltration of skin, central necrosis, invasion of blood vessels and lymphatics.[5-6]

In this case , the definitive diagnosis was made by HPE(Histo Pathological examination). It is usually misdiagnosed pre operatively. Wells et al found that pre operative diagnosis in cases was incorrect in 57% of cases where as in a series of 376 cases the pre operative and pathological diagnosis were accurate and consistent in 28.6% of cases[7]. Kumaran et al reported 46% correct preoperative diagnosis in 78 excised pilomatrixomas. Incorrect diagnosis included dermoid cyst , sebaceous cyst , foreign body[1].

On presentation a painful firm nodule was palpable in this case . 32% of cases in the case series of 346 cases had pain and tenderness . Duflo et al reported 20% incidence of pain in pilomatrixoma cases[8]. The lesion also showed tent's sign which is flattening of some portion of tumor surface which resembles a tent. It is caused due to attachment of the lesion to the overlying epidermis. There is slight discoloration over the surface of skin due to growth of vessels on the overlying skin.[9]

Although pilomatrixoma is mostly solitary sending but multiple lesions have been mentioned in literature which may be associated with myotonic dystrophy, xeroderma pigmentosa, basal cell nevus all of which are genetic disorders .[10,11,12]

Histopathology features of pilomatrixoma are well demarcated , contain basaloid cells and shadow cells. Mineralization in keratinized cells is a common feature whereas melanin pigmentation in either basaloid cells or stromal pigmentation is rare.

In our case the histopathological findings were islands of basaloid cells and ghost/shadow cells in the lower dermis and extending into the subcutaneous plane . basaloid cells show abrupt to gradual transition to ghost cells at various places by gradual loss of nuclei and leaving on unstained shadow of the last nuclei. Keratinisation is seen in both basophilic island and shadow cells with presence of melanin in some areas

Diagnostic imaging though rarely done pre operatively, has been reported and includes USG, CT , MRI. CT will show well demarcated lesion with or without calcification. USG will show superficial position , well demarcated mass , continuity of lesion in deeper planes .MRI according to Hoffman et al is inconclusive in diagnosis of pilomatrixoma.

As performed in this case the treatment involves excision of lesion with min 1 mm of tumor free margin. Tumors of extremities are managed conservatively if not associated with symptoms.

The recurrence rate varies from 0-3% and tumor needs to be re excised due to suspicion of malignancy[12]

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