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Original Research Paper

Surgery

PILOMATRIXOMA – THE GHOST A SURGEON SHOULD REMEMBER

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ABSTRACT Pilomatricoma or pilomatrixoma or calcifying epithelioma of Malherbe are commonly misdiagnosed, benign neoplasms of the skin, thought to arise from hair follicle matrix cells and represent 0.12% of all skin tumors. Pilomatricomas usually develop slowly and most frequently appear as solitary, firm nodules, exhibiting a normal to pearl white epidermis and calcium deposits are present in more than 50% of the lesions giving them the name as calcifying epithelioma. In this article along with a review of the Literature we describe a series of 4 cases of pilomatricoma arising near the left shoulder, neck, elbow and one along the left supra-clavicular region. Three were males and one was a lady and were in the age group of 16 to 62 years.

KEYWORDS : Pilomatrixoma , Calcifying Epithelioma of Malherbe, Ghost cells,

INTRODUCTION

Pilomatrixoma is an uncommon tumor with differentiation towards hair cells, especially hair cortex cells. It is benign skin appendageal tumor.They are generally asymptomatic and found commonly in the head and neck area and upper extremities, but rarely found to occur on the chest, trunk, or lower extremities and can occur in any hair bearing skin. They are reported to occur in people of all ages, however the distribution favors both children less than 20 years and the elderly above 60 years of age. An accurate preoperative diagnosis of pilomatricoma is usually only rarely made and mostly confirmed after examining the removed lesion under the microscope. Histologically, pilomatricomas present as well demarcated lesions arising from the dermis and extending into the subcutaneous fat and classically consist of islands of epithelial cells comprised of both basophilic cells with meager cytoplasm and ghost cells that have a central unstained area indicative of a lost nucleus.

CASE PRESENTATION

A 62-year-old man presented with a swelling of 3cm * 3cm in the left shoulder region along with a 0.5 cm nodule nearby of 2 years duration. It was firm in consistency, non-tender, mobile and some calcification was observed at one area of the bigger swelling. After preliminary investigations with a provisional diagnosis of Hamartoma, the patient underwent Excision Biopsy of both the lesions which was reported as Pilomatricoma.

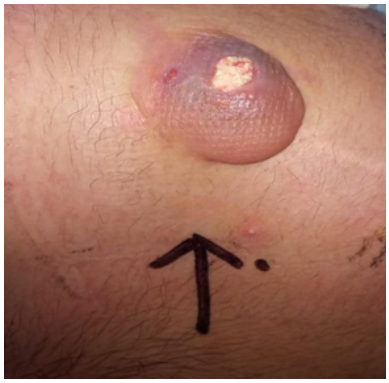

FIGURE -1

The second case involved a 20 year-old lady with a swelling of 2cm * 2cm over the middle of neck on right side since 9 months, slowly growing, painless, mobile and not associated with cough or any systemic symptoms. A provisional diagnosis of Epidermoid cyst was made and the patient underwent Excision Biopsy and the Histopathology revealed it was a Pilomatricoma.

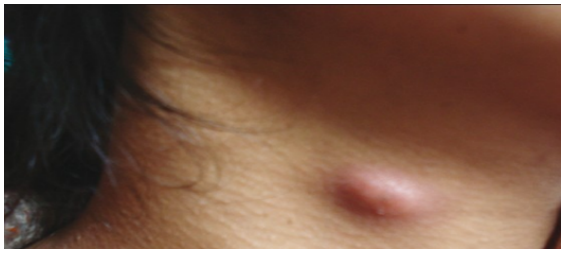

FIGURE -2

The third case involved a 16 year old boy with a soft to firm swelling of 1 year duration near the elbow of size 3cm* 2cm, mobile non-tender and a provisional diagnosis of Papilloma was made and removed and sent for Histopathologic examination which was reported as Pilomatricoma.

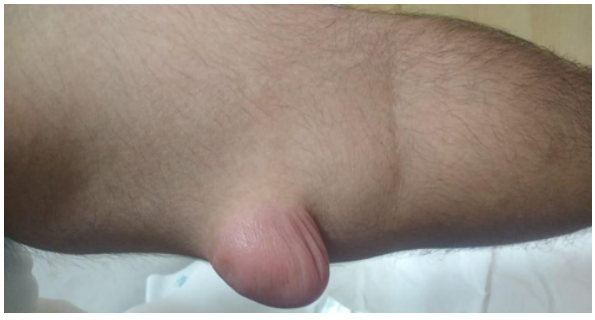

FIGURE -3

The fourth case was in a 35 year male patient who presented with a firm swelling of 8cm *6 cm over the left supra-clavicular region with restricted mobility, slow growing for the past 2 years. The overlying skin was discoloured and hyperpigmented at few areas, with mild tenderness. Ultrasound and MRI were suggestive of Dermato-fibrosarcoam protruberance and FNAC was inconclusive. The patient underwent wide excision followed by Split skin grafting of the resultant raw area. Histopathology revealed that the lesion was a Pilomatricoma. All the 4 patients are doing well and being regularly followed up for the past 1 year with no evidence of any recurrence at the operated site or the occurrence of any new swelling. A review of the literature was done and it was found that it in most of the cases a definitive pre-operative diagnosis was not established and only detected after removal except in a few cases were out of clinical suspicion FNAC was done which showed in some cases the characteristic basophilic cells and ghost cells.

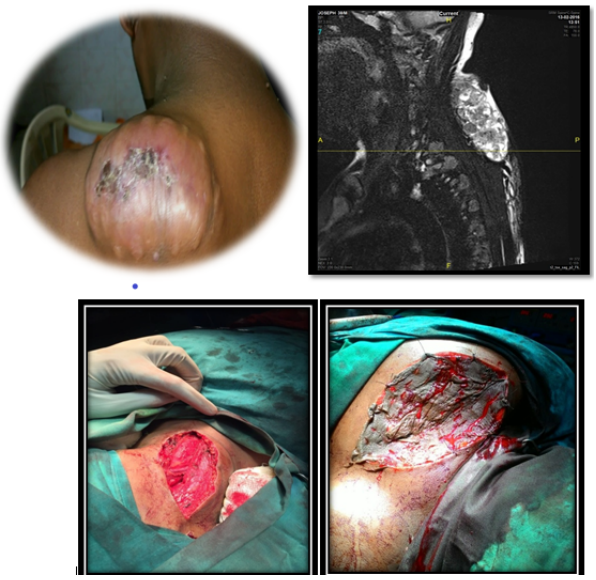

FIGURE – 4

DISCUSSION

Pilomatricoma, a benign neoplasm of the hair follicle, was initially thought to arise from sebaceous glands and was called *calcifying epithelioma of Malherbe* by Malherbe and Chenantais in 1880. In 1961, after histochemical and electron microscopic analysis of 228 such tumors, Forbis and Helwig found the cell of origin to be the outer root sheath cell of the hair follicle and proposed the name, *pilomatrixoma*, now called *pilomatricoma*.

In 1973, Moehlenbeck reviewed 140 000 skin tumors and noted that pilomatricoma represented 0.12% of cases. Although pilomatricoma can develop in patients of any age, it occurs most often in children and young adults. Among the young, 40% of cases occur before the age of 10 years and 60% of cases occur before the age of 20 years. However, a second, smaller peak of onset has been reported in the elderly. The female-male ratio ranged from 0.43:1 to 2.45:1, and a female preponderance was noted in a majority of the studies.

The most common anatomical location for the tumor is the head and neck region; between 40% and 77% of cases occurred in this area. The second most common site is the upper extremities, followed by the trunk and the lower extremities. In the head and neck region, the most frequent locations are the neck and the frontal, temporal, periorbital, and preauricular areas. The neck and cheek were the most common sites in the pediatric group (age, <18 years), while the scalp and neck were the most common sites in the adult group. No cases of pilomatricoma have been reported on the palms, soles, or genital region, probably because of the lack of hair-bearing skin in these areas.

Pilomatricomas are usually solitary nodules, but multiple occurrences have been observed in 2% to 10% of reported cases. Multiple pilomatricomas have been associated with myotonic dystrophy, Gardner syndrome, Steinert disease, Turner syndrome, and sarcoidosis Pilomatricoma is considered to be of ectodermic origin. This theory is supported by evidence that pilomatricoma originates from the hair germinal matrix. Pilomatricoma formation represents a disturbance of the hair follicle cycle in which limited cytologic differentiation of pilar keratinocytes occurs but further development into mature hair fails to take place. There is a study suggesting that apoptosis is the main mechanism leading to the development of the dead shadow cells and is most probably responsible for the banal biological behavior of pilomatricoma.. A recent study suggests that trisomy-18 is a consistent feature in pilomatricoma, suggesting that genes carried on this chromosome such as that for the Antiapoptotic Oncoprotein BCL2 may have a role in the growth and differentiation of this benign self-limited tumor. The etiology of pilomatricomas is also thought to be due to a somatic mutation in the CTNNB1 gene which regulates the protein beta-catenin. Beta-catenin is a protein involved in cell adhesion and cell signaling and when mutated, leads to uncontrolled cell division and pilomatricoma formation. There are stray reports of pilomatricoma cases occurring after vaccination at the site of vaccination.Most lesions are benign but rarely malignant transformation can occur in middle age to elderly individuals.

MORPHOLOGICAL STAGING

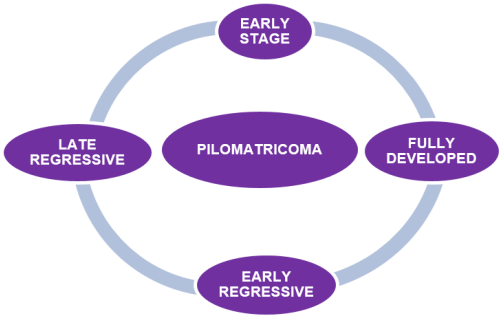

FIGURE -5

These stages reflect the evolution of pilomatricoma and how a tumour from a matrix cyst transforms into a calcified and osseous nodule with no visible epithelial component.

The diameter of a pilomatricoma ranges from 0.5 to 3 cm in most cases. The tumor slides freely over the underlying layer. Also, its irregular consistency, firm shape, and overlying skin discoloration are suggestive of its nature. The tumor is usually asymptomatic and grows slowly over a period of several months or years. Calcification is present in 70%–95%. The clinical differential diagnosis of head and neck pilomatricoma includes sebaceous cyst, ossifying hematoma, brachial remnants, preauricular sinuses, adenopathy, giant cell tumor, chondroma, dermoid cyst, degenerating fibroxanthoma, foreign body reaction, and osteoma cutis. In the preauricular area, pilomatricomas can be particularly difficult to distinguish from tumors or inflammatory conditions that originate from the superficial lobe of the parotid gland. Although diagnosis of pilomatricoma can be based solely on the clinical features, previous reviews have shown that the accuracy rate of the preoperative diagnosis of pilomatricoma ranges from 0% to 30%. The differences in accuracy rates may be attributable to the fact that most clinicians are not familiar with this tumor.

Radiologic imaging is of little diagnostic value for pilomatricoma. Imaging has been used mainly for differentiating preauricular tumors from parotid tumors and for evaluating large or aggressive tumors. The characteristic computed tomographic findings are those of a sharply demarcated subcutaneous lesion with various amounts of calcification. Magnetic resonance imaging has revealed high-signal bands on T2-weighted images that correlate with the bands formed by basaloid cells that are evident on histologic examination.14 Ultrasonography has also been described as a relatively fast and noninvasive method for estimating the depth of larger masses.

Histopathologic examination of pilomatricoma shows sharply demarcated dermal nodules surrounded by a capsule of compressed fibrous tissue located in the lower dermis and extending into the subcutaneous fat. The cells in the islands are arranged in a circular configuration, with nucleated basaloid cells on the periphery and enucleated shadow cells in the center .The basaloid cells exhibit deeply staining basophilic nuclei with scant cytoplasm that lack distinct cell borders. The shadow cells, also termed ghost cells, which evolve from basaloid cells, represent dead

cells that retain their cellular shape and show a central unstained area that corresponds to the lost nucleus. The transitional cells, which are localized between basaloid cells and shadow cells, were thought to represent apoptotic cells that were finally proceeding to shadow cells. Calcification is mostly seen in the ghost cell regions, with the incidence ranging from 69% to 85%. Foreign body giant cell reaction, which represents a granulomatous response to the shadow cells, can also be identified in regions where keratinized debris is abundant.

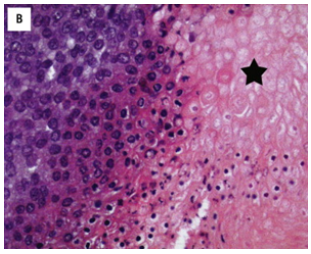

FIGURE -6: **Basaloid cells, * Shadow / Ghost Cells**

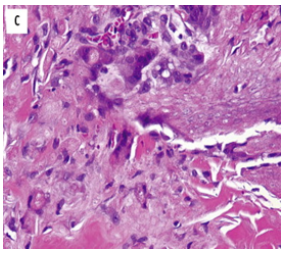

FIGURE -7: **Multi-Nucleated Giant cells**

Malignant transformation is rare, with fewer than 20 cases described in the literature. Malignant pilomatricomas tend to occur in middle-aged or elderly patients. The histologic features of pilomatrix carcinoma include active proliferating basaloid cells with atypical mitoses and nuclear pleomorphism , invasion of blood vessels, and infiltration into underlying structures. Treatment is wide local excision. Rare cases of pilomatrix carcinoma with distant metastasis have been reported.

Fine-needle aspiration cytology has been described as a preoperative diagnostic investigation. However, the results can be misleading if there are no ghost cells present in the aspirate. Since spontaneous regression is never observed, the standard treatment of pilomatricoma is complete surgical excision with clear margins. Wide excisions with margins of 1 to 2 cm are recommended for malignant variants to minimize the risk of local recurrence. Occasionally, the overlying skin will need to be excised because of tumor adherence to the dermis. Recurrences after surgery are rare, with an incidence of 0% to 3% Malignant transformation to a pilomatrix carcinoma should be suspected in cases with repeated local recurrences.

CONCLUSION:

Pilomatricoma is a benign tumor of the skin, which is not rare in occurrence but rarely diagnosed preoperatively and it is imperative that the Surgeon should have it in his mind in the differential diagnosis while evaluating any benign skin tumour. This article highlights the diagnostic pitfalls in solitary subcutaneous swellings that are commonly diagnosed as sebaceous cyst or lipoma in day to day practice and is mainly presented to create awareness among surgeons to consider the possibility of pilomatricoma in the differential diagnosis of solitary nodular lesions especially if they are in the head, neck and upper extermities.

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