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# PILOMATRICOMA – STUDY ON 27 CASES AND REVIEW OF LITERATURE

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**ABSTRACT**
*The purpose of this study was to report the clinicopathological spectrum of Pilomatricoma, an uncommon benign tumour of hair matrix. The study is a retrospective review of 27 cases of pilomatricoma reported in a private Laboratory of Lahore from 2006-2008. The clinical data of the patients was obtained from their respective files. Age range of the patients was from 8-76 years. Maximum number of cases was observed in the second decade. Female to male ratio was 2:1. The size of the tumour ranged from 4-90 mm in diameter. The most common site for the occurrence of pilomatricoma was head and neck followed by upper extremities. Majority of the specimens were submitted as unspecified lumps. Other provisional diagnoses included enlarged cervical lymph nodes, sebaceous cysts, tuberculoma, haemangioma, keloid and granulation tissue. All the patients presented with solitary lump. Preoperative diagnosis of pilomatricoma is most often confused clinically with other skin diseases due to its wide variety of clinical characteristics. Careful clinical examination would result in a more accurate diagnosis; however, definite diagnosis is only possible after microscopy of the lesion.*

## INTRODUCTION

Pilomatricoma, formerly known as Pilomatrixoma is a benign dermal and/or subcutaneous tumour. It most commonly affects children and adolescents; however, a second smaller peak of onset is also reported in the elderly. It is slightly more common in females.[1]

This tumour was first described by Malherbe and Chenantais in 1880 as a benign, subcutaneous tumor arising from hair cortex cells. Since then, this uncommon entity has been called *calcifyng epithelioma of Malherbe*[2]. In 1922, Dubreuilh and Cazenave described the characteristic histological features, i.e., islands of epithelial cells, including shadow cells and giant cells[3]. The name pilomatrixoma was proposed by Forbis and Helwig in 1961, thus avoiding the word *epithelioma,* which carries the connotation of malignancy.[4]

The most frequent anatomical location of pilomatricoma is the head and neck region, followed by upper extremities[5,6]. Most of the cases present as single nodules but multiple occurrences have also been reported[7,8]. Familial cases have been observed in association with disorders such as Gardner syndrome, Steinert disease, and sarcoidosis[5]. A few cases of malignant pilomatricoma metastasizing to the lung, bone, brain, abdominal organs, skin, and lymph nodes have also been described in the literature[9,10]

Despite being better defined, pilomatricomas continue to be frequently misdiagnosed and are not usually considered in differential diagnoses. The usual clinical differential diagnosis of pilomatricomas includes sebaceous cyst, ossifying haematoma, giant cell tumor, dermoid cyst, chondroma, degenerating fibroxanthoma, foreign body reaction, and osteoma cutis. Malignant neoplasms in the clinical differential diagnosis include basal cell carcinoma with matrix differentiation, pilomatrix carcinoma, and squamous cell carcinoma[11,12].

The usual clinical presentation of pilomatricoma is typically as a superficial, firm, solitary, slow growing, and painless mass of the dermis[5,6]. The size of the tumour rarely exceeds 3 cm. The overlying skin may exhibit a bluish discoloration or ulceration. Since this tumour does not regress spontaneously; surgical excision is both curative and a treatment of choice. Recurrence is rare after complete resection.[12]

The findings of fine-needle aspiration biopsy can be misleading, and histopathological examination is often needed for a definitive diagnosis.[13,14]

## METHODS

This retrospective study was carried in a private laboratory. The records were searched for all cases of pilomatricoma between 2006 and 2008. Patients data such as sex; age at presentation; location, and size of the tumour; preoperative diagnosis and pathological features were collected from the computerised data.

All cases were classified according to the site of the tumours in different parts of the body. The sizes of the tumours were categorised into three:

Small 20 mm or less in diameter, medium size 21-40 mm and large tumours above 40 mm in diameter.

## RESULTS

A total of 27 cases were included in our study with an age range from 8-76 years. The majority of the patients were found between the ages 21-30 years, followed by 31-40 and 1-10 year age group (Table 1). The female:male ratio was 2:1. The most common site for the tumors was head and neck (48.14%), followed by upper limb (33.33%) and the trunk (7.40%) whereas no site was mentioned in three cases. The sizes of the tumours were variable. Most of the cases (55.55%) were less than 20mm in diameter, 29.62% were between 21-40 mm in size and only 14.81% were over 40 mm in diameter. The smallest lesion was of 4mm in maximum dimension whereas the largest measured 90mm which was seen in a 22-years female in the lower back.

**Table** 1: *Size of lesion in 27 cases of Pilomatricoma.*

| Size of the lesion (mm) | Number (n) | Percentage (%) |
|---|---|---|
| 0 – 20 | 15 | 55.55 |
| 20 – 40 | 8 | 29.62 |
| > 40 | 4 | 14.81 |

None of the cases was diagnosed clinically as pilomatricoma. Most of them were submitted as unspecified lump. Other clinical diagnoses included enlarged cervical lymph nodes, sebaceous cysts, tuberculoma, haemangioma, keloid and granulation tissue. All the patients presented with solitary lump.

**Table 2:** *Provisional Diagnosis in 27 cases of Pilomatricoma.*

| Clinical diagnosis | Number | Percentage |
|---|---|---|
| Unspecified lump | 15 | 55.5 |
| Sebaceous cysts | 4 | 14.8 |
| Cervical lymph nodes | 3 | 11.1 |
| Tuberculoma | 2 | 7.4 |
| Haemangioma | 1 | 3.7 |
| Keloid | 1 | 3.7 |
| Granulation tissue | 1 | 3.7 |

Histological findings allowed definitive diagnosis in all cases. In most cases, the tumour consisted of islands of cells in circular configuration with enucleated shadow cells in the center and nucleated basophilic cells on the periphery. The basaloid cells had deeply staining basophilic nuclei with scant cytoplasm that lack distinct cell borders. The shadow cells, also termed ghost cells, evolve from basaloid cells and represent dead cells that retain their cellular shape and show a central unstained area that corresponds to lost nucleus (Fig.1). The transitional cells were localized between basaloid cells and shadow cells. Shadow cells were surrounded by foreign body-type giant cells and at times by granulomatous response. Calcification was present in 70% of our cases (Figure 2). Bone metaplasia was observed in one case only.

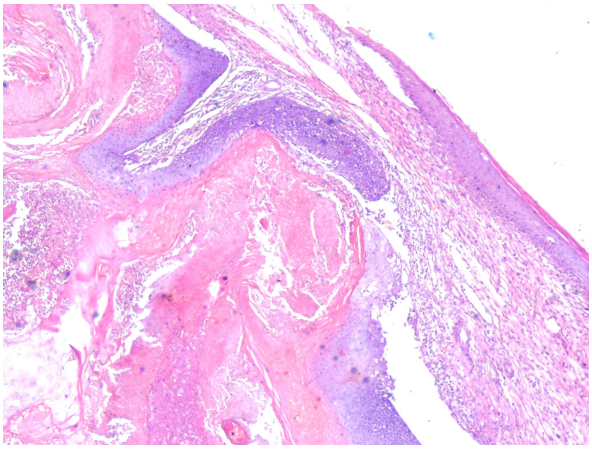

**Fig. 1:** *Histology of Pilomatricoma.*

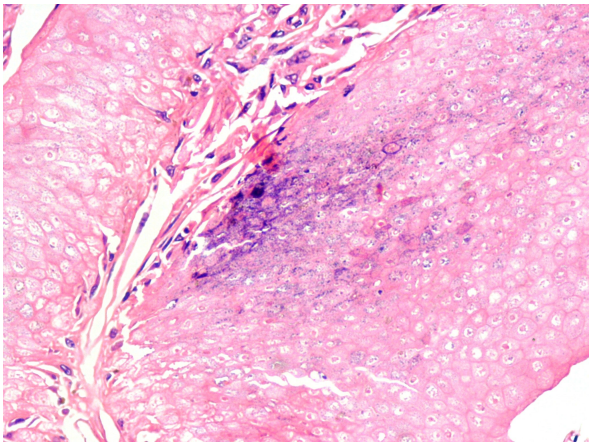

**Fig. 2:** *Calcified squamous cells in pilomatricoma.*

## DISCUSSION

Pilomatricoma, a benign tumour arising from hair cortex cells, was initially thought to be a sebaceous gland neoplasm. In 1961, Forbis and Helwig, after histochemical and electron microscopic analysis of 228 such tumours, found the cell of origin to be the outer root sheath cell of the hair follicle and, therefore, proposed the name, *pilomatrixoma,*

now called *pilomatricoma.*[4]

Although pilomatricoma can develop in patients of any age, children and young adults are most commonly affected. No study has so far been done on pilometricoma in Pakistan. In the present study, maximum number of cases was observed in second and third decades. These findings are consistent with those of Darwish et al who observed 78% of the tumours below the age of thirty[12]. In contrast, most of the studies concluded that 1-20 years was the most affected age group[5,6]. Celia et al observed bimodal presentation of pilomatricoma in 1st and 6th decades[16]. Kuddo et al studied 118 cases of pilomatricoma in adults and found that majority of the cases were above 45 years.[17]

Yaqoob et al studied the tumours of pilo-sebaceous unit at Agha Khan University Hospital and found that plomatricoma was one of the five most common skin adnexal tumour, other being nodular hidradenoma (eccrine acrospiroma) Syringocyradenoma papilleferum, eccrine poroma and eccrine spiradenoma.[15]

A female preponderance is noted in majority of the studies.[1'2'3] Our study also demonstrates 2:1 female to male ratio. However, Ming-Ying observed almost equal number of cases in both sexes. The most common anatomical location for pilomatricoma is the head and neck region followed by upper extremities, trunk and lower extremities[5,6]. Majority of our cases presented with a lump in head, neck and upper limb.

Pilomatricoma usually present as solitary nodules but multiple occurrences have been observed in 2-10% of the cases[7,8]. None of the patient in our study presented with multiple pilomtricomas. Tumour diameter in majority of the cases ranged from 0.5 to 3 cm[5,6,12], but lesions up to 15 cm have been reported. The present report regarding the size shows similar results to those reported previously.[18]

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 of the clinicians are not familiar with this entity. None of our cases has been diagnosed clinically as pilomatricoma.

Radiologic imaging is of little diagnostic value for pilomatricoma[20]. The findings of fine needle aspiration biopsy can be misleading and can be mistaken for squamous cell carcinoma due to good cellularity, high nuclear/cytoplasmic ratio and anucleated squamous cells[12,13,14]. Histopathologic examination is often needed for definitive diagnosis. It shows well-circumscribed nodule in the deep dermis and subcutaneous tissue having no epidermis connection. Tumour is composed of nodules with nucleated basaloid cells peripherally and enucleated shadow cells centrally. Early lesions tend to become cystic whereas older become solid with prominent shadow cell component, keratin debris, multinucleated giant cells and dystrophic calcification with the incidence ranging from 69% to 85%.[1,5,6,12] We observed calcification in 70% of our cases. In previous articles, bone metaplasis is noted in 15% of cases owing to conversion fibroblasts into osteoblasts[1,5]. In our study, only one case (3.7%) demonstrated osseous metaplasia. This is consistent with the findings of Zamanian et al who observed ossification in 4.4% of the cases.

Malignant transformation of pilomatricoma is rare. In the literature, 80 cases of pilomatrix carcinoma have been reported with metastases in nine cases. 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[9,10]. Yet, no evidence of malignancy or aggressiveness was observed in this study. Since spontaneous regression is never observed, the treatment of choice is surgical excision. Wide resection margins (1-2 cm) are recommended to minimize the risk of local recurrence. 12

It is **concluded** that Pilomatricoma generally present as firm subcutaneous nodules and are often misdiagnosed. They can appear at any age, with a peak presentation in the second decade. Head and neck and upper extremities are the most commonly involved regions. The physicians and surgeons should be familiar with this entity and consider it in the differential diagnosis of a superficial mass.

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