PILOMATRICOMA- ANALYSIS OF 15 CASES WITH REVIEW OF LITERATURE

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ABSTRACT: A pilomatricoma is a benign appendage tumor related to hair cells matrix. Pilomatricoma usually occurs as a solitary firm lesion with predilection for head and neck, and upper extremities. It is more common in children. AIM: The objective of this study is to know the clinical presentation and histopathology of this condition. MATERIALS AND METHODS: This is a 4 yrs retrospective and 1 yr prospective study carried in Upgraded Department of pathology, Osmania general hospital between June 2008 and May 2013. Patients data such as sex, age, site of occurrence, clinical presentation were considered. The study included 15 patients with histopathologically diagnosed pilomatricoma. RESULTS: A total of 15 cases were included in our study with an age range from 6-36 year. Out of which benign tumors were 14 (93.3%) and malignant 1 (6.7%). The female: male ratio was 3:2. The sizes of the tumour range from 2-3cm. Most of the cases included Upper extremity-7 cases (46.7%), Head and neck region- 6 cases (40.0%), Trunk-2cases (13.3%). The most common clinical presentation was sebaceous cyst. CONCLUSION: Pilomatricoma generally present as firm subcutaneous nodules in children and young adults and are often misdiagnosed. The physicians and surgeons should be familiar with this entity and consider it in the differential diagnosis of a superficial mass.

KEYWORDS: Hair cell matrix, Pilomatricoma, Sebaceous cyst.

INTRODUCTION: Pilomatricoma is a benign appendage tumor related to hair cells matrix1. This tumor 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 calcifying epithelioma of Malherbe2. It was believed to be a rare tumor, but increasing reports in the literature show that it is not uncommon. Presently, pilomatricoma is the most common hair follicle tumor. Pilomatricoma usually occurs as a solitary firm lesion with predilection for head and neck, and upper extremities. Rare cases of multiple pilomatricoma are associated with different conditions such as myotonic dystrophy, Rubinstein–Taybi syndrome and Turner syndrome. It is more common in children, but occurrence in adults is increasingly being recognized3-6. Most of the literature review about pilomatricoma is in the form of case reports with few studies.

AIM: The objective of this study is to know the clinical presentation and histopathology of this condition.

MATERIALS AND METHODS: This is a 4 yrs retrospective and 1 yr prospective study carried in Upgraded Department of pathology, Osmania general hospital between June 2008 and May 2013. Patients data such as sex, age, site of occurrence, clinical presentation were considered. The study included 15 patients with histopathologically diagnosed pilomatricoma. Only patients with
confirmed pathology report of pilomatricoma (post-surgical excision) were included. Routine processing and paraffin embedded sections were stained using haematoxylin and eosin stain and were subjected to light microscopic examination.

**RESULTS:** A total of 15 cases were included in our study with an age range from 6-36 years. Out of which benign tumors were 14 (93.3%) and malignant 1 (6.7%). Incidence among all skin tumors was 1.5%. The female: male ratio was 3:2. The sizes of the tumour range from 2-3cm. Most of the cases included Upper extremity-7 cases (46.7%), Head and neck region- 6 cases (40.0%), Trunk-2 cases (13.3%). The most common clinical diagnosis was sebaceous cyst followed by Dermoid, Lipoma, Antibioma, Neurofibroma, Calcified hamartoma, Basal cell carcinoma. (Table 1).

There were 8 patients in the pediatric age group (<16 years) and 7 in the adult age group. The most common site in both the groups was the upper extremity. In one case we had preoperative diagnosis on cytology and no imaging investigations were available for all the patients included in the study prior to surgical excision.

Excision and histological examination was performed for all cases where macroscopic appearance of the excised lesion showed grey white solid areas in most of the cases. Calcification was macroscopically described in 7 cases.

All cases more or less shared the same microscopic features of basaloid cells, ghost cells, calcification, and foreign body type giant cells, presenting features of a calcifying epithelioma of Malherbe (pilomatricoma) (Figure 1).

One case which was diagnosed clinically as basal cell carcinoma showed features of malignancy like high mitotic rate (3-5/HPF), hyperchromatic nuclei, central necrotic debris, and lymphoplasmacytic infiltrate and was diagnosed as pilomatrical carcinoma. Margins were free of the tumor. Past history of this patient revealed swelling at nasolabial angle 1 yr back, which was excised and reconstruction using forehead flap was done and was reported as having pilomatricoma. Now patient presented with recurrence of the tumor since 3 months, measuring 3 cm in diameter at the edge of forehead flap. (Figure 2)

All the patients notes were reviewed to identify any associated medical conditions or family history. There were no relevant past medical history or family history identified as a predisposing factor for the pilomatricoma to occur.

**DISCUSSION:** Pilomatricomas are one of the most common superficial masses excised in children, second only to epidermoid cysts. They are variably termed calcifying epitheliomas (of Malherbe), benign calcifying epitheliomas, and trichilemmal cysts. Pilomatricoma usually occurs as a solitary firm lesion with predilection for head and neck, and upper extremities. Although pilomatricoma can develop in patients of any age, children and young adults are most commonly affected.

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 tumors below the age of thirty. In contrast, most of the studies concluded that 1-20 years was the most affected age group. Celia et al observed bimodal presentation of pilomatricoma in 1st and 6th decades. Kuddo et al studied 118 cases of pilomatricoma in adults and found that majority of the cases were above 45 years.

A female preponderance is noted in majority of the studies. Our study also demonstrates 3:2 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. In our study, 7 cases (46.7%) presented with a swelling in upper arm followed by head and neck which is in contradiction with other studies.

Pilomatricoma usually present as solitary nodules but multiple occurrences have been observed in 2-10% of the cases. None of the patient in our study presented with multiple pilomatricomas. Tumor diameter in majority of the cases ranged from 0.5 to 3 cm, but lesions up to 15 cm have been reported. In the present study the size was ranging from 2-3 cm.

Radiologic imaging is of little diagnostic value for pilomatricoma. 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.

Histopathologic examination is often needed for definitive diagnosis. It shows well-circumscribed nodule in the deep dermis and subcutaneous tissue having no epidermis connection. Tumor is composed of nodules with nucleated basaloid cells peripherally and anucleated 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%. We observed calcification in 47% of our cases. In previous articles, osseous metaplasia is noted in 15% of cases owing to conversion of fibroblasts into osteoblasts.

Malignant change of pilomatricoma is rare. In the literature, about 60 cases of malignant transformation have been documented until date. The principal indicators of malignancy are cellular pleomorphism, frequent mitotic figures or atypia, central necrosis, and infiltration of the skin, soft tissues, and lymphatic or vascular elements. The malignant version of pilomatricoma is more frequent in males (with a ratio of 3:1) and in elderly individuals (60% were over 40 years in age). Malignant pilomatricoma had the same site distribution as the benign version of the tumor. Once the diagnosis was confirmed, management consisted of radical excision with adequate resection margins due to the high associated relapse rate in some studies. There was malignant transformation in a male patient in 4th decade. There was atypia, central necrosis and mitotic activity of 3-5/HPF. There was no infiltration of the skin, soft tissues, and lymphatic or vascular elements.

CONCLUSION: 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. Though malignant transformation of pilomatricoma is rare, it should be considered in case of recurrence.

REFERENCES:


Table 1: Clinical diagnosis in 15 cases.

<table>
<thead>
<tr>
<th>Clinical Diagnosis</th>
<th>No. of cases</th>
<th>Percentage</th>
</tr>
</thead>
<tbody>
<tr>
<td>Sebaceous cyst</td>
<td>8</td>
<td>53.3%</td>
</tr>
<tr>
<td>Dermoid</td>
<td>2</td>
<td>13.3%</td>
</tr>
<tr>
<td>Lipoma</td>
<td>1</td>
<td>6.7%</td>
</tr>
<tr>
<td>Antibroma</td>
<td>1</td>
<td>6.7%</td>
</tr>
<tr>
<td>Neurofibroma</td>
<td>1</td>
<td>6.7%</td>
</tr>
<tr>
<td>Calcified hamartoma</td>
<td>1</td>
<td>6.7%</td>
</tr>
<tr>
<td>Basal cell carcinoma</td>
<td>1</td>
<td>6.7%</td>
</tr>
</tbody>
</table>

Figure 1: Pilomatrixoma

Figure 2: Pilomatrix carcinoma
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