A STUDY ON THE CLINICO-HISTOPATHOLOGICAL FEATURES OF PILOMATRICOMA AND ITS MANAGEMENT
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ABSTRACT: Pilomatricoma is commonly a benign neoplasm, which is considered to differentiate towards hair follicular cells particularly the cortex and comprises of basaloid, eosinophilic and shadow cells. It is also described as a sack of infundibular epithelium which is above matrical and supermatrical cells of Pilomatricoma cornifying as shadow cells. It is a slow growing asymptomatic tumor originating from the outer sheath cells of hair follicle. In patients with recurrence for repeat growth of Pilomatricoma, pilomatrix carcinoma may be considered in the differential diagnosis. Clinical presentation and histopathological features of Pilomatricoma are described in this report. Wide local excision is the treatment of choice.

KEYWORDS: Pilomatricoma, Carcinoma, Infundibular, Epithelium, Calcifying.

INTRODUCTION: Malherbe was the first to define Pilomatricoma in 1880 as calcifying epithelioma. Although it was presumed that this tumor originate from sebaceous glands, optical and electron microscopic studies as well as the immuno-histochemical findings confirm this to be Pilomatricoma which was suggested by Peterson WC Jr. and Hult AM, in 1961.\(^1\) Lever and Griesemer suggested that these tumors originated from hair matrix cells. The name Pilomatricoma was suggested by Forbis and Helwing in 1961. It is a benign appendageal tumor with differentiation toward hair follicle cells.\(^2\) The development of this benign dermal subcutaneous tumor derived from the matrix of the hair follicle is associated with the mutation in the CTNNB1 gene, the gene involved in encoding beta-catenin. Normally these tumors form during the first two decades of life and there does not seem to be any predilection for either sex. While the affected sites are generally head and neck\(^3\) it was found in the upper arms as in the present case. In this study a case report of Pilomatricoma is presented along with the complete clinical and histological aspects.

CASE REPORT: A 7 year old male child presented with the swelling in the lower third of the right arm measuring 2x1cms with multiple episodes of pain for the past 6 months (Fig. 1). The swelling was firm in consistency and the skin was not pinchable. The swelling was excised in toto. The cut section showed homogeneous pearly white outer surface with a well demarcated tumor in the deep dermis. Microscopic findings of the sectioned skin was found to be covered with globular tissue. The tumor has a connective tissue capsule and has islands of shadow cells with minimal to absent basophilic cells with large areas of calcification (Fig. 2).
**Histopathological Examination:** The histopathological examination of the excised specimen displayed typical histology of pilomatricoma, consisting mainly of nests of basaloid cells, few islands of ghost/shadow (Fig. 3) cells along with many giant cells and clear areas of calcification (Fig. 4 & 5).
DISCUSSION: Pilomatricoma is also referred to as epithelioma and calcifying benign skin adnexal tumor of hair matrix origin. Generally Pilomatricoma is found as a benign dermal subcutaneous tumor primarily derived from the matrix of the hair follicle. A known mutation in the CTNNB1 gene, the gene that encodes for beta-catenin seems to be associated with its development. Pilomatricomas are relatively common tumors and they usually arise during the first two decades and they have no predilection for either sex; Gupta, Marwah. et al.4 Pilomatricoma is a benign cutaneous appendageal tumour with differentiation towards inner sheath of the normal hair follicle and cortex. Generally, it occurs in the hair bearing areas with a predilection for the head and neck region, as well as the upper extremities.5,6,7 Clinically, it presents as a solitary slow growing dermal or subcutaneous nodule commonly seen in children and young adults.

The patients usually present with a solitary nodule, growing slowly over several months or years. In most of the cases the patients are asymptomatic with a few patients complaining of pain. The lesions range from 0.5-3.0 cm but rarely, lesion as large as 5 cm have been reported so far.8 The lesion comprises of a solitary, rubbery to hard mass. In this reported case it measured 2x1cms. And on palpation it appeared as single, firm and stony hard nodule. Plain X-rays revealed nonspecific calcification while ultrasonography shows a well-defined round hyperechoic mass with post dense acoustic shadows. Computed tomography and magnetic resonance imaging reveal a sharply demarcated subcutaneous opaque lesion that does not enhance on injection of contrast media or small areas of single dropout consistent with the presence of calcification. The cytologic diagnosis is based on combination of basaloid cells and foreign body giant cells against an inflammatory background. Basaloid cells have high nucleolus: Cytoplasm (N:C) ratio and can be seen as single cells or in clusters, sheets or as bare nuclei in the background while ghost cells are usually seen in clusters. Fine needle aspiration cytology may misdiagnose pilomatricoma as round cell tumor.9

CONCLUSION: The histopathological and other studies carried out on the patient with nodular swelling in the arm, growing slowly showed to be a case of Pilomatricoma, a benign skin neoplasm of hair matrix cells. Typically it displays cellular evolution encompassing over parts of the hair follicle such as the outer and inner aspects of the root as well as the sub-cutaneous infundibular components. Calcification is a common finding. A study of the literature shows such like features are rarely traced, hence, a correct diagnosis could be missed when it is based on fine needle aspiration cytology or small biopsies. This case report highlights the importance of histopathological diagnosis of pilomatricoma with a stress on the fact fine needle aspiration cytology may be misleading diagnosis of this benign lesion

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REFERENCES:
1. Peterson WC Jr, Hult AM. Calcifying epithelioma of Malherbe. Arch Dermatol 1964; 90: 404-10.
2. Hurt MA, Kaddu S, Kutzner H, Cribier B, Schulz T, Hartschuh W. Benign tumours with follicular differentiation in Pathology and Genetics of Skin: 152-163 tumours. In: LeBoit PE, Burg G, Weedon D, Sarasin A, editors. Lyon: IARC Press; 2006.
3. Kaddu S, Soyer HP, Wolf IH, et al. Proliferating Pilomatrixcoma. A histopathologic simulator of metrical carcinoma. J Cutan Pathol 1997; 24; 228-34.
4. Gupta, Veena; Marwah, Nisha; Jain, Promil; et. al.; Diagnostic pitfalls of Pilomatrixoma on fine needle aspiration cytology; Iranian Journal of Ddermatology, Vol. 15, 2, summer 2012.
5. Sakai A, Maruyama Y, Hayashi A. Proliferating Pilomatrixcoma: a subset of Pilomatrixcoma. J Plast Reconstr Aesthet Surg 2008; 61: 811-14.
6. Nilyama S, Amoh Y, Saito N, et al. Proliferating Pilomatrixcotma. Eur J. Dermatol 2009; 19: 188-9.
7. Biernat W, Wozniak L. P53 expression in sweat gland tumours. Pol J Pathol 1996; 47: 3-6.
8. Duflo S, Nicollas R, Roman S, et al. Pilomatrixoma of the head and neck in children: A study of 38 cases and a review of the literature. Arch Otolaryngol Head neck Surg 1998; 124 (11): 1239-42.
9. Agrawal L, Kaur P, Singh J, Singh N. Pilomatrixoma misdiagnosed as round cell tumor on fine-needle aspiration cytology. Indian J Cancer 2010; 47: 483-5.

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