Hey! Not A Sebaceous Cyst, I Am the Pilomatrixoma—A Case Report

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Abstract: **With very low incidence, recurrence, and initial diagnostic accuracy, pilomatricoma is usually an isolated benign tumor of the hair follicle matrix. It is a hair matrix differentiation tumor that has historically been described as consisting of regions of calcification, multinucleated giant cells, basaloid and shadow cells, and other components. However, this tumor has a wide variety of histologic characteristics that are frequently overlooked. It affects women and children more regularly. For this kind of tumor, the cervical and face regions are the most common places. Pilomatricoma diagnosis needs to be validated histologically and clinically. We report the rare case of a pilomatricoma, in an unusual location of the right lateral forearm.**

Keywords: **Sebaceous cyst, swelling, Pilomatrixoma, Pilomatricoma, calcifying epithelioma of Malherbe, a rare tumor**

INTRODUCTION

Pilomatricoma is a rare, benign, annexic skin tumor originating from the pilar matrix's cells. It was previously known as calcifying epithelioma of Malherbe and Pilomatrixoma [1]. Malherbe and Chenantois initially reported it in 1880, although Forbes and Helwig originally suggested the term "pilomatrixoma" in 1961[2,3]. The literature indicates that pilomatrixoma affects individuals under 30 years old [4]. Pilomatrixoma tumors are known as "calcifying epithelioma of Malherbe" because they might be solid, partly solid, cystic, or contain calcium deposits [2]. Surgical excision with well-defined margins is the best course of action. Moreover, this lowers the possibility of malignant transformation [5]. It usually consists of calcified "shadow cells, eosinophilic, and basaloid cells"[6].

From a histological perspective, pilomatrixoma appears as a clearly defined lesion that originates in the dermis and spreads into the subcutaneous fat. Traditionally, they comprise islands of epithelial cells that include ghost cells, which have an unstained center region that suggests a missing nucleus, and basophilic cells, which have little cytoplasm. Dystrophic calcifications, foreign-body giant-cell responses, and shadow cells are also included [7,8].

Pilomatricomas are uncommonly detected on the chest, trunk, or lower limbs; they are typically asymptomatic and localized to the head, neck, and upper extremities. Although they are said to affect persons of all ages, the distribution seems to favor young people and the elderly [9]. After epidermoid cysts, they are the second most frequently removed superficial masses in children, excluding lymph nodes [10]. Pilomatrixoma, which is better defined but still often misdiagnosed and not taken into account in differential diagnoses, is characterized by bluish discoloration or ulceration of the overlying skin and is linked to a known mutation in the beta-catenin-encoding CTNNB1 gene. The lesion commonly appears on the skin as a single, rubbery, hard mass known as a "tent sign." These tumors have a maximum diameter of 3 cm. Their sliced surface is calcified from the tumor, making it grittier to chalkier [7,8]. It is characterized by a single, occasionally numerous, benign solid lesion that lies just beneath or within the skin and often develops slowly. [11,12] A total of 0.12% of skin tumors are pilomatrixoma.[13] Here, we describe a case of pilomatrixoma that was identified through histopathology following surgical excision.

Figure 1.0: A showing swelling on the left arm before excision & A1 shows during wide excision.

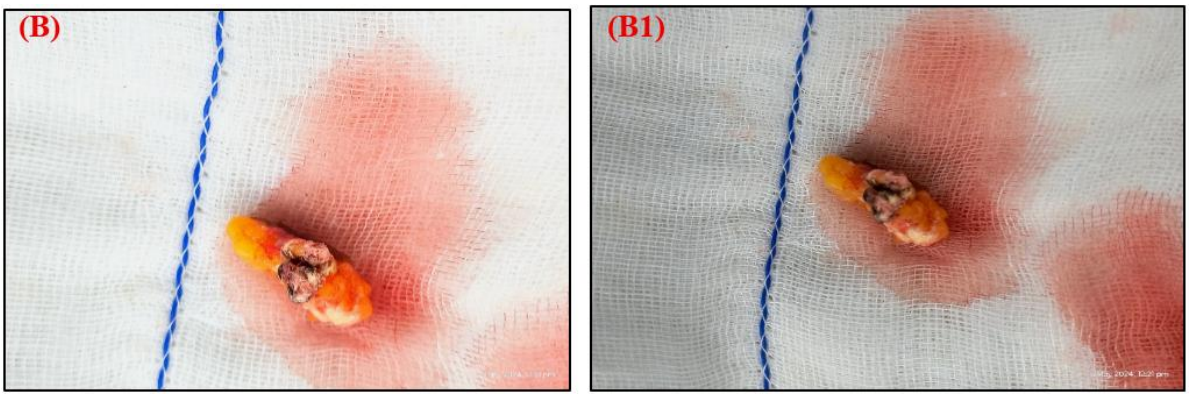

Figure 2.0: B & B1 showing specimen excised post-surgery.

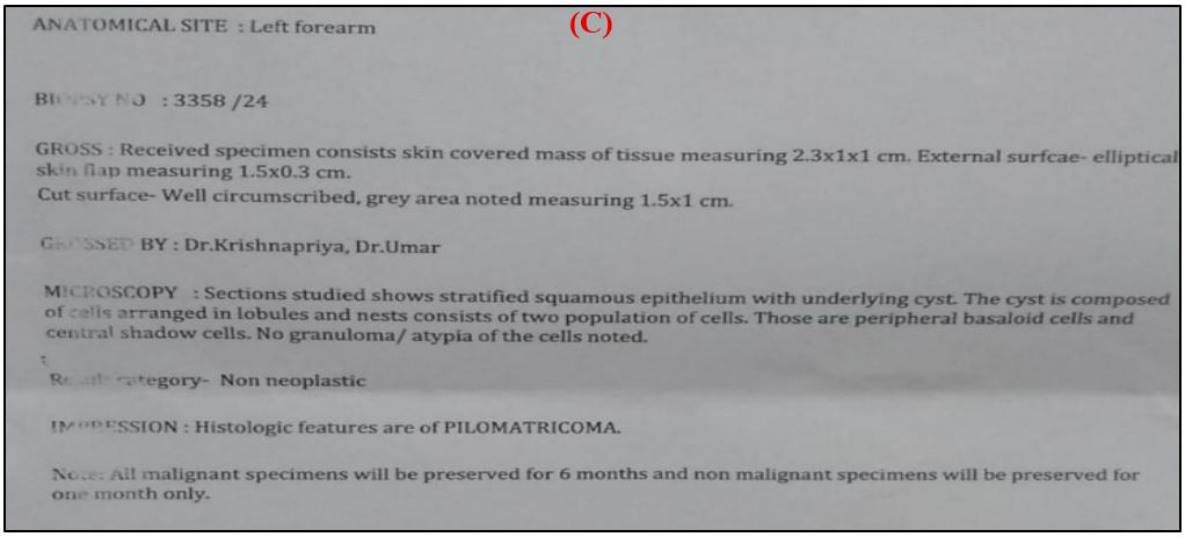

Figure 3.0: C Shows the H.P.E report of the specimen s/o PILOMATRICOMA.

CASE REPORT

A 30-year-old female presented as an outpatient in the surgery department with a complaint of swelling over the upper lateral aspect of her left forearm for 3 years. The onset was gradual and initially, the size was very small (pea-sized) but slowly over the last 1 year it had grown to the size of a big date. It was painless to begin with but there was an occasional dull ache in the swelling. There was no associated fever or malaise. The patient denied any history of trauma before onset. The family history and history of the patient were unremarkable.

On examination, there was swelling in the lateral aspect of the left upper forearm just above the elbow joint, of 3.2 × 2.7 cm size [Figure 1]. It was blue to black with central blackening and smooth on palpation on margins, but stony firm to hard in consistency at the center towards the medial aspect with the feeling of sand particles or small stones in it, and minimally tender to touch, at its center a small horn stony hard was palpable. It could be moved laterally with no restriction on any side. It appeared

to be fixed with the skin with no attachment to the underlying tissues. There was no rise in local temperature, scar, sinus, or any other remarkable feature. General physical and systemic examination was found normal. The patient's vitals were BP- 130/80mmHg, P- 78 BPM, Spo2- 99% on RA, and Temp- 98.4 F. There was no history of any comorbid conditions or history of any medication use of past surgery or any drug allergy.

A provisional clinical diagnosis of sebaceous cyst was made. A surgical excision was planned after blood investigations. The swelling was excised with a wide elliptical incision about 2 cm away from the cyst wall to secure the swelling wall and avoid it from rupturing, was done under local anesthesia, and sent for histopathological examination. After removing the swelling was examined, and it was found that the excised mass was calcified with a wall around it and some lime and sand-based hard form inside the wall, there was also a central horn made of the same material which was hard non-mouldable on pressing it firmly. Due to these unusual features, it was sent for H.P.E., The histopathology features were suggestive of pilomatrixoma.

DISCUSSION

A quite uncommon skin neoplasia is called Pilomatrixoma. It can strike people at any age, with the first and sixth decades of life seeing the highest occurrence. Among young people, it is more common in women (1.5 to 2.5:1), with 40% occurring before the age of 10 and 60% before the age of 20. [14,15] Only a portion of hair follicles are stimulated during puberty; none form after birth. Differentiation induction agents won't work on them correctly if they are in dense layers. These follicles that had partially developed would eventually produce pilomatricomas.[14] These tumors may be inherited in relation to sarcoidosis, Steinerd's disease, or Gardner's syndrome.[16]. Pilomatricomas appear as hard, single, cutaneous, benign lesions of the neck, face, and upper limbs. Pilomatrixomas can have a variable morphology and occasionally an uncommon look that resembles more common tumors, making diagnosis difficult. These subcutaneous nodules can mimic malignant diseases like squamous cell carcinoma or benign lesions like keratoacanthoma, ossifying hematoma, and fibroxanthoma in their presentation. It is crucial to diagnose this lesion because despite being incredibly uncommon—fewer than 20 cases have been reported in the literature—pilomatrixoma have the potential to malignously change into pilomatrix carcinomas. [17,18]. The majority of pilomatrix carcinomas affect patients in their medium to late years on the head and neck. Due to the general rarity of the disease and the absence of distinctive characteristics that can determine if a malignant pilomatrixoma has developed from scratch or if it is the malignant transformation of an already-existing pilomatrixoma, it is challenging to determine the rate of malignant transformation.[19] Pathological examination is still the sole method of diagnosis that is genuinely trustworthy. It is said that the presence of basophilic cells and ghost or shadow cells characterizes classical histology. The histological pattern of a well-circumscribed nodulo-cystic tumor is typically observed at low power in pilomatricomas. The lower dermis is where it is typically observed, however it can sometimes extend into the subcutaneous tissue.

With little knowledge of a potential transition to pilomatrical carcinoma, pilomatricomas are typically thought of as benign tumors. It is crucial to include these lesions in the differential diagnosis because of the clinical challenge of differentiating pilomatrixoma from more frequent skin lesions and the patient population's less-than-ideal medical access. Surgery is used as a treatment, with large 1-2 cm margins. Following excision, pilomatricoma recurrences are relatively rare, with an overall rate of 2.6%. [20-22] Little evidence supports recommendations for follow-up care following excision due to the lesion's rarity. The tumor can be completely removed surgically, which is an adequate and curative treatment with a good prognosis for both cosmetic results and preventing the tumor from turning malignant.

Conclusion: It is still difficult to identify Pilomatrixoma clinically because it has similar features of a sebaceous cyst and it is very rarely found, mostly diagnosed on histopathological examination. Pilomatrixoma is overlooked. This lesion is the most common of hair follicle tumors. The location at the level of the limbs remains exceptional. Its diagnosis is clinical, its confirmation is histological, and its treatment is surgical.

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