

## Pilomatricoma of the Neck Misdiagnosed as Sebaceous Cyst

### Sebace Kist Olarak Yanlış Tanı Konulan Boyun Pilomatrikomu

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#### ABSTRACT

Pilomatricoma is a benign skin lesion originating from the hair follicle matrix. Most cases are usually only diagnosed post-operatively through histopathological examination (HPE) mainly because of the nature and presentation that resemble benign and superficial skin lesion, hence, the misdiagnosis prior to surgery. In this case report, we would like to highlight this condition and its important characteristics which appear to be under-reported due to lack of awareness among clinicians.

**Key Words:** Pilomatricoma, pilomatricoma, neck, skin diseases, benign neoplasms

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#### ÖZET

Pilomatricoma, saç folikülü matrisinden kaynaklanan iyi huylu bir deri lezyonudur. Vakaların çoğu genellikle ameliyat sonrası histopatolojik inceleme ile teşhis edilir, çünkü esas olarak iyi huylu ve yüzeysel cilt lezyonuna benzeyen doğası ve sunumu, dolayısıyla cerrahi öncesi yanlış tanıdır. Bu olgu sunumunda, bu durumu ve klinisyenler arasında farkındalık eksikliği nedeniyle yetersiz rapor edilmiş gibi görünen önemli özelliklerini vurgulamak istiyoruz.

**Anahtar Sözcükler:** Pilomatrikom, pilomatricsoma, boyun, cilt hastalıkları, benign neoplazmlar

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**INTRODUCTION**

Pilomatricoma or also known as pilomatrixoma used to be regarded as an uncommon benign skin lesion that arises from the hair matrix cell (1). Pilomatricoma constitutes only 1% of all benign cutaneous entity (2). It can affect any age group but predominantly young adults and children. <sup>2</sup> There is a minimal sexual predilection towards male with ratio of 0.97:1 (3). Most pilomatricoma occur in the head and neck region with the highest occurrence over the neck (30.2%) followed by cheeks (16.8%), scalp (16.2%) and lastly periorbital area (14.0%) (3).

**CASE REPORT**

A 18 year-old healthy gentleman presented with 9 months history of a painless right lower neck swelling. The swelling was not associated with odynophagia, dysphagia, hoarseness and constitutional symptoms. Prior to the presentation, he was diagnosed with sebaceous cyst over the neck and an excision was performed in a nearby clinic about 6 months ago. The swelling recur approximately 2 months after the procedure.

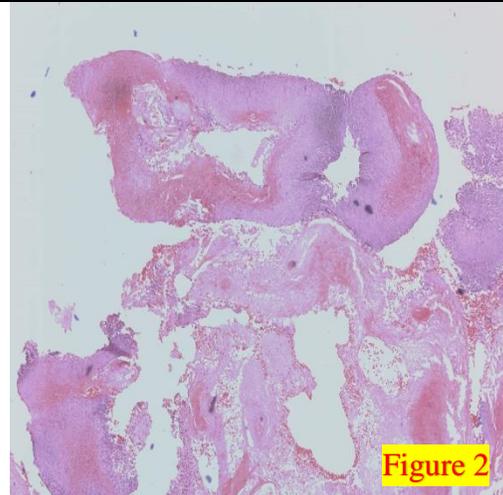
A firm, non-tender, superficial swelling size of 1 x 1 cm was palpated over the right level IV region of the neck (Figure 1). There was no other swelling over the head and neck region and other parts of the body. Other ear, nose and throat examinations including endoscopy of the nose and larynx were unremarkable.



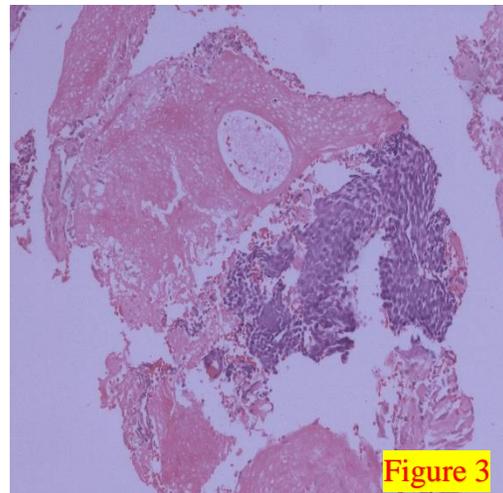
**Figure 1:** Single dark red-bluish well circumscribed swelling that is fixed to the overlying skin.

The mass was excised together with the skin. The mass was found to be gritty and adhered to the subcutaneous tissue and skin. Complete excision was done with cold instruments and sent for histopathological examination (HPE).

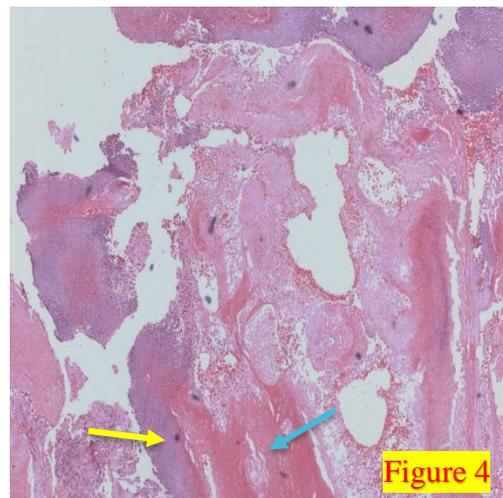
Microscopically, the specimen had biphasic population and multiple foci of basaloid cells proliferation (Figure 2, 3 and 4). The cells were undergoing abrupt trichilemmal type keratinization (Figure 5). There are presence of eosinophilic cells lacking nuclei that exhibited shadow or ghost cells (Figure 6 and 7). Multinucleated giant cells with histiocytic infiltration were also present. HPE was consistent with pilomatricoma. Post-operative period was uneventful and at 1 year follow up, he was found to be well and free of recurrence.



**Figure 2:** Biphasic population seen under microscope. (x10 magnification)



**Figure 3:** Tumor shows biphasic population with peripheral basaloid cells and central shadow cells. (x4 magnification)



**Figure 4:** Two cell types are present – nest of basaloid cells (yellow arrow) and shadow cells (blue arrow). (x4 magnification)

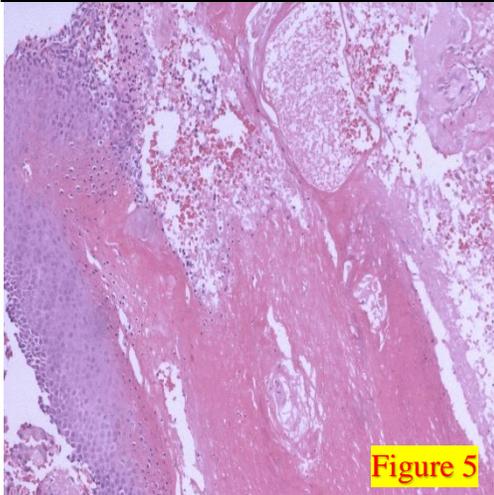


Figure 5

**Figure 5:** The basaloid cells show abrupt trichilemmal-type keratinization. (x10 magnification)

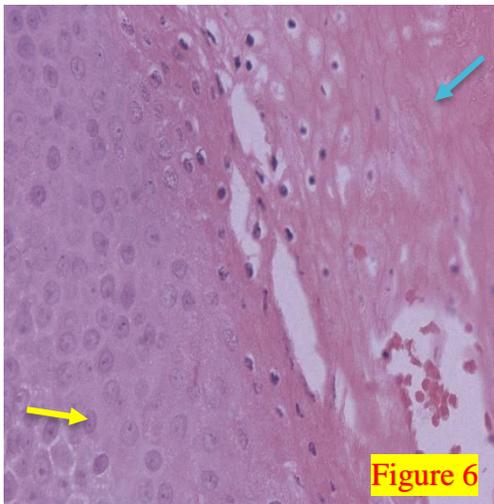


Figure 6

**Figure 6:** Basaloid cells (yellow arrow) display round to oval hyperchromatic nuclei with small nucleoli and scanty cytoplasm. The shadow cells (blue arrow) display eosinophilic cytoplasm with pyknotic or absent of nuclei. (x40 magnification)

## DISCUSSION

In the 1880, Malherbe and Chenantais described this condition as benign subcutaneous tumor of sebaceous glands and named it calcifying epithelioma of Malherbe (1). The term pilomatricoma was coined 81 years later by Forbis and Helwig to avoid confusion of nomenclature that was previously similar to malignancy (1). Approximately 75% patients with pilomatricoma present with single, painless swelling measuring less than 1.5 cm (2). Only 1 patient out of 179 cases presented with multiple lesions (3). The lesion may be painful in event of skin ulceration or associated inflammation (4).

A classical 'tent sign' described by Graham and Merwin can be exhibited by stretching of the skin over the tumor to feel the irregularity and multifaceted mass underneath the skin (5). The tent sign was not exhibited in this patient probably due to the fibrosis of overlying skin after previous surgical procedure 6 months ago. The tumor may appear to be discolored with bluish-reddish hue especially in individuals with fairer skin tone as demonstrated in this patient.

On pressing of the lesion, a 'teeter-totter' sign that causes the opposite edge of the skin to bulge can be elicited but was not present in this patient as the mass was relatively small to begin with (4). Classical characteristics of pilomatricoma are probably often overlooked in many cases due to darker skin tone or simply lack of awareness.

In the literature, some authors found association with previous regional trauma prior to development of pilomatricoma (1, 6). Patients with multiple pilomatricomas also had either Turner syndrome, Sticker syndrome, Gardner syndrome, Steinert disease, myotonic dystrophy or sarcoidosis. However, the significance and pathophysiology of these relations are still unknown due to sparsity of reported cases (6, 7). In spite of well described clinical characteristic of pilomatricoma, preoperative diagnosis remain very low, ranging between 0–30% in terms of diagnostic's accuracy (8). Some lesions may be firm, some cystic while other cases may be multiple lesions, ulcerating or infected. In terms of rarity among benign skin lesions, clinicians are more accustomed to more familiar lesions like sebaceous cyst in this case and other lesions as well such as inclusion cyst, dermoid cyst and brachial cleft remnants (4).

Imaging test and fine needle aspiration cytology (FNAC) are of little help in diagnosis. FNAC can even be misleading and cause misdiagnosis especially when aspiration do not show ghost cell which is the hallmark of pilomatricoma (9). Other microscopic characteristic of pilomatricoma include presence of basaloid cells and calcium deposition (9). Therefore surgical excision can be both diagnostic and therapeutic (2, 3). The recurrence rate is almost 0% when the lesion is completely excised (3, 7). In this case, the first procedure performed in clinic may be incomplete hence the recurrence of lesion after 2 months of surgical procedure.

## CONCLUSION

Pilomatricoma was previously thought to be a rare lesion due to lack of awareness and under-reporting. In young adult and children that present with firm and superficial head and neck masses, the list of differential diagnosis should include pilomatricoma. Careful clinical examination and high index of suspicion may improve pre-operative diagnosis of pilomatricoma thus aiding in surgical planning and also preventing unnecessary imaging and laboratory tests.

## Conflict of interest

No conflict of interest was declared by the authors.

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