

## Pilomatricoma of the Retroauricular Area

### Retroaurikuler Bölgede Pilomatrikoma

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#### ABSTRACT

Pilomatricoma is a benign skin tumor of hair matrix cells. The head and neck is the most commonly affected site of the body. This article describes 41 year-old man who referred to our clinic with an asymptomatic, slow-growing mass on the right retroauricular, suboccipital region. The tumor was excised and histopathologically diagnosed as pilomatricoma. Because of its frequency and predilection of head and neck region, otolaryngologists should be familiar of this tumor.

**Key Words:** Pilomatricoma, skin neoplasm, calcifying epithelioma.

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#### ÖZET

Pilomatrikoma, kıl folliküllerinin matris hücrelerinden köken alan benign deri tümörleridir. Bu tümörün en sık görüldüğü yerleşim yeri baş ve boyun bölgesidir. Bu yazıda, 41 yaşında sağ retroauriküler, suboccipital bölgede semptomsuz, yavaş büyüyen kitle ile başvuran bir hasta tarif edilmektedir. Bu tümör çıkarılarak histopatolojik incelemeye gönderilmiş ve pilomatrikoma tanısı almıştır. Baş ve boyun bölgesinde sık görülme eğilimi nedeniyle kulak burun boğaz hekimlerinin bu tümörün klinik sunumunun bilincinde olması ve baş boyun kitlelerinin ayırıcı tanısında bu tümörleri akla getirmesi önerilmektedir.

**Anahtar Sözcükler:** Pilomatrikoma, deri tümörü, kalsifiye epitelyoma

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#### INTRODUCTION

Pilomatricoma, also described as calcifying epithelioma of Malherbe is a skin neoplasm, which originates from hair matrix cells (1-3). This tumor is benign and the neck is the one of the most commonly affected sites of the body. This tumor is actually not uncommon neoplasia that pathologist confirmed this diagnosis for one of every 500 specimens (4). Despite the frequency of pilomatricoma, it is rarely come to mind as prediagnosis by clinicians (5). Its varying presentations may be a cause of misdiagnosis. Thus, it should be important to be aware of this tumor. This article describes a case of pilomatricoma in the retro-auricular, sub-occipital region and discuss this neoplasia.

#### CASE REPORT

A 41 year-old male patient referred to our clinic with a right retro-auricular, sub-occipital mass. The tumor has grown in 8 months. No pain, discharge or discoloration occurred in this area. There were no other masses or lymphadenopathy or significant medical history of the patient. The superficial, solid, mobile mass measured approximately 3 cm x 1,5 cm in the right retro-auricular, sub-occipital region (Figure 1). The other parts of ENT examination and systemic examination was normal. Superficial ultrasound examination revealed a 3 cmx 1,5 cm subcutaneous cystic lesion with containing solid areas. Excision surgery was performed under general anesthesia.

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Skin incision was made in the sub-occipital region and then by proceeding with blunt dissection of the subcutaneous tissue, the mass was achieved. A firm calcified mass, which is non-adherent to the surrounding tissues was excised (Figure 2). The pathologist explained the diagnosis as pilomatricoma (Figure 3).

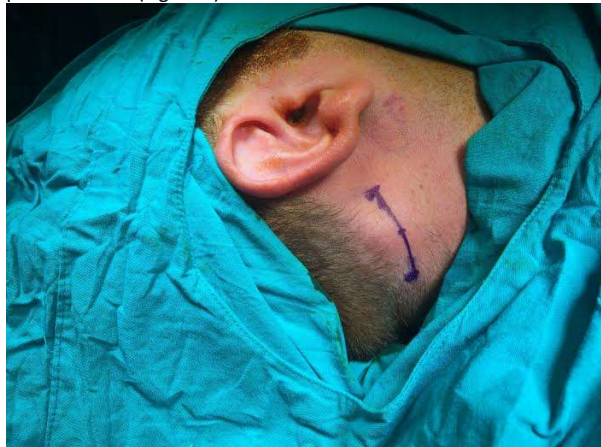


Figure 1: Photograph of a mass in the retroauricular, suboccipital region.



Figure 2: Intraoperative photograph of a firm, calcified mass

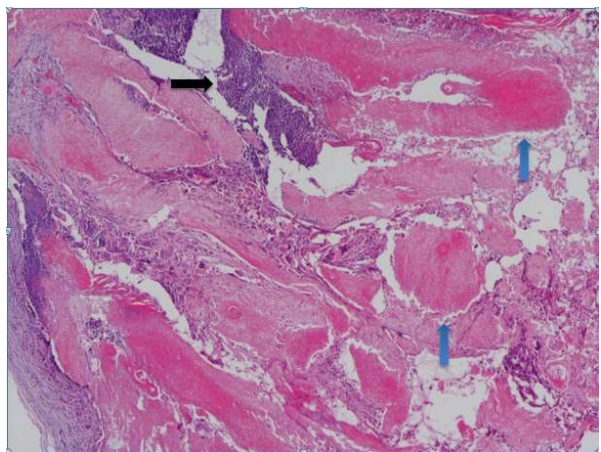


Figure 3: Photomicrograph of pilomatricoma showing the characteristic basaloid (black arrow) and (blue arrow) shadow cells. Among them, multinucleated giant cells with chronic inflammatory infiltrates are also seen (H&E, x40)

## DISCUSSION

The formation mechanism of pilomatricoma, which is the tumor of hair matrix cell, had explained by the failure of the hair follicle cycle that cause the breakdown in the differentiation of pilar keratinocytes into mature hair follicles. (6,7). Furthermore, the association of pilomatricoma with the genes for myotonic dystrophy, polyoma virüs have been demonstrated (8,9).  $\beta$ -catenin gene mutations have also been shown to be associated with pilomatricoma (10).

Clinically, pilomatricoma presents as a superficial, firm, freely mobile, cystic or solid mass. Discoloration as reddish-blue or ulceration of the skin may be seen. These tumors generally are painless, occasionally they present with pain and tenderness (5, 6, 11,12). Pilomatricoma commonly occurs at 5-15 years and at 50-65 years (13) and it has female predominance (5). Although these tumors are solitary lesions, multiple lesions reported in patients with myotonic dystrophy, Turner syndrome, Gardner syndrome, Steinert disease (5, 14). Pilomatricoma can be usually seen in haired parts of the body. Thus, they occur most frequently in the cervicofacial region and the limbs are the second common area. In the head and neck region, because of the high density of hair follicles, the cheek, neck, peri-auricular, and peri-orbital are the common areas fo this tumor. (6,11,15). To avoid misdiagnosis, benign and malignant tumors such as sebaceous and epidermoid cysts, basal cells and scuamous cell carcinoma, vascular lesion, lymph node, lipoma, branchial remnants, preauricular sinuses, chondroma and parotid region tumors should be considered before intervention to the mass (5,12,16,17).

For diagnostic investigation, ultrasonography can be performed for the determination of the position and the calcification of the mass, especially for young patients (6). Computed tomography or magnetic resonance imaging are mainly requested for certain cases such as preauricular and extensive tumors (5, 12). As a part of diagnostic investigation fine needle aspiration has been described (18).

Histopathologically, pilomatricoma is markedly limited nodules with a fibrous capsule. The islands of well-organized malpighian cells are the most frequent microscopic features. The tumor contains basaloid cells with nucleus settled in the peripheral of the island and eosinophilic shadow cells (ghost cells) without nucleus settled in the center (5,6,12). Calsification in the ghost cell region is common feature that ranges from 69% to 85% (18). Malign forms have atypical, undifferentiated cells, a large epithelial cell component and may infiltrate blood vessels and fibrous capsule (12,20).

The target of treatment is the total removal of the tumor (5). This tumor has low recurrence and malignant transformation rate (6). Malignant transformation may occur with advancing ages in males (20). Also death due to the malignant metastatic pilomatricoma was reported (21).

## CONCLUSION

Pilomatricoma is one of the most common benign skin neoplasm. Otolaryngologists should consider this tumor in a patient who presented with mobile, superficial, slowly progressive mass in the head and neck region. Clilical presentation is usually suggestive for the diagnosis. Radiologic assesment is important in certain cases. Complete resection of the mass is essential for the treatment. Recurrence and malignant transformation rates are low.

## Conflict of interest

No conflict of interest was declared by the authors.

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