



# Perforating pilomatrixoma showing atypical presentation: A rare clinical variant

*Atipik prezentasyon gösteren perforan pilomatriksoma: Nadir bir klinik varyant*

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## Abstract

Pilomatrixoma, also known as calcifying epithelioma of Malherbe, is a rare benign skin tumor arising from hair follicle stem cells. The most common localization is the head and neck region. Female/male ratio is 3/2. It shows deep subcutaneous placement and occurs in the first two decades of life. Its diameter ranges from 0.5 cm to 3 cm. Multiple lesions are rarely seen. Histopathologically it is characterized by basoid and ghost cells. Perforating type is a rare clinical variant. Treatment is surgical excision. Our case is presented to draw attention to a rare clinical variant of pilomatrixoma.

**Keywords:** Pilomatrixoma, atypical presentation, bilateral localization

## Öz

Malherbe'nin kalsifiye epitelyoması olarak bilinen pilomatriksoma kıl folikülünden kaynaklanan nadir, selim bir deri tümörüdür. En sık baş boyun bölgesinde yerleşim göstermektedir. Kadın/erkek oranı 3/2'dir. Derin subkutan yerleşim gösterir ve hayatın ilk iki dekadında ortaya çıkar. Çapı 0,5 cm ile 3 cm arasında değişmektedir. Birden fazla lezyon görülmesi nadirdir. Histopatolojik olarak bazoloid hücreler ve gölge hücreler ile karakterizedir. Perforan (ekzofitik) tipi nadir bir klinik varyanttır. Tedavisi cerrahi eksizyondur. Olgumuz pilomatriksomanın az rastlanan bir klinik varyantına dikkat çekmek amacıyla sunulmuştur.

**Anahtar Kelimeler:** Pilomatriksoma, atipik prezentasyon, bilateral yerleşim

## Introduction

Pilomatrixoma (calcifying epithelioma of Malherbe) is a benign tumor, which originates from matrix cells of the hair follicles. It was first described by Malherbe<sup>1</sup>. It accounts for 0.1% of skin tumors. It occurs most often in the first two decades of life<sup>2</sup>. The mass is localized in the head and neck region in approximately 50% of cases and rarely in the trunk and extremities<sup>3</sup>. Clinically, pilomatrixomas are solitary painless well-defined subcutaneous tumors. Frequently, it is seen as a firm, bluish red color cutaneous or subcutaneous single mass. It is fixed to the overlying skin but sometimes may be mobile. Epithelial thinning or even ulceration may occur. It is more common in Caucasians. Female/male ratio

is 3:2<sup>4</sup>. It shows slow progression. Multiple lesions have been reported in 3.5% of cases<sup>5</sup>. Treatment is surgical excision. The definite diagnosis is made histopathologically. The aim of this article was to evaluate clinical and histopathological characteristics of the tumor.

## Case Report

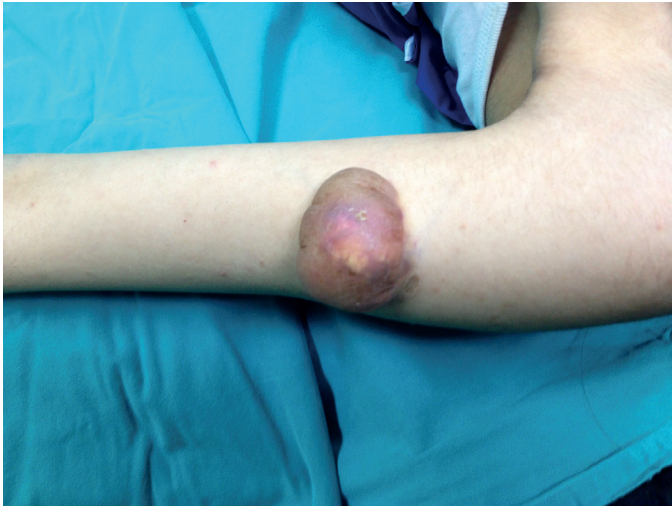
A 17-year-old female patient presented to our clinic with the complaint of masses on both arms. In her history, she declared that she palpated small mobile masses that have increased in diameter in the past 6-7 years. Laboratory findings were normal. In her physical examination, there were two hard mobile masses disrupting the epithelial architecture

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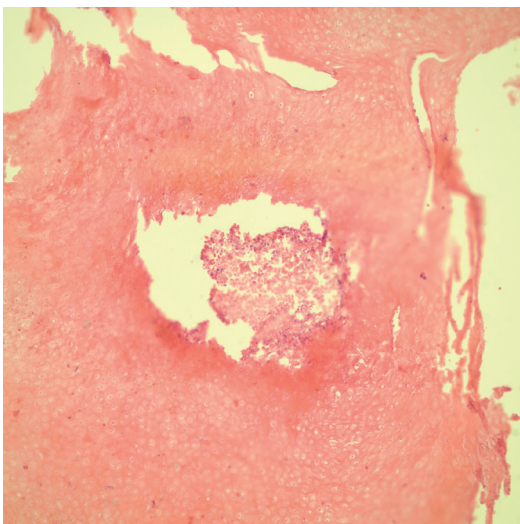
symmetrically localized on the extensor site of the right and left forearms (Figure 1, 2). There was purple discoloration of the overlying skin. The masses were totally excised together with the surrounding



**Figure 1.** Preoperative appearance of the mass localised on the left arm

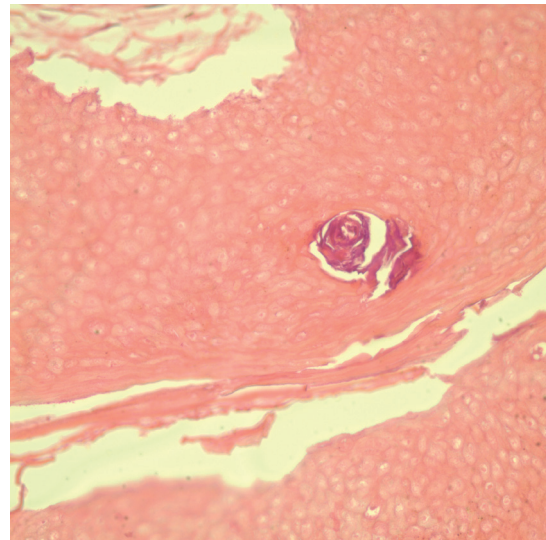


**Figure 2.** Preoperative appearance of the mass localised on the right arm



**Figure 3.** Microscopic view of pilomatrixoma revealing ghost cells (hematoxylin and eosin x200)

healthy tissue with a safety border of a 2-3 cm and the incisions were closed with sutures. The surgical specimens were sent to pathologic examination for definitive diagnosis. In microscopic examination, a neoplasm with large eosinophilic ghost cells and peripherally and centrally localized basaloid cell islands with calcification, and foreign body giant cells were observed. Histopathology, the result was reported as pilomatrixoma (Figure 3, 4). Recurrence was not observed in one-year follow-up period.



**Figure 4.** Microscopic view of pilomatrixoma revealing psammom calcification (hematoxylin and eosin x200)

## Discussion

Pilomatrixoma is a slow growing asymptomatic benign tumor. It may occur at any age but 60% of patients are younger than 20 years. The diameter ranges between 0.5 cm and 3 cm<sup>6</sup>. Progression is slow and it may take months to years. Besides, the most commonly seen head and neck region, the tumor may also be observed in the upper extremities, trunk and the lower extremities with increasing order of frequency. It is multiple in 2-3% of cases. Multiple familial pilomatrixomas are associated with myotonic dystrophy, Gardner syndrome and Rubinstein-Taybi syndrome<sup>7</sup>. The incidence of bilateral localization is 2-3%. Although there are various factors, the etiology is unknown. In recent studies, beta-catenin gene has been found responsible from the repeating mutations<sup>8</sup>. Pilomatrixoma is a deep subepidermal tumor that comprises irregular epithelial cell islands. Histopathologically, it contains ghost cells in the center and cells with basophylic nucleus localized peripherally. Also foreign body giant cells are seen. There is calcification around ghost cells in 70-95% of cases. There are ghost cells in the center. If there is pleomorphic high mitotic activity, local or vascular invasion, the diagnosis is made as pilomatrix carcinoma predicting that pilomatrixoma has underwent malign transformation

The incidence of recurrence is low. It is a well-defined solitary subcutaneous tumor on ultrasonography, magnetic resonance imaging and computed tomography. Direct radiography showing calcification in the epidermal cyst, calcified lymphadenopathy, calcified or ossified hematoma, foreign body and other benign and malign soft tissue tumors must be considered in differential diagnosis<sup>9</sup>. Some of the components

of pilomatrixoma, which disrupt the epidermis and become superficial is called as perforating pilomatrixoma (transepidermal elimination)<sup>10</sup>. Since to observe spontaneous regression is not possible, suggested treatment method is surgical excision. The lateral surgical margin must be 1 cm. The rate of recurrence after surgical intervention is 2-6%<sup>11</sup>. If the tumor is fixed to the dermis, it must be excised with the overlying dermis.

Since our case was a rare perforating type and symmetrically localized in the upper extremities symmetrically, it represents a rare clinical variant of pilomatrixoma.

#### Ethics

**Informed Consent:** Inform consent was taken.

**Peer Review:** External and internal peer-reviewed.

#### Authorship Contributions

Surgical and Medical Practices: N.S., İ.M., Concept: N.S., Design: N.S., Data Collection or Processing: N.S., Analysis or Interpretation: N.S., Literature Search: N.S., Writing: N.S.,

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