

Pilomatrixoma case report: A very rare localization

Pilomatriksoma: Çok nadir bir lokalizasyon

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ABSTRACT

Pilomatrixoma (pilomatricoma) is a benign neoplasm of hair follicle matrix cells of the head and neck. Pilomatrixoma, firm mass found most commonly on the face, neck, back and upper extremity, however is rarely found in the lower extremity. We report a case of a 12-year-old female patient with a rare localisation of pilomatrixoma on the lower extremities.

Key words: Pilomatrixoma, neoplasms, lower Extremity

ÖZET

Pilomatriksoma, baş ve boyundaki kıl foliküllerinin matris hücrelerinden kaynaklanan iyi huylu bir tümördür. Pilomatriksoma, sert bir kitle olarak; sıklıkla yüz, boyun, sırt ve üst ekstremitede bulunur ancak alt ekstremitede nadiren yerleşir. Biz 12 yaşındaki kız hastada alt ekstremitede pilomatriksomanın nadir bir lokalizasyonunu olgu olarak sunuyoruz.

Anahtar kelimeler: Pilomatriksoma, tümörler, alt ekstremit

INTRODUCTION

Pilomatrixoma is a rare, benign appendageal tumour originating from the hair-follicle matrix cells. It was first described by Malherbe and Chenantais in 1880 and therefore it is also known as Malherbe's calcified epithelioma. The tumour is in the form of a subcutaneous nodule and is blue-red or dark red in colour. Usually, it is painless on palpation. It is more frequently encountered in the first two decades and in women. It is mostly seen in the head and neck area. Less frequently it is localized in the torso and the lower extremities [1,2].

Our aim of presenting this case report is to emphasize the very rare localization of the pilomatrixoma in the lower extremities.

CASE REPORT

A 12-year-old female patient was referred to us with the complaint of a painful mass in her left leg for 6 weeks. She denied any changes in the size of the

mass during this period. Her medical and family history was unremarkable.

On admission, physical examination showed a hard, painful, mobile mass measuring 1.5x1 cm at the distal region of the left thigh. The skin covering the mass was blue-red colored.

Superficial ultrasonography revealed a heterogeneous hyperechoic lesion measuring 15x8 mm with excessive bleeding. The mass was totally excised under general anaesthesia and sent to the pathology for tissue diagnosis and treatment.

In the microscopic examination, the tumor is composed of islands of epithelial cells made up of varying amounts of uniform basaloid matrical cells and often shows cystic change. Foreign body giant cells, keratin debris, and central calcifications are also characteristic (Picture 1). Ghost cells characteristically retain their cell and nuclear borders, however, the nuclei lose their basophilic staining leaving a "ghost-like" remnant (Picture 2). The patient was diagnosed with pilomatrixoma. She has been followed with a good clinical condition so far.

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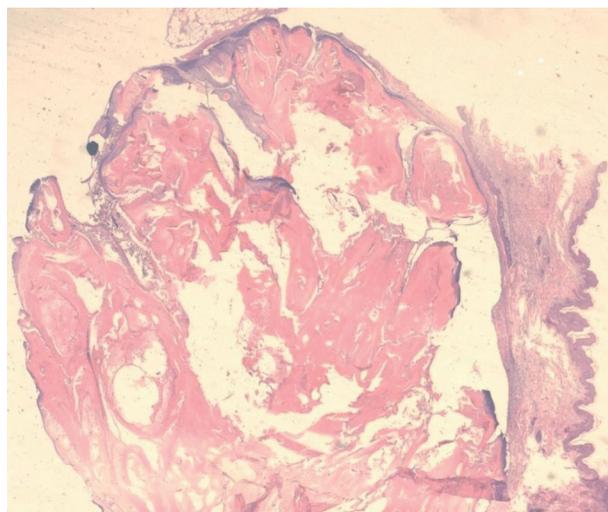
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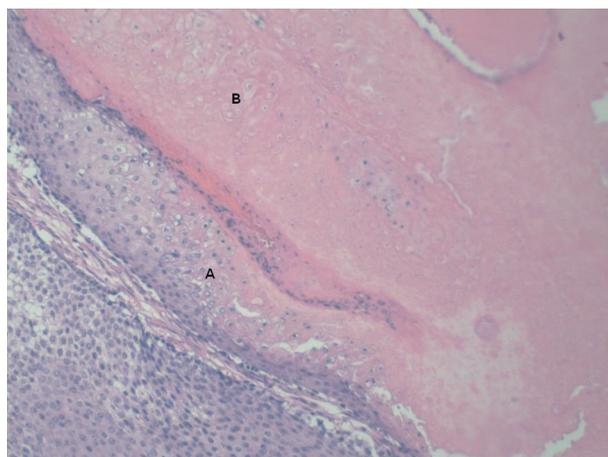
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Picture 1. On the surface, cutaneous tissue part covered with keratinized multi-layered flat epithelium and a tumoral lesion with regular rim localized in the dermis is seen (HE×10).



Picture 2. In the microscopic view of the pilomatrixoma two cell types are evident. The first one is the layer formed by basal cells (A) and the other is the layer formed by shadow 'ghost' cells (B). In between, calcification and bleeding regions are seen (HE×200).

DISCUSSION

In various studies, the incidence of pilomatrixoma was reported to be between 0.03 and 0.1%. Most of the cases belong to the pediatric age group and approximately 90 % of the cases are younger than age of 10. Pilomatrixomas are usually localized in the head and neck area (40-77%), followed by the torso and the upper extremities. Localization in the lower extremities is very rare [2,3]. In our case, the localization of the tumor is in the lower extremity.

Even though trauma and infection were thought as possible triggers of tumor, its etiology remains unknown. We were not able to take any history of trauma and infection in our case.

Clinically, the pilomatrixoma is usually detected as asymptomatic, mobile, subcutaneously localized nodule with possible discoloration of the skin [2]. In our case, however, a lesion was painful with palpation and raised from the skin and there was a blue-red appearance on the skin.

β -catenin is responsible for adhesion between epithelial layers and among the cells. Pilomatrixoma is thought to arise from mutation of the β -catenin gene. There have also been immunohistochemical studies associating the BCL2 proto-oncogene over-expression to pilomatrixoma [4].

In the literature, although rarely, malignant pilomatrixoma cases with distant metastases have also been reported [5].

The local recurrence rate is stated as 0-3% in the literature and it is recommended that the tumour is excised along with 1-2 cm of the substantial tissue. Complete surgical excision is curative and can be done with skin excision [6,7]. We removed the tumour by surgical excision leaving substantial tissue around it.

In conclusion, we suggest that the patients with painful mass on the lower extremity should be evaluated in terms of the pilomatrixoma.

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