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Case Report / Vaka Sunumu

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Pilomatrixoma localized in vulva

Vulvada lokalize pilomatriksoma

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ABSTRACT

Pilomatrixoma is a benign lesion generally found at the back of the head. A 44-year-old woman presented to our outpatient unit with the complaint of a palpable hardness in her vulva. On examination a palpable hard, mobile, painless mass approximately 2.0x1.0 cm in size was detected about 2.5 cm below the left labium major. The overlying skin was intact. After surgical removal of the mass pathologic findings were found compatible with "pilomatrixoma." No recurrence was noted during one year follow-up. Pilomatrixoma should be kept in mind as a diagnosis for dermal or subcutaneous nodules localized outside the head and neck region.

Keywords: *Pilomatrixoma, vulva, nodule*

ÖZ

Pilomatriksoma genellikle kafanın arkasında bulunan benign bir lezyondur. Kırk dört yaşındaki kadın hasta vulvasında ele gelen sert kitle yakınmasıyla başvurdu. Muayenede sol labium majusun yaklaşık 2,5 cm alt kısmında ortalama 2.0×1.0 cm ağrısız, mobil, sert kitle saptandı. Üzerindeki cilt sağlamdı. Çıkarıldıktan sonra patoloji sonucu pilomatriksomayla uyumlu saptandı. Bir yıllık takibinde yineleme olmadı. Kafa ve boyun bölgesi dışındaki dermal veya subkutanöz nodüllerde de pilomatriksoma ayırıcı tanıda akılda bulundurulmalıdır.

Anahtar kelimeler: *Pilomatriksoma, vulva, nodül*

INTRODUCTION

Pilomatrixoma (PM) was first described by Malherbe and Chenantais in 1880. It is a benign tumor originating from the sebaceous gland and has an incidence between 0.03, and 0.1%[1]. Although PM is most common among children, it can be observed in every age group[2]. Clinically, it is detected as a slowly growing subcutaneous or intradermal solitary, asymptomatic, firm nodule. Its size ranges between 0.5, and 3.0 cm[3-6]. Nodules larger than 5.0 cm are rarely seen.

Pilomatrixoma is generally detected as a mobile, hard, elastic mass; symptoms such as pain and precision can also be observed. It can interfere clinically with various benign and malignant skin lesions. After removal, recurrence is very rare. Clinical types of PM, like bullous, giant, perforating, or multinodular PM were also defined[7].

Approximately 50% of PM cases are found to be localized at the head and neck. Rarely, it can localize on the trunk, arm or legs[8]. These tumors can have familial inheritance and they are associated with Gardner syndrome, Steinert disease, and sarcoidosis. In the literature, malignant forms-although less frequent-were also shown. The malignant form is referred to as pilomatrix carcinoma, and it is reported that they can cause metastasis to the lungs, bones, brain, skin, lymph nodes, and abdominal organs[9].

We are presenting the surgical method, histological findings, and literature review concerning a subcutaneous PM case located in the vulva, which is an atypical localization.

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CASE

A 44-year-old woman applied to Dr. Sami Ulus Women Health Care, Training and Reserch Hospital for routine control. She complained of a hardness in the vulva that existed for about 15 years. In the examination, there was no lesion present by inspection, but when palpated, a hard, mobile, painless mass approximately 2.0x1.0 cm in size was detected about 2.5 cm below the left labium major. The overlying skin was normal. Fine needle aspiration of the lesion was performed for differential diagnosis, and hemorrhagic fluid was drained. The diagnosis of the material obtained from this needle aspiration after pathological investigation was "fibrin mass." After one month, the mass was removed by dissecting it from the surrounding tissue with the patient under general anesthesia. In the pathological examination of the 1.5x1.0x0.7 cm mass, there were several shadow cell layers ("ghost cells"), wide dystrophic calcification, surrounded by foreign-body-type giant cells and foreign-body-type granulation tissues consisting of very rare mononuclear inflammatory cells, vascular proliferation, and fibrosis (Figure 1-2). In this area, instances of keratinization were noted in a scattered pattern. Findings were found to be compatible with "pilomatrixoma." Dysplasia or malignancy was not detected. Disease recurrence was not observed during one year follow-up examination.

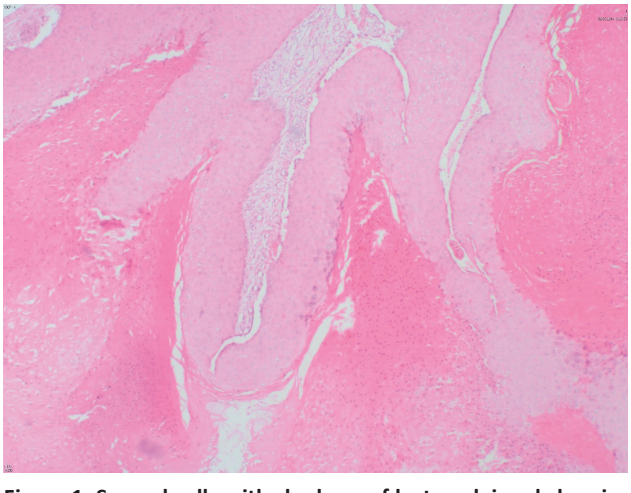

Figure 1. Several cells with shadows of lost nuclei and clear intercellular limits, or "ghost cells."

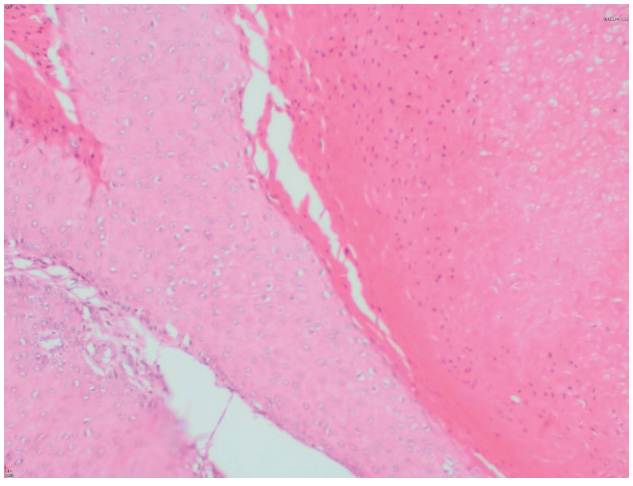

Figure 2. Foreign-body-type giant cells and foreign-body-type granulation tissues consisting of very rare mononuclear inflammatory cells, vascular proliferation, and fibrosis.

DISCUSSION

Pilomatrixoma is a rare neoplasm of the hair follicle that is also referred to as Malharbe calcified epithelioma, trichomatrixoma, and pilomatricoma[1]. In a large sample size of 346 pilomatrixoma cases, most of them were reported as being localized on the head and neck region, with 15.3% of the cases being reported as localized on the upper extremities[7]. Presentation of pilomatrixoma in the vulvar region is unusual. As far as we know, there are only three cases of vulvar pilomatrixoma were reported in the literature[10,11].

PM generally presents as a singular, asymptomatic nodular mass. Patients generally describe a solitary nodule that will gradually grow over several years. There can be episodes of inflammation or ulceration.

Clinical diagnosis is very difficult to make. The overlying skin is generally observed to be normal. When the superficial epidermis is normal, hard subcutaneous tissue can result in herniation from atrophic tissue when squeezed with fingers[3]. In this particular case, any lesion was not detected during inspection. Besides its classical appearance, lesions can be keratotic or telangiectatic, mimicking squamous cell

carcinoma and basal cell carcinoma[12]. There can be blue-black skin discoloration resembling hemangioma. These lesions are generally well-defined, spherical or oval, and sometimes can be encapsulated.

Its exact etiology is not known. In the latest studies, it was shown that repeated mutations in a gene, the β-catenin gene (CTNNB1), can be responsible for the development of PM and pilomatrix carcinoma[13]. Bremnes et al.[14] reported that, in literature, there have only been 55 pilomatrix carcinoma cases.

Diagnosis is generally made via incisional biopsy and surgical excision in case of need. Histopathologic examination of tissue showed the characteristic properties of PM as described by Souto MP at al.[7]. In our case we could not see the basophilic cells that are usually present in older lesions. Some authors suggest a 2 cm safety margin, but as malignant transformation is very rare, this will not be necessary. Recurrence rates are generally low (0-3%)[15]. Diameter of the lesion is not related to the prognosis. Episodes of recurrence can be related to incomplete resection[16].

This case shows us that PM should be kept in mind as a valid diagnosis for dermal or subcutaneous nodules localized outside the head and neck region. As this condition can cause diagnostic difficulty, histopatologists should be very attentive.

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