



A case of atrophic dermatofibroma

Atrofik dermatofibrom olgusu

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Abstract

Dermatofibromas are benign tumors caused by fibroblasts and histiocytes, which are quite common in middle-aged adults. It is characterized by hard, single or multiple, papule, plaque or nodul-shaped lesions that are localized to the lower extremity. Many clinical and pathological variants of dermatofibroma have been identified. One of these variants is atrophic dermatofibroma. Atrophic dermatofibroma is generally seen in middle-aged women as lesions that collapse inward, showing placement on the upper part of the body and arms. It can be confused with morphea, atrophoderma, neurofibroma, localized lipoatrophy, healing panniculitis lesions, anetoderma, steroid atrophy, and basal cell carcinoma. The definitive diagnosis is made by histopathological examination as well as clinical findings. Positive immunohistochemical staining for factor XIIIa, as well as a negative reaction for CD34, support a diagnosis of dermatofibroma. A 38-year-old woman diagnosed with atrophic dermatofibroma is presented here.

Keywords: Dermatofibroma, atrophic dermatofibroma, benign

Öz

Dermatofibromlar orta yaş erişkinlerde oldukça sık görülen, fibroblastlar ve histiyositlerden kaynaklanan benign tümörlerdir. Genellikle alt ekstremitelerde yerleşen sert, tek veya multipl, papül, plak ya da nodül şeklindeki lezyonlarla karakterizedir. Dermatofibromun birçok klinik ve patolojik varyantları tanımlanmıştır. Bu varyantlardan biride atrofik dermatofibromdur. Atrofik dermatofibrom genel olarak orta yaşlı kadınlarda, gövde ve kolların üst kısmına yerleşim gösteren, içeri çökük lezyonlar olarak görülür. Morfea, atrofoderma, nörofibrom, lokalize lipoatrofi, iyileşmekte olan pannikülit lezyonları, anetoderma, steroid atrofisi ve bazal hücreli karsinom ile karışabilmektedir. Kesin tanı klinik bulguların yanı sıra histopatolojik inceleme ile konulmaktadır. İmmünohistokimyasal incelemede faktör XIIIa ile pozitif boyanma izlemesinin yanı sıra CD34 için negatif boyanma görülmesi dermatofibrom tanısını desteklemektedir. Burada 38 yaşında atrofik dermatofibrom tanısı konulan bir kadın olgu sunulmuştur.

Anahtar Kelimeler: Dermatofibrom, atrofik dermatofibrom, benign

Introduction

Typical dermatofibromas are most commonly observed as hard, darker pink-brown papules located in the lower extremities of young and middle-aged women. Atrophic dermatofibroma is

a newly defined rare type of dermatofibroma that does not have classical clinical characteristics. Generally, it is seen as depressed or flat lesions that collapsed on palpation on the upper body or upper extremities in middle-aged women¹⁻⁴.

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Herein, we present the case of a female patient diagnosed with atrophic dermatofibroma based on clinical and histopathological examination findings.

Case Report

A 38-year-old female patient presented to our outpatient clinic with the complaint of collapsed lesions that had formed spontaneously for 3 years in the anterior aspect of the trunk. Systemic examination of the patient, whose personal and family histories were unremarkable, was normal. Dermatological examination revealed an atrophic lesion with a diameter of 2x2 cm in the right clavicular region, with soft hyperpigmented marked depressions on the skin (Figure 1). In the histopathological examination of the skin biopsy taken from the lesion with atrophic scar, atrophic dermatofibroma, and morpheic basal cell carcinoma, there was a basket-weave orthokeratosis in the epidermis, epidermal hyperplasia, dermal atrophy, a thin intact grenz zone in the dermis, leaving a tummy-type fibrous tumor covering the entire dermis (Figure 2). In the immunochemical examination, intravascular staining was observed with CD34, but no staining was observed in neoplastic cells (Figure 3). No staining with factor XIIIa was observed. Dermoscopic examination could not be performed during the examination because of technical reasons. A diagnosis of atrophic dermatofibroma was made according to the clinical and histopathological findings. The patient was consulted with the plastic surgery department for the total excision of the lesion. Histopathological examination of the excision material also revealed findings compatible with dermatofibroma, and no lesion was observed within the surgical margins.



Figure 1. Close-up view of a soft, hyperpigmented, atrophic-appearing lesion (2x2 cm in diameter) with prominent depression of the skin in the right subclavicular region

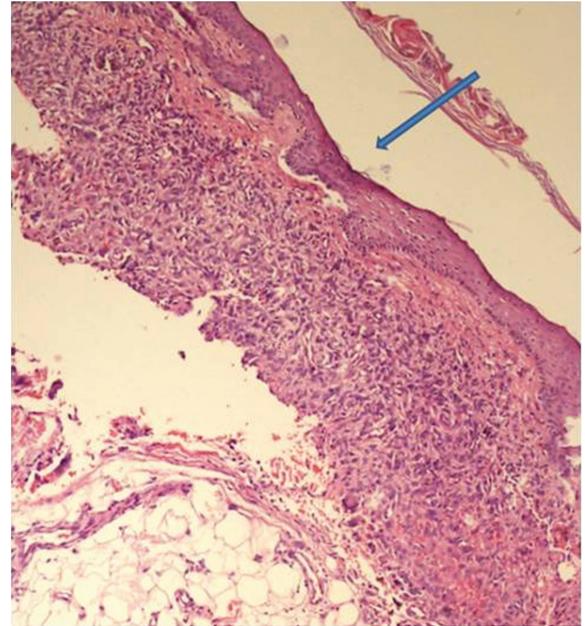


Figure 2. Basket-weave orthokeratosis of the epidermis, epidermal hyperplasia, dermal atrophy, and fibrohistiocytic neoplasia containing Touton-type giant cells that cover the entire dermis, leaving a thin intact grain zone between the epidermis and dermis and depression on the middle of the lesion indicated by the arrow (hematoxylin and eosin, x100)

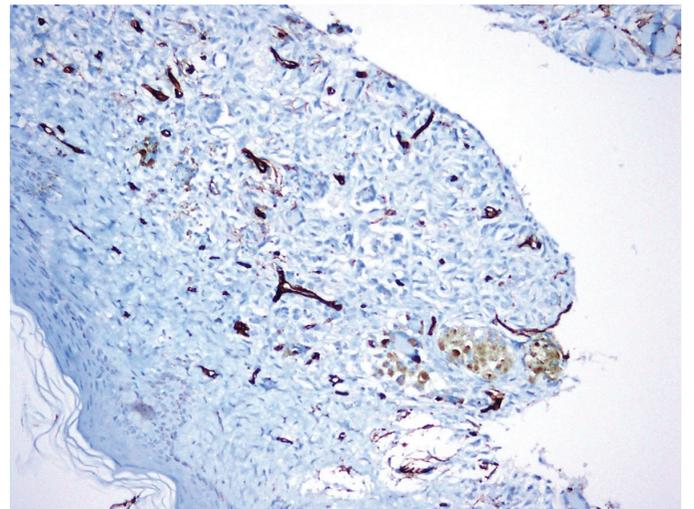


Figure 3. Intravascular positive staining with CD34 and negative staining in neoplastic cells (CD34, x10)

Verbal consent was obtained from the patient for publication of this case report and accompanying images.

Discussion

Numerous clinical subtypes of dermatofibroma such as atypical polypoid, generalized eruptive histiocytoma, grouped palmoplantar histiocytoma, multiple histiocytoma, and erosive dermatofibroma have been identified. Atrophic dermatofibroma, one of the atypical clinical forms, was first reported by Page and Assaad in 1987¹. Despite its rarity, it was reported to account for approximately 2% of

all dermatofibromas³. In a review of all reported cases with atrophic dermatofibroma, a total of 102 patients were identified, and it was seen most frequently in women aged 45-65 years and mostly located on the upper parts of the arms and back, as in the present case⁴. Although our patient is younger, the location of her lesion is compatible with the literature.

The definitive diagnosis of atrophic dermatofibroma, which can be confused with morphea, atrophoderma, neurofibroma, localized lipoatrophy, healing panniculitis lesions, anetoderma, steroid atrophy, and basal cell carcinoma, is usually made after pathological examination, and the likelihood of having a correct clinical diagnosis before a biopsy is low¹. Although histopathological findings are decisive for a definitive diagnosis, dermoscopic examination also provides an advantage in the diagnosis of dermatofibroma. Some of the atrophic dermatofibromas have a network structure around them and a white scar-like structure in the center, which can be seen in classical dermatofibromas⁴. A dermal nodule formed by collagen, fibroblasts, capillaries, and histiocytes, which are typical findings of dermatofibroma, is observed in atrophic dermatofibroma during a histopathological examination. In addition, findings such as epidermal hyperplasia, basal pigmentation, and peripheral sclerosis may accompany an atrophic dermatofibroma. Dermal atrophy, which manifests itself with a 50% reduction in the thickness of the dermis compared with the surrounding tissue, may be noted. In the immunohistochemical examination, positive staining for factor XIIIa and negative staining for CD34 were observed. This finding is particularly important in differentiating atrophic dermatofibroma from atrophic dermatofibrosarcoma. However, it can rarely be seen in positive CD34 staining^{1,4,5}. Zelger et al.³ examined 26 patients with atrophic dermatofibroma, of which two patients had positive staining for CD34, which is described as the "background-border" phenomenon at the periphery of the lesion. This study also emphasized that it should be differentiated from dermatofibrosarcoma according to other clinical and histopathological criteria if there is abnormal CD34 staining. Subcutaneous tissue, as well as the dermis, is also involved in dermatofibrosarcoma. In addition, the cell type is more uniform, but epidermal hyperplasia, giant cells, inflammatory cells, and xanthomatous structures are not observed in dermatofibrosarcoma⁵. In the histopathological examination of the presented case, epidermal hyperplasia and dermal atrophy supporting atrophic dermatofibroma

were detected in addition to the classical findings of dermatofibroma. CD34 immunostaining and subcutaneous tissue involvement observed in dermatofibrosarcomas were not detected. The diagnosis of atrophic dermatofibroma was made according to the clinical and histopathological examinations.

The presented case emphasizes that atrophic dermatofibroma, which is a rare clinical type of dermatofibroma, should also be considered in the differential diagnosis of depressed or flat lesions that felt collapsed on palpation and located mostly on the upper body parts and upper extremities in middle-aged women.

Ethics

Informed Consent: Verbal consent was obtained from the patient for publication of this case report and accompanying images.

Peer-review: Externally peer-reviewed.

Authorship Contributions

Concept: A.Ş.A., D.B., Design: A.Ş.A., D.B., Supervision - A.Ş.A., D.B., Resources - A.Ş.A., Ö.K., Materials - A.Ş.A., G.T., Data Collection or Processing: A.Ş.A., Analysis or Interpretation: A.Ş.A., D.B., Ö.K., G.T., Literature Search: A.Ş.A., D.B., Ö.K., G.T., Writing: A.Ş.A., D.B., Ö.K., G.T., Critical Review: A.Ş.A., D.B., Ö.K., G.T.

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