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# A Case Of Linear Scleroderma Following Blaschko's Lines

## Blaşko Çizgisini Takip Eden Lineer Sklerodermalı Bir Vaka

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**ABSTRACT**
Linear scleroderma is a rare variant of localized scleroderma typically seen in children and localized in the extremities. A 21-year-old female patient had a well-defined, hyper-pigmented, sclerotic plaque on right half of the frontal trunk, starting from the breast and arcing up to the right arm, then continuing linearly with anti-nuclear antibody positivity in her laboratory results. Histopathology was compatible with scleroderma. She was diagnosed linear scleroderma. There is an ongoing argument on whether localized scleroderma follows Blaschko's lines or not. By presenting this case, we aimed to contribute to literature claiming that localized scleroderma may follow Blaschko's lines.
**Key words:** Blaschko's Lines; Linear Scleroderma

**ÖZET**
Lineer skleroderma lokalize sklerodermanın nadir bir varyantıdır, tipik olarak çocuklarda görülür ve ekstremite yerleşimlidir. 21 yaşında bayan hastanın gövdesinin sağ üst yarısında meme üzerinden başlayıp ark çizerek sağ kola uzanan ve sonra lineer devam eden belirgin sınırlı, hiperpigmente, sklerotik bir plağı vardı ve antinükleer antikoru pozitifti. Histopatolojisi skleroderma ile uyumlu idi. Hastaya lineer skleroderma tanısı konuldu. Lokalize sklerodermanın Blaşko çizgisini takip edip etmediği tartışmalı bir konudur. Bu vakayı sunmakla lokalize sklerodermanın Blaşko çizgilerini takip edebileceğini düşünerek literatüre katkıda bulunmayı amaçladık.
**Anahtar kelimeler:** Blaşko çizgileri; Lineer Skleroderma

## INTRODUCTION

Localized scleroderma is a connective tissue disease confined to skin and subcutaneous tissue with unknown etiology. While it is observed most frequently in lower extremities, localized scleroderma is also found in upper extremities, frontal scalp and front wall of chest less frequently (1). Linear scleroderma, a rarely observed variant of localized scleroderma, is observed typically in children and localized in the extremities (2).

Here, we present a case of linear scleroderma following Blaschko's lines.

## CASE

A 21-year-old female patient referred to our department complaining about hardening of her skin for the last 11 years. Dermatologic examination revealed well-defined, hyper-pigmented, sclerotic plaque on right half of the frontal trunk, starting from the breast and arcing up to the right arm, then continuing linearly (Fig-1). It was learned that this plaque started in an erythematous form with edema and then hardened with time. Laboratory parameters were within normal limits, excluding

the anti-nuclear antibody positivity (1/320 E.P.). Lesional skin biopsy showed thinning of epidermis, loss of fat tissue surrounding the cutaneous appendages in the middle dermis and increase in the fibrous tissue leading down to the subcutaneous tissue (Fig-2). Diagnosis of linear scleroderma was made to the patient clinically and histo-pathologically.

Discussion

Blaschko's lines defines the distribution pattern of many congenital and acquired skin diseases like epidermal nevus, sebaceous nevus, linear lichen planus, lichen striatus, linear porokeratosis, incontinentia pigmenti (3, 4). Although yet the process of formation of these lines is not clear, it is believed that it reflects the embryonic migration pattern and is formed by genetic mosaism (5, 6). Also linear scleroderma is considered to occur after the exposure of genetically susceptible cells localized along the Blaschko's lines during the embryonic life, to certain autoimmune and environmentally triggering factors (7). Frequently Blaschko's lines are confused with dermatomes. Since these lines follow the shape of arc on the upper chest, that of an S on the abdominal area and that of a V when they are closer to the median line, it is easy to diagnose them at these areas. However, it is harder to diagnose them on the extremities, since they follow a linear pattern at those areas.

Clinically, localized scleroderma is divided into three sub-groups, well-defined plaques and bands, linear scleroderma and fronto-parietal lesions (8). There is an ongoing argument on whether localized scleroderma follows Blaschko's lines or not and there have been 6 cases reported in the literature supporting this idea (7, 9, 10). Four of these cases are localized on fronto-parietal, one on lower extremities and the other on trunk. Since linear scleroderma localized on the extremities is in the form of wide bands and doesn't show lesion continuity, it is hard to determine its pattern. However in our case, there was a narrow, well-defined plaque of which the histo-pathology was in compliance with scleroderma, starting from the body and extending out over the arm and clinically following Blaschko's lines in a very apparent form.

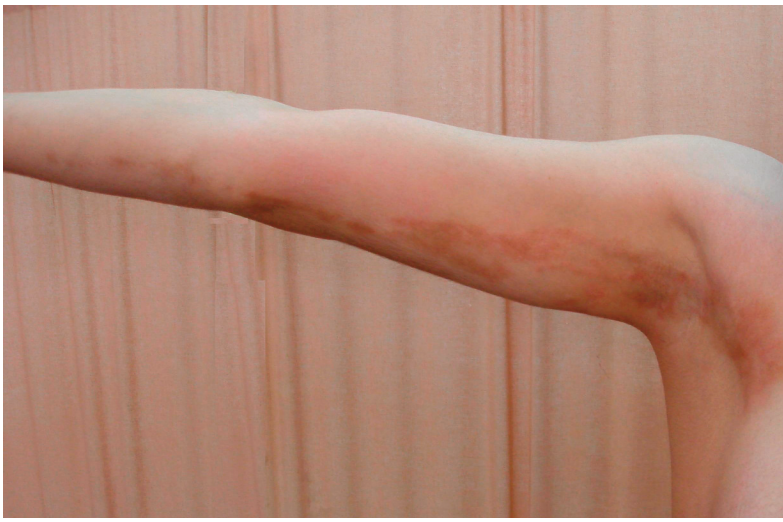

Fig. 1. Well-defined, hyper-pigmented, sclerotic plaque following Blaschko's lines

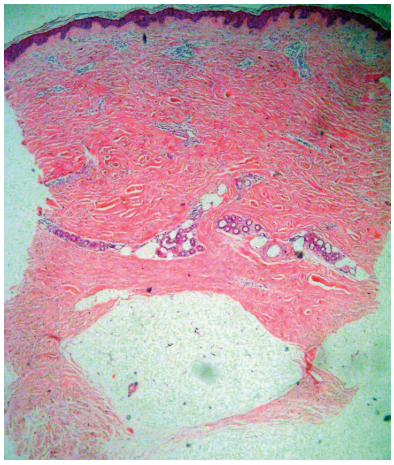

Fig. 2. Thinning of epidermis, loss of fat tissue surrounding the cutaneous appendages in the middle dermis and increase in the fibrous tissue (H&E X 100)

Linear scleroderma is observed more frequently in children, it is typically localized on the extremities, in the form of single unilateral bands and progresses with high antinuclear antibody titrations (11). Also in our case the lesions started when she was 10 years old and laboratory analysis detected antinuclear antibody positiveness at 1/320 titration.

To conclude, by presenting this case with linear scleroderma, we aimed to contribute to the literature claiming that localized scleroderma may follow Blaschko's lines.

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