

Case Report

Morphea in a Zosteriform Distribution: A Rare Clinical Entity

Berna Aksoy,^{1*} MD Aslı Hapa,² MD, Müzeyyen Astarıcı,³ MD, Mahi Balcı,⁴ MD,
Hasan Mete Aksoy,⁵ MD, Hüseyin Üstün,⁶ MD

Address: ¹TDV 29 Mayıs Private Ankara Hospital, Dermatology Clinic, Ankara and Private Konak Hospital, Dermatology Clinic, Kocaeli, Turkey; ²Bolu İzzet Baysal State Hospital, Dermatology Clinic, Bolu, Turkey; ³Ankara Research and Training Hospital, Pathology Clinic, Ankara, Turkey; ⁴Private TDV 29 Mayıs Hospital, Pathology Clinic, Ankara, Turkey; ⁵Private Konak Hospital, Plastic and Reconstructive Surgery Consultant, Kocaeli, Turkey; ⁶Ankara Research and Training Hospital, Pathology Clinic, Ankara, Turkey.

E-mail: draltaykan@yahoo.com

* *Corresponding Author:* Aslı Hapa, MD, Turan Güneş Bly, Sedir Sitesi, 9. Durak, B2/2 Oran 06450, Ankara, Turkey

Published:

J Turk Acad Dermatol 2009; **3** (4): 93401c

This article is available from: <http://www.jotad.org/2009/4/jttad93401c.pdf>

Key Words: morphea; zosteriform; Blaschko's lines

Abstract

Observations: Morphea, which is also known as localized scleroderma, comprises a group of distinct conditions that causes sclerosis of the skin and subcutaneous tissues but they do not affect internal organs with some exceptions. Here, we describe a case of morphea presenting with a zosteriform distribution that does not seem to be a linear morphea following Blaschko's lines.

Introduction

Morphea, which is also known as localized scleroderma, comprises a group of distinct conditions that causes sclerosis of the skin and subcutaneous tissues but they do not affect internal organs with some exceptions. A recent classification has divided morphea clinically into five subtypes: circumscribed morphea, linear morphea, generalized morphea, pansclerotic morphea, and mixed subtype (combination of one or two of the types above) [1]. Here, we describe a case of morphea presenting with a zosteriform distribution that does not seem to be a linear morphea following Blaschko's lines.

Case Report

A 58-year-old female applied to dermatology outpatient clinic with asymptomatic, rough and brown colored plaques of 3 years' duration on the left side of her trunk with a zosteriform distribu-

tion of T12-L1 dermatomes. There was no history of herpes zoster and any other precipitating factor affecting the same location. Her past medical history was unremarkable except a diagnosis of breast cancer 3 and half years ago. She was treated with radical mastectomy followed by chemotherapy. Since then she has been receiving tamoxifen therapy for breast cancer. Dermatological examination revealed light- brown to slate- gray colored sclerotic patches over the left side of her abdomen extending from the umbilicus anteriorly to the left lumbovertebral region conforming with a zosteriform pattern (Figure 1A, B).

Histopathologic examination of the skin biopsy specimen revealed thickened collagen bundles affecting mid and deep reticular dermis along with collagen replacing the fat around the sweat glands and extending to subcutaneous fat tissue. Under a thinned epidermis, perivascular mononuclear cell infiltration and atrophy of epidermal appendages were present (Figure 2A, B). With these histological and clinical findings, the diagnosis of zosteriform morphea was concluded. Routine blood tests, urine analysis and autoantibody in-



Figure 1A, B. Light- brown to slate- gray colored sclerotic patches over the left side of her abdomen extending from the umbilicus anteriorly to the left lumbovertebral region

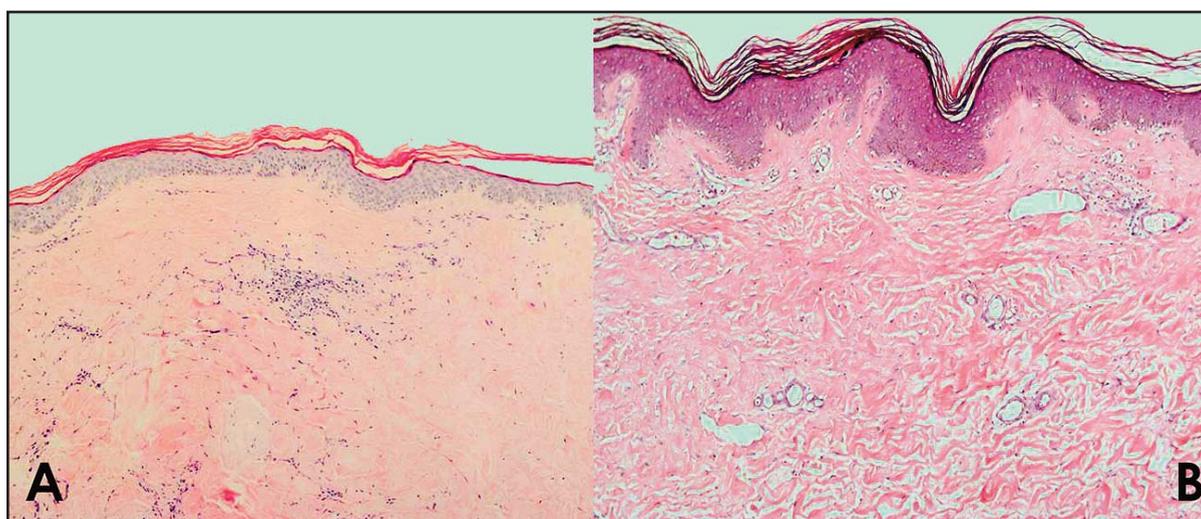


Figure 2A. Under a thinned epidermis, thickened collagen bundles affecting whole dermis (HE x 40); **2B.** Perivascular mononuclear cell infiltration and atrophy of epidermal appendages within the thickened collagen bundles (HE x 100).

vestigations revealed no abnormalities. Topical calcipotriene 0.005% ointment twice daily was prescribed for the treatment of the lesions with a partial response after 6 months.

Discussion

Linear type of morphea is a rare form which is typically observed in pediatric group of patients and especially involves the extremities. There are reports indicating that the linear morphea follows the *Blaschko's* lines but not dermatomes. However, distribution of linear morphea is still a controversial subject [2]. To the best of our knowledge only three cases of morphea presenting with a zosteriform distribution have been previously reported.

Lopez et al [3]. described a 45-year-old man presenting with a zosteriform morphea follo-

wing a herpes zoster infection at the same localization. Moreover, morphea with features of lichen sclerosus et atrophicus at the site of C2-C4 dermatomes of herpes zoster scar has been described [4]. These two cases were regarded as examples of the isotopic response which has been described by *Wolf et al* [5] as the occurrence of a different type of skin disorder at a previously involved site after resolution of the prior condition. Another case of a 21-year-old male with zosteriform morphea on the left flank region of his abdomen has been reported [6]. However; this patient had no history of herpes zoster affecting the same location like the case presented here.

Additionally, *Wakelin et al* [7] also described a 53-year-old man with a history of unilateral atrophoderma of *Pasini* and *Pierini* with loca-

lized areas of morphea affecting the left side of his trunk in a zosteriform distribution.

Dermatomes are the segments of the skin that are defined by sensory innervations while *Blaschko's* lines reflect the lines of embryonal development of the epidermis and dermis. It sometimes seems difficult to determine whether linear morphea follows *Blaschko's* lines or dermatomes especially in the extremities [8]. When discontinuous, unilateral, and wide pattern skin lesions are taken into consideration, our case seems to be one of the rare and new clinical form of morphea in a zosteriform distribution.

References

1. Laxer RM, Zulian F. Localized scleroderma. Curr Opin Rheumatol 2006; 18: 606-613. PMID: 17053506
2. Weibel L, Harper JI. Linear morphea follows Blaschko's lines. Br J Dermatol 2008; 159: 175-181. PMID: 18503590
3. López N, Alcaraz I, Cid-Manas J, Camacho E, Herrera-Acasto E, Matilla A, et al. Wolf's isotopic response: zosteriform morphea appearing at the site of healed herpes zoster in a HIV patient. JEADV 2009; 23: 90-92. PMID: 18355190
4. Forschner A, Metzler G, Rassner G, Fierlbeck G. Morphea with features of lichen sclerosis et atrophicus at the site of a herpes zoster scar: another case of an isotopic response. Int J Dermatol 2005; 44: 524-525. PMID: 15941448
5. Wolf R, Brenner S, Ruocco V, Filioli FG. Isotopic response. Int J Dermatol 1995; 34: 341-348. PMID: 7607796
6. Joshi A, Al-Mutairi N. Zosteriform morphea. Acta Derm Venereol 2005; 85: 279-280. PMID: 16040427
7. Wakelin SH, James MP. Zosteriform atrophoderma of Pasini and Pierini. Clin Exp Dermatol 1995; 20: 244-246. PMID: 7671424
8. Soma Y, Kawakami D, Yamasaki E, Sasaki R, Mizoguchi M. Linear scleroderma along Blaschko's lines in a patient with systematized morphea. Acta Derm Venereol 2003; 83: 362-364. PMID: 14609105