Morphea in a Zosteriform Distribution: A Rare Clinical Entity

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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 distribution 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-
vestigations revealed no abnormalities. Topical calcipotrione 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 following a herpes zoster infection at the same localization. Moreover, morphea with features of lichen sclerosis 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