A Child With Morphea in a Zosteriform Distribution

Perihan Öztürk¹, Harun Çıralık²

ABSTRACT

Morphea is a localized form of scleroderma which is characterized by sclerotic plaques, limited to the skin. Although its cause is unknown, various (genetic, infectious and autoimmune) mechanisms have been suggested. It is more common among children and young women. Although clinical outcome is good, sometimes it can be prominent. Morphea have five subtypes, which are known as plaque, generalized, bullous, deep and linear forms. Zosteriform morphea is a recently described pattern and it is rarely observed. A six years old female, was admitted with multiple brown and white asymptomatic plaques arranged in a zosteriform pattern, confined to the right back since two months, without history of herpes zoster at the same location. We aimed to report the first pediatric case of morphea with zosteriform pattern in the absence of herpes zoster history.

Key words: Zosteriform, Morphea, child

INTRODUCTION

Morphea involves a group of diseases that progress with sclerosis in skin and subcutaneous tissue. Morphea was first described in 1854 by Thomas Addison and named as “keloid syndrome of Alibert”. The idiopathic disease manifests as a sclerotic dermal plaque with violaceous borders and central hypopigmentation. Although it generally cause small, limited lesions; It may cause wide and deep lesions lead to functional or cosmetic deformities. Approximately, 75% of the patients with morphea is between the ages of 20-50. It is 2.6 times higher in women than men. Incidence is 27/1,000,000 per year. In the beginning of the disease, environmental factors can be effective. Onset may be “slow and insidious” or “rapid and progressive”. Although morphea is classified in 5 groups classically as plaque, generalized, bullous, linear and deep, zosteriform morphea is a newly described pattern (1-3). The term zosteriformis is commonly used to describe the morphological pattern of a skin dermatosis resembling the distribution of herpes zoster (4). We hereby report the first pediatric case of morphea, with zosteriform pattern, in the absence of prior history of skin herpes zoster infection.

Zosteroform Şeklinde Morfealı Bir Çocuk

ÖZET


Anahtar kelimeler: Zosteriform, Morphea, çocuk

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CASE

A 6-year-old female was admitted with multiple brown and white asymptomatic plaques arranged in a zosteriform pattern, confined to the right side of the back since two months. There was no history of the herpes zoster affecting the same location. General physical and systemic examinations were normal. Cutaneous examination was revealed a zosteriform band composed of brown-white indurated-depressed plaque confined to the right back with a strict midline demarcation and 6 cm in width (Figure 1). There was not erythema in and around the lesion. Routine baseline investigations including complete blood cell count, ESR, clinical chemistry and urinalysis were normal. Serology for antinuclear antibodies and Borrelia burgdorferi was negative. Biopsies were taken from the active border and the deep region, with a prediagnosis of lichen sclerosis atrophicus and morphea. Histopathological examination of the biopsy specimens was revealed orthokeratosis on the tissue that covered by squamous epithelium and homogenized in the connective tissue with mononuclear inflammatory cell infiltration around the blood vessels in lower and upper dermis. Adnexial structures were normal (Figure 2). The patient was diagnosed with concordance of clinical and histopathologic findings.

DISCUSSION

Morphea is a localized form of scleroderma characterized by sclerotic plaques. Fibrotic reaction is limited to the skin and visceral involvement is uncommon (2). The 75% of morphea patients are between the ages of 20-50 and 2.6 times more common in women. Our patient was a 6-year-old girl. Morphea is an idiopathic disorder (4). Usually, lesions appear without any specific precipitating factors, but also may follow chickenpox, BCG vaccination, radiotherapy, injury and as a consequence of hypoplastic breast (5). There was not any triggering factor in our patient. Eosinophilia and antinuclear antibody positivity are common in patients with generalized morphea. Laboratory findings were normal in our patient. Histopathological findings of morphea varies according to the stage of the disease and where the biopsies are taken. Biopsies must be taken from the subcutaneous tissue and peripheral edge of an active lesion which contains lymphocytes, macrophages, plasma cells, mast cells, and sometimes intense inflammatory infiltrate with eosinophilia (1). In our patient, biopsies were taken from the active margin of the lesion. Histopathologic examination was consistent with morphea. There is not effective treatment for morphea yet (6). Topically steroid cream may be helpful in some cases. Inflammatory lesions can be treated with intralesional triamcinolone injection. For rapidly progressing symptomatic disease, prednisone 20-40 mg once daily for 6-8 weeks can be considered (1). Highly-potent topically steroid was given to our patient and control was recommended.

In the pediatric age group the linear lesions are predominant, especially on the limbs; however, the zosteriform pattern of morphea has not been reported yet. Joshi et al. have been reported a 21-year old male with zosteriform morphea (4). Wakelin et al. have been described a 53-year-old man who had a 12-year history of unilateral idiopathic atrophoderma of Pasini and Pierini affecting
the right side of his trunk in a zosteriform distribution (7). According to our knowledge only two cases of morphea as a post herpetic reaction have been reported by Lopez et al (8) and Forschner A et al (9). But there was not any clinical evidence of herpes zoster in this patient. The present case illustrates that morphea could be seen in zosteriform pattern without herpes zoster history.

Zosteriform morphea is a newly pattern of morphea. Although other cases were presented adult age group, our zosteriform pattern of morphea patient was the first pediatric case without prior history of herpes zoster.

REFERENCES


