Unilateral generalized morphea: A case report

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Morphea is a localized form of scleroderma characterized by sclerotic plaques limited to the skin. Although its cause is unknown, genetic, infectious and autoimmune mechanisms have been suggested in the pathogenesis of the morphea. It is more common among childen and young women. Although the prognosis is generally good, it sometimes causes significant morbidity. Morphea has five subtypes as plaque, generalized, bullosa, deep and linear. Unilateral forms of generalized morphea have rarely been reported in the literature. Our case has been presented because it is very rarely seen.

Keywords: generalized, morphea, unilateral

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INTRODUCTION

Morphea includes a group of diseases that show sclerosis of skin and subcutaneous tissue. Although it generally shows small limited lesions, it may also shows deep lesions causing functional or cosmetical deformites [1]. About %75 of morphea patients are 20-50 years of age and it is 2.6 times more common in women than men. Environmental factors may play a triggering role in the onset of the illness. Thirteen percent of the patients with morphea have generalized morphea. If morphea plaques are seen in at least two of seven anatomical sites (head-neck, right upper extremity, left upper extremity, right lower extremity, left lower extremity, body, face, trunk), the diagnosis is generalized morphea [1]. "Unilateral generalized morphea" form is seen quite rarely.

CASE REPORT

A 14 year-old male patient came to our clinic with spottling lesions starting on the left side of the body. He only complained of the stiffness of the lesions and pruritus, and had no prior history of trauma. On dermatologic examination, hard hyperpigmented indurated lesions with diameters ranging from 0.5 to 10 cm were present on the

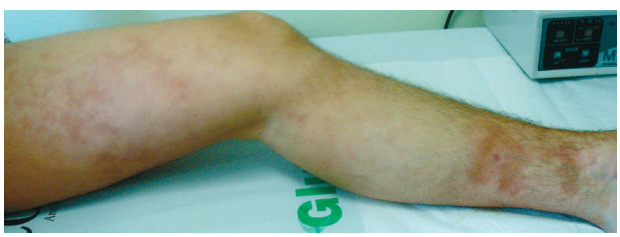

Figure 1. Hyperpigmented indurated plaques with diameters ranging from 0.5 to 10 cm on the left thigh and left leg.

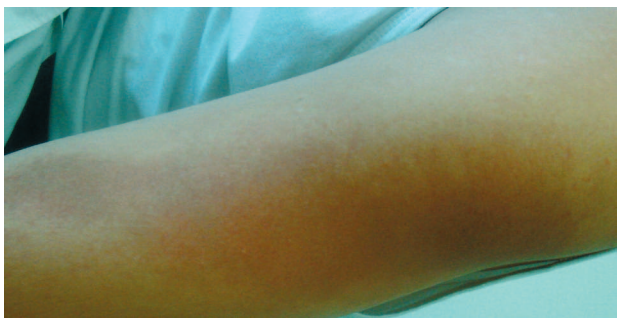

Figure 2. Hyperpigmented indurated on the extensor surface of the left arm.

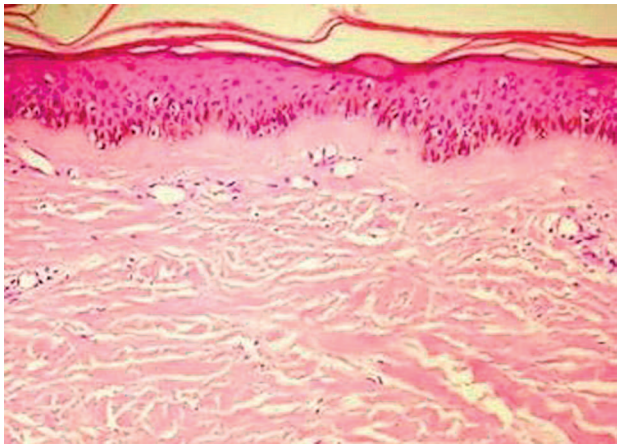

Figure 3. The histopathological view: atrophic epidermis with diffuse dermal fibrosis (H&E*10).

front of the left shoulder, left arm extensor surface, the left surface of the back, left thigh, left leg and left foot (Figure 1,2). Laboratory tests including antinuclear antibody, complete blood count, sedimentation analysis, rheumatoid factor levels were normal. Initial diagnoses were lichen sclerosus et atrophicus, lupus panniculitis and morphea. Deep punch biopsy specimens were taken from one of the lesions. The histopathological examination of the samples revealed orthokeratosis with an atrophic squamous epithelium. In the dermis, diffuse fibrotic tissue was seen with mononuclear inflammatory infiltrative cells around the blood vessels. Adnexal structures were normal (Figure 3). Considering clinical and histopathological findings, the patient was diagnosed with morphea and received potent topical steroids twice daily and intra-lesional triamcinolone acetonide injection three times per month. At the end of the third month, the lesions improved significantly.

DISCUSSION

Morphea is a localized form of scleroderma and is characterized by sclerotic plaques. The fibrotic reaction is limited to the skin and visceral involvement is uncommon [2]. Morphea has five subtypes as plaque, generalized, bullous, linear and deep [3]. About 75% of morphea patients are 20 to 50 year-old women and it is 2.6 times more common in women. Thirteen percent of morphea patients have generalized morphea [1]. Morphea may be triggered by environmental factors including trauma, infections (measles, varicella, Epstein-Barr virus, Borellia Burdorgferi), malignancies and radiation therapy [4]. Our patient had no triggering factors. Morphea pathogenesis is similar to systemic scleroderma as endothelial cells, inflammatory cells and fibroblasts are involved [1]. If morphea plaques are seen in at least two of seven anatomical sites, then the diagnosis of generalized morphea was made [1]. In our patient, morphea plaques were seen on the left arm, left leg, left side of the body. 'Diffuse morphea' covers large areas of the body and has an insidious onset.

The chest wall involvement in patients with severe thoracic deformity may cause difficulty in breathing. Despite widespread cutaneous involvement in generalized morphea, internal organs involvement is rare. Arthralgia is seen by 9% of the patients [1]. There was no systemic signs or symptoms in our patient. Circulating auto-antibodies can be detected in morphea like other autoimmune connective tissue diseases [1]. Laboratory findings such as serum ANA, ssDNA antibodies, eosinophilia, antihistone antibodies and hypergammaglobulinemia are more common in linear and generalized morphea [1]. Laboratory tests were normal in our patient. Histopathologic findings of morphea vary according to the stage of the disease and the biopsies taken. Biopsy should be taken to include the subcutaneous tissue. In the biopsies from the peripheral edge of an active lesion, lymphocytes, macrophages, plasma cells and mast cells can be seen; sometimes, intense inflammatory infiltrates with eosinophilia may also be seen [1]. In our patients, biopsies were taken from the active margin of the lesion. Histopathologic examination was consistent with morphea. In contrast to localized morphea, generalized morphea lesions usually do not tend to regress spontaneously. The effect of strong topical corticosteroids can be increased with intralesional triamcinolone injection. Systemic glucocorticoids, antimalarials, azathioprine and phototherapy are not usually very effective [5]. Although controversial results exist about inhibitory influence of salazopyrin (sulfasalazine) on the fibroblast proliferation, some authors have reported good results with salazopyrin (Sulfasalazine) in the treatment of generalized morphea [6]. Our patient received strong effective topical steroids twice daily and intra-lesional triamcinolone acetonide injection three times per month. At the end of

the third month, a significant improvement was seen in the lesions.

Unilateral generalized morphea is a newly described type of generalized morphea. It has been very rarely reported in the literature. Nagayi et al, reported a 6 year-old boy with unilateral generalized morphea on the right side of his lower leg, trunk, and upper arm. The levels of antinuclear antibodies, rheumatoid factor, and anti single-stranded DNA antibody were elevated. No severe deformities or functional disabilities were noted. With topical corticosteroid therapy, the sclerotic skin became gradually softer, and no progression of sclerosis was noted for one year [7]. Kraigher et al, reported a 20-year-old healthy Jewish woman of Yemenite origin who presented with a 2-year history of a linear eruption on the right shoulder and thorax, upper and lower arm, dorsal surface of the hand, and lower leg [8]. Gerçeker Turk et al, presented a case of unilateral generalized morphea that was triggered by vibration. They proposed that development of ipsilateral generalized morphoea without pulmonary involvement in a left handed marble worker indicated exposure to hand–arm vibration rather than to silica as an aetiological factor in this condition [9]. Appelhans et al, reported four cases of unilateral generalized morphea [10]. We decided to present this case because of the rarity of the disease and for the purpose of literature review.

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