

Generalized morphea - a case report

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Abstract Generalized morphea is a subtype of morphea in which widespread sclerotic plaques are seen without systemic involvement. We are reporting a case of 10-year-old boy with generalized morphea for 6 years.

Key words

Morphea, localized scleroderma, generalized morphea.

Introduction

Morphea, sometimes referred to as localized scleroderma is a group of related disorders characterized by varying degrees of sclerosis, fibrosis and atrophy in skin and subcutaneous tissue. It can be classified into various groups. Generalized morphea presents as plaques four or more in number larger than 3 cm that become confluent and involve at least two out of seven anatomical sites (head, neck, right upper extremity, left upper extremity, right lower extremity, left lower extremity, anterior trunk and posterior trunk).

Case Report

A 10-years-old boy was brought to Dermatology OPD, Mayo Hospital, Lahore by his parents who were concerned about multiple hyperpigmented plaques on right foot developed initially then involving extensor surface of right leg, medial side of left leg, lower abdomen and extensor surface of right arm for six years. There was difficulty in flexion of right knee which was gradual over two years. He was born full term with unremarkable antenatal history. His parents are not related and there is no family history of such disease.

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On examination, anthropometric measurements of weight and length were within normal limits. There were multiple hyperpigmented atrophic linear plaques with well-defined margins present on dorsal and medial side of right foot extending upto right iliac crest. Similar plaques starting from dorsal surface of right hand and extending upto shoulder were also present. More plaques were also seen on medial side of left thigh, front of chest and left side of lower abdomen. There was limitation in the flexion at right knee joint.

Scalp, mucous membranes, and nails were normal. Rest of the systemic examination was also normal. Laboratory examination including complete blood count, serum electrolytes, liver function tests, renal functions test were normal in range. Radiographic examination of chest, right knee joint and right leg (A.P and lateral view) showed no abnormality. The histopathological examination showed normal stratum corneum, focal areas of atrophy in stratum spinosum, effaced rete ridges, hyperpigmented basal layer, eosinophilic amorphous collagen fibres in dermis, perivascular and periappendageal lymphohistiocytic infiltrate.

On the basis of widespread disease, absence of systemic involvement and suggestive histopathology, he was labelled as generalized morphea. After test dose, he was started on 7.5mg methotrexate weekly along with topical betamethasone valerate twice daily. After two



Figure 1 Atrophic, sclerotic plaque on medial aspect of right leg and right foot.



Figure 2 Ivory-coloured atrophic plaque on right deltoid region.



Figure 3 Ivory-coloured atrophic plaque affecting medial aspect of left thigh.



Figure 5 Ivory-coloured atrophic linear plaque on right hand.

Months the plaques started becoming softer. Patient is currently under follow-up.

Discussion

Morphea represents a wide variety of clinical entities that share a common underlying pathophysiology of increased collagen deposition in an autoimmune setting. They are largely confined to skin, subcutaneous tissue, underlying fat, fascia, muscle, bone and joints and occasionally with involvement of eye and brain. Paterson *et al.*² divided morphea into five classes, plaque, generalized, linear, bullous and deep.

Generalized morphea is induration of skin starting as four or more individual plaques larger than 3 cm, that become confluent and involve at least two of the seven anatomical sites (head, neck, right upper extremity, left upper extremity, right lower extremity, left lower extremity, anterior trunk and posterior trunk).^{3,4} Within this group there are distinct clinical presentations which are: disseminated plaques morphea, pansclerotic morphea and eosinophilic fasciitis.

Overall incidence of morphea is 4-27 per million per year occurring at any age and female to male ration between 7:1 and 2.6:1.⁵ Aetiology of morphea is poorly understood. Trauma, radiation, medication and injections all have been proposed as triggering events in its development in susceptible individuals. Autoimmune mechanisms are thought to play an important role in induction of morphea.³ Arif and Hassan reported a case of generalized morphea associated with hypothyroidism.⁶

All subtype of morphea share similar findings of an early active inflammatory phase in which newer lesions demonstrate a lymphocytic infiltrate with a variable number of plasma cells and eosinophils. As lesions evolve the intensity of inflammatory cells is reduced as collagen bundles thicken and skin sclerosis increases in later fibrotic phase.^{3,5,6}

ANA antibodies are present in approximately 50% of morphea patients. Antihistone antibodies are detected in 42% of localized scleroderma patients and 87% of patients with generalized morphea.⁷ Anti-single stranded DNA antibodies are present in 25% of plaque type morphea and 75% of those with generalized morphea. There has been a debate about a possible association between *Borrelia burgdorferi* and morphea.⁸

Treatment of generalized morphea is challenging. Topical glucocorticoids, tacrolimus and vitamin D analogues can be used. Systemic medication which can be used include corticosteroids, methotrexate, ciclosporin, infliximab, UVA phototherapy may also be helpful.^{4,9,10}

Our patient was responded to oral methotrexate and topical steroids.

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