ORIGINAL RESEARCH PAPER

Dermatology

MORPHEA: LOCALIZED SCLERODERMA PUZZLE

KEY WORDS:

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INTRODUCTION:

Morphea, also known as localized scleroderma, is an uncommon chronic inflammatory connective tissue disorder characterized by excessive collagen deposition in the skin and underlying tissues.[1] Unlike systemic sclerosis, morphea does not usually involve internal organs.[2] The disease presents in various clinical forms, including plaque, linear, generalized, and deep morphea.[1,2] Although its exact etiology remains unclear, autoimmune mechanisms, genetic predisposition, trauma, and infections have been implicated.[2,3] Morphea may result in cosmetic disfigurement, functional impairment, and reduced quality of life.[4] Early diagnosis and appropriate management are essential to prevent disease progression and long-term complications.[1]

CASE REPORT:

A 35-year-old female presented with a six-month history of asymptomatic, gradually enlarging hyperpigmented plaques over the right lower leg. Physical examination revealed well-demarcated plaque with a violaceous border and central sclerosis (Figure 1). There was no history of Raynaud phenomenon, dysphagia, or systemic complaints. Routine laboratory investigations were within normal limits, and antinuclear antibody testing was negative. Skin biopsy demonstrated thickened collagen bundles in the dermis with perivascular lymphocytic infiltration, consistent with morphea. The patient was treated with topical corticosteroids and phototherapy, resulting in significant softening of lesions and clinical improvement.

DISCUSSION:

Morphea is a rare fibrosing disorder characterized by localized sclerosis of the skin and subcutaneous tissues.[1] The disease predominantly affects females and can occur at any age, with peaks in childhood and middle adulthood.[1,2] The pathogenesis involves a complex interplay of immune dysregulation, vascular abnormalities, and fibroblast activation, leading to excessive collagen synthesis and tissue fibrosis.[2] Several triggering factors have been proposed, including trauma, radiation exposure, infections, and autoimmune conditions.[2,3]

Clinically, morphea manifests in different subtypes. Plaque morphea is the most common form and typically presents as circumscribed indurated plaques with a characteristic lilac ring during the active inflammatory phase.[1] Linear morphea commonly affects children and may involve deeper tissues, resulting in limb length discrepancies, joint contractures, or facial deformities.[2] Generalized morphea is characterized by multiple widespread plaques affecting larger body surface areas, while deep morphea extends into fascia, muscle, or bone.[1,2]

Diagnosis is primarily clinical but can be supported by histopathological examination. Skin biopsy usually demonstrates thickened and densely packed collagen bundles, atrophy of skin appendages, and inflammatory infiltrates in early lesions.[2] Laboratory investigations are generally nonspecific, although positive autoantibodies may be detected in some patients.[3] It is important to distinguish morphea from systemic sclerosis, which is associated with visceral organ involvement and a different prognosis.[1]

Treatment depends on disease severity, subtype, and extent of involvement.[3] Limited superficial lesions may respond well to topical corticosteroids, calcineurin inhibitors, or vitamin D analogues.[3] Phototherapy, particularly ultraviolet A1 and narrow-band ultraviolet B, has demonstrated efficacy in reducing inflammation and fibrosis.[1,3] Patients with extensive, rapidly progressive, or deep disease often require systemic therapy, including methotrexate, corticosteroids, mycophenolate mofetil, or other immunomodulatory agents.[2,3] Early intervention is crucial in preventing irreversible fibrosis and functional disability.[3]

The prognosis of morphea is generally favorable compared with systemic sclerosis. However, residual pigmentation changes, skin atrophy, and cosmetic concerns may persist even after disease activity subsides.[1] Relapses can occur, particularly in generalized and linear variants.[2] Long-term follow-up is therefore recommended to monitor disease activity, treatment response, and complications.[3] A multidisciplinary approach involving dermatologists, rheumatologists, physiotherapists, and other specialists may be beneficial in complex cases.[1] Continued research into the molecular mechanisms of fibrosis may provide opportunities for the development of targeted therapeutic interventions and improved patient outcomes in the future.[2]

CONCLUSION:

Morphea is a chronic localized fibrosing skin disorder with variable clinical presentations and outcomes.[1] Although generally limited to the skin and underlying tissues, the disease can cause significant cosmetic and functional impairment, especially in deep and linear forms.[2] Early recognition, accurate diagnosis, and timely initiation of therapy are essential for preventing complications and improving quality of life.[3] Histopathological confirmation may aid diagnosis in uncertain cases.[1] Current treatment strategies include topical agents, phototherapy, and systemic immunosuppressive medications for severe disease.[3] Regular follow-up remains important for monitoring disease progression, recurrence, and treatment effectiveness.[4,5]

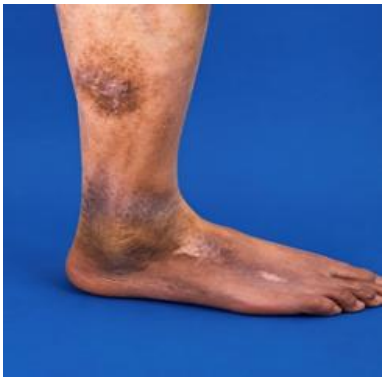

Figure 1 : well-demarcated plaque with a violaceous border and central sclerosis over right lower leg

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