Morphea: A Case Of Ignorance

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Abstract : Morphea is a chronic autoimmune disease characterized by hardening (skleros) of the skin (derma) also referred as localized scleroderma. It is great cause of morbidity, aesthetic disfigurement & psychological stress. We report a case of 11 year old male child who presented to outpatient department of dermatology with dark colored, shrunken skin lesion on right thigh since 6 to 8 months, initially ignored by parents and inspite of empirical treatment from general practitioner, lesion keeps on progressing so decided to seek specialist advice. The boy was diagnosed as a case of linear morphea clinically and was confirmed by skin biopsy. Medical line of treatment was started and regular follow up was advised to evaluate the outcome. The purpose of this case report is to raise awareness about this disease among clinicians and also parents for early diagnosis and treatment.

Key words : Morphea, Scleroderma, Localised scleroderma

Introduction : Morphea is a clinically distinct chronic inflammatory, autoimmune disease primarily of the dermis and subcutaneous fat that ultimately leads to a scar like sclerosis.[1] Morphea itself has a spectrum of manifestations ranging from skin only to multiple organ involvement.[2] The estimated incidence of morphea is 2.7 per million with a female to male ratio of 2-3:1, and it is more common in caucasians. [3] The morphea is very rarely life threatening disease however in about 10% of the patient scar formation may lead not only to usually disfigurement but also to significant contractures or growth retardation and handicapped the affected individual for their entire life.[1] The etiology and pathogenesis of morphea is poorly understood but is thought to involved three major closely connected components i.e. vasuclar damage, activated T cell and altered connective tissue production by fibroblast.1 The onset of morphea is generally insidious and manifestation varies according to disease subtype such as Plaque, Linear, Generalised, Bullous and Deep type.[4] The initial skin manifestation of morphea includes slightly elevated erythematous or violaceous some what oedematous plaque that undergoes centrifugal expansion followed by indurated, hyperpigmented or hypopigmented, atrophic lesion.[3,5,6] Here we report a case of child affected by morphea, ignored by parents.

Case report : An 11 year old male child came with dark colored, shrunken skin lesion of right thigh since 6-8 months with mild discomfort and aesthetic problem. The patient was on empirical treatment advised by general practitioner with no improvement and lesion keeps on progressing. There was no history suggestive of trauma, infection, systemic involvement and associated autoimmune disease. There was no history of similar presentation in family. On general examination child was healthy, Systemic examination were normal and Cutaneous examination revealed linear, hyperpigmented, indurated atrophic plaque. Routine blood investigations were within normal limits. Skin biopsy showing atrophic epidermis and dermis shows abundant collagen bundles with nonspecific inflammatory infiltrate. The child was diagnosed as a case of morphea or localized scleroderma of linear type.

The parents were properly counseled and medical line of treatment was started and regular follow up was advised to evaluate the outcome.

Discussion : Linear morphea is a most common type of localized scleroderma accounting for twenty-five to eighty-seven percent of pediatric cases, with limb or trunk involvement in 70%-80%, the lesion may follow Blaschko's line.[7] It can also affect the face fronto-parietal region (en coup de sabre), orbital area and scalp.8 Twenty to thirty percent of morphea begins in childhood but it can occur at any age. Extracutaneous manifestations develop in 22%-56% of morphea patients and some have concomitant autoimmune disease.[2,3,5,6] The period of disease activity vary from 3-6 years but reactivation after periods of remission occurs in 20%.[3,5,6,9,-12] Morphea does not usually progress to systemic sclerosis and the survival rate is not significantly different from the general population.

Morphea probably arises from a genetic background that increases disease susceptibility; combine with other causative factors (infections, enviornmental exposures) that modulate disease expression. Like many other autoimmune connective tissue disease morphea is likely a complex genetic disease. Familial clustering is rarely seen.6,13,14 The triggers of

morphea include trauma, viral exanthems, vitamine K1, radiation, valproic acid, pentazocine, paclitaxel, borrelia burgdorferi and silicon implants.[15]

Routine blood investigations may show peripheral eosinophilia, hyper gamma globulinemia and increase ESR/ CRP with active disease of any type but particularly deep morphea. Auto antibodies reported in a patient with morphea include ANA, Anti ss DNA, anti ds-DNA, Anti histone, anti-phospholipid, anti-centromere, anti-Scl70 and RF (MMP-1). (5,16, 26, 28-34 Fitz) In children, the prevalence of ANA is higher than in adults and is more frequently in linear and deep types of morphea.[5]

Assessment of disease course may be done with 20 MHz ultrasound, which can visualize skin to a depth of 1 cm. MRI provides a complete assessment of the extent of disease, including depth of involvement and disease activity.[15]

The treatment depends on the type of morphea, extent of disease, severity and evidence of activity. Management of morphea includes topical (corticosteroid, calcineurin inhibitor, calcipotriol, and PUVA), systemic therapy (corticosteroids, methotrexate, mycophenolate mofetil and cyclosporin etc.) and physiotherapy, surgical release of contractures and surgical reconstruction where indicated.[15]

Conclusion : Early diagnosis & effective management will prevent the disabilities and disfigurement associated with morphea.

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Fig 1 : Sclerodermatous lesion on right thigh

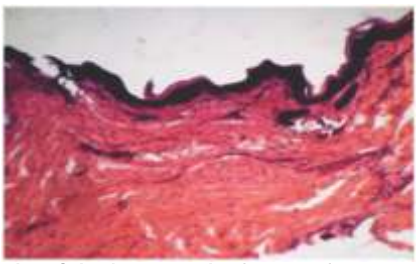

Fig 2: Skin biopsy showing features of morphea