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Linear Morphea in a Child: A Case Report

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ABSTRACT

Linear morphea is a morphological variant of morphea. It typically occurs in children . Linear morphea most often affects the lower extremities, upper extremities, frontal portion of the head, and anterior trunk. Linear morphea, includes the trunk/limb, en coup de sabre(ECDS), and Parry-Romberg variants The etiology of linear morphea is unclear but may be autoimmune in origin. Current evidence supports first-line treatment with methotrexate and oral glucocorticoids followed by mycophenolate mofetil and phototherapy.

Key words: Linear Morphea, glucocorticoids.

INTRODUCTION

Linear morphea, a variant of morphea, is characterized by induration of the skin, often with pigmentary changes, and it frequently occurs in children in the first or second decade[1].The extremities are most commonly affected followed by the face.

Case report

12 year old female presented to skin opd with hyperpigmented ,linear ,macular and thickened lesions extending from the lateral aspect of left knee joint to lateral malleolus for the past 5 years. The lesion first appeared on the lateral aspect of left knee joint and gradually progressed along the lateral border of the leg upto the lateral malleolus within last 5 years. There was no history suggestive of Raynauds phenomenon and there were no systemic complaints. Physical examination revealed multiple, hyper-, hypopigmented , indurated plaques with atrophic, shiny surface along lateral border of left leg. The lesions ranged from 1cm to 30cms in diameter. The lesion was atrophic in the proximal part and demonstrated induration in the distal part [Figure 1]. Hair was sparse on the affected parts.There was no notable joint contractures or limb shortening. Systemic examination was normal.Punch biopsy was taken from the proximal part of leg and sent for histopathological examination . Histopathology showed epidermal atrophy, a sparse superficial and predominantly deep, dermal and subcutaneous perivascular lymphocytic infiltrate and plasma cells, and the collagen bundles appeared thickened and closely packed with sparse adnexal structures[Figure 2]. The complete blood count with differential analysis, liver function tests, chemistry panel, and urinalysis were normal. Antinuclear antibodies and anti-Scl-70 antibodies were negative.

DISCUSSION

Peterson et al. divide morphea into five categories based on the morphologic findings:

plaque, generalized, bullous, linear, and deep. Plaque morphea constitutes 56%,linear morphea 20%,genearalised morphea 13% and deep morphea 11% of total cases.[2]Linear morphea may account to considerable morbidity, especially in children, when they interfere with growth. Substantial disability and deformity may result from joint contractures, limb-length discrepancy, and prominent facial atrophy. Extracutaneous involvement is frequent, with ophthalmologic and neurologic findings[3].

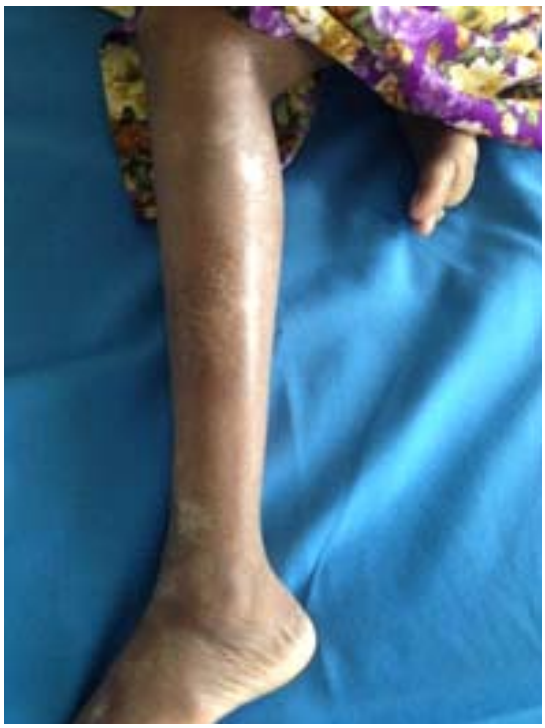

Fig. 1: Multiple, hyper to hypopigmented and brown-colored indurated plaques with atrophic, shiny surface along lateral border of left leg

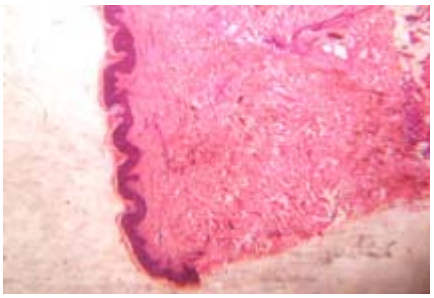

Fig. 2: Histopathology shows epidermal atrophy, a sparse superficial and predominantly deep dermal and subcutaneous perivascular infiltrate of lymphocytes and plasma cells, and the collagen bundles appears thickened

The cause of morphea is unknown. An autoimmune mechanism involving an increased frequency of autoantibody formation , which possibly is initiated by an environmental trigger such as local trauma to the skin and a higher prevalence of personal and familial autoimmune disease in affected patients are suggested etiologies. Elevated titers of one or more autoantibodies, most commonly anti-nuclear antibody (ANA) and anti-single stranded DNA (ssDNA) antibody are seen. Less commonly, rheumatoid factor, antiphospholipid antibodies, anti-histone antibodies, anti-Fcγ receptor antibody, and anti-topoisomerase IIα antibody are elevated.

Histology

In the early inflammatory phase of morphea there is an interstitial lymphocytic infiltrate distributed among deep dermal collagen bundles. Collagen bundles appears minimally swollen.

Over time, collagen bundles become thickened, hypocellular, and swollen. Lymphocytic infiltrates separate collagen strands, surround eccrine coils in the deep dermis, and are associated with loss of adipocytes around eccrine apparatus.

In an established lesion of morphea appendages, such as pilar apparatus are absent. Signs of inflammation is sparse and localized at the dermal–subcutaneous interface. The trabeculae dividing the subcutaneous fat are thickened and there is a patchy lymphocytic infiltrate. Pale, thickened collagen bundles appear arranged parallel to each other[4].

The treatment options include initial use of methotrexate and systemic glucocorticoids, followed by PUVA photochemotherapy, UVA1, narrow-band UVB phototherapy[5], or mycophenolate mofetil[6]. Other treatment options supported by randomized trials include topical vitamin D analogs topical tacrolimus, in combination with imiquimod and topical glucocorticoids[7].

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