

CASE REPORT

Disabling Pansclerotic Morphoea of Children Treated with Methotrexate

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Abstract

Disabling pansclerotic morphoea of children is a rare, aggressive, and mutilating variant of deep morphea. It usually begins before the age of 14 years and has relentless progression and severe disability. In children, the localized form of scleroderma is more frequent than the systemic but both of them are rare, constituting less than 3% of rheumatic diseases in childhood. Approximately 1.5% of all scleroderma cases occur before 10 years of age (1, 2). We present a rare case of disabling pansclerotic morphoea that responded well to methotrexate.

Keywords: Disabling pansclerotic morphoea, childhood, Methotrexate

Introduction:

Disabling pansclerotic morphoea is a rare atrophying and sclerosing disorder that involves epidermis, dermis, subcutaneous tissue, muscle and bone (3). It begins with the extensor of extremities and progresses to involve the trunk, flexors, face, and scalp, sparing fingertips and toes. Diagnosis is based on the history, physical examination and often skin biopsy. Though there are no specific therapies, various medications are used. Variable course of the disease with spontaneous remission in some cases and lack of standardization of outcome measures make evaluation of the effectiveness of these treatments challenging. We present a rare case of disabling pansclerotic morphoea that was treated with methotrexate and responded well.

Case Report:

A 6 years old male child presented with tightness of skin of right leg since 1 year and difficulty in walking and straightening of right leg since 6 months. There was history of development of multiple hyperpigmented lesions over trunk, forearm, and face since 2 months. Cutaneous examination showed unilateral multiple hide bound hyperpigmented indurated plaques involving face, arm, forearm, abdomen, and lower limb of right side [Fig. 1A, 1B]. Investigation showed normal hemogram, liver, renal function tests; with ESR of 32 mm. Radiological survey was normal. Antinuclear antigen was positive in the titre of 1:40 while anti double stranded DNA; anti-scleroderma antibodies (anti-Scl 70) were negative. Skin biopsy from indurated plaque showed atrophic epidermis and thickened dermis with dense collagen [Fig. 2].

Patient was started on weekly Methotrexate in the dose of 5 mg/m²/week (7.5mg/week) with monitoring of hematological, liver and renal parameters. After 10

cycles there were no new lesions and no increase in the old lesions and few lesions showed decreased induration. At present, patient has completed 20 cycles of the pulse

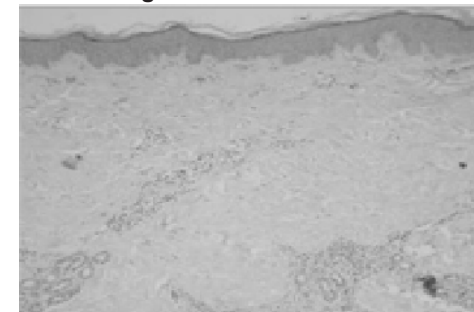
Figure: 1 A Hyperpigmented depressed plaque present on right side of the face.



Figure: 1 B Hyperpigmented depressed plaque on right thigh and lower limb.



Fig.2- Skin biopsy showing thickened dermis with dense collagen



Discussion:

Disabling pansclerotic morphea is a variant of localized scleroderma characterized by a rapid progression of cutaneous fibrosis with extension to joints and fascia. Various modalities of treatment are mentioned in the literature but with little effect. The response is often poor and in most patients the condition is progressive (4) and occasionally fatal (3). High dose of parenteral penicillin or ceftriaxone along with oral corticosteroid given in several courses over a time span of several months has beneficial effect in early cases (5). Phototherapy with UVA-1 (340-400 nm) has been found to be effective because of induction of interstitial collagenase [matrix metalloproteinase-1 (MMP-1)] and immunomodulation through induction of cytokines (6). Other therapeutic modalities tried with varying results are calcitriol (7), D-penicillamine (8), cyclosporine (9), and topical calcipotriene (10). Several therapies that may hold promise are Thalidomide and newer anti-tumor necrosis factor treatments (etanercept, infliximab). Low dose methotrexate had also being tried in widespread morphea (11, 12, 13). It inhibits dihydrofolic acid reductase enzymes involved in the synthesis of ribonucleic and deoxyribonucleic acids. The exact mechanism of action in localized scleroderma has not been well evaluated (11). Uziel et al. used methotrexate along with methyl prednisolone pulse (14). Combination of MTX and pulse methylprednisolone is well tolerated and appears to be effective in the treatment of localized scleroderma. In our case response to Methotrexate suggests its efficacy in reducing skin induration although further trials are needed. Surgical treatment like bone grafting, tissue expansion and microsurgical reconstruction may be required in cases of progressive facial hemiatrophy (15).

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