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Dermatology

DISABLING PANSCLEROTIC MORPHEA OF CHILDHOOD – A CASE REPORT

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ABSTRACT

INTRODUCTION: Disabling pansclerotic morphea (DPM) of childhood is a rare generalized type of localized scleroderma (LS) known to follow an aggressive course with pansclerotic lesions leading to severe joint contractures and consequent immobility. It is a chronic disease of unknown etiology, seen mostly in children less than fourteen years, involving sclerosis of all the layers of skin extending rapidly through the dermis and subcutaneous tissue to involve muscle, tendon and bone. It is distinguished from generalized scleroderma by its lack of systemic involvement. Mortality is due to complications of the disease such as bronchopneumonia, sepsis or gangrene. There is no specific laboratory finding. Treatment protocols are still evolving.

CASE REPORT: 14 years old boy presented with chief complains of tightness of the skin involving all the four limbs and trunk since 1 year. There was no history of Raynaud's phenomenon, or exposure to chemicals and dyspnea or dysphagia. On examination multiple sclerotic plaques with atrophy present on the trunk and all four limbs and a single non- healing, painful ulcer on the left foot and distal left leg. Skin biopsy showed thickening & homogenization of collagen bundles in dermis and subcutaneous issue. He was treated with oral steroids & methotrexate without any significant benefit.

CONCLUSION: Disabling pansclerotic morphea of childhood is exceedingly rare and severe form of Morphea. It can be very disabling and sometimes fatal. Treatment continues to present a therapeutic dilemma with only sporadic remission despite multimodality therapy. Early diagnosis is essential for initiation of proper therapy. An interdisciplinary approach is indispensable for proper management.

KEYWORDS : Disabling pansclerotic morphea of childhood, localized scleroderma, Dexamethasone pulse therapy, Methotrexate

INTRODUCTION:

Morphoea is a term used in preference to 'localized scleroderma', encompasses a group of related conditions characterized by varying degrees of cutaneous fibrosis and in the later phase atrophy of skin. It can sometimes extend deeply into muscle and bone.

Localized scleroderma has a wide clinical spectrum, ranging from superficial, circumscribed sclerotic plaques to severe, generalized and pansclerotic forms. The numerous clinical variants include: plaque-type morphea-including superficial, guttate and nodular variants; generalized morphea; linear scleroderma-including "en coup de sabre" lesions and Parry-Romberg syndrome (progressive hemi facial atrophy); and deep morphea-including morphea profunda, eosinophilic fasciitis and disabling pansclerotic morphea of childhood (DPMC). [1,2]

DPMC is a chronic disease, of unknown etiology which is seen mostly in children less than fourteen years, involving sclerosis of all the layers of skin extending rapidly through the dermis and subcutaneous tissue to involve muscle, tendon and bone. It is distinguished from generalized scleroderma by its lack of systemic involvement. [3] Progression of DPMC is explosive, with severe disablement causing mutilating contracture deformities, increased susceptibility to recalcitrant ulcers and malignant transformation with development of non-melanoma cancers.[4]

In isolated cases, it is found in association with hypergammaglob ulinemia,[5] thrombocytopenia, squamous cell carcinoma[6] and a highly unusual pleomorphic acid fast bacterium[7]

Mortality in DPMC is due to complications of the disease such as bronchopneumonia, sepsis or gangrene. There is no specific laboratory finding for this disease and treatment protocols are still evolving.

CASE REPORT:

A 14 years old boy presented with chief complains of tightness of the skin involving all the four limbs and trunk since 1 year. There was no history of sclerodactyly, Raynaud's phenomenon or exposure to chemicals and dyspnea or dysphagia. Family history was insignificant. On examination multiple, sclerotic and partially atrophic plaques of size ranging from 2 to 5 cm were present on the trunk, and both extremities.[Figure -1a,b,c,e,f] A single painful ulcer of size about 4× 2.5 cm with irregular borders, undermined edge and base covered with red granulation tissue was present over the left foot and distal left leg.[Figure -1d] Boy was severely handicapped with sclerosis involving all the four limbs. On general clinical examination no signs of visceral involvement was found. Patient's peripheral pulses were palpable and there was characteristic sparing of the tip of the fingers and face. Gait was difficult and there was a marked muscular atrophy of all limbs but the patient could write, hold objects in hands or care for himself unassisted. Laboratory investigations showed moderate hypochromic anaemia, thrombocytopenia and elevated ESR. Immunological profile showed no antinuclear, anti-centromere or anti scl-70 antibody. Culture of swab from the ulcer showed Staphylococcus aureus. On H&E stain, histopathology of the skin showed flattened and atrophic epidermis with loss of the rete ridges; homogenous and eosinophilic collagen deposition with diminished space in between seen extending from dermis up to subcutaneous tissue.[Figure-2] Patient was started on dexamethasone pulse therapy at every 28 days interval & intravenous antibiotics were started according to the culture sensitivity of swab. The patient received physiotherapy on a daily basis. The patient also received oral supplements of iron, folic acid, multivitamin and protein. During his two weeks stay, his wound improved but there was no response in the sclerosed skin. This therapy was given for 3 months then patient was shifted to tablet methotrexate 7.5mg/week.

DISCUSSION:

Disabling pansclerotic morphea of childhood (DPMC) is exceedingly rare and severe form of Morphea. It can be very disabling and sometimes fatal.DPMC may resemble at onset the plaque-type morphea or linear scleroderma and hence the diagnosis of the complete form, with its ominous prognostic implications might be established only late, once that pansclerosis with joint ankylosis and muscle atrophy occur. [8]

Anemia, thrombocytopenia and eosinophilia present in our case are usually rare in plaque-type morphea, but are described relatively frequently in pansclerotic morphea. [9,10,11] These parameters correlate well with the activity of the disease, and may be used to monitor the evolution.

During the diagnostic work-up of DPMC, it is important to differentiate this aggressive form of localized scleroderma from

progressive systemic sclerosis. Absences of Raynaud's phenomenon and of proper sclerodactyly, with characteristic sparing of the tip of the fingers are useful clues for the diagnosis of this disorder in the group of circumscribed scleroderma.

The prognosis of DPMC is poor and the complications to fear are: the possible generalization of cutaneous sclerosis, with irreversible ankylosis; progression of lesions to the face, with secondary mutilation due to scarring alopecia, ectropion, spontaneous amputation of ears, dental malpositions; osteolysis with distal amputations of phalanges; cachexia; transformation into systemic sclerosis; the development of cutaneous trophic ulcers with superinfection and possible septicemia; neuro-compressive syndromes and development of squamous cell carcinoma.

The lethal risk is related to chronic restrictive pulmonary disease in the event of extension of sclero-atrophic lesions of the thoracic wall; to superinfection of cutaneous ulcers with secondary septicemia; to secondary cancers developing on the areas with cutaneous sclerosis and ulcers and to progressive cachexia. [12]

Pansclerotic morphea, with all possible complications represent a challenge for the management. To date, there is no standard strategy of treatment, as no therapy has been consistently proven to be efficient in stopping the progression of the disease. [12,13,14,15] Corticosteroid pulse therapy is important for the strong anti-inflammatory and immunosuppressant effect, in highly active phases of the disease, with less of the risks of adverse reactions associated to long term corticoid therapy. Methotrexate is assumed to inhibit the synthesis of proinflammatory cytokines (IL-2, 4, 6, 8) and the fibrosis process.[16] Many studies have reported the successful use of PUVA and UVA1 in treating DPM.[17,18] UVA irradiation has been shown to reduce procollagen synthesis and induce the expression of collagenase-1, an enzyme, that cleaves collagen bundles.[19]

CONCLUSION:

DPMC is a rare and severe form of Morphea and can be very disabling and sometimes, fatal. Treatment of such disorder continues to present a therapeutic dilemma with only sporadic remission despite multimodality therapy.[20]

This article underlines the importance of a severe disorder, with cutaneous onset but with implications at systemic level, way beyond the "skin-depth", and with a potential lethal prognosis. The early diagnosis of such disorders is essential for initiation of proper therapy and interdisciplinary approach is required for the proper management of these difficult cases. Because of the lack of established efficient therapy and facing a disease with unpredictable evolution, a major role belongs to attentive care, physio-kinetic rehabilitation and careful monitoring of such patients for prevention of complications, especially those who bear a vital risk like skin cancers, ulcers, superinfections, and restrictive pulmonary disease.

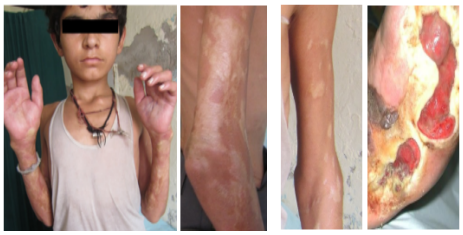


FIGURE-1 (a,b,c,d,e,f): Multiple sclerotic plaques coalescing together on extremities & multiple discrete sclerotic plaques over back. Left foot showing a non healing ulcer**.**

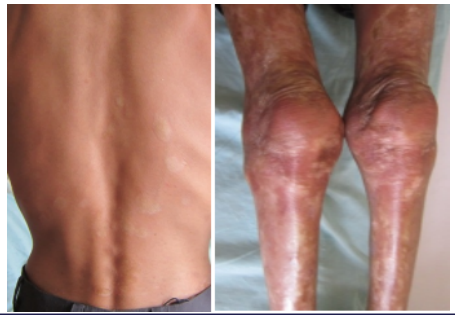
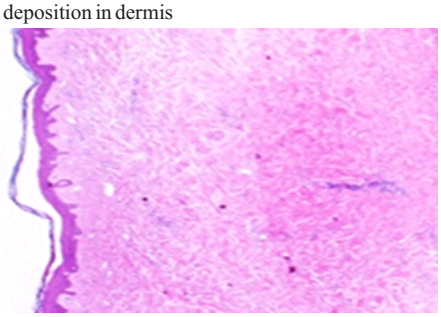


Figure-2: H&E stain showing atrophic epidermis with increased collagen deposition in dermis



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