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Case Report

Panscleroticmorphea: A case report

Anum Sadia, Javeria Zulfiqar, Nadia Ali Azfar, Muhammad Nadeem

Department of Dermatology, Sir Ganga Ram Hospital/ Fatima Jinnah Medical University, Lahore.

Abstract Generalized disabling pansclerotic morphea is a rare and mutilating form of morphea involving the dermis, subcutaneous tissue, fat, muscle, and even bone. It most often occurs in young children but adults can also be affected. We report a case of rapidly progressing and disabling adult onset generalized pansclerotic morphea, which responded well to methotrexate and steroid pulse therapy.

Key words
Adult, generalized pansclerotic morphea, steroid pulse therapy.

Introduction

Generalized disabling pansclerotic morphea is a rare form of morphea involving the dermis, subcutaneous tissue, fat, muscle, and even bone.[1] It causes severe functional and psychological impairment due to its aggressive and mutilating course. It is usually seen before the age of 14 years, with the patient complaining of arthralgia and stiffness at the time of onset.[2] Contractures, osteoporosis, and other bony changes are commonly seen.[3] It may be associated with raised erythrocyte sedimentation rate, eosinophilia, hypergammaglobulinemia[4] and squamous cell carcinoma.[5]

Case report

A 22 year old male presented on 7th August 2020, with diffuse thickening of skin for the last one year. It started from left forearm and rapidly spread to involve the whole body within a period of six months. Painful contracture deformities of hands, elbows, knees and hip joints also developed, impairing the patient's normal activities. There was also history of marked weight loss over a period of one year. There was no history of Raynaud's phenomenon. On examination, the patient had a thin cachexic look with multiple contractures, diffusely hard skin and multiple ulcerations over palms, soles and joint surface. There were thickened hyperkeratotic plaques over palms and soles and mottled hyperpigmentation of skin of the whole body including the face. There were claw like contracture deformities of the fingers and bilateral flexion deformities of elbow, knee and hip joints. Systemic examination was unremarkable. ESR was raised. Rest of the haematological parameters were normal. Antinuclear antibody, rheumatoid factor, antistreptolysin O titer and HIV screening were negative. Chest X ray was normal. Radiological examination of bilateral elbow and knee joints showed almost complete obliteration of joint spaces and generalized decrease in muscle bulk. The histological examination of skin was suggestive of pansclerotic morphea. The patient was started on low dose oral steroids and oral methotrexate along with intravenous antibiotics and other symptomatic treatments. Due to gastric side effects, oral methotrexate was shifted to subcutaneous route. Later on, it was combined with intravenous steroid pulse therapy. Along with these treatments, regular physiotherapy of the patient was also done.

Address for correspondence
Dr. Anum Sadia
Department of Dermatology,
Sir Ganga Ram Hospital/
Fatima Jinnah Medical University, Lahore.
Email: anumsaadia@gmail.com

Figure 1 Before disease onset.

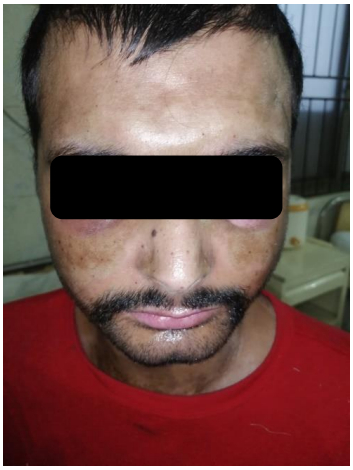

Figure 2 After disease onset; mottled hyperpigmentation of face and marked sclerosis below lower eyelids.

Figure 3 Panscleroticmorphea affecting whole body.

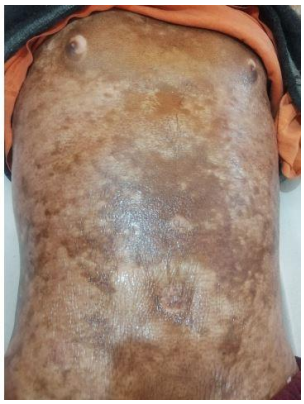

Figure 4 Abdomen.

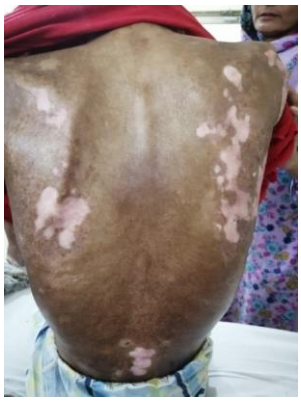

Figure 5 Back.

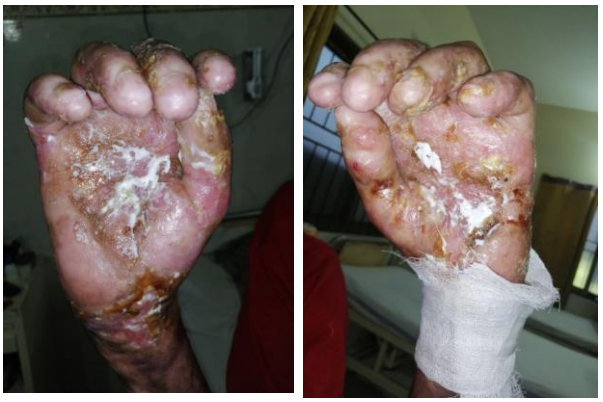

Figure 6,7 Right and left hands showing claw deformity with hyperkeratotic plaques and ulceration.

There was remarkable improvement in pain, ulcerations and range of movements.

Discussion

Panscleroticmorphea is a very rare disorder with extensive involvement of skin, subcutaneous tissue, muscle and bones. It starts from the extremities and may extend to the trunk, face and scalp. It results in significant morbidity due to its aggressive and mutilating course and can cause severe complications such as muscle atrophy, contractures, irreversible ankylosis, alopecia, ectropion, spontaneous amputation of ears, dental malpositions, osteolysis with distal amputations of phalanges, cachexia, development of cutaneous trophic ulcers with superinfection and possible septicemia, neurocompressive syndromes and development of squamous cell carcinoma.[6,7]

It is important to differentiate this aggressive form of localized scleroderma i.e. panscleroticmorphea from progressive systemic sclerosis. Sclerodactyly with characteristic sparing of the tip of the fingers and absence of Raynaud's phenomenon are findings that help us to differentiate between the two.[8]

Diagnosis of panscleroticmorphea is made mostly on clinical grounds, but skin biopsy may also help.[9]

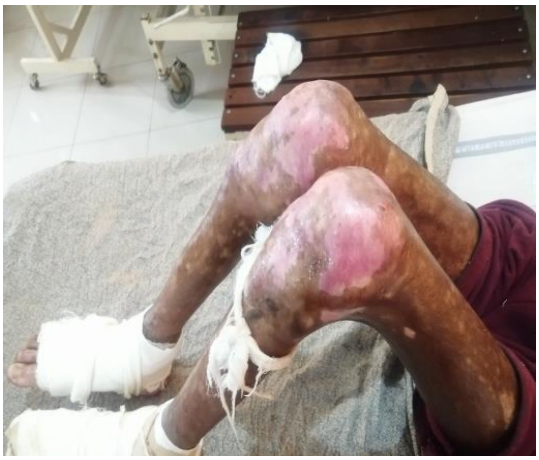

Figure 8 Flexion deformity of knee and hip joints.

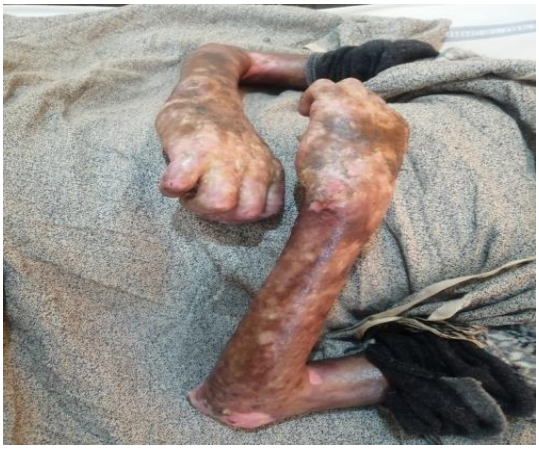

Figure 9 Felxion of deformity of elbow joints.

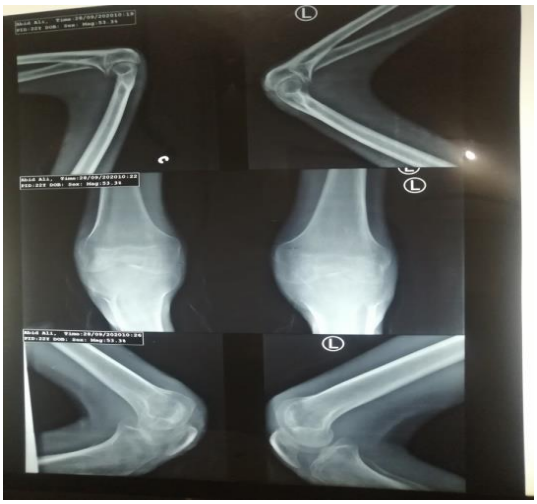

Figure 10 X-ray knee and elbow joints.

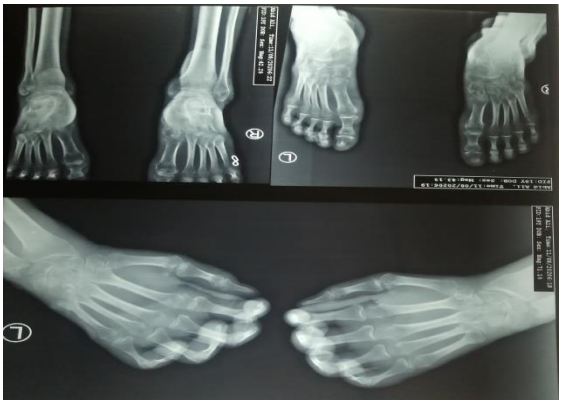

Figure 11 X-ray hands and feet.

Etiology of morphea is still unknown, however, several triggers have been implicated like trauma, BCG vaccination, previous radiotherapy or infection.[10,11] In our case, no triggering factor was identified.

No standard treatment strategy for the management of pansclerotic morphea has been developed. It is found that combined treatment with systemic steroids, methotrexate and UVA is more effective than monotherapy.[12] The use of low doses of methotrexate along with pulse therapy with methylprednisolone has the most favorable risk/benefit ratio.[13,14]

The aim of treatment in our patient was to halt the progression of the disease and improve the quality of life.

After giving steroid pulse therapy, there was marked improvement in his dependency on others for his normal daily activities and his mental well-being also improved.

Many new treatment modalities have been tried for pansclerotic morphea, with variable success rates. They include extracorporeal photopharesis, mycophenolate mofetil with IV immunoglobulins, interferon-gamma and recombinant human relaxin. Abatacept and tocilizumab have been reported effective in treating juvenile pansclerotic morphea.[15] Bosentan and sildenafil have been reported to show good results in patients with cutaneous ulcerations.[16,17]

Conclusion

As pansclerotic morphea has a poor prognosis, there is a need for regular follow up to keep a check on the complications and disease progression and to introduce further treatment modalities if needed. Absence of organ involvement must be regularly checked, as late progression to systemic fibrosis in the course of disabling pansclerotic morphea has been reported. In treating such a disease with unpredictable evolution and prognostic dilemma, a major place belongs to good nursing care, physio-kinetic rehabilitation and psychological counseling.

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