A CASE REPORT OF GENERALISED MORPHEA

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Summary

A female case of generalised cutaneous morphea is reported. The incidence as reported by A. Rook[2] is of the ratio 3:1 for F:M. Christiansen H.B. et al[1] in his study of 44 cases, reported that the incidence of generalised morphea was 1:44.

The onset of symptoms is generally between the age of 30 and 50 years. 80% showed manifestation between the age of 11 and 50[1]. Our case showed skin manifestation at the age of 63 years.

No familial nature of the similar skin involvement was seen in our case.

Indurated, shiny, whitish, and skin coloured plaque and scarring alopecia of scalp and occasionally bullae and ulcerations are the typical lesions of generalised morphea[2]. Our case shows the identical clinical picture.

Systemic manifestation and Contractures, joint-pains, rheumatoid arthritis were not present in our case.

Natural resolution have been seen in such cases, though corticosteroids, chelating agents, potassium P-amino-benzoate and relaxin have been advocated.

Generalised morphea is a rare condition in which sclerosis of the skin occurs in a widespread manner, usually starting on the trunk and unassociated with systemic disturbances. Aetiology of this condition is unknown. The age distribution of onset is between 30 and 50 years and 80% develop between the age of 11 and 50[1]. About 3 females are affected for every male[1]. No familial cases have been reported.

Clinically, the onset is usually insidious with development of plaques, resembling those of localised morphea. A lilac coloured border surrounding the indurated, ivory white, shiny lesions is usually seen in the early stages. The plaques are larger, usually start on the trunk, gradually increase in size with the development of new plaques. The areas involved are the upper trunk, breasts, abdomen, upper thighs. The arms, hands may also be involved like those seen in the tumid phase of systemic sclerosis with spindling of the fingers, binding of the skin to the underlying tissues and semiflexion of the hands and fingers. The legs, face, neck and scalp may also be involved. Scaring alopecia can result from involvement of the scalp. In some cases the whole of the body may be involved from the top of the head to the feet. Only one out of 44 cases showed such generalised involvement[1]. If the chest wall is markedly involved there may be difficulty in breathing due to constriction of the thorax.

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The face is expressionless, the skin being shiny, often brown and indurated. Telangiectasia of the face is not a feature and although the mouth opening may be restricted, the radial furrowing of systemic sclerosis is not seen. Raynaud's phenomenon occurs occasionally but is not a characteristic feature. After trauma, the hands may develop whitlows, but there is no atrophy as seen in systemic sclerosis. The tissues of the trunk and limbs sometimes show a browny non-pitting oedema. Bullae may develop in localised areas, particularly around the abdomen. A lilac border may not be seen in generalised morphea. Pigmentation is common and in some patients may be generalised.

Contractures occur in limbs, which become thin and hard. In acute phase, there may be soreness of the trunk and breasts. Joint pains occur in about 40% of patients. Definite rheumatoid arthritis may be found. Very occasionally severe contractures, atrophy and infection may be associated with intractable pain in the limbs and amputation may be required.

Sometimes, generalised morphea may develop as an extension of localised morphea or be associated with the lesions of lichen sclerosis et atrophicus.

Investigations for systemic disease is usually negative, as are blood counts and serum-proteins. L.E. cells have occasionally been found in generalised morphea[2]. *Histopathology*: the epidermis may be normal or flattened and atrophic with loss of the rete pegs. At first the dermis is oedematous, with swelling and degeneration of the collagen fibrils, which become homogenous and eosinophilic. A slight perivascular lymphocytic infiltrate may be present. Later the dermis is markedly thickened with dense collagen and relatively few recognizable fibroblasts. The elastic tissues are reduced. The dermal appendages and subcutaneous fat are progressively lost. Small blood vessels may show intimal thickening.

Electron microscopic studies have failed to demonstrate any consistent abnormality in the collagen fibres of the ground substance[3].

Case Report

A female patient aged 65 years attended the skin O. P. D. of the Nair Hospital, Bombay with complaints of itching, darkening and peeling of the skin of both legs since last 18 months, gradually spreading all over the body within 3 months. Thereafter she started developing thickening of the skin all over the body and infiltrated areas at places. She also used to develop bullae and ulcerations over trunk and extremities. There was no history of taking any medication before the complaints.

Patient had attacks of hypertension off and on since 15 years. No history of similar illness or other major illness in the past. No history of V.D.

Family history revealed death of parents, elder brother and elder sister due to unknown cause. Husband died 4 years ago due to Gastroenteritis. Only one son, developed high fever and mental disturbances at the age of 25 years and died subsequently at the age of 32 years.

General examination

Averagely built and nourished.

Bilateral immature cataract.

Systemic examination

No abnormality.

Local examination

The face was expressionless. Depigmented macular lesions (patches of leucoderma) were seen over upper eyelids. Shiny, whitish, tiny lesions were

seen-inner canthus of both the eyes. Telangiectasia were not noted on the face. There was no restriction of mouth opening. No radial furrowing seen around the mouth. Scarring alopecia seen over the scalp (Fig. 1). Indurated, waxy, skincoloured shiny infiltrated plaques and hyperpigmented scaly lesions and atrophic areas at places were seen over both upper and lower extremities (Fig. 2).

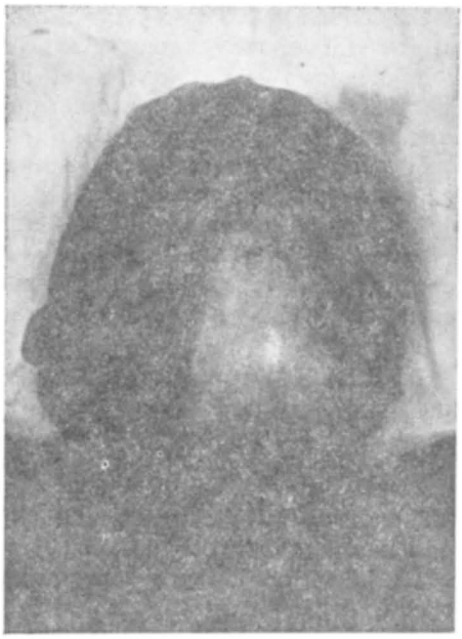

Fig. 1

Scarring alopecia on the scalp

Investigations

Complete haemogram: within normal limits; Urine and stool: N.A.D.; Liver function tests: within normal limits; E.S.R. 70 mm/hr; V.D.R.L.: negative, Blood-Sugar: F. 100 mg. %; P.P. 130 mg %; Serum cholesterol: 196 mg. %; Urea Nitrogen: 11 mg. %; Creatinine: 2 mg %; Serum amylase done on 7th June '72, 17 units and on 1st July '72, 33 units. Bleeding time, clotting time, Prothrombin time, platelet count were within normal limits.

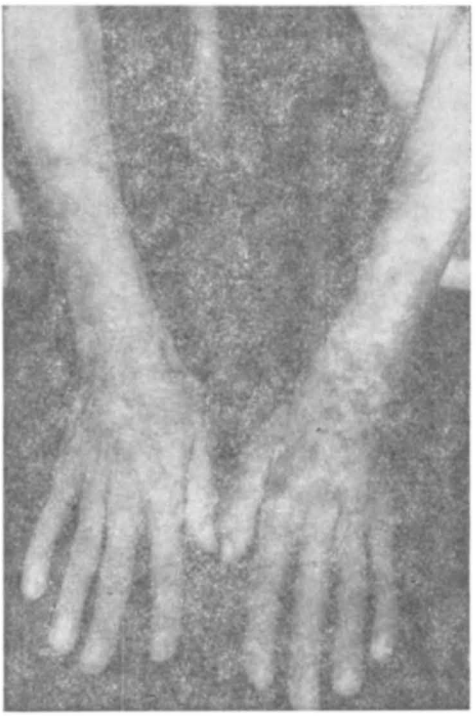

Fig. 2

Obophic areas over the upper extremitis

B.M.R: plus 6, E.C.G. and E.E.G: N.A.D., L.E. Cell: negative. Plain X-ray–chest, abdomen, skull and barium swallow, meal for oesophagus, stomach and duodenum and Barium enema showed no abnormality. Skin-biopsy the section stained with H–E stain (Fig. 3) showing normal stratum corneum with mild hyperkeratosis. The rete pegs are lost and the upper dermis is oedematous and the collagen swollen, homogenous and eosinophilic in staining. A slight lymphocytic infiltration is present between the collagen bundles and perivascularly. Sweat glands are atrophic.

Discussion

Indurated, shiny, white plaques over trunk, scarring alopecia over scalp and bullae, ulcers over trunk and extrimities presented in our case, were the typical features of generalised morphea.

Fig.

H E of the skin biopsy

Practically whole of the body was involved in our case which is otherwise rare as described by Christiansen et al[1]. In his study of the cases only one out of 44 had extensive involvement of body.

Contractures of limbs and joint-pain, rheumatoid arthritis were not found in our case. These findings may be present in some cases of generalised morphea[2].

L.E. cells have occasionally been found in generalised morphea[2]. In our case L.E. Phenomenon was negative.

In differentiating generalised Morphea from systemic sclerosis and scleredema : Frequent Raynand's Phenomenon, main involvement of face, hands and to a lesser extent feet, frequent systemic involvement occur more common in sclerosis than generalised morphea. Scleredema is usually a more acute and less widespread disorder.

Generalised Morphea lasts for years but some improvement is usually seen in the course of 3 to 5 years[2]. One patient still shows changes 33 years after diagnosis — Zarafonetis et al[2]. The patient usually remains in good health and if death occurs, it is due to other causes. The skin slowly softens and pigmentation decreases, tendency to ulcerations with trauma and blistering decreases. Some patients may be severely disabled while others may remain surprisingly active.

Histopathology study in our case is also compatible with that of generalised morphea.

There is no specific treatment for the condition. Corticosteroids, chelating agents, potassium P-amino-benzoate and relaxing have been tried with variable results, therefore their indications are still to be assessed. Contractures can be prevented by physiotherapy.

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