CALCINOSIS IN GENERALISED MORPHEA
(Case report)

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Summary

A rare case of generalised morphea with extensive calcification in a female patient is reported. The literature on the same is briefly reviewed.

Wilson is credited to be the first to use the term 'morphea'. He felt that the affected areas were vestiges of true leprosy[1]. Hilton Fagge in 1968 wrote an extensive essay on 'Keloid, Scleriasis and Morphea' and clearly delineated the keloid of Alibert (true keloid) from that of Addison (morphea[1]).

Sclerosis of the skin occurs in a variety of conditions, such as dermatomyositis and lupus erythematosus. The term 'scleroderma' should strictly be confined to sclerosis of the skin either localised or generalised, occuring in patients as the only or prominent feature. In such circumstances it is better to use the term 'morphea[2]'. Scleroderma is a disease of unknown cause. It has many synonyms and varies from a mild localised to a severe generalised form with systemic manifestations. "Standard Nomenclature of Diseases and Operations" has classified scleroderma into generalised, progressive and localised types. O'Leary classified the entity as follows: (i) localised scleroderma (Morphea) with guttate variety, (ii) linear scleroderma, (iii) generalised morphea, (iv) acrosclerosis (v) systemic scleroderma and (vi) hemiatrophy[3].

Generalised morphea is a rare disease, in which there is widespread sclerosis of the skin characterised by circumscribed or diffuse, hard, smooth, ivory colored areas that are fixed to underlying tissues and give the feeling of hide-bound skin. It is not associated with systemic involvement.

The occurence of calcinosis in the lesions of generalised morphea is rare. The available literature on the subject is very scanty. We are reporting a case of generalised morphea with extensive calcification in a female patient.

Case Report

A hindu female patient aged 38 years attended the skin OPD on 3-2-79 complaining of painful, hard lesions with ulcerations at places and discharge of white chalky material from the lesions for 4 years. History revealed that the patient had started to develop gradually progressive tightening and hardening of the skin of the face, forearms, hands, thighs and legs 13 years earlier. No illness preceded the development of the

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Received for publication on 4—5—1979

lesions. Since the patient was asymptomatic, she had not taken any treatment for her skin problem. There was no history of Raynaud's phenomenon, fever, difficulty in swallowing, abdominal complaints, urinary trouble or joint pain. 4 years prior to her hospital visit plaques were noticed on the hard and tight skin. First plaque was seen on the left arm and forearm. Within one year additional plaques of varying sizes appeared on the medial aspects of both thighs and on the lateral aspect of left leg. These plaques were painful. She took treatment from local doctors with no improvement. At times, the plaques ulcerated discharging chalky material. With the development of these plaques patient had restricted movements of thighs.

Family history was non-contributory. Systemic examination did not show any abnormality.

Fig. 1 A large plaque with ulceration on right thigh

The skin of the face, arms, forearms, hands, thighs and legs was smooth, shiny, indurated and could not be pinched up. The hair follicles in the affected area were prominent. There were tender plaques of irregular shape and sizes on the extremities (Fig. 1). Some of the plaques showed ulcerations from which white chalky material could be squeezed out. The plaques were so hardened, it gave an impression of wooden pieces kept in the plaques.

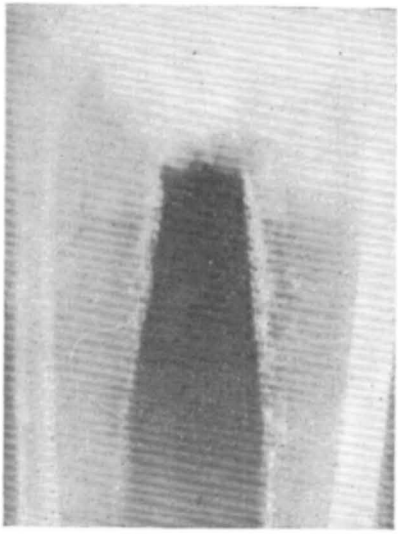

Fig. 2 Cutaneous and subcutaneous calcinosis of both thighs

Blood for TLC, DLC, HB%, E.S.R., L.E.Cells, urea : serum creatinine and serum calcium ; L,F.T; routine examination of urine and stool were all within normal limits. Radiological examination of all the affected areas revealed extensive cutaneous and subcutaneous calcification (Fig. 2 and 3) Radiological survey of rest of the skeleton did not show any pathology. X-ray chest was normal. Barium meal and barium enema were also normal. E.C.G. and fundoscopy of both eyes did not disclose any abnormality. Biopsy of the skin from the plaque showed features of calcifying morphea. Muscle biopsy of the affected area did not show any pathology.

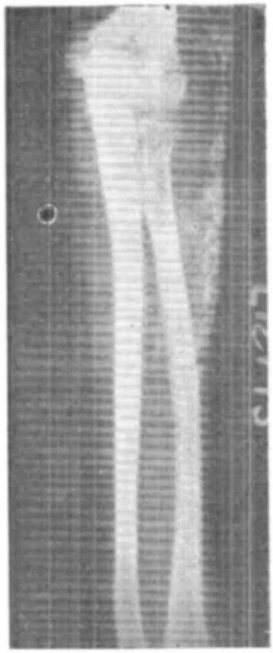

Fig 3 Cutaneous and subcutaneous calcinosis of left arm and forearm

Discussion :

Generalised morphea is a rare entity. The precise etiology of this condition is not known. It usually begins in the 3rd decade. About three females are affected for every male. The onset is insidious. Contracture, keratoses, calcinosis and squamous cell carcinoma can occur in the lesions of very long duration[2,4]. Regarding the relationship between morphea and scleroderma, some investigators opine that morphea is a limited form of scleroderma[5] or that it is associated with systemic scleroderma[6,7].

Calcinosis occuring in lesions of generalised morphea is very rare. In the studies on 235 cases of localised scleroderma[3] by Christianson et al in 1956 and 106 cases of scleroderma by Curtis et al[1] in 1958 no mention has been made regarding calcification. Muller et al[8] in 1959 reported the results of an elaborative study on 381 patients with all types of scleroderma. The female : male ratio was 10:1. Out of 132 cases with acrosclerosis 5 had calcinosis. Among 18 cases with widespread morphea one showed calcinosis and amongst 36 cases of linear scleroderma one revealed calcification. Remaining 195 cases did not reveal calcification. Calcification occured almost seven times more in patients with acrosclerosis than in patients with other forms of scleroderma. In 1959, Muller et al published a report on "Calcinosis in dermatomyositis". In this report, they pointed out the differentiating features of calcification occuring in dermatomyositis and scleroderma (Table 1).

TABLE 1

Calcinosis	Dermatomyositis	Scleroderma
1. Incidence :	Both children and adults; especially children.	Only adults.
2. Distribution :	Chiefly in proximal muscles of shoulder and pelvic girdle.	Usually in upper extremities, particularly hands.
3. Amount :	Usually large	Small to moderate.
4. Onset :	2 to 3 years after onset of the disease.	Usually in the 11th year of the disease.
5. Prognosis :	Very good for survival but poor for functional recovery.	Indicate chronicity. Functional impairment usually not great.

In the cases reported of morphea and/or systemic sclerosis by Shah et al[10], Handa et al[11], Goldberg et al[12], Laser et al[13], Paul et al[14] and Umbert et al[15] no mention has been made regarding occurence of calcification. In 12 cases reported of calcinosis cutis circumscripta by Syamala et al[16] none had underlying scleroderma.

The treatment of this entity is usually unsatisfactory. The physicians can only give symptomatic relief. General measures comprise of protection from cold, rest and physiotherapy. Calcinosis which may be a major problem may be managed by a combination of low calcium diet, cellulose phosphate-15 grams daily-an ionic exchange substance with an affinity for bivalent cations, and probenecid in dosage of 2 grams daily[17].

Our patient had generalised morphea for 13 years and had no involvement of any internal organ. This leads us to believe that generalised morphea and systemic scleroderma are two different diseases. In our case, the patient developed extensive calcification 9 years after the onset of the condition.

Acknowledgement :

We are thankful to Dr. M. N. Jindal, Dean and Superintendent, M. P. Shah Medical College and Irwin Group of Hospitals, Jamnagar for allowing us to publish this paper.

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