LOCALIZED SCLERODERMA
(Case Report of 23 Cases)

B. S. N. REDDY, * AND GURMOHAN SINGH †

Summary

Localized scleroderma is an uncommon disorder. The clinical and histological features in 23 cases of localized scleroderma are presented. A rare familial instance of en coup de sabre is described. The concurrent lesions of en coup de sabre and discoid lupus erythematous in a 16 years female is reported. It is felt important to differentiate between the facial hemiatrophy associated with en coup de sabre and Romberg's syndrome (progressive facial hemiatrophy).

KEY WORDS : Circumscribed Morphea, Linear Scleroderma Encoup De Sabre, Facial Hemiatrophy.

Localized scleroderma is a relatively uncommon, chronic benign disorder of the connective tissue of unknown etiology. It is recognized by localized sclerosis of the skin and subcutaneous tissue. On morphological basis it is further subdivided into three types namely, morphea, linear scleroderma and en coup de sabre with or without facial hemiatrophy. The etiology of localized scleroderma is not clear but autoimmunity, trophoneurosis, trauma, heredity, metabolic abnormalities, infections due to virus or acid fast bacilli have all been considered as possible etiological factors. Familial cases were reported in a few instances supporting role of heredity in this disease. The development of localized scleroderma like lesions in two children after successful bone marrow implantation for aplastic anemia, has been reported. This was therefore considered as a possible chronic graft versus host reaction and was quoted to support the autoimmune etiology.

Several interesting case reports and reviews of this disorder have been published from different places[1-5]. A few case reports have appeared in recent Indian literature[6-8]. We felt it relevant to review the clinical aspects of 23 patients with localized scleroderma. They were studied in the outpatient clinic of the Skin and V.D. department, Sir Sunderlal Hospital, Institute of Medical Sciences, Varanasi.

* Assistant Professor,
Skin & V. D. Department
Maulana Azad Medical College
New Delhi-110002
† Professor and Head
Skin & V. D. Section
Institute of Medical Sciences
Banaras Hindu University
Varanasi-221005
Request for reprints :
Dr. B. S. N. Reddy, Assistant Professor, Deptt of Skin & V.D., Maulana Azad Medical College, New Delhi.
Received for publication on 18—4—1981

Case Summaries

Twenty three patients were studied. Six had circumscribed morphoea, eight had linear scleroderma and nine had

frontoparietal lesions. Females were affected more commonly than males, the M : F ratio being 2 : 1. The cases were observed in the age group of 6-40 years, the majority being in the first and second decades of life. The linear type of lesions were predominant in children. History of trauma prior to the onset of lesions was elicited in four cases. In one family, the disease affected mother and daughter both of whom had en coup de sabre lesions. (Fig. 1).

In circumscribed morphoea the lesions started initially as hyperpigmented macules and gradually the skin in the centre of the lesion became thickened, smooth and taut with a violaceous hue at the border (Fig. 2). Hair was absent over the lesions. The common sites affected were chestwall, abdomen, back and limbs in that order of frequency. In linear scleroderma the lesions were distributed unilaterally in the form of ivory-coloured, sclerotic, ribbon-like bands on the extensor aspects of extremities. The lower extremities were more commonly involved. In one case the lesions were homolateral affecting both lower and upper extremities on right side of the body (Fig. 3A and B). Arthralgia, contractures and flexion deformities were observed in three cases. In one instance ulceration of the lesion was noted (Fig. 3B).

Fig. 2 Circumscribed morphoea over the waist, showing a whitish sclerotic plaque with peripheral violaceous hue.

Among the 9 patients with en coup de sabre, 4 had facial hemiatrophy of varying degrees. All these patients were males. All of them had a history of trauma prior to the onset of lesions. The lesions started as hyperpigmented

Fig. 1 Familial en coup de sabre occurring in mother and daughter.

Fig. 3 A

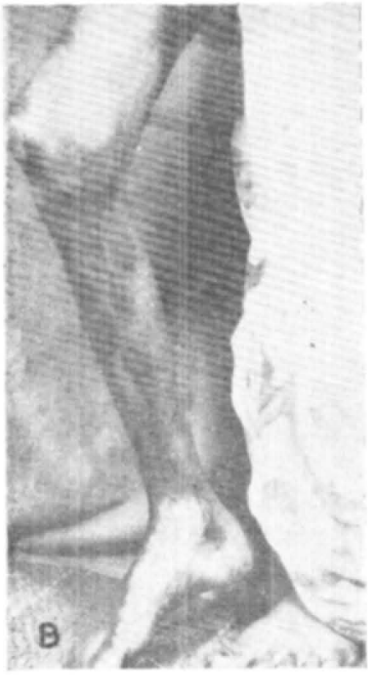

Fig. 3 B
Homolateral linear scleroderma. Note the flexion deformities and ulceration over the dorsum of the foot.

linear streaks on the frontoparietal region and gradually irregular linear depression appeared in the centre extending upwards producing cicatricial alopecia and in some cases extending downwards to the chin resulting in varying degrees of facial hemiatrophy (Fig. 4A). Telangiectasia of the vessels was noted at the border of the lesions. In one instance, patient in addition to en coup de sabre lesion had chronic discoid lupus erythematosus. The en coup de sabre lesion on the forehead was present for 6 years and atrophic, scaly, depigmented and scarred lesion of DLE on the mucous membrane of the lower lip was present for 2 years (Fig. 4B). The diagnosis on both lesions were confirmed histologically.

Fig. 4 A En coup de sabre with facial hemiatrophy on the right side of the face. Note the loss of hair on the scalp and eyebrows with an atrophic sclerotic furrow on the chin over the right side.

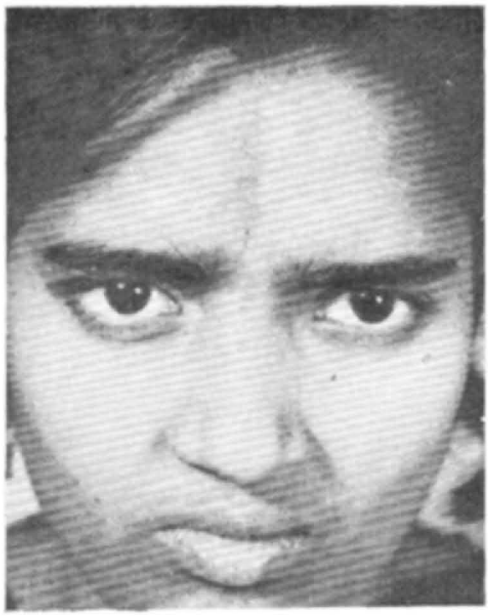

Fig. 4 B En coup de sabre over frontal region and DLE on the lower lip.

None of the patients had evidence of systemic involvement. Blood counts, erythrocyte sedimentation rate, electrophoretic pattern of serum proteins and VDRL test were normal. L.E. cells and antinuclear factor were absent. Radiological examination revealed no abnormality. The histologic picture varied according to the stage of the disease process and the site of biopsy taken. The sections obtained from the active border of early lesion revealed flattening of reteridges with lymphohistioytic infiltrate around blood vessels and between the collagen fibrils in the lower two-thirds of the reticular dermis and fibrous trabeculae of the subcutis. Thickening of the vessel wall and narrowing of the lumen was noted at places. Dermal collagen bundles appeared swollen, homogeneous and oedematous with increased eosinophilia. Specimens obtained from the older lesions revealed the collagen bundles markedly thickened, sclerosed and packed closely with little inflammatory infiltration. The appendages were seen in varying degrees of atrophy and destruction.

Repeated intralesional injections of triamcinolone acetonide (4 mg/ml) at weekly intervals for 9 to 12 weeks were found to be most effective in the resolution of localized plaques (Fig. 5A and B). The response was not satisfactory with this treatment in linear scleroderma involving extensive areas on the limbs. However, physiotherapy with Vitamin E 10 mg daily for a period of 2–3 months was found to be of some benefit.

Discussion

The preponderance of lesions in females, gradual onset and the nature of clinical and histological features observed in our patients are all similar to the findings reported by other workers [1,2].

Familial scleroderma is rare. Review of the limited literature available on this subject by Burge et al[9] in 1962 revealed only five convincing reports of familial scleroderma and two more of

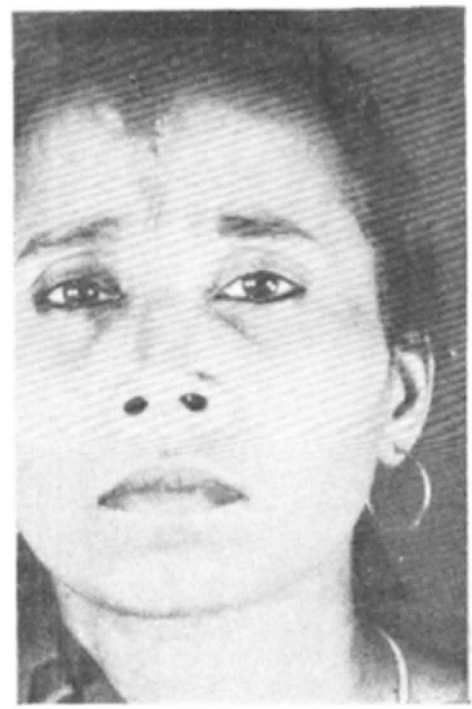

Fig. 5 A En coup de sabre

Fig. 5 B Same patient as in Fig 5A after treatment with intralesional triamcinolone acetonide.

probable familial cases which were incompletely documented. Christianson et al[1] in their study of 235 cases of localized scleroderma observed only three instances of familial disease. Reporting two cases of familial localized scleroderma, Wuthrich et al[4] stated that this is exceedingly rare. Franschetti et al[10] noted a few lesions of frontal scleroderma occurring in families and suggested[9] genetic basis for this disorder. We observed one instance of familial scleroderma in our series. The disease manifested as en coup de sabre in mother and daughter. To our knowledge this is the first documented case of familial en coup de sabre in Indian literature.

The association of one or more systemic collagen diseases in the same patient is well known but the occurrence of localized scleroderma and discoid lupus erythematosus (both are cutaneous disorders) in the same patient is exceedingly rare. Stating that only two such instances were available in literature till then. Umbert and Winkelmann[11] reported four more such cases and proposed a rare cutaneous disease similar to systemic "Mixed" or "Overlap" connective tissue disease. We had one patient with such a "mixed" disease. The occurrence of such a case to our knowledge is so far unrecorded in Indian literature.

Linear scleroderma may be associated with congenital anomalies of the vertebral column like spina bifida and cervical rib[1], a phenomenon not observed by us. Ulceration of the lesions is an uncommon feature in localized scleroderma (5 out of 108 in Christianson's series[1]). In one of our cases of linear scleroderma chronic ulceration of the lesion was seen over the dorsum of the foot.

Fronto-parietal scleroderma (en coup de sabre) is sometimes associated with varying degrees of facial hemiatrophy (4 in our series). This may be confused with progressive facial hemiatrophy (Parry-Romberg Syndrome). It has been stated that both these entities are different manifestations of a single disease process and that it is impossible to distinguish the two[12]. We believe these diseases to be separate entities and may be differentiated despite similarities. Scleroderma manifesting as en coup de sabre is a relatively superficial and localized process. The skin over the lesion is hyperpigmented, fibrotic and fixed to the underlying structures with prominent loss of hair on the scalp and eyebrows, resulting in cicatrical alopecia. On the other hand, progressive facial hemiatrophy is a more diffuse process often involving deeper structures such as muscles and bones. The overlying skin is freely movable and appears normal with no loss of hair. Other workers also consider this entity to be distinct from en

coup de sabre. We feel this differentiation is important because scleroderma presenting as en coup de sabre being a relatively mild disease process, if detected early may be arrested by local infiltration of intralesional corticosteroids at the active border of the lesion.

The relationship between localized scleroderma and systemic sclerosis is not clear. Occasionally patients with localized scleroderma have been reported to develop systemic sclerosis later [1,2]. Visceral localization and immunologic abnormalities similar to systemic sclerosis were reported in a few cases of localized scleroderma[14]. Ikai et al[15] reported a case of acrosclerosis developing morphoea like cutaneous plaques later. Certain observations like detection of dysproteinemias and antinuclear factors in some cases of morphoea, transformation of localized scleroderma into a systemic process and similar results of chronaxy, histology, histochemistry, X-ray defraction, historadiographic and electron microscopic studies in both the conditions lead some authors to speculate that both these entities are basically one disease process[3]. Inspite of these observation, it is important to note that the clinical picture and course of the disese is distinct and different in both these conditions. Systemic sclerosis may lead to a fatal termination unlike localized scleroderma which is a relatively benign self-limiting disorder. Variation in the clinical picture, absence of Raynaud's phenomenon and lack of serological abnormalities in most cases of localized scleroderma may serve a useful purpose in differentiating these two entities. None of our patients demonstrated systemic involvement or serological abnormalities. Since the prognosis is entirely different in these two conditions, it is appropriate to consider them as separate entities for therapeutic purposes.

1. Christianson HB, Dorsy CS and O' Leary PA : Localized scleroderma, Arch Derm, 1956; 74 : 629-639.
2. Curtis AC and Jansen TG : The prognosis of localized scleroderma, Arch Derm, 1958; 78 : 749-757.
3. Irgang S : Scleroderma, a comprehensive review, Cutis, 1973; 12 : 848-859.
4. Wuthrich RC, Roenigk HH Jr. and Stick WD : Localized scleroderma, Arch Derm, 1970; 111 : 98-100.
5. Bonafe JL, Lasseri J, Oksman F et al : Localized scleroderma in children, Rev Med Toulouse, 1976; 12 : 825-830.
6. Shah RN, Marquis L and Mehta TK : A case report of generalized morphoea, Ind J Derm Vener Leprol, 1973; 39 : 199-202.
7. Handa F, Agarwall RR and Singh RL : Familial disseminated morphoea, Ind J Derm Vener Leprol, 1974; 40 : 108-111.
8. Pavitran K : Linear morphoea with hypertrichosis and entrapment neuropathy, Ind J Derm Vener Leprol 1981; 47 : 45-49.
9. Burge KM, Perry HO and Stickler GB : Familial scleroderma. Arch Derm, 1979; 99 : 681-687,
10. Franceschetti A and Koenig H : quoted by 9.
11. Umbert P and Winkelmann RK : Concurrent localized scleroderma and DLE. Arch Derm, 1978; 114 : 1473-1478.
12. Wartenburg R : Progressive facial hemiatrophy, Arch Neur Psychiat, 1945; 54 : 75-96.
13. Dawson TAJ : Facial hemiatrophy (Parry-Romberg syndrome), Brit J Derm, 1966; 78 : 545-546.
14. Bourgeois-Droin C and Touraine R : Scleroderma in plaques-Immunological and visceral disturbances, Fra Ann Med Intern, 1978; 129 : 107-112.
15. Ikai K, Tagami H, Imamura S et al : Morphoea-like cutaneous changes in a patient with systemic scleroderma, Dermatologica 1979; 158 : 438-442.