DIFFUSE SYSTEMIC SCLERODERMA: A RARE VARIETY OF PROGRESSIVE SYSTEMIC SCLEROSIS—A CASE REPORT

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Summary

A middle aged women presented with unusual weakness, multiple joint pain & tiny boil like eruptions over her upper part of the body. Her skin was diffusely hyper pigmented with area of depigmentation and dry rough & hidebound. She had no systemic involvement. Diagnosis was made by skin biopsy. Prognosis was good as she had only skin involvement.

Introduction

Progressive systemic sclerosis (PSS) is a multi systemic disease of unknown etiology rarely seen in Indian subcontinent[1]. Its diagnosis is easy when typical skin lesion is evident and a physician carefully examine the skin. This is a rare varierty of PSS, for this reason we reporting this case.

Case Report

A 30 years old Hindu female patient was admitted in medical unit of Sher-E-Bangla Medical College Hospital with weakness, multiple joint pain & multiple acneform eruption over the back and chest. She was a house wife, mother of one child & coming from poor socio-economic background.

She felt unusual tiredness during her household work for the last one year. She had occasional joint pain in both knee joints and all fingers of both hands for the last ten months but her joints were never swollen and no history of morning stiffness. She noticed whitish area over her forearms & legs and it was gradually increasing for last two years. She had no problem of numbness sensation & pallor of hands & feet after exposure to cold. She gave no history of dysphagia, epigastric fullness, retrosternal burning, bloating and abdominal pain, diarrhoea or constipation. She had no cold intolerance, loss of appetite, unusual weight gain or hoarseness of voice. She had no respiratory distress, cough, urinary problem or oedema.

Her face, trunk and dorsum of hands & feet were hyper pigmented (fig. 1) and skin over both shin area & forearms were depigmented (fig. 2). She had multiple acneform eruptions over her chest, back & upper limbs. Skin over the face, trunk, upper & lower limbs was dry, coarse, thick & firmly bound to the underlying structures. She failed to open her mouth fully & able to make her fist only partially. Her body build was average & her pulse, temperature, respiration, blood pressure & other systems were found normal.

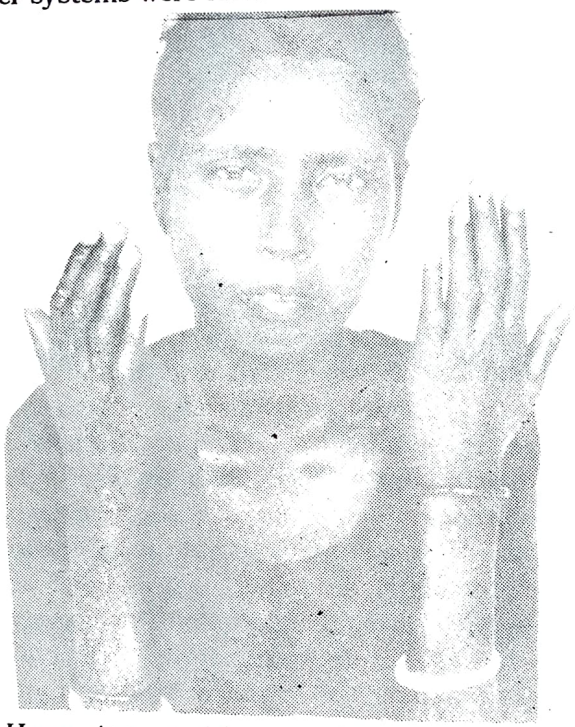

Fig-1-*Hyperpigmented face, trunk & dorsum of hands.*

Laboratory examination :

1. Blood Count-Neutrophilic Leucocytosis.
2. ESR - 67mm 1st. hour (westergren method).
2. HB%-52%
4. Routine urine analysis-No abnormality.
5. X-ray Chest, X-ray of hands, fluoroscopic examination of Barium swallow of oesophagus, Barium meal of stomach & follow through were normal.
6. Peak Expiratory Flow Rate-350 1/min

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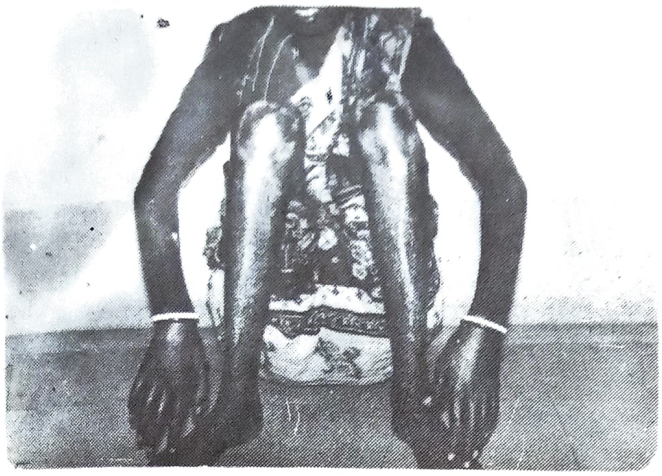

Fig-2 : *Depigmented area over shin & forearms.*

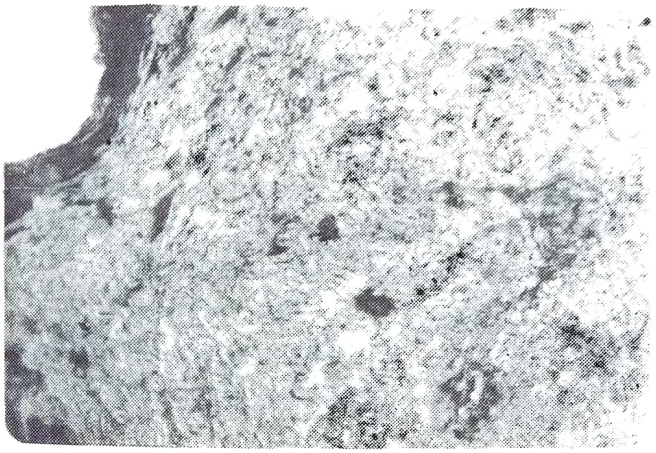

Fig-3 : *Paraffin section of skin (hematoxylin-eosin stain X40) showing atrophy of skin with dense sclerosis of dermal tissue.*

7. ECG-Normal.

8. Skin Biopsy - (H & E stain) showed atrophy of skin with dense sclerosis of dermal tissue and atrophy of the adnexa, (Fig-3)

Discussion

On the basis of cutaneous involvement PSS is of 2 types. In 95% cases hands & face affected first, then gradually extends centripetally is termed as "acrosclerosis"[2] It is associated with Raynaud's phenomenon and which may precede other manifestations by months or even years. In about 5% cases the cutaneous lesions first appear on the trunk and is termed as "diffuse systemic scleroderma." Raynauds phenomenon is absent in such patients[3]. Diffuse hyper pigmentation is usual in this variety. Macular telangiectasis on the face & hands, calcinosis cutis & ulceration on the tips of the fingers and over the knuckles occur predominantly in acrosclerosis[4].

In the both form of PSS the skin in the involved areas is diffusely indurated and as a result of diffuse fibrosis of the subcutaneous fat, they are firmly bound to the underlain structures[5]. The skeletal muscles are affected resulting in weakness and atrophy.

This patient had no features of Raynaud's phenomenon, telangiectasis, calcinosis cutis, ulceration of skin or dysphagia. As the patient was coming from poor socio-economic background she ignored her pigmented and depigmented skin. She was examined for unusual tiredness and accidentally during examining anaemia it was found that skin of lower eye lid was firmly bound to the underlying structures. The skin was examined thoroughly & ultimately diagnosis was confirmed by skin biopsy.

As her internal organs were not affected her prognosis was good.

References

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