Scar Depigmentation in Systemic Sclerosis

Sir,

Pigmentary changes are commonly seen in systemic sclerosis and usually occur in 3 patterns (1): (i) a diffuse brown melanoderma that mimics Addison's disease, (ii) patchy areas of depigmentation interspersed with perifollicular pigmentation, so-called "salt and pepper" pigmentation, and (iii) focal hypopigmentation and hyperpigmentation on sclerosed skin. We have recently observed depigmentation in the scars of patients with systemic sclerosis, a clinical feature that appears not to have been described previously.

CASE REPORTS

The sign was observed in 5 patients with advanced systemic sclerosis. There were 4 women and one man, whose ages ranged from 22 to 58 years. All patients fulfilled the American Rheumatism Association criteria for the diagnosis of systemic sclerosis and had disease for periods ranging from 2.5 to 7 years. All the patients had Raynaud's phenomenon and binding down of the skin of the extremities, face and trunk. Four patients had sclerodactyly, while all had finger tip ulcers and/or scars. Pulmonary functions were deranged in all the patients and 4 had clinical dyspnoea. Barium swallow revealed decreased esophageal motility in 4 patients. One patient had proteinuria.

All patients had patchy depigmentation with residual perifollicular pigmentation within the macules ("salt and pepper pigmentation"). Diffuse hyperpigmentation of the skin was seen in 2 patients. In

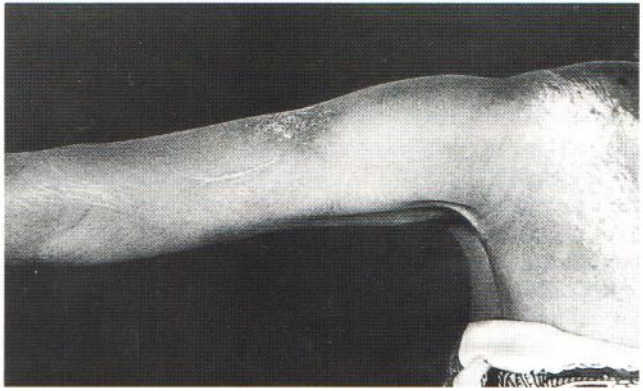

Fig. 1. Depigmentation in muscle biopsy scar on arm. Note "salt and pepper" pigmentation on adjacent skin and on the chest.

addition, all patients had depigmentation in the scars following muscle biopsy (1) (Fig. 1), skin biopsies (2), smallpox vaccination (1), a healed axillary sinus (1) and in long-standing scars (2), whose cause the patients were unable to recall. Three scars were normally pigmented before the onset of systemic sclerosis and depigmented subsequently; 3 scars developed after the onset of disease and were depigmented de novo, while one patient was unable to date the onset of depigmentation in the scar. In no patient did the depigmentation of scars precede the onset of the "salt and pepper" depigmentation of systemic sclerosis.

COMMENT

The patchy depigmentation of systemic sclerosis has many clinical, histological, histochemical and ultrastructural similarities to vitiligo (2). Koebner's phenomenon is known to occur in vitiligo, and a similar process may explain the depigmentation that developed in the fresh scars following trauma, skin and muscle biopsies in our patients. However, depigmentation of the pre-existing scars of smallpox vaccination and previous injury cannot be thus explained.

An alternative explanation is suggested by the clinical observation that focal depigmentation develops on sclerosed skin (2). If this finding indicates that depigmentation supervenes when a critical level of dermal sclerosis has been reached, it could explain the predilection of depigmentation for fibrotic scars, new or old.

While our finding is unlikely to be of any diagnostic value, since it appears in patients with well-established, unmistakable systemic sclerosis, it may provide clues to the pathogenesis of this ill-understood disease.

REFERENCES

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