LETTERS TO THE EDITOR

Systemic sclerosis and porphyria cutanea tarda

Sir: The appearance of sclerodermoid changes in porphyria cutanea tarda is well known; cases of porphyria cutanea tarda associated with systemic sclerosis have rarely been described, however.² A further case of this association is reported here. Repeated photobothy led to a slight improvement in the cutaneous sclerosis, but visceral involvement remained unchanged.

A 48-year-old man was admitted for study of generalised cutaneous thickening. He was a heavy smoker and drinker. He had received no drugs and had not been in contact with toxic products. Outbreaks of vesicular lesions on the body, including between the toes of the feet and intermittent dark urine had been evident for the past three years. Generalised skin thickening, particularly on his hands, and Raynaud’s phenomenon had developed nine months before admission. Other clinical data included constitutional symptoms, pyrosis, impotence coeundi of six months’ duration, and diffuse cutaneous hyperpigmentation lasting, in the past three months, before admission.

Physical examination showed erosions, crusts, scars, and milium cysts in areas exposed to the sun and on the soles of his feet. There was diffuse hyperpigmentation with vitiligo-like patches. Skin thickening of the chest, back, abdomen, face, and arms made pinching impossible and greatly limited articular function. The legs were relatively unaffected. The finger tip showed pitting scars. There were no telangiectasias or subcutaneous calcifications. The remainder of the physical examination, blood and urine analysis were unremarkable. Alpha-fetoprotein, carcinoembryonic antigen, cortisol, and adrenocorticotrophic hormone were normal; iron 10 µmol/l (normal 10–28 µmol/l), transferrin 1.7 g/l (normal 1–6–2.7 g/l), saturation index 22% (normal 20–40%), ferritin 350 µg/l (normal 18–300 µg/l), the urinary uroporphyrin level was 440 µg/mol/24 h (normal <48 µmol/24 h) with presence of faecal isoporphyrins; antinuclear antibody titre was 1/160 homogenous pattern and 1/10 240 speckled pattern; and ScI-70 and other antinucleus antibodies were negative.

A chest radiograph showed slight emphysema. Functional respiratory tests showed slight restrictive ventilatory disturbance, with impairment of both the transfer factor of the lung for carbon monoxide and the transfer coefficient. Pulmonary gammagraphy with gallium-67 was normal. A hand radiograph disclosed lysis of the distal phalanges. Nailfold capillaroscopy showed capillary dilatation in several fingers. Abdominal echography showed slight liver enlargement. Liver biopsy showed moderate hepatic siderosis, but no needle-like inclusions in the hepatocyte cytoplasm were seen under polarised light microscope. Skin biopsy disclosed epidermal hyperpigmentation of the basal layer, a thickened basement membrane positive for periodic acid-Schiff reagent, superficial perivascular hyalinisation, and an increase in dermal collagen. Oesophageal manometry showed hypotension of the lower oesophageal sphincter and severe hypomobility of the distal two thirds of the oesophagus.

The patient was diagnosed as having porphyria cutanea tarda and systemic sclerosis. Alcohol abstinence was recommended and treatment with repeated phlebotomy (300 ml every two weeks) was started. At six weeks of treatment urinary porphyrins were normal and there was a slight improvement in the dermatological and cutaneous sclerosis was seen over several months. Oesophageal manometry results after six and 24 months of treatment remained unchanged.

Our patient had porphyria cutanea tarda before the clinical onset of systemic sclerosis. We wonder whether this was a chance coexistence of two admittedly different pathogenic entities or whether there is a causal relation between them.

Sclerodermoid lesions may develop in 18 to 27% of patients with porphyria cutanea tarda. They are usually late lesions, which appear in patients with untreated porphyria cutanea tarda of several years’ duration and tend to have a characteristic appearance and location.² The face may take on a systemic sclerosis-like appearance and the fingers may show sclerodactyly not usually associated with Raynaud’s phenomenon.⁵

Experimental studies have shown that uroporphyrin I increases collagen synthesis in human skin fibroblast cultures but skin cyanosis is a characteristic feature of porphyria cutanea tarda. In our case, the appearance of lesions was immediate, and lesions were confined to the skin. This suggests the existence of a characteristic porphyria cutanea tarda skin lesion even in the presence of pigmentary changes and normal levels of normal and abnormal porphyrins.²³

Increased concentrations of soluble interleukin-2 receptor in the serum of patients with systemic sclerosis

Sir: The extent to which the immune system is important in the pathogenesis of systemic sclerosis is still poorly understood. In a recent issue of the Annals Gustafsson and colleagues showed the presence of increased numbers of HLA-DR positive T lymphocytes in the peripheral blood of patients with systemic sclerosis. These findings were healthy controls.¹ Expression of HLA-DR antigens as well as the interleukin-2 receptor on a cellular surface is acquired by T cells after stimulation by antigens or mitogens.

It has been reported that a soluble form of interleukin-2 receptor is released in culture supernatants by activated lymphocytes,² derived mostly from T cells,³ and that it can be measured in the sera of patients with various chronic disorders in which the immune system is activated. Therefore, the soluble form of interleukin-2 receptor has been regarded as a new marker of T cell activation: increased concentrations have been noted in patients with various autoimmune diseases, including systemic lupus erythematosus, rheumatoid arthritis, and Sjögren’s syndrome.⁴—⁶

We evaluated the concentration of the soluble form of interleukin-2 receptor in the sera of patients with systemic sclerosis by an enzyme linked immunosorbant assay (ELISA) based on the sandwich principle (Cellfree, T Cell Sciences, Cambridge, MA). Results were expressed in units/ml relative to a set of standards supplied with the test kit.

Twenty eight patients (27 women, one man; median age 52 years) were studied. They met the American Rheumatism Association preliminary criteria for the classification of systemic sclerosis:¹¹ 21 had typical sclerodermatous skin changes proximal to the metacarpophalangeal joints and ScI-70 was present in nine of them and antitopomerin in three); sclerodactyly and at least three of the other four signs of the CREST syndrome (calcinosis, Raynaud’s phenomenon, oesophageal dysmotility, sclerodactyly, and telangectasia) were present in the other seven patients (six of them were antitopomerin positive).

Serum concentrations of the soluble form of interleukin-2 receptor were markedly higher in patients with systemic sclerosis than in 19 controls matched for age and sex (mean (SD) 846 (387) U/ml vs 301 (90); p<0.005, Wilcoxon’s test). Twenty four of 28 patients (86%) had concentrations of the soluble form of interleukin-2 receptor more than 3 SD above the mean concentration of the controls. Patients with the CREST syndrome had non-significantly lower concentrations than those with diffuse scleroderma (679 (148) U/ml vs 902 (427)). We agree with those of other previous studies showing the presence of activated lymphocytes in systemic sclerosis.⁴—⁶ Our results support the findings of Gustafsson et al.,¹ however, as we did not note a significant inverse correlation between disease duration and the marker of T cell activation (R=−0.04; Spearman’s test).


8 Ann Rheum Dis, first published as 10.1136/ard.50.4.270-a on 1 April 1991. Downloaded from http://ard.bmj.com/ on June 18, 2022 by guest. Protected by copyright.