Sclerodactyly, CREST syndrome, proximal scleroderma

Sir, In their interesting paper Furst et al. define CREST as patients with sclerodactyly and PSS as those with ‘proximal scleroderma’. It is surprising that the authors found differences in the internal organs of the two subsets only in the case of lung involvement. It is reasonably well known that patients with limited skin sclerosis (so-called CREST syndrome) have a less severe prognosis compared with patients with ‘diffuse scleroderma’. One possible explanation of this unexpected finding could be that the PSS subset with proximal scleroderma constitutes too broad a group of patients, including also cases in whom skin sclerosis is confined to a few areas of the skin (face, neck). Such cases could be closer to sclerodactyly than to diffuse scleroderma. Furthermore, the clinical findings in Table 1 of the paper by Furst et al. show that not all cases of CREST syndrome presented all five features of CREST, and that other patients with proximal scleroderma also had features of CREST. What in effect distinguishes the two groups is that the so-called CREST group presents skin sclerosis confined to the fingers, whereas the other group has skin sclerosis also in other areas. This being so, would it not be better to speak of patients with sclerodactyly rather than CREST?

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References

Anti-Ro positive rheumatoid arthritis

Sir, The high incidence of side effects in patients with anti-Ro positive rheumatoid arthritis treated with d-penicillamine noted by Moutsopoulos will be of interest to clinicians. A 61% incidence of side effects compared with 8.5% in the anti-Ro negative group would question the use of d-penicillamine in anti-Ro positive patients. Our own observations on a small group of patients with rheumatoid arthritis treated with d-penicillamine have not confirmed this striking difference. A group of 17 rheumatoid arthritis patients, most of whom had developed serious side effects, was studied retrospectively. The results are shown in Table 1. Of the 13 who developed side effects, four had anti-Ro antibodies. A further nine patients had their anti-Ro status determined before commencing d-penicillamine. Two of these have developed side effects, and both were anti-Ro negative. The two patients who were anti-Ro positive have not developed side effects after more than six-months’ treatment. Of the 15 patients with significant side effects, only four...