86 Correspondence


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 the 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


Sir, Professor Giordano's letter points out a source of misunderstanding that has existed for some time in the rheumatic disease literature. For some groups the diagnosis of CREST has separated patients specifically from those with PSS. In most cases, however, patients with criteria for CREST syndrome can also have PSS. Thus the presence of CREST criteria per se, does not separate patients in terms of prognosis.

Those few patients who have CREST alone (without criteria for PSS) may, in fact, have a different prognosis than those with the 'usual' diagnosis of CREST syndrome. However, the patients with CREST alone are relatively infrequent. As stated in our paper, over 150 prospectively followed-up patients with systemic sclerosis, we only found five CREST patients who did not meet criteria for PSS. Thus all of our CREST patients had skin involvement confined to the fingers, and we referred specifically to that difference in our paper. On the other hand, we had five CREST patients who did not have PSS by any criteria, and they did not appear different in any way from the 12 CREST patients who met the minor criteria for PSS (also as mentioned in our paper).

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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 in 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
Sclerodactyly, CREST syndrome, proximal scleroderma.

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