

Skin Scoring in Systemic Sclerosis: A Modification – Relations to Subtypes and the Aminoterminal Propeptide of Type III Procollagen (PIIINP)

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Forty-one patients with systemic sclerosis were investigated with a new and simple skin score method measuring the degree of thickening and pliability in seven regions together with area involvement in each region. The highest values were, as expected, found in diffuse cutaneous systemic sclerosis (type III SS) and the lowest in limited cutaneous systemic sclerosis (type I SS) with no lesions extending above wrists and ankles. A positive correlation was found to the aminoterminal propeptide of type III procollagen, a serological marker for synthesis of type III collagen. The skin score is considered simpler than previous methods and is recommended for more general use. *Key words:* fibrosis; evaluation; serological markers.

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MATERIAL AND METHODS

The body is divided into seven regions (Fig. 1): head and neck, trunk, fingers, hands, arms, feet, and legs. The degree of thickening and pliability is quantified as by Kahale et al. (7) by numerical units: 0 for normal skin, 1 for thickened skin, 2 for decrease in possibility to pinch and/or move skin, and 3 for skin that the examiner is unable to pinch and/or move (hide-bound skin). The worst part of the region determines the score. Involvement in each area is then determined by the counts: 0 for non-involvement, 1 for <33%, 2 for 33–67%, and 3 for >67%. The sum of the numerical units is the scleroderma skin status score. We studied serum PIIINP and scleroderma skin status score in 41 patients with SS. Serum PIIINP was assayed by radioimmunoassay from Orion Diagnostica, Oulunsalo, Finland, according to a previously described method (10).

The American Rheumatism Association established in 1980 "preliminary criteria for the classification of systemic sclerosis (scleroderma)" (1). Since then these criteria have been the common reference when patients are studied. Systemic sclerosis (SS) is, however, a rather heterogenous disease, and therefore many attempts have been made to subdivide the disease into different types. The most common method today is to distinguish between limited cutaneous systemic sclerosis (lSSc), with type I representing an acrosclerosis with no lesions extending above the wrists and ankles, and type II having lesions extending above the wrists and ankles. Type III is so-called diffuse cutaneous systemic sclerosis (dSSc) with sclerotic lesions involving the trunk. The head and neck may be involved in all three types (2). Although the frequency and severity of new organ system involvement is the most important measure of disease progression, it has been common to use skin scoring as well, and now and then a skin score alone for the evaluation of prognosis and therapy (3–6). The original scoring system proposed by Medsger and coworkers in 1980 (7) was found to have an unacceptable inter-observer variation by Kahale et al. (8), who therefore in 1986 modified it. These scoring systems were based on measurements of both extent and severity. Another method (4) calculates only the extent of skin involvement.

In this brief communication we suggest a simpler version of the early scoring system (6, 7), based on the experience in dermatology from the PASI-score used in psoriasis (9). We have related scoring to the different subtypes of SS as well as to the aminoterminal propeptide of type III procollagen (PIIINP), which is a serological marker for type III collagen synthesis (10) that has been reported to correlate with skin involvement and internal organs in SS (11, 12).

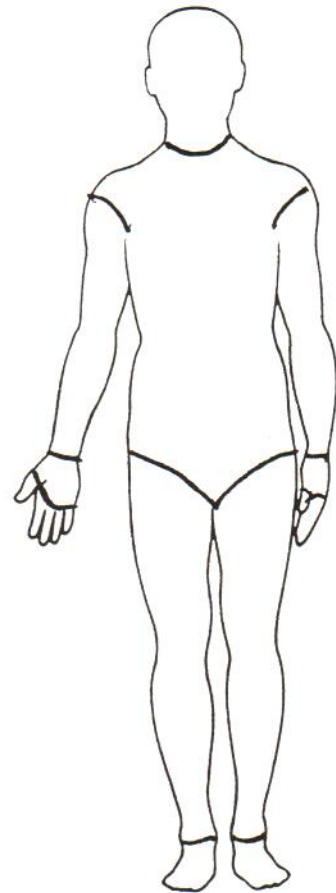


Fig. 1. Schematic representation of the seven regions used for scoring.

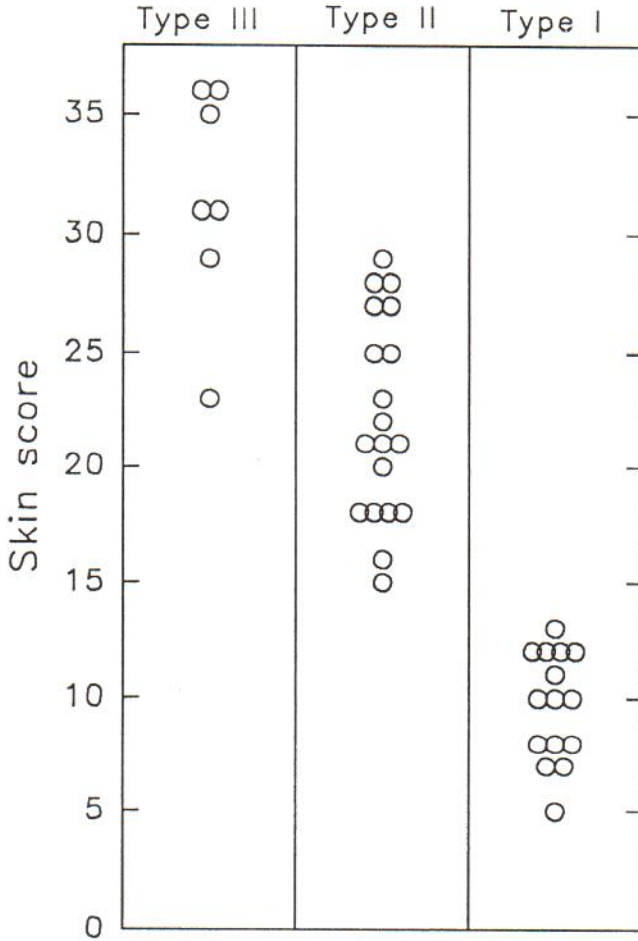


Fig. 2. Data of individual skin scores for patients suffering from systemic sclerosis divided into the three common subdivisions.

RESULTS

The data shown in Fig. 2 are scleroderma skin status score for the 41 patients divided into the three accepted subdivisions of SS. The mean scores for the different subtypes appear from Table I. Looking for a correlation between scleroderma skin status score and PIIINP (Fig. 3), we found a positive correlation with a linear regression ($r=0.69, p<0.005$). Fig. 4 shows an example of scleroderma skin status score and PIIINP followed in a 54-year-old male with RNP antibodies from the start of the disease and for 2 years during different types of therapy. The patient was originally classified as type I, later as type II, and finally as type III.

Table I. Mean skin scores for three types of systemic sclerosis

Type	No	Mean score	25-75 percentiles
Type I	15	10	8 -12
Type II	19	21	18 -26.5
Type III	7	31	29.5-35.75

The differences in the median values among the groups are greater than would be expected by chance ($p<0.001$) using chi-square approximation.

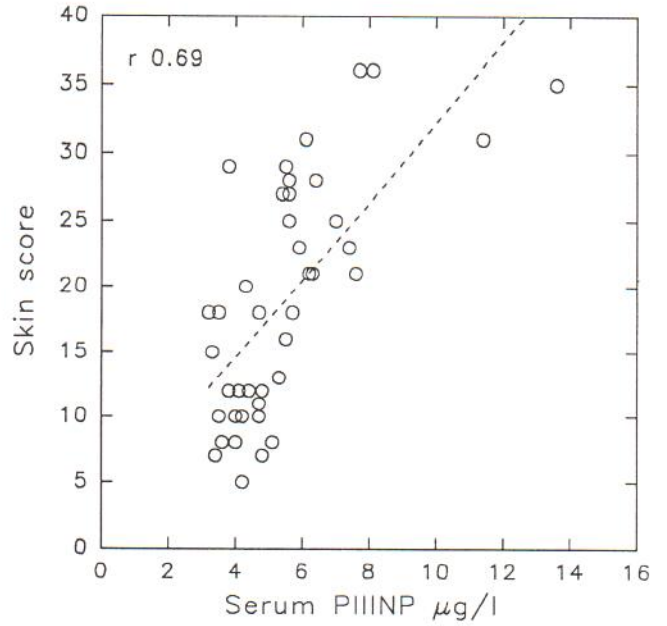


Fig. 3. Relationship between skin scores and PIIINP in patients with systemic sclerosis.

DISCUSSION

We have found the skin scoring of Kahale and coworkers (7) too time-consuming for daily clinical use. This also applies to the version reported by Brennan et al. (12). The present and even simpler modification, where the regions have been cut down from 22 over 17 to 7, takes into account the progression in the extension of the disease by assessing percentage of involved skin within each region. It thereby combines the advantages of the two types of skin scoring systems: the Medsger and Rodnan type (7, 8) and the so-called Manniken type used by Jimenez & Segal (4), which only measures areas.

Our data show the expected difference between the subgroups with the highest mean score in type III SS. They also demonstrate a correlation between scleroderma skin status score and PIIINP as serological marker for collagen synthesis. Fig. 4 gives

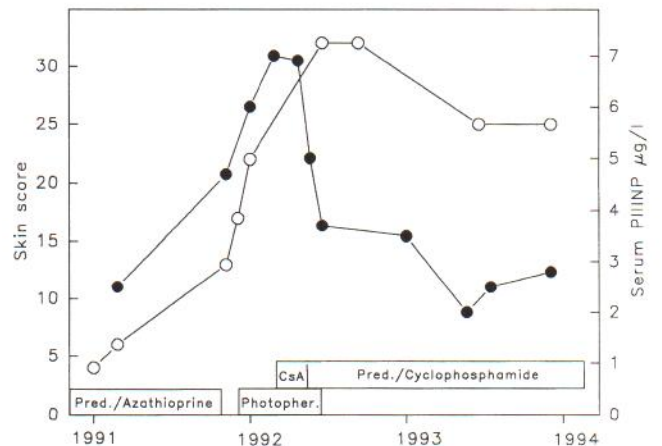


Fig. 4. Skin scores (○) and PIIINP (●) in the serum of a 54-year-old male during the first 3 years following the diagnosis of his disease. The therapy given during this period appears in the figure.

an example of an increase in PIIINP later followed by a similar increase in skin score, which is the logical sequence of events.

Since the present scoring method, as has been the case with previous skin scores, does not take into account joint mobility, ulcerations, and calcinosis, our data for pulpa/vola distance, mobility of hands and elbows, number of digital ulcers, and number of calcifications on fingers, are always included in the same chart as the skin score. We hereby recommend this simpler skin score for more general use.

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