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This is the most comprehensive report in the English-language literature on nail unit BCC. Because a number of extensive reviews regarding carcinoma of the hand lack specific tumor location, however, it is possible that other cases, not included here, exist. There is further difficulty in establishing the exact number of cases because the validity of the histopathologic diagnoses in several cases has been questioned by Mehregan. Also notable but not included in the summary is a case report by Ashby of a basosquamous carcinoma that clinically mimicked paronychia.

Treatment of BCC of the nail unit has ranged from radiation to amputation. The use of Mohs micrographic surgery in 87 cases of nail unit squamous cell carcinoma documented this as an excellent approach for cutaneous carcinomas lacking bone involvement. That report noted that healing by second intention gave superb results. With emphasis on achieving a permanent cure while preserving function and cosmetic appearance of the nail unit, Mohs micrographic surgery followed by second-intention healing should be regarded as the current treatment of choice for nail unit BCC. Full- or split-thickness skin grafting is an alternative to second-intention healing when more rapid wound coverage is desired and when the wound base is sufficiently vascular to support a graft. Functionally and cosmetically, the results are essentially equivalent.

All reported cases of nail unit BCC have lacked classic clinical features, resulting in initial clinical misdiagnosis. The presentation often mimics such benign processes as chronic paronychia, chronic dermatitis, or onychomycosis. A unique presentation of longitudinal melanonychia was reported in one case. Bowen's disease is a malignant mimic. We stress the need for biopsy of all nail unit lesions atypical in appearance, course, or response to therapy.

REFERENCES
nation with a quickly performed objective method to assess thickness.

Several methods have been advocated but have never been validated.1-3 Objective measurements with ultrasonography,4,5 laser Doppler flowmetry,6 skin torsion,1 and serum type III procollagen aminoterminal peptide7 have been described. Rodnan, Lipinski, and Luksick8 measured skin thickness and collagen content in biopsy specimens. This method, however, has the disadvantage of being invasive, time-consuming, and unsuitable for patients with widespread morphea.

A device to measure skin hardness (the durometer) was first described by Falanga and Bucalo9 and was found to be useful in the assessment of patients with systemic sclerosis when used as a comparative test.10 For morphea, however, the use of the durometer has not been described.

Clinical scores of skin involvement have been used extensively in the assessment of patients with systemic sclerosis.11-16 For morphea, clinical skin scores have been used in a few cases but have never been evaluated for their reliability.1,3,17-19 An easy and quickly performed modified skin score (MSS) for systemic sclerosis has been described by Zachariae et al.20 This scoring system has the advantage of dividing the body into only seven regions (rather than 17 to 26 regions as in previous methods).8,11,15,17 The MSS measures the degree of thickening and pliability in combination with area of involvement.

Neither the durometer nor the MSS has been used before in the assessment of morphea, and the interobserver and intraobserver variability of both methods have not been determined. In this study, these were investigated. The degree of agreement between the two methods was also calculated.

PATIENTS AND METHODS

Patients

Thirteen consecutive patients with histologically confirmed morphea were included in this study. All patients were scored twice at a 1-week interval by three independent observers with variable experience: a consultant dermatologist, a consultant rheumatologist, and a resident in dermatology. Each observer first conducted the MSS as described by Zachariae et al.20 and then assessed skin hardness in the most indurated lesion in each of seven regions by means of the durometer.

Clinical score

The MSS is a clinical skin score in which the body is divided into seven regions (R): head and neck, trunk, arms, hands, fingers, legs, and feet. The degree of thickening and pliability (T) was assessed on a 0 to 3 scale: 0, normal skin; 1, thickened skin; 2, decreased ability to pinch or move skin; and 3, skin that is unable to be pinched or moved (hidebound). The most affected part of the region determined the score. In addition, involvement in each area (A) was determined by estimation and given the following counts: 0, no involvement; 1, less than 33%; 2, 33% to 67%; and 3, more than 67%. The sum of the numeric units for thickening and the percentage of area surface involved is the MSS. Thus the MSS is \( \Sigma(T + A)_{r1-r7} \). The possible minimum score is 0, representing no affected skin, and the maximum score is 42, for extreme involvement in all areas.20

Durometer scores

The hardness of the lesions was examined by means of a handheld durometer (model 1600-00; Rex Gauge Co., Glenview, Ill.) with an affixed weight of approximately 400 gm. In this way measurements were carried out with a constant weight that did not allow additional pressure.9,10 The durometer is fitted with a calibrated gauge that registers linearly divided units on a scale from 0 to 100. The durometer readings were taken at the intersection of two imaginary lines drawn through the largest horizontal and vertical diameters of the most indurated lesion in each of the seven regions. Four consecutive readings were taken at the same site while the patients were lying flat, and contraction and muscle tension were avoided by putting a pillow under the extremities. The total durometer score is the sum of the means of the four determinations at each site.

Statistical analysis

The total durometer score (the sum of the means of all body sites) was used for statistical analysis. Observer agreement for both methods was analyzed by considering the total amount of variation present in the ratings and estimating the amount of variability from the patients (\( \sigma^2_p \)), the amount from the repeating of the measurements (\( \sigma^2_m \)), the amount from the observers (\( \sigma^2_o \)), and the amount from random error (\( \sigma^2_e \)). In a reliable method, the interobserver variability (clinician variation) and the intraobserver variability (variation from repeated measurements) should be low, whereas the variability caused by diversity of patients should be high.

All variances mentioned were estimated with the maximum likelihood variance components estimation procedure of the SAS package (SAS Institute Inc., Cary, N.C.). Results for the various sources of variation are presented as the percentage of the total variance.
Table I. Percentage of total variance from observers, repetition, patients, and random error

<table>
<thead>
<tr>
<th>Variance (% of total)</th>
<th>MSS</th>
<th>Durometer score</th>
</tr>
</thead>
<tbody>
<tr>
<td>Variance from patients</td>
<td>80.0</td>
<td>93.9</td>
</tr>
<tr>
<td>Variance from observers</td>
<td>2.2</td>
<td>0.5</td>
</tr>
<tr>
<td>Variance from repetition</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Variance from random error</td>
<td>17.8</td>
<td>5.6</td>
</tr>
</tbody>
</table>

\( (\sigma^2_p + \sigma^2_o + \sigma^2_r + \sigma^2_e) \). For analysis of correlations between the MSS and the durometer scores, the Spearman correlation coefficient was used.

RESULTS

Thirteen patients (four men and nine women) participated in this study. Their mean age was 49.2 (± 4.57 SEM) years; the youngest was 20 years old and the oldest was 68 years old. The duration of morphea ranged from 9 months to 7 years, with a mean of 3.9 (± 0.8 SEM) years. The number of body sites affected ranged from one to four; the fingers, hands, and feet were not affected in any patient.

In the MSS, the percentage of variance from the patients was high, 80.0% (Table I). This means that the amount of variation from the other variables (observers, repeated measurement, and random error) was low. The interobserver variability, expressed as the percentage of variance from observers, was 2.2% (Table I). The intraobserver variability, which is the variation from repetition of measurements by the same observer, was estimated to be 0 (Table I). The interobserver variability in the durometer score was 0.5, and intraobserver variability was estimated as 0.

Correlations between the total MSS and the durometer score were approximately 0.5. If only the pliability scores in the MSS were correlated with the durometer scores, a correlation of more than 0.8 was found. The correlations between MSS (total MSS and pliability alone) and the durometer score for the different observers and both time points are summarized in Table II.

DISCUSSION

To assess disease activity and efficacy of therapy in scleroderma, it is important to use standardized outcome measures that are reliable, clinically meaningful, and feasible to use.16 Both the durometer and the MSS proved reliable in the assessment of morphea. The interobserver variability of the durometer score was low, even lower than the interobserver variability of the MSS (0.5% vs 2.2%). Correlations between the total MSS and the durometer score were 0.5, which is not high. This is because in the MSS not only pliability but also area of involvement is represented. This area of involvement, which accounts for half the total MSS, cannot be correlated with hardness measured by the durometer. If only the pliability scores in the MSS were correlated with the durometer scores, a correlation of 0.8 was found. In all analyses, the correlation coefficient never reached 1, which indicates that the highest durometer scores are not always found in lesions with the highest score for pliability. An explanation might be that the durometer scores vary with the localization of the lesions on different body parts. The durometer values thus generally increase with clinical severity scores but do not always do so. This is in accord with the results found by Aghassi, Monoson, and Braverman10 and Falanga and Bucalo9 in systemic sclerosis. It is recommended to use both methods simultaneously for the most accurate assessment of patients with morphea and for monitoring therapeutic effects in clinical trials.

REFERENCES

assessed by computerized image analysis on 20 MHZ B
5. Serup J. Localized scleroderma (morphoae): thickness of 
sclerotic plaques as measured by 15 MHZ pulsed ultra
6. Klyszcz T, Hahn M, Junger M. Laser Doppler flowmetry 
in treatment follow-up of circumscribed scleroderma. 
Serum aminoterminal propeptide of type III procollagen in 
progressive systemic sclerosis and localized scleroderma. 
8. Rodnan GP, Lipinski E, Luksick J. Skin thickness and 
collagen content in progressive systemic sclerosis and 
9. Falanga V, Bucaio B. Use of a durometer to assess skin 
10. Aghassi D, Monoson T, Braverman I. Reproducible mea
surements to quantify cutaneous involvement in sclero
thickness in systemic sclerosis: an assessment of interobserver variability in 3 independent studies. J 
intraobserver variability of total skin thickness score 
(modified Rodnan TSS) in systemic sclerosis. J 
scores and clinical measurements in scleroderma. J 
scleroderma skin scoring method. Clin Exp Rheumatol 
16. Pope JE, Bellamy N. Outcome measurement in sclero
33. 
17. Steen VD, Medsger TA Jr, Rodnan GP. D-penicillamine 
therapy in progressive systemic sclerosis (scleroderma): 
18. Falanga V, Medsger TA Jr. D-penicillamine in the treat
ment of localized scleroderma. Arch Dermatol 1990;126: 
609-12. 
tochemotherapy in systemic sclerosis and severe mor
score in systemic sclerosis: a modification—relations 
to subtypes and the aminoterminal propeptide of type III 
procollagen (PIIINP). Acta Derm Venereol (Stockh) 

Single patch of hair at a denervated site in a patient with alopecia 
universalis

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Alopecia areata and its variants, alopecia totalis and alopecia universalis, are diseases of unknown cause. We describe an unusual presenta

tion of alopecia universalis that spared an area denervated during a lymph node biopsy.

CASE REPORT

A 36-year-old man with long-standing alopecia univer

salis was examined to evaluate recent nail changes. 
Nail pits and loss of lunulae had been present since 17 
years of age. He also reported loss of sensation in the 
right axilla after a lymphadenectomy also at 17 years of 
age, during which cutaneous branches of the intercos-
tobrachial nerve were severed. Alopecia universalis 
subsequently developed, with the exception of the right 
axilla.

Examination revealed complete hair loss except for

Fig. 1. Patch of axillary hair adjacent to linear scar.