The Challenge of Very Early Systemic Sclerosis

To the Editor:

We read with great interest the article by Blaja, et al. on the challenge of very early systemic sclerosis (SSc). We have been addressing this topic for some years. Actually, VEDOSS (very early diagnosis of systemic sclerosis), subsequently renamed very early SSc, was proposed before the development of the 2013 American College of Rheumatology/European League Against Rheumatism (ACR/EULAR) classification criteria. The careful analysis of these criteria prompted us to emphasize the need to clearly define the boundaries of the condition. In addition, the subsequent detection of an evolution into SSc satisfying criteria in only 50% of strictly defined very early/early SSc led to the proposed term undifferentiated connective tissue disease at risk for SSc (UCTD-risk-SSc).

Defining which of the patients satisfying VEDOSS entry criteria already fulfill SSc classification criteria requires a careful assessment of each patient by history, physical examination, and physiologic and imaging investigations.

Predicting the evolution into definite SSc by those strictly labeled UCTD-risk-SSc has long been considered an accomplished task. In that regard, based on the disease course of 102 patients, we have developed a weighted score (Table 1). By receiver-operation characteristic curve analysis, patients with a score at admission ≥ 2.75 evolved into SSc, with a sensitivity of 91.3% and a specificity of 73.2%.

The analysis of disease evolution in the VEDOSS cohort patients who at enrollment did not fulfill ACR/EULAR criteria for SSc and did not present any manifestation consistent with SSc sine scleroderma might validate or disprove the use of the criteria.

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Table 1. Variables independently predictive of development of SSc based on multivariate regression. Relative weight in a 10-point score.

<table>
<thead>
<tr>
<th>Variable</th>
<th>HR</th>
<th>95% CI</th>
<th>p</th>
<th>Weight</th>
</tr>
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<tbody>
<tr>
<td>Anti-Scl-70</td>
<td>11.57</td>
<td>3.12–42.88</td>
<td>0.0003</td>
<td>3.00</td>
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<tr>
<td>ANA ≥ 1:320</td>
<td>6.93</td>
<td>1.81–26.51</td>
<td>0.0049</td>
<td>2.75</td>
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<tr>
<td>Avascular areas</td>
<td>5.03</td>
<td>1.36–18.59</td>
<td>0.0158</td>
<td>2.25</td>
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<tr>
<td>ACA</td>
<td>3.92</td>
<td>1.13–13.59</td>
<td>0.0319</td>
<td>2.00</td>
</tr>
</tbody>
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REFERENCES