Background: Disease activity as measured by the European Scleroderma Study Group Activity Index (EScSG-AI) (1) has been recently found to predict the development of damage over time in an early systemic sclerosis (SSc) cohort (2). The European Scleroderma Trials and Research Group Task Force for the Development of Revised Activity Criteria for SSc recently succeeded in identifying a preliminarily revised activity index (EUSTAR-AI) that had a greater construct validity than the EScSG-AI i.e. performing better in identifying patients with active disease (3).

Objectives: To assess in patients with SSc the predictive value of the EUSTAR-AI for disease severity accrual.

Methods: SSc patients from the EUSTAR database with a disease duration from the onset of the first non-Raynaud sign/symptom/5 years were first extracted. Patients were considered for the study if they presented the following features: a) availability of data included in the EUSTAR-AI, in the EScSG-AI and in the Medsger severity scale at baseline and yearly for 2 consecutive years; b) availability of vital status at the last observation; c) availability of other disease features known to predict disease progression and development of severe organ involvement in SSc and works better than EScSG-AI.

Table 1. Predictive features of severity progression by multivariate regression analysis

<table>
<thead>
<tr>
<th>Predictive factors</th>
<th>OR (IC 95%)</th>
<th>severity deterioration</th>
<th>p-value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Adjusted mean EUSTAR-AI age</td>
<td>1.41 (1.23-1.61)</td>
<td>Δ summed severity score ≥ 1</td>
<td>&lt;0.0001</td>
</tr>
<tr>
<td>Adjusted mean EUSTAR-AI peripheral vascular</td>
<td>1.32 (1.14-1.54)</td>
<td>Δ lung severity ≥ 1</td>
<td>0.0002</td>
</tr>
<tr>
<td>Adjusted mean EUSTAR-AI heart</td>
<td>1.40 (1.15-1.70)</td>
<td>Δ heart severity score ≥ 1</td>
<td>0.0005</td>
</tr>
<tr>
<td>Adjusted mean EUSTAR-AI Peripheral Vascular</td>
<td>1.48 (1.21-1.82)</td>
<td>Δ skin severity score ≥ 1</td>
<td>0.0002</td>
</tr>
<tr>
<td>Adjusted mean EUSTAR-AI Peripheral Vascular</td>
<td>0.97 (0.95-0.99)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Adjusted mean EUSTAR-AI</td>
<td>1.45 (1.24-1.70)</td>
<td>Δ Peripheral Vascular severity score ≥ 1</td>
<td>&lt;0.0001</td>
</tr>
</tbody>
</table>

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The results of this study confirm the data on the positive effect of RTX in patients with SSc (decrease of skin induration, increase of FVC, stabilization of DLCO). The decrease of skin score is accompanied by the improvement of lung function indicators. In our study, there was a significant increase of DLCO associated with long-term treatment (over 36 mo) and a cumulative dose of RTX over 3,45±1 gr. Patients with initially lower DLCO can achieve a significant improvement by the improvement of lung function indicators. In our study, there were 135 cases (86.4%) with positive ARS; 29 (25.7%) showed a cytoplasmic pattern and 99 (87.6%) with a nuclear pattern. On the other hand, 99 of the 113 cases (87.6%) with negative ARS presented positive IIF: 29 (25.7%) showed a cytoplasmic pattern (21 of them with an associated nuclear pattern) and 42 cases (37.2%) presented only a nuclear pattern. Correlating the ARS positivity, IF pattern and the diagnosis criteria fulfillment:

- 13 of 15 cases (86.6%) with positive ARS and Solomon’s criteria fulfillment presented a cytoplasmic pattern; and 2 of 15 cases (13.3%) presented only a nuclear pattern.
- 13 of 19 cases (68.4%) with positive ARS and cytoplasmic pattern fulfilled Solomon’s criteria; and 6 only fulfilled those from Connors’.

Conclusion: One-fifth of the immunoblots requested by Rheumatologists presented positive ARS; almost all these cases fulfilled Connors’s criteria, and more than a half fulfilled the stricter Solomon’s criteria. All patients with positive ARS, and a high rate of those without ARS, presented positive IIF. The presence of a cytoplasmic pattern was considerably higher in patients with ARS positivity and in those that met Solomon’s criteria. Thus, our results suggests that in patients evaluated by a Rheumatologist, with clinical suspicion of ASSD or myositis and with ARS positivity, the probability of fulfilling Solomons’s criteria is higher when the IIF presents a cytoplasmic pattern than when only a nuclear pattern is observed. Nevertheless, presenting only a nuclear pattern does not exclude the detection of ARS in the myositis immunoblot and the fulfillment of Solomon’s criteria.

REFERENCES


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**SAT0275**

**PERFORMANCE OF THE ANTISYNTHETASE ANTIBODIES AND THEIR INDIRECT IMMUNOFLOUORESCENCE PATTERNS IN THE ANTISYNTHETASE SYNDROME DIAGNOSIS**

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**Background:** The antisynthetase syndromes (ASSD) are characterized by the presence of an anti-aminoacyl transfer RNA synthetase (ARS) autoantibodies; which difficult the binding of amino acids to the transfer RNA during the protein synthesis. ARS can be detected by indirect immunofluorescence (IIF), and can be identified by immunoblot assay and ELISA (Enzyme-Linked ImmunoSorbent Assay) and immunoblotting. The main clinical features of the ASSD are myositis, arthritis, interstitial lung disease, Raynaud’s phenomenon, mechanic hands, and fever. Two ASSD diagnosis criteria have been developed; those proposed by Connors, and the stricter criteria proposed by Solomon (1, 2).

**Objectives:** To evaluate the performance of the ARS and their IIF patterns in the ASSD diagnosis.

**Methods:** We performed an observational retrospective study in one center during the period 06/2006-06/2018. We searched all the myositis immunoblots (Eurimmum assay) requested by the Rheumatologists under suspicion of ASSD or myositis. We assessed: 1) the rate of cases with positive ARS; 2) the rate of cases with Connors’ or Solomon’s diagnosis criteria fulfillment; and 3) their relation with the IIF patterns (Hep-2 cells; >1/80) evaluated by an expert in autoimmune tests.

**Results:** A total of 140 myositis immunoblots were searched. Twenty-seven cases (19.3%) presented positive ARS: anti-Jo1 (n=13), anti-PL-12 (n=7), anti-PL-7 (n=1), anti-EJ (n=2), and anti-OJ (n=4). Twenty-five of these (17.9%) fulfilled Connors’ criteria, and 15 (10.7%) additionally met Solomon’s criteria. Thus, the fulfillment of Connors’ and Solomon’s criteria in patients with a positive ARS was of 92.6% and 55.5%, respectively.

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**SAT0276**

**STUDY OF THE EPIDEMIOLOGICAL, CLINICAL AND ANALYTICAL CHARACTERISTICS IN PATIENTS WITH SYSTEMIC SCLEROSIS AND CANCER IN VALL D’HEBRON HOSPITAL**

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**Background:** Scleroderma or systemic sclerosis (SSc) is a systemic, chronic, autoimmune disease characterized by great clinical heterogeneity. In recent years, studies have proven that there is a relationship between SSc and neoplasia. SSc is associated with an increased risk of certain types of cancer, particularly liver, hematological, non-melanoma skin and urothelial cancer. Despite this increase, the relative risk of developing cancer is still low in these patients. In the literature, neoplasms have been described in 3-11% of patients with SSc.

**Objectives:** Our objective is to analyze the epidemiological, clinical and analytical characteristics previously described as possibly linked to the development of a cancer in patients with systemic sclerosis (SSc) in the Vall d’Hebron Hospital cohort.

**Methods:** We analyzed 559 patients in the Vall d’Hebron Hospital cohort of SSc. The inclusion criteria were age > 18 years and the diagnosis of SSc limited, diffuse and SSc sine scleroderma. The different variables were analyzed by univariate statistical analysis with SPSS v21.

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