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SAT0273

PREDICTIVE VALUE OF THE REVISED EUROPEAN SCLERODERMA TRIALS AND RESEARCH GROUP ACTIVITY INDEX (EUSTAR-AI)

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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 items 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 (male sex, clinical and serological subset).

To capture the disease activity variations over time, we calculated the EUSTAR-AI and EScSG-AI adjusted mean (area under the curve over time divided by time interval). Disease progression was based on the Medsger severity scale and included accrual of a $\Delta \geq 1$ summed severity score and of a $\Delta \geq 1$ severity in each organ systems at the 2 year follow-up visit compared with the initial visit. To explore specific determinants of disease progression, logistic regression analysis was carried out.

Results: A total of 549 patients satisfied the entry criteria. At univariate logistic regression analysis (among sex, age, clinical and serological subset, EScSG-AI and EUSTAR-AI adjusted mean and baseline severity score), EScSG-AI adjusted mean (OR 1.41 95%CI 1.20-1.67), antiScl70 antibody positivity (OR 1.72 95%CI 1.20-2.47), diffuse subset (OR:1.46 95%CI 1.01-2.10) and EUSTAR-AI adjusted mean (OR 1.41 95%CI 1.23-1.61) predicted disease severity accrual. Multivariate analysis revealed that the EUSTAR-AI adjusted mean was the best predictor of disease progression (Table1). Moreover, at multivariate analysis the EUSTAR-AI adjusted mean also predicted severity accrual of lung (OR 1.32), heart (OR 1.40), skin (OR 1.48) and peripheral vascular disease (OR 1.45).

Conclusion: The adjusted mean EUSTAR-AI has a distinct predictive value for disease progression and development of severe organ involvement in SSc and works better than EScSG-AI.

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

Predictive factors	OR (IC 95%)	severity deterioration	p- value
Adjusted mean EUSTAR-AI	1.41 (1.23-1.61)	Δ summed severity score ≥ 1	<0,0001
Adjusted mean EUSTAR-AI	1.32 (1.14-1.54)	Δ lung severity score ≥ 1	0,0002
Adjusted mean EUSTAR - AI age	1.40 (1.15-1.70)	Δ heart severity score ≥ 1	0,0005 0,0223
	1.03 (1.01-1.05)		
Adjusted mean EUSTAR-AI age	1.48 (1.21-1.82)	Δ Skin severity score ≥ 1	0,0002 0,0359
	0.97 (0.95-0.99)		
Adjusted mean EUSTAR-AI	1.45 (1.24-1.70)	Δ Peripheral Vascular severity score ≥ 1	<0,0001

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SAT0274

THE EFFECT OF RITUXIMAB ON LUNG FUNCTION AND SKIN SCORE IN SYSTEMIC SCLEROSIS-ASSOCIATED INTERSTITIAL LUNG DISEASE. LONG-TERM OBSERVATION

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Background: There is a large clinical experience about the efficiency of rituximab (RTX) for the treatment of systemic sclerosis (SSc). There are several studies showing the decrease in skin induration and interstitial fibrosis in the lungs as the effect of therapy. However, there are not many long-term observations.

Objectives: To describe the efficacy of RTX on lung function and skin score in patients with systemic sclerosis-associated interstitial lung disease, in long-term follow-up.

Methods: This study included 71 patients (pts) with SSc. Data were collected prospectively. The mean follow-up period was 42 mo (12-72). Mean age was 46 years (17-66), female-59 (83%), diffuse cutaneous subset of the disease had 42 (59%), Scl-70 positivity-73% of pts. Duration of the disease was 5,6±4,4 yrs. All pts received concomitant treatment with low dose prednisolone and 45% - with immunosuppressants. The following indicators were evaluated: forced vital capacity,% predicted (FVC), diffusing capacity for carbon monoxide,% predicted (DLCO) and Rodnan skin score (mRss) over a periods of 12-18 months (point 1), 24-30 months (point 2), 36-42 months (point 3), 48-54 months (point 4) and 60-72 months (point 5) after the start of therapy. The results are presented in the form of mean values, delta, median, upper and lower quartile.

Results: In point 1 (n=71) the cumulative mean dose of RTX was 1,43 ±0,6 gr. The mRss decreased from 11,3±9,6 to 8±6,6 (p = 0,000007). FVC increased from 77,4±19,9 to 82,6±20,7% (p=0.0001). DLCO remained stable (from 47,0±18,5 to 47,23±16,7%). In point 2 (n=55) the cumulative mean dose of RTX was 2,97±0,8 gr. ΔmRss - 5,4 (median 3; 25th% 0; 75th% 10). ΔFVC - 7,5% (median 8,2; 25th% -1,1; 75th% 14,4). ΔDLCO - 0,21% (median -0,2; 25th% -6,2; 75th% 6,6). In point 3 (n=36) the cumulative mean dose of RTX was 3,45±1,3 gr. ΔmRss - 5,1 (median 3,5; 25th% 0; 75th% 9). ΔFVC - 9,3% (median 7,7; 25th% 1,45; 75th% 15,3). ΔDLCO - 3,4% (median 3,6; 25th% 2,45; 75th% 7,76), p=0,02. In point 4 (n=24) the cumulative mean dose of RTX was 3,96±1,1 gr. ΔmRss - 5,3 (median 3; 25th% 0; 75th% 10). ΔFVC - 12,2% (median 7,9; 25th% 1,1; 75th% 24,2). ΔDLCO - 3,9% (median 0,45; 25th% -0,95; 75th% 6,8). In point 5 (n=17) the cumulative mean dose of RTX was 5,15±1,7 gr. It should be noted that this group included patients with initially the lowest DLCO (below 40%). ΔmRss - 7,3 (median 5; 25th% 1; 75th% 14). ΔFVC - 13,2% (median 11; 25th% 8,7; 75th% 23,4). ΔDLCO - 5,9% (median 1,6; 25th% -4,8; 75th% 13,8).

Conclusion: 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±1,3 gr. Patients with initially lower DLCO can achieve a significant improvement by the 60th month of RTX therapy. Our work shows that patients with SSC-associated interstitial lung disease are required long-term treatment with RTX (at least 3-5 years) to achieve an obvious improvement of the lung function.

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SAT0275

PERFORMANCE OF THE ANTISYNTHEASE ANTIBODIES AND THEIR INDIRECT IMMUNOFLUORESCENCE PATTERNS IN THE ANTISYNTHEASE SYNDROME DIAGNOSIS

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Background: The antisynthetase syndromes (ASSD) are characterized by the presence of 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 06/2008-06/2018. We searched all the myositis immunoblots (Euroimmun 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 Connor's 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 Connor's and Solomon's

criteria in patients with a positive ARS was of 92.6% and 55.5%, respectively.

All cases (100%) with positive ARS presented positive immunofluorescence: 19 (70.4%) showed a cytoplasmic pattern (10 of them with an associated nuclear pattern) and 8 cases (29.6%) presented only 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, IIF 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 Connor'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 Solomon'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.

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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 lung, 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 583 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.