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 ANTISYNTHETASE ANTIBODIES AND THEIR INDIRECT IMMUNOFLUORESCENCE PATTERNS IN THE ANTISYNTHETASE 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 increasement, 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.