THU0420 AN EXTENT OF INTERSTITIAL LUNG DISEASE IS A POTENTIAL PREDICTOR OF RESPONSE TO A-B-CELL THERAPY IN THE PATIENTS WITH SSC

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Background: Systemic sclerosis (SSc) is a connective tissue disease associated with chronic polyclonal B-lymphocytic activation and immunological tolerance disturbance. Several research and clinical studies showed that B-cell depletion is potentially efficacious in SSc treatment. However, neither strong evidence of RTX efficacy for treatment of interstitial lung disease (ILD) associated with SSc, no potential predictor of response to a-B-cell therapy.

Objectives: To evaluate rituximab (RTX) therapy efficacy in the patients with systemic scleroderma (SSc) differing in extent of interstitial lung disease (ILD) based on multispiral computed tomography (MSCT) findings.

Methods: 42 patients (average age 48±2 years; male/female 1:6, diffuse/limited disease 1.5:1 (25 and 17), disease duration since the first non-Raynaud syndrome (~6.6±5.9 years) with definitely diagnosed SSc and ILD signs evidenced by MSCT were enrolled into the study. During the observation period 29±15.3 months the patients received rituximab (RTX) total dose of 2±1.3 grams in combination with glucocorticoids at average dose of 11.7±3.9 mg. 10 (24%) patients concurrently took immunosuppressants. The therapy efficacy was evaluated both in the general study population and in the patient subgroups with interstitial lesion extent up to 20% (Group A, n=13) and greater than 20% (Group B, n=29) of total pulmonary tissue area.1

Results: In the general population significant FVC increase from 73.2±18.8% to 82±21.8% (p=0.0003) and stabilisation of DLCO (42.6%±15.7% vs 44.7±14.6%, p=0.2) were observed. Median FVC increment was 6% (25%-75%=3.3%; 75%>15%). FVC-based parameters increased by >10% in 16 (38%) patients and decreased in 7 (17%) patients.

Average FVC values in Group A were significantly higher compared with Group B both at the baseline (88.8%±18.6% vs 65±4.15%, p=0.002) and after the treatment (103.3%±15.9% vs 74±18.5%, p<0.0009) with statistically significant FVC increase in both groups during the treatment period (p=0.016 and p=0.0014, respectively). Median FVC increment in Group A and Group B was 10.2% (25%-75%=4.7%; 75%>21.9%) and 5.9% (25%-75%=2.75%; 75%>14.7%), p=0.05, respectively. FVC-based parameters increased by >10% in 6 (46%) patients in Group A, and in 10 (34%) patients in Group B, and decreased in 1 (8%) and 2 (7%) patients, respectively. Average DLCO values were also significantly higher in Group A compared to Group B both before and after treatment (58.4%±16.4% vs 36±3.10%, p=0.025; 59.3±15.2% vs 38±9.97%, p=0.005); DLCO values did not change over time during RTX therapy.

Conclusions: RTX therapy resulted in significant FVC increase. FVC increment in the patient group with ILD extent up to 20% achieved clinical significance level in contrast to the patients with ILD extent greater than 20%, where FVC increment was 5.9%. Obtained data suggest that initial lung lesion area is a potential predictor of response to a B-cell therapy in the patients with SSc.

REFERENCE:


THU0422 PERFORMANCE OF EULAR/ACR 2017 IDIOPATHIC INFLAMMATORY MYOPATHIES CLASSIFICATION CRITERIA IN A REAL WORLD COHORT

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Background: Idiopathic Inflammatory Myopathies (IIM) are an heterogeneous group of multisystemic diseases. It includes Polymyositis (PM), Dermatomyositis (DM) with its clinically amiotic variant (CADM), the antisynthetase syndrome (ASS), the inclusion body myositis (IBM), Immune-mediated necrotising myopathy (IMNM), the juvenile variants of DM/P and the connective tissue disease-myositis overlap (CTD-OM).

Distinction between subtypes is made on grounds of clinical features, histologic findings at muscle biopsy and presence of certain autoantibodies. Multiple classification criteria had been proposed through time, EULAR/ACR been the most recent. However, their performance in patients from common practice in Latin America had not been widely evaluated.

In our practice, access to muscle biopsy and electromyogram (EMG) is not always available.

Objectives: To evaluate the performance of the EULAR/ACR 2017 IIM classification criteria in a real world cohort and compare it with the performance of other classification criteria.

Methods: Retrospective study. IIM patients defined by expert opinion followed in our centre between October 2007 and November 2017 were included. The patients were classified clinically in DM, CADM, PM, ASS and CTD-OM. Patients with positive antisynthetase antibodies were reclassified as ASS. Availability of EMG, muscle biopsy and anti-Jo-1 antibodies was evaluated. Bohan & Peter (1975) Tanimoto (1995) y EULAR/ACR (2017) criteria were applied to the population.

Results: 60 patients were included, DM 20 (33.3%), CADM 4 (6.6%), PM 4 (6.6%), ASS 10 (16.6%) y CTD-OM 22 (36.6%). Muscle biopsy available 14/ 60 (23.3%), EMG available 33/60 (55%) and anti-Jo-1 determination available in 57/60 (95%).

In general, 12/60 (20%) classified as defined disease by Bohan and Peter criteria, 29/60 (48.3%) by Tanimoto criteria and 34/60 (56.6%) by EULAR/ACR 2017 criteria.