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

O. Koneva, O. Desinova, O. Ovasyannikova, L. Garzanova, M. Starovoytova, L. Ananieva. *Nasonova Research Institute of Rheumatology, Moscow, Russian Federation*

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 sclerosis (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 2.5±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 patient population significant FVC increase from 73.2±18.8% to 82±21.8% (p=0.000031) and stabilisation of DLCO (42.6%±15.7% vs 44.7±14.6%. p=0.2) were observed. Median FVC increment was 6% (25th% >3.3%; 75th%>16%). FVC-based parameters increased by ≥10% in 16 (38%) patients and decreased in 3 (7%) 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±14.5%, p=0.0002) and after the treatment (103.3%±15.9% vs 74.1±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% (25th% >4.7%; 75th%>21.9%) and 5.9% (25th%>2.75%; 75th%>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±10.1%. p=0.025; 59.3±15.2% vs 38.9±9.7%. 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.

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THU0421 PREVALENCE AND CLINICAL CORRELATES OF SMALL AIRWAY OBSTRUCTION IN PATIENTS WITH SYSTEMIC SCLEROSIS

P. Ostojic^{1,2}, M. Vujovic², A. Blanka Protic³. ¹*Institute of Rheumatology*; ²*School of Medicine, University of Belgrade*; ³*Clinical Center of Serbia, Clinic for Pulmology, Belgrade, Serbia*

Background: Small airways are usually defined as non-cartilaginous airways with an internal diameter <2 mm. Small airway obstruction (SAO) may be result of a primary bronchiolar disorder, or secondary to a disease that also affects large airways (like asthma or chronic obstructive pulmonary disease - COPD), or related to an interstitial lung disease with bronchiolar involvement.

Objectives: To assess prevalence and clinical correlates of SAO in patients with systemic sclerosis (SSc).

Methods: 69 consecutive patients with SSc (63 women and 6 men) were included in this study. Patients with previously diagnosed bronchiectasis, COPD or asthma were excluded. Forty two (60.9%) patients had limited cutaneous SSc, whilst 27 (39.1%) of patients had diffuse form of the disease. Seventeen patients (24.6%) were tobacco-smokers, 52 (75.4%) were nonsmokers. Lung function tests, including assessment of lung diffusing capacity, were performed in all patients. Patients were considered to have small airway obstruction (SAO) when Maximum Expiratory Flow at 25% of the forced vital capacity (MEF₂₅) was lower than 60% as predicted. We assessed the relationship of SAO in our patients with large airway obstruction, decreased lung diffusing capacity, disease duration, disease subtype, scleroderma-specific antibodies and smoking.

Results: SAO was noticed in 46/69 (66.6%) of patients with SSc. Restrictive lung disease was found in 4/69 (5.8%), obstruction of large airways in 18/69 (26.1%) and decreased lung diffusing capacity in 47/69 (68.1%) of patients. No difference in gender, age, disease duration, disease form and scleroderma-specific antibodies was found between patients with and without SAO. 18/46 (39.1%) patients with SAO had decreased FEV₁ and FEV₁/FVC, indicating presence of coexistent large airway obstruction. Indeed, all patients with signs of obstructive lung disease on spirometry, had associated SAO. Moreover, MEF₂₅ correlated significantly with FEV₁ (ρ=0.54, p<0.001), FEV₁/FVC (ρ=0.74, p<0.001), PEF (ρ=0.29, p=0.02) and MEF₅₀ (ρ=0.80, p<0.001) in our patients with SSc. However, 28/46 (60.9%) SSc patients with SAO did not have signs of concomitant large airway obstruction. 20/46 (43.5%) patients had associated decreased lung diffusing capacity, whilst 8/46 (17.4%) of patients had isolated SAO. We have found that tobacco-smokers among SSc patients with SAO have more common associated obstructive lung disease on spirometry (58.8% vs 15.4%, p=0.004), and have a significantly lower mean value of MEF₂₅ compared to non-smokers (32.1%±7.1% vs. 45.4±11.6%, p=0.001). On the other hand, isolated SAO, and SAO associated with impaired diffusing capacity was equally frequent among smokers and non-smokers.

Conclusions: Patients with SSc have commonly SAO. It can be considered as clinical feature of undiagnosed asthma or COPD, if associated with large airway obstruction, especially in tobacco-smokers. On the other hand, isolated SAO or associated with decreased lung diffusing capacity was found to be not related to smoking, and may indicate a possible prominent bronchiolar involvement within SSc related interstitial lung disease.

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

R.A. Gómez, A. Brailard Pocard, M. Pino, G. Medina, D. Dubinsky. *HTAL DE CLÍNICAS, Ciudad Autonoma de Buenos Aires, Argentina*

Background: Idiopathic Inflammatory Myopathies (IIM) are a heterogeneous group of multisystemic diseases. It includes Polymyositis (PM), Dermatomyositis (DM) with its clinically amyopathic variant (CADM), the antisynthetase syndrome (ASS), the inclusion body myositis (IBM), Immune-mediated necrotising myopathy (IMNM), the juvenile variants of DM/PM 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 (6.66%), PM 4 (6.66%), ASS 10 (16.66%) y CTD-OM 22 (36.66%). Muscle biopsy available 14/60 (23.33%), 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.

Extending the classification to probable besides defined, 38/60 (63.3%) met Bohan and Peter criteria and 51/60 (85%) met EULAR/ACR criteria.

Table 1 shows percentage of patients meeting each criteria set by clinical subtype.

Patients with available muscle biopsy (n=14) were subanalyzed: 11/14 (78.6%) muscle biopsy were compatible with IIM (3 not compatible, but previously treated). DM 3/11 (28.6%), ASS 1/11 (7.14%) y CTD-OM 7/11 (50%). 7/11 (50%) met Bohan and Peter criteria, 8/11 (78.6%) met Tanimoto criteria and 8/11 met EULAR/ACR criteria. If extended to probable cases, 11/11 (100%) met Bohan and Peter criteria and 10/11 (91%) EULAR/ACR criteria.

Table 1 Fulfillment of different criteria set by IIM subtype

	Bohan & Peter (%)			Tanimoto (%)	EULAR/ACR (%)		
	Probable	Definite	Total		Probable	Definite	Total
DM (n=20)	60	25	85	35	10	85	95
CADM (n=4)	0	0	0	0	25	75	100
PM (n=4)	75	0	75	50	50	0	50
ASS (n=10)	50	10	60	80	30	70	100
CTD-OM (n=22)	27,3	27,3	54,6	54,50%	41	31,8	72,8
DM/CTD-OM (n=4)	25	50	75	75	0	100	100
PM/CTD-OM (n=18)	27,8	22,2	50	55,5	50	16,6	66,6

Conclusions: EULAR/ACR²⁰¹⁷ performed especially well for DM, and were the only criteria set met by CADM patients.

PM patients failed to meet Bohan and Peter and EULAR/ACR criteria for definite disease, probably due to low availability of EMG and muscle biopsy. This was similar for pure PM and PM/CTD-OM.

Tanimoto criteria were the most sensitive for classification of ASS patients, which could be explained because they consider arthritis among clinical features.

When considering probable and defined cases, EULAR/ACR criteria were highly sensitive in this real world cohort

Disclosure of Interest: None declared

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THU0423 EPIDEMIOLOGY AND SURVIVAL OF SYSTEMIC SCLEROSIS-SYSTEMIC LUPUS ERYTHEMATOSUS OVERLAP SYNDROME

S. Alharbi^{1,2,3,*}, Z. Ahmad², A. Bookman¹, Z. Touma^{1,4}, J. Sanchez-Guerrero^{1,2}, N. Mitsakakis^{4,5}, S.R. Johnson^{1,2,4}. ¹Toronto Scleroderma Program, Division of Rheumatology, Toronto Western Hospital, University Health Network; ²Division of Rheumatology, Mount Sinai Hospital, University of Toronto, Toronto, Canada; ³Taibah University, Medina, Saudi Arabia; ⁴Institute of Health Policy, Management and Evaluation; ⁵Toronto Health Economics and Technology Assessment Collaborative, University of Toronto, Toronto, Canada

Background: Systemic sclerosis (SSc) is an immune disorder characterised by vasculopathy and fibrosis that may overlap with another disease such as systemic lupus erythematosus (SLE). Little is known about the epidemiology, clinical characteristics, and survival of SSc-SLE overlap (also called lupoderma). We evaluated the prevalence of SSc-SLE overlap syndrome, differences in SSc clinical characteristics and survival compared with SSc without SLE.

Methods: A cohort study was conducted including subjects who fulfilled the ACR-EULAR classification criteria for SSc and/or the ACR criteria for SLE. The primary outcome was the time from diagnosis to death from all causes. Survival was evaluated using Kaplan Meier curves and Cox Proportional Hazard models.

Results: We identified 1252 subjects (SSc n=1166, SSc-SLE n=86) with a SSc-SLE prevalence of 6.8%. SSc-SLE were younger at diagnosis (37.9 years versus 47.9 years, p<0.001), more frequently had lupus anticoagulant (6% versus 0.3%, p<0.001), anticardiolipin antibody (6% versus 0.9%, p<0.001), and pulmonary arterial hypertension (PAH) (52% versus 31%, p<0.001). SSc-SLE less frequently had calcinosis (13% versus 27%, p=0.007), telangiectasia (49% versus 75%, p<0.001) and diffuse subtype (12% versus 35%, p<0.001). There were no significant differences in the occurrence of renal crisis (7% versus 7%), interstitial lung disease (40% versus 34%), and digital ulcers (38% versus 32%). SSc-SLE had better survival (median 26.1 versus 22.4 years), but this was not statistically significant (log rank p=0.06). Female sex and diffuse subtype attenuated survival differences between groups (Hazard Ratio 0.70, 95% CI 0.45, 1.11).

Conclusions: SSc-SLE are younger at diagnosis, more frequently have PAH, and less frequently have cutaneous manifestations of SSc. SSc-SLE patients should be monitored for pulmonary hypertension, interstitial lung disease, renal crisis and digital ulcers.

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THU0424 ARTICULAR INVOLVEMENT IN SYSTEMIC SCLEROSIS: COMPARISON OF CLINICAL, RADIOGRAPHIC AND SONOGRAPHIC FINDINGS

S. Thietart¹, K. Louati², E. Gaigneux², M. Gattfosse¹, S. Riviere¹, J. Sellam², F. Berenbaum², O. Fain¹, A. Mekinian¹. ¹Internal medicine; ²Rheumatology, Hôpital Saint Antoine, AP-HP, Université Paris 6, 75012 Paris, France., Paris, France

Background: Joint involvement in Systemic Sclerosis (SSc) is frequent and varied.^{1,2}

Objectives: We study US synovitis and its correlation with clinical synovitis, radiological erosions and organ involvement.

Methods: In a prospective cohort of SSc patients, tender and swollen joint counts, DAS28-CRP, hand US sonographies, X-ray hand views, as well as respiratory, cardiac, cutaneous and renal characteristics were assessed.

Results: 54 patients were included with a median age of 59 years,²⁷⁻⁸¹ 45 women (83%), with a diffuse cutaneous subtype in 13 patients (24%). 23 patients (52%) presented with arthralgia, 9 had clinical synovitis (16%) and DAS28-CRP of 3.7 (2.98-5.90). US sonography (34 patients) found at least one synovitis in 23 patients: 14 patients with grade 1 (66%), 6 patients grade 2 (29%), 1 patient grade 3 (5%), with a positive power Doppler signal in one case (3%). Among the patients having US-synovitis, 4 had clinical synovitis (17%), and 4 had X-ray erosions (17%). Radiological erosions were present in 8 patients (15%), without any correlation with clinical or US synovitis. Articular involvement (defined as clinical synovitis, US-synovitis and/or articular erosions) were found more frequently in limited SSc (n=28, 72%) than in diffuse SSc (n=4, 31%) (p<0.001), with a more frequent positivity of anti-centromere antibodies (n=23, 60% versus n=3, 20%). No correlation was found with disease severity or other organ impairment.

Conclusions: US synovitis were found more frequently than clinical synovitis, which are merely active, and did not correlate with articular destruction.

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THU0425 18F-FLUORODEOXYGLUCOSE POSITRON EMISSION TOMOGRAPHY/COMPUTED TOMOGRAPHY AND LUNG INVOLVEMENT IN SYSTEMIC SCLEROSIS

S. Bellando-Randone¹, L. Tartarelli², E. Cavigli³, L. Tofani⁴, C. Bruni¹, G. Lepri¹, J. Blagojevic¹, A. Moggi Pignone⁵, C. Miha⁶, J. Avouac⁷, A. Passeri², M.T. De Cristofaro², O. Distler⁶, Y. Allanore⁷, S. Guiducci¹, M. Matucci Cerinic¹. ¹Dept of Experimental and Clinical Medicine, University of Florence, Italy and Dept of Geriatric Medicine, Div Rheumatology AOUC; ²Department of Biomedical, Experimental and Clinical Sciences, "Mario Serio", Nuclear Medicine Unit, University of Florence; ³Department of Radiodiagnostic and Emergency, Careggi University Hospital, Florence; ⁴Department of Neurosciences, Psychology, Drug Research and Child Health, University of Florence, Florence; ⁵Internal Medicine of Careggi University Hospital, Florence, Italy; ⁶Department of Rheumatology, University Hospital Zurich, Zurich, Switzerland; ⁷Department of Rheumatology A, Cochin Hospital, INSERM U1016, Paris Descartes University, Paris, France

Background: At early stages, SSc lung involvement is characterised by Ground Glass Opacities (GGO) at High Resolution Computed Tomography (HRCT). However, HRCT scan is not able to provide functional information and to discriminate between an "active inflammatory" and an "established fibrotic" GGO. 18Fluoro-Deoxy-d-Glucose (18F-FDG) Positron Emission Tomography/Computed Tomography (PET/CT) is able to detect metabolic activity picking up inflammation and provides both morphologic and metabolic data.

Objectives: The aim of this study was to evaluate, if 18F-FDG PET/CT scan may identify the inflammatory component of GGO in SSc interstitial lung disease.

Methods: Seven patients with SSc (1 male and 6 females; mean age 59.56 y ±9.15 SD; median disease duration 5 years,²⁻¹¹ who underwent 18F-FDG PET/CT scan because of cancer screening, were retrospectively analysed. HRCT