diseases. PM and MG present both with muscle weakness and other similar features; electrophysiological and laboratory features of each of them are distinct. In Literature there are only few reports about PM and MG association

Objectives: To evaluate the prevalence of MG in patients with PM in our case series

Methods: We enrolled patients with PM/DM visited in our Centre in past 10 years; diagnoses of PM/DM were based on Bohan and Peter criteria. The follow-up was conducted until December 2017

Results: We made 17 PM/DM diagnosis:14 F and only 3 M aged 41–85 years (mean 65 y). We found 6 patients (42.8%) with association of PM and MG. They are all F, aged 51–78 years (mean 61 y) who developed inflammatory myositis confirmed by increasing of CK, EMG, autoantibodies pattern positivity and deltoid biopsy. Paraneoplastic, post-infectious or post-vaccinal syndromes was excluded. At the onset all presented progressive proximal muscle weakness and pain and asthenia. After initial treatment with oral corticosteroids (0.5–1 mg/kg/day methylprednisolone) a minimal response was observed only as improvement of asthenia and decreasing of CK levels. After one month of therapy 2 patients developed a bilateral palpebral ptosis, one dysphagia and mild dyspnea, one a severe intestinal pseudo-obstruction, 2 a mild dysarthria, ipovision and a worsening of muscular tone. Pyridostigmine test was positive in all patients; antiAChR antibodies levels were high. We started high doses corticosteroids (methylprednisolone 500–1000 mg/day for 5 days) and pyridostigmine (180–240 mg/day) with smart improvement. After this we introduced an immunosuppressant: azathioprine in 3 patients, mycophenolate mofetil in 4 patients. At the same time, because of the severity of the disease, monthly cycles of high doses IgV (20 g/kg in 5 days) were performed in 5 patients. A stable remission was achieved and maintained in all patients

Conclusions: This is one of the largest case series of patients with PM/DM-MG overlap. Our findings suggest that this association is not so rare and that patients affected by PM presenting palpebral ptosis, diplopia, gastrointestinal or oral symptoms, bulbar symptoms, weakness, asthenia, should be evaluated to exclude a concomitant MG, despite the absence of thymic pathology. A patient with PM-MG overlap should allow us for proper management of both conditions. This may include a more adequate therapy providing simultaneous association between immunosuppressant, pyridostigmine and, if necessary, a short time of high doses IgV therapy

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THU0412

ABSOLUTE REDUCTION OF PERIPHERAL CD4 + REGULATORY T CELL SUBSET OF PATIENTS WITH SYSTEMIC SCLEROSIS AND ITS RESTORATION BY SHORT-TERM AND LOW DOSE IL-2 TREATMENT

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Background: Systemic sclerosis (SSc) is a chronic inflammatory disease with complex pathogenesis. The role of regulatory T cells (Tregs) in the development of SSc has started being studied during the last decade with new aspects being disclosed continuously. Although there is a general agreement in the medical literature regarding the decreased functional capacity of circulating Tregs in SSc, the alteration of absolute number of Treg cells as well as other CD4 + T lymphocyte subsets in SSc is still unclear.

Objectives: The aim of the present study was to explore the absolute number status of CD4 + T subsets in peripheral blood of patients with SSc using our modified flow cytometric method and investigate whether the imbalance of Th17 and Treg cells can be corrected by supplementing low dose interleukin -2 (IL-2).

Methods: The peripheral CD4 + T subsets from 54 patients with SSc and 30 healthy control subjects were analysed. The patients were divided into the untreated group (n=29) and treated group (n=25). The patients were also divided into group A (n=28) who only used low-dose glucocorticoids (GCS), and group B (n=26) who also used immunosuppressant cyclophosphamide (CYC), methotrexate (MTX), leflunomide (LEF) or mycophenolate mofetil (MMF). In addition, 21 patients from 54 patients, given a small dose of IL-2 (50WU) treatment for a 5 day

course, divided into pre-treatment group and post-treatment group. Directly using the results from flow cytometry combined with internal standard beads, absolute number of peripheral CD4 + T subsets from the subjects in each group were calculated.

Results: Although there were some changes among CD4 + T cell subsets in peripheral blood from these SSc patients, the major alteration was the reductions of Treg cell absolute number. Compared with the normal controls, the number of CD4 + CD25 + FOXP3 + Treg cells were significantly decreased in all patients (p=0.001), in untreated group (p=0.029), in treated group (p=0.029), in group A (p=0.006) and in group B (p=0.004). The ratio of Th17/Treg in total patients (p=0.008) and group B (p=0.001) were significantly higher than that in normal control group, and the group B was significantly higher than the group A (p=0.032). Moreover, the number of Th17 cells in group B was significantly higher than that in group A (p=0.023). After IL-2 treatment, the absolute number of CD4 + CD25 + FOXP3 + Treg cells (p<0.001) were significantly increased, while the number of Th1, Th2 and Th17 cells were slightly higher than those before treatment with IL-2 (p>0.05). Since Th17 cells increases significantly lower than Treg cells, so their ratio decreases significantly (p=0.001) to get re-balance.

Abstract THU0412 – Figure 1. Comparison of the levels of CD4+ T lymphocyte subgroups among different groups. (A and D) The percentage of Th2 cells were significantly increased in untreated group and Group A compared with healthy controls. (B and E) The number of CD4+CD25+FOXP3+ Treg cells decreased in patients with SSc, whether or not they use immunosuppressive agents. (C and F) The ratio of Th17/Treg cells was found to be significantly increased in Group A, as well as between Group B and Group A. *P<0.05; **P<0.01; ***P<0.001.

Abstract THU0412 – Figure 2. Comparison of the levels of CD4+ T lymphocyte subgroups among different groups as well as before and after IL-2 treatments. (A) The percentage of Th2 cell were significantly increased before and after IL-2 treatment compared with healthy controls, and the percentage of Treg cells significantly increased after IL-2 treatment. (B) The number of CD4+CD25+FOXP3+ Treg cells decreased in patients before IL-2 treatment, but increased significantly after IL-2 treatment (p<0.05); while Th1, Th2 and Th17 cells after IL-2 treatment were slightly higher than those before treatment (p>0.05). (C) The ratio of Th17/Treg cells were found to be significantly increased after IL-2 treatment. *P<0.05, **P<0.01, ***P<0.001.

Conclusions: The absolute number of peripheral CD4 + CD25 + FOXP3 + Treg cells decreased in untreated patients, indicating that this reduction arising from the disease itself. Our findings suggest that SSc progression is associated with the absolute decrease of peripheral Treg cells. Although there was no statistically

significant different, absolute number of the Treg cells trended to decrease in patients treated with immunosuppressive agents. The use of IL-2 can effectively up-regulate Treg cells as well as increase Th17 to a certain extent, which restores the balance of Th17/Treg cells, but the long-term effect needs further investigation.

Disclosure of Interest: None declared

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THU0413 NORMALISATION OF NAILFOLD MICROVASCULATURE AFTER HEMATOPOIETIC STEM CELL TRANSPLANTATION IN SEVERE SYSTEMIC SCLEROSIS

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Background: Microangiopathy in Systemic Sclerosis (SSc), as visualised by nailfold capillary microscopy (NCM), is a dynamic and sequential process. Uncontrolled case-reports and a case-series report possible improvement of nailfold microangiopathy after hematopoietic stem cell transplantation (HSCT).

Objectives: To evaluate differences in nailfold microvasculature in patients with severe SSc treated with HSCT and those treated otherwise.

Methods: Severe SSc was defined as fulfilling criteria of the ASTIS trial.¹ For included patients two treatment groups were defined: 1. Patients treated with HSCT, 2. Patients treated otherwise (including cyclophosphamide, methotrexate, mycophenolate mofetil and rituximab). All available capillaroscopy images collected prior to and after treatment were scored independently by two trained observers, blinded for treatment. Scoring included categorical scores (0=no changes to 3≥66% alterations per millimetre) for capillary loss, neoangiogenesis, haemorrhages, dilatations, giants, disorganisation, together with a qualitative pattern and a VAS score for overall severity of microangiopathy.

Results: In total 55 patients were included, of whom 33 were female, mean age was 50±13 years and median mRSS of 20.¹⁵⁻²⁸ Twenty-nine patients were treated with HSCT. After treatment categorical score of capillary loss was lower with HSCT (median score 1.0 vs. 2.0, p=0.04) and VAS scores for severity of microangiopathy were better with HSCT (50.0 vs. 67.3, p=0.03). Compared to controls, a typical SSc pattern was less frequent in patients treated with HSCT (n=12/22, 55%, vs. n=18/20, 90% p=0.01).

In 25 patients (HSCT n=11, control n=14) images prior to treatment were available. Improvement in capillary loss (HSCT -0.5 [IQR -1 to 0] vs. control +0.3 [IQR -1.0 to 1.0] p=0.01;), dilatation (HSCT -1.5 [IQR -2.0 to -1.0] vs. control 0.0 [IQR -1.0 to 0.5] p<0.01) and VAS scores (HSCT median -32.5 [IQR -80.5 to -4.5] vs. - controls +2.0 [IQR -35.5 to +46.0] p<0.01; improved significantly more in the HSCT group.

Conclusions: In patients with severe SSc, microangiopathy as reflected by nailfold capillaroscopy images, is less severe after treatment with HSCT and shows more improvement over time compared to patients with severe SSc treated otherwise. In specific, improvement seems to be characterised by normalisation of the capillary diameter and capillary density. To our knowledge, this is the first study to report on changes in microangiopathy after HSCT in a controlled study design.

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THU0414 ASSOCIATION OF THE ESOPHAGEAL DILATION AND INTERSTITIAL LUNG DISEASE ON CHEST HIGH-RESOLUTION COMPUTED TOMOGRAPHY IN PATIENTS WITH IN SYSTEMIC SCLEROSIS: A CROSS-SECTIONAL STUDY

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Background: Esophageal dilation and dysfunction has been implicated in the pathogenesis of interstitial lung disease (ILD) in systemic sclerosis (SSc).

Objectives: The aims of this were to explore the association of the esophageal dilation and ILD on chest high-resolution computed tomography (HRCT) in patients with SSc and to establish a cut-off point for the esophageal dilation suggestive for the presence of a significant SSc-ILD.

Methods: The widest esophageal diameter (WED) was obtained on axial HRCT images. The parenchymal abnormalities on HRCT were coded and scored according to Warrick method. Patient-centred measures, pulmonary function tests (PFTs) and the single breath carbon monoxide diffusing capacity of the lung (DLco) were also obtained. Multivariate regression analyses were performed to identify factors associated with esophageal diameter on HRCT.

Results: 126 subjects with SSc were included in the analysis. The mean (±SD) WED was 13.5 (±4.2) mm, and in 76 (60.3%) participants WED was ≥11 mm. ILD was diagnosed in 86 SSc patients (Warrick score ≥7), while in 40 subjects the lung findings were normal. SSc patients with ILD had larger mean esophageal diameter than those without lung disease (19.4 mm vs. 14.1 mm, p<0.001). We observed a high correlation between WED and Gastroesophageal reflux disease questionnaire (GerdQ) (r=0.886, p<0.001), Borg score (r=0.705, p<0.001), Warrick score (r=0.614, p<0.001). WED negatively correlated with DLco (r=-0.508, p<0.001). Multivariate analysis demonstrated positive associations between mean esophageal diameter and GerdQ (p<0.0001), Borg index (p<0.0005), and total Warrick score (p=0.019).

Conclusions: In patients with SSc, an increased esophageal diameter (>11 mm) on chest HRCT is associated with pulmonary and esophageal symptoms, more severe ILD, and lower DLco.

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THU0415 ANTI-PM/SCl ANTIBODY CLINICAL ASSOCIATIONS IN PATIENTS WITH SYSTEMIC SCLEROSIS. ANALYSIS OF THE MULTICENTER EUSTAR COHORT

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Background: Antibodies to the PM/ScL complex are found in patients with Systemic Sclerosis (SSc), but also with Polymyositis, Dermatomyositis and overlap syndromes. Historically, the main clinical association of anti-PM/ScL antibodies in SSc, which include calcinosis, articular and muscle involvement, and interstitial lung diseases, were described by some large single-centre studies, or by multicenter studies which recruited a relatively small number of positive patients. Therefore, some unresolved issue deserves further research. In particular, scleroderma renal crisis was recently identified in a sizeable number of anti-PM/ScL +SSc patients in a large monocentric British cohort (4 out of 70; 5.7%), a somewhat unexpected finding since this antibody type is generally considered not to be associated with renal crisis.¹

Objectives: To evaluate clinical associations of anti-PM/ScL in patients with SSc in the large multicenter EUSTAR database, with specific focus on scleroderma renal crisis.

Methods: Patients from the EUSTAR database were included when the item anti-PM/ScL was fulfilled in at least one visit; clinical data were collected from the last visit available.

Results: Anti-PM/ScL status was available in 8,287 SSc patients from EUSTAR database: 295 were anti-PM/ScL +. After exclusion of 145 patients with multiple autoantibody positivity, 150 anti-PM/ScL+patients were compared with 7992 anti-PM/ScL -negative patients. Among these, 2530 were positive for anticentromere, 1933 for anti-topoisomerase I, 186 for anti-RNA polymerase III, and 220 for anti-U1Rnp antibodies.

Renal crisis was identified in 8 of 150 anti-PM/ScL+SSc patients (5.3%), and was significantly more frequent than in anti-PM/ScL-negative SSc patients (1.6%; p=0.0015). Positivity for anti-PM/ScL was also associated with male sex, diffuse cutaneous subsets, joint and muscle involvement, lung fibrosis at chest X-rays, heart conduction blocks, stomach and intestinal symptoms (table 1). However, in multivariate analysis, adjusted for age at disease onset, sex, and disease duration, the association of anti-PM/ScL with renal crisis was not significant, whereas