AB0588

INFECTION AGGRAVATED DECREASE OF THE LEVEL OF TH17 AND TREG CELLS AND LOW-DOSE IL-2 REBALANCED TH17/TREG IN THE PERIPHERAL BLOOD OF PATIENTS WITH IDIOPATHIC INFLAMMATORY MYOPATHY

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Background: Idiopathic inflammatory myopathies (IIM) are featured by a series of clinical presentation such as proximal muscle weakness, increased serum levels of creatine kinase and other muscle enzymes and involvement of other organs and systems^[1, 2], which results in high morbidity and early mortality^[3]. We have known the changes of the level of Th17 and Treg cells in IIM in previous studies^[4-6]. However, whether infection affects lymphocyte subsets or not and whether the effect of low-dose interleukin-2 (IL-2) can be influenced by the use of immunosuppressants or not are still unclear.

Objectives: The study aimed to explore the changes of lymphocyte subsets in patients of IIM with or without important organ infection, and the restoration of Th17/Treg after receiving low-dose IL-2.

Methods: A total of 118 IIM patients were enrolled and classified into infection group and non-infection group based on the important organ infection. Of them, 48 cases were treated with low dose IL-2 (5.0×10^5 IU for 5 days). The absolute number of peripheral total T, B, CD4⁺ T, CD8⁺ T, NK, Th1, Th2, Th17 and Treg cell subsets were analyzed by flow cytometry combined with absolute counting beads. Clinical data, laboratory examinations and the levels of peripheral lymphocyte subsets were analyzed retrospectively.

Results: In these patients, especially in the infection group, the absolute number of T, CD4⁺ T, CD8⁺ T, NK, Th1, Th2, Th17 and Treg cells were significantly decreased as compared with that in the healthy controls, which were significantly increased by low dose IL-2 (especially Treg cells) treatment. The levels of ESR, LDH and HBDH and the ratio of Th17/Treg were significantly lower than those before IL-2 treatment ($Z = -2.237, -2.083, -2.140, -3.663, P = 0.025, 0.037, 0.032, 0.000$). The 48 cases who received IL-2 treatment were divided into 2 groups according to whether they used immunosuppressants. There was no significant difference in the absolute number of T, B, CD4⁺ T, CD8⁺ T, Th1, Th2, Th17 and Treg cells, the proportion of Th17 and Treg cells and the ratio of Th17/Treg between the 2 groups ($P > 0.05$).

Conclusion: Global decrease in lymphocyte subsets was found in IIM patients, especially those who had important organ infection. A significant re-balance of Th17/Treg was observed after receiving treatment with low-dose IL-2. Furthermore, the restoration of lymphocyte subsets showed similar degree after treatment with or without immunosuppressants. Low-dose IL-2 may become a potential therapy for IIM patients. The mechanism of lymphocyte decrease in IIM is required further to study.

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Acknowledgments: Thanks for the support of my teachers, classmates and my family.

Disclosure of Interests: None declared

DOI: 10.1136/annrheumdis-2020-eular.5255

AB0589

THE IMPORTANCE OF A SYSTEMIC SCLEROSIS CLINIC IN A TERTIARY REFERRAL CENTER – A PORTUGUESE EXPERIENCE

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Background: Systemic sclerosis (SSc) is a rare systemic rheumatic disease (SRD) characterized by small vessel inflammation and fibrosis of skin and internal organs. Pulmonary and cardiac involvement contribute to both morbidity and mortality associated with the disease. A multidisciplinary approach with strict monitoring is therefore key to attain clinical success.

Objectives: To describe the organization and patient pathways of our SSc outpatient clinic.

Methods: Observational study using data extracted from Reuma.pt/SSc (a subset of the Rheumatic Diseases Portuguese Register). Data extracted included demographic variables and clinical and immunological manifestations. The disease was classified according to the 2013 ACR/EULAR criteria. Our SSc clinic is managed by two dedicated Rheumatologists and up to two Rheumatology residents on a weekly basis, but it is a dynamic multidisciplinary clinic where various medical specialties collaborate closely. There are two associated subspecialty clinics (pulmonary hypertension and pulmonary fibrosis) where the Rheumatologists engage with pneumologists and cardiologists, allowing greater collaboration in the management of these patients. Patients' data is systematically registered in Reuma.pt/SSc as a part of the routine activity of this clinic, contributing to real-world data on SSc.

Results: A total of 220 patients were registered between July 2011 and June 2019. 196 (89.1%) were female, with a mean age of 58.9 ± 14 years and a mean disease duration of 14.6 ± 9 years. Ninety-seven patients (44.1%) had limited cutaneous SSc, 52 (23.6%) had diffuse cutaneous SSc, 35 (15.9%) had overlap SSc, 24 (10.9%) had preclinical SSc and 12 (5.4%) had SSc sine scleroderma. Raynaud phenomenon was present in 92% of the SSc patients and 40% had a history of digital ulcers. Gastrointestinal manifestations included esophageal dysmotility in 39.5% of patients, gastric disease in 24.4% and intestinal involvement in 15.5%. Pulmonary involvement was found in 47.6% of SSc patients, heart disease in 43.6% and kidney involvement in only two patients. Antinuclear antibodies were positive in 92.2% of the patients, anti-centromere in 44.1%, anti-topoisomerase I antibodies in 39.1%, anti-U1RNP in 4.5% and only three patients had anti-PM-Scl and one had anti-RNA polymerase III. 31 patients were lost to follow-up and 32 died. 18 patients are currently being followed up in the pulmonary hypertension clinic and seven in the pulmonary fibrosis clinic.

Conclusion: The implementation of a standardized approach with regular multidisciplinary work has proven very helpful in evaluating patients with SSc. The continuous registry of patients in Reuma.pt/SSc has been essential for patient care, research and healthcare planning.

Disclosure of Interests: None declared

DOI: 10.1136/annrheumdis-2020-eular.2958

AB0590

CLINICAL AND DEMOGRAPHIC CHARACTERISTICS OF PATIENTS WITH ANTISYNTHEASE AUTOANTIBODIES: DATA FROM A PORTUGUESE TERTIARY OUTPATIENT CLINIC

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Background: Antisynthetase syndrome (AS) may have different clinical phenotypes and outcomes associated with different anti-aminoacyl RNA-synthetase (anti-ARS). Patients may also present with incomplete/early phenotypes that do not fulfil the classification criteria.

Objectives: To evaluate the clinical and demographic characteristics of patients positive for anti-ARS in our Myositis clinic.

Methods: Observational study using data from the Portuguese Rheumatic Diseases Register (Reuma.pt/Myositis protocol). Data extracted included demographic variables, clinical features and immunological expression of the disease.

Results: 17 patients were identified. All met the criteria for AS according to Connors criteria, while 3 did not met according to the Solomon criteria. Mean age at diagnosis was 60.1 years (26 - 80) and 76.5% were female. Mean follow-up time was 2.8 (0.5-9) years. Only 3 patients had history of smoking in the past. The autoantibodies expressed were anti-Jo1 (n=12), anti-PL12 (n=2), anti-OJ (n=2) and anti-PL7 (n=1). 4 patients positive for anti-Jo-1 also expressed anti-Ro52. The clinical information and treatment are described in table 1. One patient presented as a paraneoplastic syndrome associated with anti-Jo-1.

Table 1. Demographic and clinic characteristics of our cohort. AZA – azathioprine; CYC – Cyclophosphamide; DM – dermatomyositis; GS – Gottron's sign; ILD – Interstitial lung disease; PM – Polymyositis; NSIP - Nonspecific interstitial pneumonia; MH – mechanic hands; MMF – Mycophenolate mofetil; MTX – Methotrexate; PDN – prednisolone; RP – Raynaud phenomenon; RTX – Rituximab; UIP - Usual interstitial pneumonia; Y – yes; N – no. Connors criteria – anti-ARS plus one or more of RP, MH, arthritis, ILD, fever. Solomon criteria – anti-ARS plus 2 major criteria or 1 major and 2 minor criteria. *induction therapy.

Case	Serology	Major criteria (ILD) [NSIP/UIP/ non-specific] or PM/ DM	Minor criteria (Arthritis, MH, RP)	Other manifestations	Connors (2010)/ Solomon (2011) criteria	PDN	DMARDcs (MTX/MMF/ AZA)	CYC/ RTX
1	Jo1	PM	MH, arthritis	GS	Y/Y	Y	MMF	-
2	Jo1	NSIP	RP, MH	Asthenia	Y/Y	Y	AZA	-
3	Jo1	NSIP	MH, arthritis	Calcinosis, asthenia	Y/Y	Y	AZA	-
4	Jo1	NSIP; DM	-	GS	Y/Y	Y	-	-
5	Jo1/Ro52	PM	MH, arthritis	GS, asthenia	Y/Y	Y	MTX	-
6	Jo1	UIP	RP, arthritis	Asthenia	Y/Y	Y	-	RTX
7	Jo1/Ro52	NSIP; PM	arthritis	-	Y/Y	Y	MMF	CYC*
8	Jo1	NSIP; PM	-	-	Y/Y	Y	MMF	-
9	Jo1/Ro52	NSIP; PM	-	-	Y/Y	Y	AZA	-
10	PL7	-	RP, arthritis, MH	Dysphagia	Y/N	Y	-	-
11	Jo1	-	RP	Asthenia	Y/N	-	-	-
12	Jo1/Ro52	PM	MH, arthritis	GS, asthenia	Y/Y	Y	MMF	-
13	PL12	PM	RP, arthritis	GS, asthenia	Y/Y	Y	MMF	-
14	Jo1	UIP	RP, arthritis	Weight loss, asthenia	Y/Y	Y	-	RTX
15	OJ	Non-specific pattern	-	-	Y/N	Y	MMF	-
16	OJ	NSIP	-	-	Y/Y	Y	-	-
17	PL12	PM	RP, MH	-	Y/Y	-	-	-

Conclusion: The most frequent autoantibody was anti-Jo-1, which is consistent with the literature. Interestingly, patients with anti-PL, usually described as having severe lung disease, in our series do not have it. Additionally, we found a trend for a younger age at diagnosis in Jo1 positive patients and remarkably more than half of these patients have been diagnosed with ILD, being the NSIP pattern the most frequently reported.

Disclosure of Interests: None declared

DOI: 10.1136/annrheumdis-2020-eular.4857

AB0591 ANALYSIS OF A COHORT OF PATIENTS WITH SYSTEMIC SCLEROSIS AND INTERSTITIAL LUNG DISEASE

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Background: Interstitial lung disease (ILD) is a common manifestation of systemic sclerosis that is associated with early mortality. The development or progression of ILD can occur at any time, so patients should be monitored regularly, particularly in the first years after diagnosis.

Treatment should be considered when the disease is clinically significant, particularly when there is evidence of progression based on a decrease in lung function, progression of fibrosis on the HRCT or worsening of respiratory symptoms.

Objectives: -To relate the type of systemic sclerosis (SS) with pulmonary involvement with the radiological pattern.

-To study if there is a relationship between the antibodies and the aforementioned affection.

Methods: Retrospective descriptive study of patients treated in our Hospital (2009-2019) by the Rheumatology and Internal Medicine department diagnosed with systemic sclerosis and interstitial lung disease.

The data were obtained through the review of medical records.

We have included data from patients who have a diagnosis of limited or diffuse systemic sclerosis or overlap with interstitial lung involvement.

Results: Of the 213 patients with systemic sclerosis in our database 43 had interstitial lung involvement (20.2%). 79% of the patients with ILD (34) had a non-specific interstitial pneumonia type (NSIP) radiological pattern and 21% of the patients (9) had a pattern of usual interstitial pneumonia (UIP)

Among the patients with ILD with a NSIP -type radiological pattern, 19 patients were diagnosed with diffuse SS, 9 patients with overlap syndrome and 6 with limited SS. Of the patients with ILD with radiological pattern type UIP, 5 patients were diagnosed with diffuse SS, 3 patients with overlap syndrome and 1 patient was diagnosed with limited SS.

TABLE 1. RELATIONSHIP BETWEEN SS TYPE AND RADIOLOGICAL PATTERN

	NSIP	UIP
LIMITED SS	6 (17.6%)	1 (11.1%)
DIFFUSE SS	19 (55.8%)	5 (55.5%)
OVERLAP	9 (26.4%)	3 (33.3%)

Among the patients with the NSIP pattern, 17 had positive SCL70 antibody, 3 positive ANA patients and 1 patient had positive anti-centromere antibody.

Of the patients with UIP type interstitial pneumopathy, 8 patients had anti-SCL70 antibody, 3 patients ANA positive antibody and 2 patients anti-centromere positive antibody.

TABLE 2. RELATIONSHIP BETWEEN TYPE OF AB AND RADIOLOGICAL PATTERN

	NSIP	UIP
Anti SCL70	17	8
Anti centromere	1	2
ANA	3	3

Regarding treatment, 21 patients were taking Mycophenolate, 16 patients required cyclophosphamide and 6 patients rituximab.

No patient in our cohort died due to interstitial lung disease.

Conclusion: The data obtained are consistent with what is collected in the medical literature.

The subtype of scleroderma most related to ILD was diffuse SS. The most frequent antibody was anti-SCL 70.

Regarding the treatment, the most used in ILD in our center was the mycophenolate.

From our sample analyzed when applying the likelihood ratio (RV) a value of 47,186 is obtained, which has an associated probability of 0, which is less than 0.05, leads to reject the null hypothesis (there is no dependence between antibodies and type of radiological pattern of ILD in SS), concluding that there is dependence between the analyzed variables.

After this analysis, we can conclude that in our sample there is a relationship between the type of interstitial pneumopathy pattern and the antibody present in patients with SS.

Disclosure of Interests: None declared

DOI: 10.1136/annrheumdis-2020-eular.2673

AB0592 NAILFOLD CAPILLARY ABNORMALITIES PREDICT INTERSTITIAL LUNG DISEASE (ILD) COMPLICATION IN SYSTEMIC SCLEROSIS PATIENTS

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Background: Systemic sclerosis (SSc) have various organ involvements including pulmonary hypertension (PH), digital ulcers (DU), and interstitial lung disease (ILD). On the other hand, Nailfold capillary (NFC) abnormalities (enlarged/giant capillaries, fresh or old hemorrhages, avascular areas, ramified/bushy capillaries) detected by capillaroscopy are included in ACR/EULAR classification criteria for SSc as one of important findings. In addition, many studies have reported the relationship between NFC abnormalities and organ involvements (DU, PH) [1][2]. However, there are a few reports about the relationship between NFC abnormalities and ILD.

Objectives: We clarify the association with NFC abnormalities and ILD in SSc patients.