support from: Actelion, Roche, MSD, Consultant of: GlaxoSmithKline, VivaCell Biotechnology, Emerald Health Pharmaceuticals, Boehringer Ingelheim, Roche, Speakers bureau: Actelion, GlaxoSmithKline, Roche
DOI: 10.1136/annrheumdis-2020-eular.6566

AB0603 PDGFA AS A POTENTIAL BLOOD MARKER IN DSSC

K. Romanowska-Prochnicka^{1,2}, A. Wajda³, A. Paradowska-Gorycka³, A. Felis-Giemza², E. Walczuk³, M. Olesińska² on behalf of No. ¹Medical University of Warsaw, Department of General and Experimental Pathology with Centre for Preclinical Research and Technology (CEPT), Warsaw, Poland; ²National Institute of Geriatrics, Rheumatology and Rehabilitation, Warsaw, Poland; ³National Institute of Geriatrics, Rheumatology and Rehabilitation, Department of Molecular Biology, Warsaw, Poland

Background: PDGF is a potential important factor in the pathogenesis of scleroderma. PDGF is almost undetectable in healthy skin or lung. Immunohistochemical studies have revealed increased presence of PDGF α and PDGF β receptors in scleroderma skin biopsies.

Objectives: The aim of this study was to determine the mRNA level of *IFN α 1*, *IL-4*, *TGF β 1*, *TGF β 2*, *PDGF α* , *PDGF β* , *TNF α* in whole blood in SSc patients in the aspect of clinical

Methods: A group of 14 patients (50% were women) with systemic sclerosis based on EULAR / ACR 2013 criteria was included in the study. The modified Rodnan Skin Score (mRSS) was evaluated by same assistant at the beginning of the study and six months later. DLCO, HRCT, echocardiography and NFC were measured. Gene expression was determined using validated TaqMan probes in qPCR. Constitutive mRNA level of selected genes was analyzed using Δ Ct method. Comparison between different groups of patients was determined using non-parametric Mann-Whitney U test. Correlation was analyzed using non-parametric Spearman test.

Results: The mean age of the patients was 60 ± 15.66 . 100% of patients had organ involvement as pulmonary fibrosis. 78% - had active changes -features of ground glass. 64% of patients had mild mRSS-1-10 skin involvement, 36% had moderate to severe skin involvement. In SSc patients *TGF β 1* and *IFN α 1* revealed the highest level of expression in comparison to other analyzed genes. Additionally, very high and significant correlation between *TNF α* and *TGF β 1* ($r=0.7$ $p=0.004$) has been noted. High and significant correlation between mRNA *PDGF β* and *TNF α* levels have been observed. We did not reveal significant differences in analyzed genes expression when compare limited and diffuse SSc. Nevertheless, patients with dSSc were characterized by higher level of *IFN α 1* (almost 2 times) and *TGF β 1*. On the border of significance higher *PDGF α* mRNA level was observed in dSSc patients when compared to ISSc. Average *PDGF α* expression is higher in SSc patients with Scl70 positive than in patients without Scl70 ($p=0.04$).

In the aspect of clinical parameters, patients with ESR ≤ 12 mm/h revealed almost 6 times higher level of *IFN α 1* ($p=0.01$) in comparison to the patient with ESR >12 mm/h. Patients with mRSS above 10 points revealed significantly higher of *PDGF α* expression in comparison to patients with mRSS ≤ 10 ($p=0.04$). In these group of patients CRP and ESR were not different significantly.

In the case of patients with active fibrosis (ground glass) in HRCT *IFN α 1* expression was almost 2.5-times higher than in patients with HRCT non-active. Significantly higher *PDGF α* has been revealed in patients with active HRCT when compared with patient with non-active HRCT. Nevertheless these two groups did not differ in ESR or OB parameter.

SSc patients in active phase of NFC revealed almost 3 times higher level of *IFN α 1* expression in comparison with the patient in late phase.

Conclusion: The mRNA level of *PDGF α* may be a potential blood marker to predict worse prognosis in scleroderma

References:

[1] Rheumatology (Oxford). 2008 Oct;47 Suppl 5:v2-4. doi: 10.1093/rheumatology/ken265.

Role of PDGF in fibrotic diseases and systemic sclerosis. Trojanowska M1.

Acknowledgments: No

Disclosure of Interests: None declared

DOI: 10.1136/annrheumdis-2020-eular.5590

AB0604 EVALUATION OF PATIENTS WITH ANTISYNTHEASE SYNDROME AND INTERSTITIAL LUNG DISEASE BASED ON THE RADIOLOGICAL PATTERN

A. Ruiz Roman¹, J. A. Rodriguez Portal². ¹Hospital Virgen del Rocío, Rheumatology, Seville, Spain; ²Hospital Virgen del Rocío, Pneumology, Seville, Spain

Background: Antisynthetase syndrome is an autoimmune rheumatologic disease characterized by the presence of specific antibodies that are known as antisynthetase antibodies and with a varied clinic, including arthritis, myositis or

interstitial lung disease (ILD), among others. ILD is the manifestation that associates the highest morbi / mortality and can occur with different characteristic radiological patterns: usual interstitial pneumonia (UIP), non-specific interstitial pneumonia (NSIP), organized pneumonia (OP)

Objectives: Describe the clinical, serological and prognostic characteristics of patients with antisynthetase syndrome with interstitial pulmonary involvement (ILD) based on their radiological pattern.

Methods: Retrospective descriptive study of patients with a clinical diagnosis of antisynthetase syndrome in which they show positive for some antisynthetase antibody and with clinical follow-up of at least 6 months in a reference center consultation between the period from January 2008 to September 2019. In those patients who presented interstitial lung involvement clinical, analytical and prognostic variables were evaluated (including spirometry) based on the radiological pattern presented by high-resolution computed tomography (HRCT) of the chest

Results: 32 patients (24 women and 8 men) were included in the study. 7 cases (21.9%) did not present pulmonary involvement, while the remaining 25 cases (78.1%) presented with interstitial lung involvement. Of the patients diagnosed with ILD, 4 cases (12.5%) had an interstitial pattern UIP, 17 cases (53.1%) had an interstitial pattern of NSIP and another 4 (12.5%) interstitial pattern of OP. From an analytical point of view, the most frequent antisynthetase antibody in our sample was antiJo1 with 29 cases (96.9%) and less frequently the antiPL12 antibody with 2 cases and antiPL7 with 1 case. The specific characteristics depending on the type of EPID pattern are summarized in Table 1.

Table 1.

CHARACTERISTICS	ILD PATTERN		
	Pattern UIP N=4	Pattern NSIP N=17	Pattern OP N=4
Gender (Female) n (%)	4 (100)	11 (64.7)	3 (75.0)
Age at diagnosis	47.5 $\pm$ 12.9	52.9 $\pm$ 10.5	42.0 $\pm$ 12.9
Myositis, n (%)	2 (50.0)	13 (76.5)	4 (100)
Arthritis, n (%)	3 (75.0)	15 (88.2)	3 (75.0)
Fever, n (%)	1 (25.0)	9 (52.9)	2 (50.0)
Mechanic's hands, n (%)	2 (50.0)	7 (41.2)	2 (50.0)
Raynaud's phenomenon, n (%)	1 (25.0)	8 (47.1)	1 (25.0)
Skin involvement, n (%)	2 (50.0)	10 (58.8)	4 (100)
Treatment, n (%)			
Glucocorticoids	4 (100)	16 (94.1)	4 (100)
Immunosuppressants	4 (100)	16 (94.1)	4 (100)
Biological therapy	1 (25.0)	8 (47.1)	2 (50.0)
Drug's number (mean, SD)	3 $\pm$ 0.8	5 $\pm$ 2.9	4.7 $\pm$ 1.8
Antisynthetase antibodies, n (%)			
Anti Jo1	3 (75.0)	15 (88.2)	4 (100)
PL7	0 (0)	1 (5.9)	0 (0)
PL12	1 (25.0)	1 (5.9)	0 (0)
Ro52	3 (75.0)	14 (82.4)	2 (50.0)
RF positive	1 (25.0)	4 (23.5)	1 (25.0)
CK increase	2 (50.0)	11 (64.7)	4 (100)
Aldolase increase	2 (50.0)	12 (70.6)	4 (100)
Functional respiratory tests: mean (CI95%)			
Variation DLCOc (%)	-4.4 (-12.5, 3.7)	-2.5 (-6.1, 1.0)	7.4 (-10.6, 25.3)
Variation FVC (%)	-1.1 (-12.8, 9.1)	6.0 (3.2, 8.8)	7.5 (-9.5, 24.6)
Variation FEV1 (%)	0 (-0.2, 2.5)	5.0 (1.9, 8.3)	7.35 (-18.0, 32.7)
Variation FEV1/FVC (%)	0.5 (-7.1, 8.2)	-1.3 (-2.22, -0.37)	-0.4 (-12.88, 11.98)

Conclusion: ILD is a frequent and serious manifestation that can occur in patients with antisynthetase syndrome, which may have different radiological patterns. In our series, the most observed radiological pattern has been the NSIP pattern with 68%. The pattern most associated with the Ro 52 antibody was the UIP pattern and the NSIP pattern. As for the group that required the largest numbers of drugs (including biological therapy) it was the NSIP pattern. When evaluating changes in respiratory function tests, the pattern that shows a tendency to improve over time is organized pneumonia with improvement of the DLCO, FVC, FEV1, FEV1 / FVC, while the UIP pattern showed a worsening of the DLCO and the FVC.

Disclosure of Interests: None declared

DOI: 10.1136/annrheumdis-2020-eular.3584

AB0605 CLINICAL PROFILE AND CHEST HIGH-RESOLUTION COMPUTED TOMOGRAPHY (HRCT) FINDINGS IN PATIENTS WITH CONNECTIVE TISSUE DISEASES AND INTERSTITIAL LUNG DISEASE: EXPERIENCE OF A SINGLE REFERENCE RHEUMATOLOGY CENTER

I. Rusu¹, L. Muntean^{1,2}, M. M. Tamas^{1,2}, I. Felea², L. Damian¹, I. Filipescu^{1,2}, S. P. Simon^{1,2}, C. Pamfil², C. M. Marinescu³, M. A. Man^{2,4}, C. Csutak^{2,3}, S. Rednic^{1,2}. ¹Emergency Clinical County Hospital, Rheumatology, Cluj-Napoca, Romania; ²"Iuliu Hatieganu" University of Medicine and Pharmacy, Cluj-Napoca, Romania; ³Emergency Clinical County Hospital, Radiology,