SSc pattern prevailed (79%) in this study group, while unfavourable (21%) mainly consisted of SSc-PM/DM cases. Favourable overlapping SSc evolution was observed in patients with the onset before the age of 40 y, while unfavourable course was documented in pts with the late SSc onset at >40 y, with prevailing SSc-PM/DM. Fatal outcomes in 10% of cases mostly belong to SSc-PM/DM pts (8%). The specific features of overlapping SSc evolution included augmentation of SSc-characteristic symptoms – both, peripheral – teleangiectasias, calcification, osteolysis and digital trophic lesions, mainly in SSc-PM/DM pts, and visceral – involving heart, lungs, and oesophagus, which determined the unfavourable prognosis. RA manifestations (articular syndrome) in overlapping SSc pts tended to decrease, while signs of PM tended to resolve.

Conclusions: Timely detection of overlapping SSc pathological symptoms with administration of adequate therapy and dynamic monitoring of patients will improve the prognosis and outcomes of the disease.

Disclosure of Interest: None declared

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FRI0459

MICROVASCULATURE CHANGES AND ANGIOGENIC FACTORS IN SYSTEMIC SCLEROSIS – A SINGLE CENTRE STUDY

R. Bečvář¹, H. Hulejová², I. Půtová², M. Tomčík². ¹Department of Rheumatology, ²Institute of Rheumatology, Praha 2, Czech Republic

Background: In systemic sclerosis (SSc) low capillary density in acral parts leads to a reduced blood flow, to tissue ischemia. Tissue hypoxia usually initiates the formation of new blood vessels from the pre-existing microvasculature. Despite the reduced blood flow and partial oxygen pressure levels, there is no evidence for a sufficient angiogenesis in the skin of patients with SSc. Nailfold capillaroscopy is a safe, noninvasive routine way for the microvascular investigation. At the same time different cytokines and angiogenic factors are produced.

Objectives: The aim of this study was to assess whether blood levels of angiogenic biomarkers are associated with microvasculature changes in SSc patients.

Methods: Microvasculature changes were assessed using nailfold videocapillaroscopy (NVC) which was performed by two independent examiners. The obtained images were analysed anonymously by two investigators blinded for the clinical and serum status of SSc patients and classified as early, active and late pattern.¹ Serum or plasma levels of soluble vascular adhesion molecule-1 (sVCAM-1) and soluble intercellular adhesion molecule-1 (sICAM-1) were measured by ELISA, big endothelin-1 (BET-1) concentrations using competitive enzyme-immunoassay and von Willebrand factor antigen (vWFAg) concentrations using ELISA kit were measured. As potential disease activity markers soluble receptor of interleukin-2 (sIL-2r) and interleukin-6 (IL-6) serum levels using commercial kits were assayed. For statistical evaluation Pearson's correlation coefficient and univariate analysis were used.

Results: Total 40 patients (38 females) were investigated: 30 individuals with limited form, 5 with diffuse, 3 patients with scleroderma sine scleroderma, 1 with overlap syndrome and 1 with undifferentiated connective tissue disease. The mean age $\pm$ standard deviation (SD) of the whole cohort was 51 $\pm$ 22 years and the mean disease duration $\pm$ SD was 10 $\pm$ 7 years. 3 patients (7.5%) had early NVC pattern, 12 patients (30%) had active, 10 (58%) late pattern, and 15 (37.5%) had nonspecific changes or normal picture.

The patients with late NVC pattern exhibited higher vWFAg levels than patients with active pattern ($p < 0.01$). BET-1 and sICAM-1 serum levels were higher in the active pattern compared with late patterns ($p < 0.01$ and $p < 0.05$, respectively). When correlating these potential biomarkers with SSc-related clinical characteristics we found only these associations: vWFAg levels with heart arrhythmias and modified Rodnan skin score ($p < 0.01$, $p < 0.05$, respectively).

Conclusions: vWFAg and ET-1 increase in the late NVC pattern can be considered as an attempt to support deficient vasculogenesis. Further studies are needed to determine the role of other potential biomarkers of endothelial injury and repair in SSc.

REFERENCES:

- [1] Cutolo M, et al. Nailfold videocapillaroscopy assessment of microvascular damage in systemic sclerosis. *J Rheumatol* 2000;27:155–60.
- [2] Hasegawa M. Biomarkers in systemic sclerosis: Their potential to predict clinical courses. *J Dermat* 2016;43:29–38.

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FRI0460

THE COMPARISON BETWEEN PATIENTS WITH SYSTEMIC SCLEROSIS, POSITIVE FOR FOR ANTIBODIES TO RIBONUCLEOPROTEIN AND CLASSICAL SUBTYPES OF SYSTEMIC SCLEROSIS

R. Shayakhmetova¹, L. Ananieva¹, O. Koneva², M. Starovoitova², O. Desinova², O. Ovsyannikova², L. Garzanova². ¹V.A. Nasonova Research Institute of Rheumatology, Moscow, Russian Federation; ²Kashirskoe Shosse 34A, V.a. nasonova Research Institute of Rheumatology, Moscow, Russian Federation

Background: Different clinical subtypes of systemic sclerosis (SSc) vary in frequency and severity of symptoms, progression of internal organ involvement and outcomes. The frequency of antibodies to ribonucleoprotein (RNP) in SSc varies from 5% to 30% according to reports of different studies and can be connected with separate clinical subtype of SSc.

Purpose: to characterise the main clinical features of patients with SSc positive for antibodies to RNP and to compare them with ACA and Scl-70 positive subtypes of SSc.

Objectives: to characterise the main clinical features of patients with SSc positive for antibodies to RNP and to compare them with ACA and Scl-70 positive subtypes of SSc.

Methods: The study included 330 patients (289 women and 41 men, mean age 50 $\pm$ 13 years) meeting the criteria of the SSc (ACR/^{EULAR} 2013) observed between 2011 and 2017. The level of anti-nuclear antibodies was determined by ELISA. The normal level of antibodies to RNP (U1-RNP-70) was 0–25 U/ml. The level greater than 3 times the upper limit of normal considered as highly positive.

Results: In study group of 330 patients 49 (15%) had antibodies to ACA, 154 (46%) – to Scl-70, 67 (20%) – to RNP antibodies. Also 4 patients simultaneously had antibodies to Scl-70 and RNP, 4 patients – to ACA and RNP, 1 patient to ACA, Scl-70 and RNP. Among RNP +group 85% of patients were highly positive and 15% – low-positive.

The vast majority of patients were female (91%), mean age 44,2 $\pm$ 15 years. RNP +group was similar to ACA +group by predominance of a limited form of the disease which was 67,3% and 97% correspondingly. At the same time RNP +group was similar to Scl-70 +group by frequent involvement of internal organs – intestinal lung disease 67,3 and 69% correspondingly, involvement of cardiovascular system – 21% and 34%, esophagitis 61% and 44,5%.

RNP +group had sclerodema frequently – 40%, in comparison with ACA+ (10%, $p < 0,005$) and Scl-70+ (6,5%, $p < 0,005$), involvement of joints (arthralgia/arthritis) – 65% in comparison with ACA+ (24%, $p < 0,005$) and Scl-70+ (24%, $p < 0,005$), muscle weakness/pain – 43%, in comparison with ACA+ (0,2%, $p < 0,005$) and Scl-70+ (10,4%, $p < 0,005$). All 3 groups did not differ significantly by the presence of Raynauds syndrome, telangiectasia and vascular manifestations (fingertip pitting scars or digital tip ulcers).

Furthermore patients with SSc highly positive for antibodies to RNP, met the criteria of mixed connective tissue diseases^{Kasukawa et al., 1987} in 71%.

Conclusions: Our cohort of patients with SSc have high frequency of highly positive level of antibodies to RNP – 17%. Combination of specific SSc anti-nuclear antibodies (ACA, Scl-70) and antibodies to RNP was uncommon (2,7%) and predominantly in low-positive for antibodies to RNP patients.

The distinctive features of RNP +group in contrast to the "classical" subtypes of SSc were more mild skin involvement (puffy fingers) and more severe muscle and joints involvement.

Despite the limited form of the disease and mild skin involvement, RNP +group is more similar to Scl-70 +group by frequent involvement of internal organs.

We propose that well known relation between skin involvement in diffuse form of SSc with Scl-70 +and organ damage is not so evident in RNP +group of SSc.

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FRI0461

EFFICACY AND SAFETY OF ANTIFIBROTIC AGENTS IN IDIOPATHIC PULMONARY FIBROSIS

L. Nuñez-García¹, C. Valenzuela¹, T. Alonso¹, E.F. Vicente², J. Ancochea¹, S. Castañeda². ¹Pneumology; ²Rheumatology, Hospital Princesa, Madrid, Spain

Background: Antifibrotic (AF) agents are a family of drugs that improve the survival and quality of life of patients with idiopathic pulmonary fibrosis (IPF). Given that pulmonary fibrosis is also a common manifestation of many autoimmune diseases, we think of interest to know the efficacy and safety data of these agents in real life, which will likely soon reach the therapeutic arsenal of the rheumatologist.

Objectives: To analyse the efficacy and safety of treatment with AF pirfenidone (Pi) and nintedanib (Ni) at one year in patients with mild-moderate IPF treated in our hospital according to clinical practice.

Methods: Retrospective observational study in which all patients diagnosed with mild-moderate IPF who started treatment with Pi and/or Ni between January 2012 and May 2017 in our Hospital were included. The response was evaluated according to the results obtained in the Respiratory Function Tests: forced vital capacity (FVC) and carbon monoxide diffusion test (DLCO), which were carried out every 3