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 – telangiectasias, calcification, osteoarthrosis 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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FR0459

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

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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 microvascular changes in SSc patients.

Methods: Microvascular 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. 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 correlation analysis 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 standard deviation (SD) of the whole cohort was 51±22 years and the mean disease duration ±SD was 10±7 years. 3 patients (7.5%) had early NVC pattern, 12 patients (30%) had active, 10 (25%) late pattern, and 15 (37.5%) had overlap syndrome and 1 with undifferentiated connective tissue disease. The patients with late NVC pattern exhibited higher vWFAg levels than patients with early NVC pattern, 12 patients (30%) had active, 10 (25%) late pattern, and 15 (37.5%) had overlap syndrome and 1 with undifferentiated connective tissue disease.

Conclusions: Our cohort of patients with SSc have high frequency of highly positive levels of antibodies to ACA, Scl-70 and RNP. Among RNP+ group 85% of patients were highly positive and 15% low-positive.

Disclosure of Interest: None declared


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FR0460

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

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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.

Methods: The study included 330 patients (289 women and 41 men, mean age 50±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) 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%) patients had ACA antibodies (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 44,4±15 years. RNP+ group was similar to ACA+ group by predominance of a limited form of disease which was 63.9% and 97% correspondingly. At the same time RNP+ group 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 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.

Disclosure of Interest: None declared


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FR0461

EFFICACY AND SAFETY OF ANTIFIBROTIC AGENTS IN IDIOPATHIC PULMONARY FIBROSIS

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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 gefitinib (Pi) and nintedanib (N) 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 N 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), who were carried out every 3