months), continued the treatment for further 4 years (T4) (LIO group). Other 19 patients, although they continued the same cyclic intravenous iloprost treatment as the previous group, also received bosentan 125 mg twice a day for 4 years (LIO + BOS group), due to digital ulcers. DT was yearly evaluated by both US (18 MHz probe, MyLab 25, ESAOTE, Italy) and mRSS at the level of the usual seven skin areas (zygoma, fingers of hands, forearms, upper-arms, chest, abdomen, thighs, legs, and feet). 1,3,4 Non-parametric tests were used for the statistical analysis.

Results: A statistically significant decrease of DT, measured by US, was observed in the ILO+BOS group from T0 (median DT 1.135 mm) to T4 (median DT 1.088 mm) (p<0.01). No statistical significant variation of mRSS was observed during the follow-up in this group of patients (median mRSS at T0 125/1 and at T4 11/51, p=0.70). Conversely, in ILO group, a statistically significant increase of DT was observed after four years, as measured by US (median DT at T0 1.070 and at T4 1.258, p<0.001), as assessed by mRSS (median mRSS at T0 4/51 and at T4 8/51, p<0.001). Transient increase of transaminases was managed by temporary bosentan discontinuation.

Conclusions: In this open study, the long-term treatment with ET-1 receptor antagonist in combination with iloprost seems to be associated with a decrease of DT in SSc patients, in contrast to the treatment with iloprost alone. DT evaluated by US over long term seems to be more susceptible to change than by mRSS.

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LONGITUDINAL ASSESSMENT OF NAILFOLD CAPILLARY NUMBER, PERIPHERAL BLOOD PERFUSION AND DERMAL THICKNESS IN SYSTEMIC SCLEROSIS PATIENTS OVER A PERIOD OF 5 YEARS

B. Ruoaro1, A. Sulli1, C. Pizzorni2, V. Smith3, E. Gotelli4, J. Alshayeb5, A. C. Trombetta1, M. Cutolo1, 1Research Laboratory and Academic Division of Clinical Rheumatology, Department of Internal Medicine, University of Genova, San Martino Polyclinic Hospital, Genoa, Italy; 2Department of Rheumatology, Ghent University Hospital, Department of Internal Medicine, Ghent University, Ghent, Belgium;

Background: Several studies demonstrated the relationship between blood perfusion (BP), dermal thickness (DT) and nailfold microangiopathy (Early, Active and Late) vascularcapillaroscopic patterns) in patients with systemic sclerosis (SSc).1–3

Objectives: The aim of this study was to evaluate the changes of BP and DT, during a follow-up of 5 years, in SSc patients with persistent Late NVC pattern at baseline, as well as to confirm the correlations among microvascular damage extent, absolute nailfold capillary number (CN), BP and DT, during the follow-up.

Methods: Twenty-three female patients affected by SSc according to the LeRoy criteria (mean age 63±4 SD years, mean disease duration 6±4 years), displaying the most advanced Late NVC pattern of microangiopathy at baseline (T0), were enrolled and followed for five years (T5), after informed consent. Laser speckle contrast analysis (LASCa), skin high frequency ultrasound (US), modified Rodnan skin score (mRSS), and nailfold videocapillaroscopy (NVC) were yearly performed. Blood perfusion (BP), assessed by LASCa at the level of fingertips, periangular areas, dorsum and palm of both hands, was calculated as perfusion units (PU).1–3 Dermal thickness (DT) was assessed by both US and mRSS in the same above reported areas.2 The microangiopathy evolution score (MES) and the CN per linear millimetre at first distal row were evaluated by NVC.2,5 Patients were receiving a wide range of drugs, including vasodilators, immunosuppressive agents and endothelin-receptor antagonists. Statistical analysis was performed by non-parametric tests.

Results: A progressive statistically significant decrease of both BP (p<0.0001) and nailfold CN (p<0.0001) values was observed from T0 to T5 at the level of all areas, as well as a progressive statistically significant increase of DT (p<0.0001), mRSS (p<0.0001) and MES (p<0.01) values. The progressive decrease of BP positively correlated over time with the worsening of nailfold CN (p=0.05, r=-0.62), MES (p=0.05, r=-0.62), mRSS (p<0.001, r=-0.72) and DT (p<0.001, r=-0.64).

Conclusions: Microvascular damage with progressive reduction of nailfold capillary number was found, in a five-year follow-up, associated with progressive functional microvascular damage and DT worsening in the present cohort of SSc patients showing a persistent Late NVC pattern.

REFERENCES:

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EVALUATION OF HAND DERMAL THICKNESS AND PERIPHERAL BLOOD PERFUSION IN SYSTEMIC SCLEROSIS PATIENTS

B. Ruoaro1, A. Sulli1, T. Santiago2,3, J.A. Pereira da Silva2,3, C. Pizzorni2, Y. Tomatis1, M. Cutolo1, 1Research Laboratory and Academic Division of Clinical Rheumatology, Department of Internal Medicine, University of Genova, San Martino Polyclinic Hospital, Genoa, Italy; 2Rheumatology Department, Centro Hospitalar e Universitário de Coimbra; 3Faculty of Medicine, University of Coimbra, Coimbra, Portugal

Background: Systemic sclerosis (SSc) is characterised by progressive skin involvement. The modified Rodnan Skin Score (mRSS) is the gold standard to assess skin damage, but it has significant limitations. Recently, several studies demonstrated the utility of skin high frequency ultrasound (US) as an alternative.

Objectives: The aim of this study was to identify possible correlations between peripheral blood perfusion (BP) and ultrasound dermal thickness (US-DT) at DT of hand and finger in SSc patients.

Methods: Sixty-seven patients, satisfying the 2013 ACR/EULAR SSc criteria (mean age 64±9 SD years, mean disease duration 6±4 SD years) were enrolled. BP was measured as perfusion units (PU) by laser speckle contrast analysis (LASCa) at the level of dorsal region of hands. In a second time, different regions of interest (ROIs) were created at level of dorsal of the middle phalanx of 3rd finger and dorsum of hand bilaterally and the average BP was scored as perfusion units (PU), as previously reported.1,2 Both skin high frequency US and mRSS were used to evaluate DT at the above mentioned skin sites.2 The same examinations were performed in 65 healthy subjects.

Results: BP was negatively correlated with both US-DT (p=0.0005) and mRSS (p=0.007) in SSc patients at the finger sites, but not at the level of dorsum of hands. In healthy subjects, there was no statistically significant correlation between BP and DT as evaluated by both US and mRSS at each skin sites. SSc patients showed a statistically significant lower BP at the finger sites than healthy subjects (p<0.0001). No statistically significant difference in BP values was observed between SSc and healthy subjects at the dorsum of hands.

Conclusions: This study demonstrates a negative correlation between BP, as evaluated by LASca and DT, as evaluated by both US and mRSS, in the 3rd finger of SSc patients. Additionally, the results confirm a reduced finger BP in SSc patients when compared to healthy subjects.

REFERENCE:

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T-REG AND TH17 LEVELS IN PATIENTS WITH SYSTEMIC SCLERODERMA

D. Üsküdar Çansu1, H. Üsküdar Tekë1, Ç. Korkmaz2, 1Internal Medicine, Rheumatology; 2Internal Medicine, Hematology, Eskişehir Osmangazi University, Eskişehir, Turkey

Background: Regulatory T-cells (T-reg) may play an inhibitory role in the development of autoimmune diseases (AID) by suppressing the immune response to autoantigens and endothelin receptor antagonists. Impaired or decreased T-reg and/or increased Th17 cells may be responsible for the development of AID. However, studies about the association of T-reg and systemic scleroderma (SSc) are limited and conflicting.

Objectives: Our aim is to determine whether there is a relationship between T-reg and Th17 levels and disease activation in patients with SSc.