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AB0599 COMBINATION OF CAPILLAROSCOPIC AND ULTRASONOGRAPHIC EVALUATIONS OF THE HAND TO DETECT SEVERE VASCULOPATHY IN SYSTEMIC SCLEROSIS: RESULTS OF A CROSS-SECTIONAL STUDY

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Background: Although micro-vessel alterations have been largely described, macrovascular involvement is also frequent in Systemic Sclerosis (SSc). Macrovascular damages specifically involve narrowing or occlusions of proper palmar digital arteries and ulnar artery. On the contrary, radial artery is rarely concerned. Ulnar artery occlusion (UAO) assessed by power doppler ultrasonography (PDUS) has proven to be predictive of the onset of new ischaemic DUs in longitudinal studies (1). PDUS could also be a reliable tool to evaluate finger pulp blood flow (FPBF). Only few studies have explored the association between macrovascular damages evaluated by PDUS and microvascular involvement assessed by nailfold capillaroscopy (NC) (2). The association between macrovascular disease and calcinosis or Acro-osteolysis is still to be determined in SSc.
Objectives: to confront microvascular damages on NC with macrovascular manifestations evaluated by PDUS in SSc patients. Micro and macro-vascular damages were confronted with the main digital manifestations of the disease: digital ulcers (DU), Acro-osteolysis and Calcinosis.
Methods: NC, hand X-Rays and PDUS were systematically performed in 64 unselected SSc patients. PDUS evaluation with assessment of Ulnar Artery Occlusion (UAO) and finger pulp blood flow (FPBF) was performed blinded for the results of X-Rays and NC.
Results: UAO and pathologic FPBF were associated with severe capillary loss (<4 capillaries/mm) on NC (respectively OR=4.04 (1.23–13.29); $p<0.05$ and OR=3.38 (1.03–11.05); $p<0.05$). UAO was significantly associated with Cutolo's late NC pattern (OR=3.80 (1.31–11.01); $p<0.05$). A DU history was associated with UAO (OR=10.71 (3.36–34.13); $p<0.0001$), pathologic FPBF (OR=7.67 (2.52–23.28); $p<0.0001$), late pattern (OR=6.33 (2.03–19.68) and severe capillary loss (OR=8.52 (2.15–33.78); p=0.001). Acro-osteolysis was also associated with UAO (OR=15.83 (3.95–63.54); $p<0.001$), pathologic FPBF (5.52 (1.71–17.90) p=0.003), late NC pattern (OR=6.86 (2.18–21.53); p=0.001) and severe capillary loss (OR=7.20 (2.16–24.02), p=0.001). Calcinosis on X-rays were associated with late NC pattern (5.41 (1.82–16.12); p=0.002), severe capillary loss (OR=12.69 (3.14–51.26); $p<0.0001$) and UAO (3.19 (1.14–8.92); 0.025) but not with pathologic FPBF. Combination of UAO and severe capillary loss in a same patient was especially associated with DU history (OR=18.60 (2.24–154.34); p=0.001) and Acro-osteolysis (OR=10.83 (2.56–45.88); p=0.001).
Conclusions: The combination of macro and microvascular evaluations by PDUS and NC may help to detect patients with a more severe vasculopathy.
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AB0600 CLINICAL RELEVANCE OF AUTOANTIBODY PROFILES IN SYSTEMIC SCLEROSIS

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Background: Systemic sclerosis (SSc) is a connective tissue disease accompanied by immune abnormalities. A number of autoantibodies such as anti-centromere, anti-topoisomerase I, anti-RNA polymerase III and anti-U3 (fibrillarin) antibodies were proven to be of great diagnostic and prognostic factors in patients with scleroderma. Certain studies have reported the presence of antinuclear, anti-SSA (Ro) and/or anti-SSB (La), anti-Pm/Scl, anti-endothelial cell and anti-nucleosome antibodies in patients with systemic sclerosis. However, the clinical relevance of these autoantibodies is yet to be fully elucidated.
Objectives: Our aim was to assess clinical features, capillaroscopic abnormalities and autoantibody titers in patients with systemic sclerosis as well as analyze relationships between these parameters.
Methods: We conducted a prospective observational study on 36 adult patients who satisfied ACR/EULAR 2013 criteria for systemic sclerosis.We recorded disease duration. current symptoms and classified patients as limited cutaneous (lc) SSc and diffuse cutaneous (dc) SSc. Skin involvement was assessed using the modified Rodnan skin score (mRSS). We performed nailfold videocapillaroscopy using a FEDMED Digitale 100N at a magnification of 200X. Thoracic Xrays were done to establish the presence of pulmonary fibrosis. Ultrasounds were performed by a single examiner to evaluate pulmonary artery pressure. Blood samples were drawn to measure anti-topoisomerase 1, anti-centromere, anti-SSA (Ro), anti-SSB (La), anti-U1RNP and anti-nucleosome antibody titers (ELISA). Patient characteristics were included in a database and analyzed using IBM SPSS Statistics v20.
Results: Our study group was composed of 20 dc SSc (55.6%) and 16 lc SSc (44.4%) patients. Severity of capillary changes correlated with mRSS values (p,0.01), anti-topoisomerase I (p=0.006), anti-SSA (p=0.01) and anti-nucleosome antibodies (p=0.02). We found positive associations between the presence of dysphagia, anti-centromere (p=0.009) and anti-SSA ($p<0.01$) titers in patients with lc SSc. Anti-centromere antibody positivity also correlated with pulmonary hypertension (p=0,011) and pulmonary fibrosis (p=0,04) in these patients. Anti-SSA antibodies correlated with pulmonary hypertension ($p<0.01$) and capillaroscopic changes (p=0.024) in dc SSc.
Conclusions: Our findings support the relationship between autoantibody titers, systemic involvement and microvascular changes in scleroderma patients. Non-specific autoantibodies such as anti-nucleosome and anti-SSA antibodies were associated with microvascular changes in our study group. Further studies in this field may provide new information on SSc pathogenesis and possibly novel targets for treatment in scleroderma patients.
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AB0601 ANATOMIC AND FUNCTIONAL ASSESSMENT OF PERIPHERAL PERFUSION IN PATIENTS WITH SYSTEMIC SCLEROSIS. IS THERE ANY CORRELATION BETWEEN CAPILLAROSCOPIC FINDINGS AND ERGOSPIROMETRY?

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Background: Microvasculopathy of systemic circulation in patients with Systemic Sclerosis (SS) is widely assessed by digital capillaroscopy (DC), a method that evaluates the architecture of capillary network and reveals changes of vascular anatomy. On the other hand, ergospirometry (ERG) reveals functional impairment of microcirculation by assessing indirect measures of peripheral tissue ischemia (most suitable VE/VCO2). Since today, there have been studies correlating DC findings with spirometric parameters (respiratory volumes)[1,2]; nevertheless, no reports have been published correlating the same DC findings with ERG parameters (functional microvascular perfusion).
Objectives: To propose the use of DC as a screening tool for impaired functional microvasculopathy by investigating correlations between patterns of capillaroscopic findings with ergospirometric values of peripheral tissue blood perfusion.
Methods: 11 patients (11 women mean age 43+/- 12 ys) with SS were evaluated contemporary with High Resolution Computed Tomography of the chest, ERG, and DC. Parameters were correlated with multiple regression analysis. Statistic significance was considered $p<0.05$.
Results: Patient data are shown in the table.
Patients with late pattern in capillaroscopy had less endurance in exercise test ($P<0.05$) but no correlation was found between capillaroscopic pattern and VE/VCO2 ($P>0.05$). Age was a crucial confounding factor.
Conclusions: The correlation of late pattern capillaroscopy findings with reduced ergospirometric endurance in 11 patients indicates that in a larger cohort, specific parameter associations between DC and ERG are probable to emerge.

Diffuse scleroderma	6/11
Limited scleroderma	5/11
Pulmonary hypertension	4/11
History of digital ulcers	5/11
Time of endurance in minutes (ergospirometry)	5':32" (±65")
Diffusion capacity (%pred)	82±15
Pulmonary fibrosis	7/11
VE/VCO2 (mean values ± sd)	32±5
Patterns of capillaroscopy	1/11 normal 1/11 early 4/11 active 5/11 late

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AB0602 CAROTID ATHEROESCLEROTIC PLAQUES DETECTED BY ULTRASOUND IN SYSTEMIC SCLEROSIS PATIENTS UNDER 55 YEARS OLD

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Background: In Systemic sclerosis (SSc), as in other autoimmune diseases such as rheumatoid arthritis or systemic lupus erythematosus, cardiovascular events are one of the most frequent causes of mortality not attributed to the disease itself.
Objectives: The aim of this study was to assess the presence of subclinical atherosclerosis by carotid Doppler ultrasound in SSc patients under 55 years.
Methods: We conducted a cross-sectional study that included 78 SSc patients without cardiovascular events from H. Vall d'Hebron cohort (Barcelona). Carotid Doppler ultrasound was performed to measure the Carotid Intima Media Thickness (CIMT) of common carotid artery (CCA) and detection of cholesterol plaques in CCA, bulb and internal and external carotid arteries, according to Mannheim consensus criteria. The results were compared to a healthy cohort from Barcelona, adjusted to age and sex[1]. We used SCORE for populations with low risk and REGICOR as cardiovascular risk assessment charts.
Results: Risk factors and SSc related features are described in table 1. Twenty three patients (29.5%) had carotid plaques (CP) being the presence of CP statistically significant compared to the healthy cohort (29.5% vs 15.6%; $p<0.05$; IC 0.04–0.24). None of the patients were catalogued as high risk according to SCORE chart. According to REGICOR chart, 4 patients (5.1%) were catalogued as intermediate risk and none as high risk. The four intermediate risk patients had CP. Carotid Intima Media Thickness mean (CIMTm) and maximum (CIMTmax), were statistically significant increased compared to the healthy cohort (CIMTm 0,57 vs 0,53; $p<0,05$ and CIMTmax 0,74 vs 0,61; $p<0,05$).
We performed multivariate regression analysis. Age, CIMTmax, low High Density Lipid (HDL), the presence of pulmonary hypertension, and Diffuse cutaneous SSc patients were independent factors for the presence of CP.

Female	62 (79.5%)
Age	44 (20–55)
Disease duration	14 (0–38)
Limited cutaneous SSc	41 (57.7%)
Diffuse cutaneous SSc	17 (23.9%)
Sine Scleroderma SSc	8 (11.3%)
Initial Scleroderma	5 (7%)
Arterial Hypertension	18 (23.1%)
Diabetes mellitus	2 (2.8%)
Statin treatment	14 (17.9%)
Smokers	15 (19.2%)
Digital ulcers	30 (42.3%)
Pulmonary Hypertension*	4 (5.1%)
Interstitial lung disease	32 (41%)

*Mean pulmonary artery pressure >25 mm Hg at right heart catheterization.

Conclusions: SSc patients under 55 years old often have subclinical atheromatous disease which could be detected by carotid ultrasound. Risk charts do not correlate with the presence of subclinical atheromatous disease. The presence of carotid plaque is more frequent in SSc patients with pulmonary hypertension, diffuse cutaneous subtype and patients with low HDL. CIMT could be useful although it is currently not recommended in cardiovascular risk guidelines.
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AB0603 QUANTITATIVE VIDEOCAPILAROSCOPY STUDY IN SYSTEMIC SCLEROSIS

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Background: Systemic sclerosis (SSc) is an autoimmune disease characterized by immunological features, microvascular disturbances and fibrosis as a consequence of a massive production of extracellular matrix.
Objectives: The aim of this study was to analyse the alterations of the capillary nailfold bed and describe their correlation with clinical variables.
Methods: 134 patients were selected with SSc according to LeRoy and Medsger diagnostic criteria from the Hospital Universitari Vall d'Hebron cohort. Patients underwent an echocardiogram, pulmonary function test and nailfold videocapillaroscopy (NVC) within a 3 months period. NVC was performed from 2nd to 5th finger of both hands. NVC features were quantitatively measured. Informed consent was obtained by all the participants. SPSS 20.0 were used for statistical analysis. A P-value <0.05 was considered as significant.
Results: Female was the predominant gender (113, 84.3%). The most common cutaneous subtypes were limited SSc, diffused SSc (88, 65.7% and 28, 20.9%, respectively). Almost 80% of patients met the ACR/EULAR 2013 classification criteria. The age (± standard deviation) at NVC was 38.5 (± 15.7) years, and the interval from the first SSc symptom and NVC was 16 (± 12.6) years. Anticentromere antibodies (ACA) were the most frequent in 36.6% of patients, followed by anti-topoisomerase I in 23.1%. Regarding organ involvement, 62 (46.3%) patients suffered from digital ulcers, 58 (43.3%) interstitial lung disease (ILD), 11 (8.2%) pulmonary hypertension, 110 (82.1%) gastrointestinal disturbances and 103 (76.9%) some cardiac alterations. The forced vital capacity was 80.8 (± 20.1) % and diffusion capacity for carbon monoxide (DLCO) was 66.2 (± 23.7) %. Regarding echocardiography results, the tricuspid regurgitation velocity (TRV) was 2.8 (± 0.3) m/s and the right ventricular systolic pressure 30.7 (± 12.0) mmHg. NVC findings were described as follows: the median of number of capillaries was 5.4 /mm, enlarged capillaries 0.8/mm, giant capillaries 0.2 /mm, microhemorragies 0.1 /mm, ramified capillaries 0.3 /mm, tortuous capillaries 0.6 /mm and disorganized capillaries 0.0 /mm. SSc-ILD patients presented lower capillary density 4.8 /mm compared with 5.8 /mm in the other group (P=0.005), and also more frequent ramified capillaries 0.3 /mm compared with 0.2 /mm (P=0.013). With respect to the correlations, the number of capillaries was related with the DLCO (rho =0.26, P=0.003) but negatively with the RVSP (rho = - 0.21, P=0.03). Giant capillaries were correlated with LVEF (rho =0.27, P=0.001) and greater tricuspid annular plane systolic excursion (TAPSE) (rho =0.21, P=0.01). Microhemorraghies were associated with LVEF (rho =0.29, P=0.001) although negatively with age at NVC (rho = - 0.25, P=0.003) and time from first symptom (rho = - 0.25, P=0.003). Disorganized capillaries were related with age at NVC (rho =0.17, P=0.04) but negatively with DLCO (rho = - 0.18, P=0.035).
Conclusions: The existence of a correlation between NVC features and clinical variables suggests that microvascular alterations may play a role as a pathogenic link with the cardiopulmonary SSc manifestations.
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AB0604 RITUXIMAB EXPERIENCE IN PATIENTS WITH LONGSTANDING SYSTEMIC SCLEROSIS-ASSOCIATED INTERSTITIAL LUNG DISEASE: A SERIES OF 14 PATIENTS

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Background: Interstitial lung disease (ILD) is the leading cause of mortality in systemic sclerosis (SSc) patients. Treatment options are rather limited in SSc associated ILD (SSc-ILD).
Objectives: Objective of this study was to report the experience of RTX treatment in a series of patients with longstanding SSc-ILD in whom unsatisfactory response in lung functions was noted under conventional treatments.
Methods: We retrospectively reviewed charts of 197 SSc patients evaluated between April 2015 and November 2016. 14 patients who received rituximab (RTX) for SSc-ILD participated in this analysis. The severity of ILD based on PFTs was defined as follows; mild (FVC between 71% and 80% of predicted), moderate (FVC between 51% and 70% of predicted) and severe (FVC $\leq$50%). The extent of skin disease was clinically measured by using Modified Rodnan Skin Score (mRSS) tool. End of follow-up was considered as six months after the last RTX dose.
Results: Median (IQR, interquartile range) age was 53.2 (46.8–55.5) and median disease duration was 9.1 (5.1–13.6) years. Median FVC was 52.5 (41.5–64.0)