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AB0628 MACROVASCULAR DYSFUNCTION OF UPPER AND LOWER LIMBS CORRELATES WITH DIGITAL ULCERATIONS IN PATIENTS WITH SYSTEMIC SCLEROSIS

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Background: Systemic sclerosis (SSc) is a chronic connective tissue disorder of unknown etiology characterized by tissue fibrosis and vascular damage [1]. Digital ulcerations (DUs) are common manifestations of vascular involvement [2]. Althoughthe vascular dysfunction of SSc has been considered mainly to affect microvasculature [3], there is recent evidence showing that SSc is associated with the prevalence of large vessel disease [4]. However, only a few studies investigated the relationship between macrovascular disease and its role in the clinical manifestations of SSc such as ulcers.
Objectives: To assess the relationship between the Macrovascular dysfunction of upper and lower limbs with digital ulcerations in patients with systemic sclerosis.
Methods: Ninety patients with SSc (45 cases with DUs and 45 cases without DUs) enrolled in this study. Patients with other rheumatologic diseases and diabetics patients were excluded from the study. Data which were collected from the patients, included, age, past medical history of cardiovascular disease and dyslipidemia, SSc disease duration, type of SSc (lcSSc or dcSSc), Raynaud's phenomenon (RP), RP duration and digital ulcerations (DUs), Body weight (BW), Height, Waist circumference (WC), Body mass index (BMI), Blood pressure (BP), serum levels of SCL-70 antibody and Anti β2 microglobulin antibody. Then Doppler sonographies were performed. The outcome variables were the peak systolic velocity (PSV) and resistance index (RI) of ulnar, radial, popliteal, dorsalis pedis and tibia artery.
Results: The SSc patients with DUs have significantly lower PSV and higher RI in the ulnar (PSV:52.1±7.9 vs 55.7±4.1, p=0.006 and RI: 0.58±0.15 vs 0.52±0.06, p=0.003), dorsalis pedis (PSV: 33.9±1.9 vs 34.5±0.4, p=0.027 and RI: 0.54±0.11 vs 0.50±0.04, p=0.045) and tibial artery (PSV: 32.6±3.1 vs 34.4±0.9, p<0.001 and RI: 0.62±0.16 vs 0.51±0.07, p<0.001) in comparison to SSc patients without DUs. PSV and RI of ulnar artery were significantly correlated with age (p=0.012 and p=0.019), disease duration (p=0.001and p=0.001) and Raynaud's phenomenon (RP) duration (p=0.048 and p=0.028). PSV and RI of tibial artery had significant correlation with age (p=0.038 and p=0.009), systolic blood pressure (p=0.022 and p=0.037) and diastolic blood pressure (p=0.015 and p=0.010).
Conclusions: We concluded that digital ulceration in patients with SSc might be frequently related to the macrovascular dysfunction in below the elbow and knee.
References:
[1] Walmsley D, Goodfield MJ. Evidence for an abnormal peripherally mediated vascular response to temperature in Raynaud's phenomenon. Br J Rheumatol 1990;29:181–4.
[2] Chung L, Fiorentino D. Digital ulcers in patients with systemic sclerosis. Autoimmun Rev 2006;5:125–28.
[3] Ho M, Veale D, Eastmond C, Nuki G, Belch J. Macrovascular disease and systemic sclerosis. Ann Rheum Dis 2000;59:39–43.
[4] Veale DJ, Collidge TA, Belch JJ. Increased prevalence of symptomatic macrovascular disease in systemic sclerosis. Ann Rheum Dis 1995;54:853–5.
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AB0629 FROM LOCALIZED SCLERODERMA TO SYSTEMIC SCLEROSIS: A POSSIBLE EVOLUTION

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Background: Systemic Sclerosis (SSc) is a connective tissue disease characterized by skin fibrosis and visceral organ involvement. Localized Scleroderma (LoS), also known as morphea, is a fibrosing condition limited to the skin, subcutaneous tissue, underlying bone, and rarely central nervous system if present on face and head. SSc and LoS may share some aspects, such as histopathological findings, presence of autoantibodies and systemic symptoms, especially Raynaud phenomenon (RP). In this perspective they may represent two ends of a spectrum of disease.
Objectives: The aim of our study is to evaluate the evolution from LoS to SSc in our case series of SSc patients.
Methods: We retrospectively investigated 330 patients fulfilling the SSc-ACR/EULAR criteria referred to our University-based Rheumatology Unit. The occurrence of LoS preceding the SSc diagnosis was evaluated for each patient on the basis of medical records; clinical, laboratory, and instrumental features were analyzed, from the presenting symptoms at the disease onset to the first visit and during the follow-up, with particular attention to very early cutaneous manifestations.
Results: Five SSc patients (1.5% of our SSc series) had a clinical history of LoS prior to SSc diagnosis. All were women with mean age at time of LoS onset of 39±16.1SD years and time interval between LoS and SSc diagnosis of 19.2±16SD months. Skin biopsy was performed in 4/5 patients showing nonspecific inflammatory infiltrate, collagen fiber deposition and dermis sclerosis. In all patients RP was the first extra-dermal symptom, preceding LoS in 2/5 patients. Cutaneous involvement was represented by patches of skin sclerosis localized in limbs, trunk and face; while scleroderma was classified as cutaneous limited SSc in 4/5 patients and sine scleroderma in one. Following the SSc onset 2/5 patients had a history of digital ulcers, 1/5 esophagopathy, 1/5 interstitial lung disease; while capillaroscopy evidenced a SSc pattern in 4/5 patients. ANA were detected in all patients with anti-ENA positivity in 3/5 (1 ACA, 1 anti-Scl70, 1 anti-U1RNP); the presence of autoantibodies was observed in 3/5 individuals before SSc onset. None referred exposure to toxics or cigarettes smoke, while autoimmune thyropathy was the most frequent comorbidity. No local treatments had been employed for LoS but only low dosage of systemic steroids.
Conclusions: LoS and SSc are two distinct clinical entities that may share some clinical features; however, LoS is characterized by the absence of sclerodactyly, RP, digital ulcers, and typical SSc capillaroscopic changes; while possible internal organs involvement is much less frequently observed and the transition to SSc is exceptional and reported in only pediatric population. At our knowledge, this is the first observation of well-documented evolution from LoS to SSc in adult population as shown by updated review of the literature. The presence of RP and ANA positivity observed before the SSc onset can be considered as red flags of LoS evolution towards SSc, as reported in literature in pediatric population. SSc following LoS seems to be characterized by higher prevalence of vasculopatic symptoms compared to fibrotic complications. Finally, a careful clinical and laboratory monitoring of patients with LoS is recommendable to early identify the possible evolution to overt SSc.
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AB0630 CLINICAL SPECTRUM TIME COURSE COMPARISON BETWEEN PL-7, PL-12 AND EJ POSITIVE ANTISYNTHETASE SYNDROME

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Background: Arthritis, myositis and Interstital lung disease (ILD) represent the classic clinical triad of antisynthetase syndrome (ASSD). In anti Jo-1 positive patients, these findings may appear also during the follow-up. Even if a similar cumulative trend has been showed also in non anti Jo-1 positive ASSD, a head to head comparison of clinical spectrum time course in these patients is still lacking
Objectives: To assess the clinical spectrum time course in non anti Jo-1 positive ASSD, according to different underlying non anti Jo-1 specificities
Methods: Clinical, laboratory and instrumental data collection of anti PL-7, PL-12, and EJ positive patients from an international database of ASSD
Results: We identified 63 (42%) anti PL-7, 66 (44%) anti PL-12 and 20 (14%) anti EJ positive patients, reporting their characteristics in table 1 (disease onset) and 2 (last follow-up). At disease onset, no substantial differences were observed. At the end of follow-up, we observed some differences between anti PL-12 and both anti PL-7 and anti-EJ positive patients. In particular, anti PL-12 positive patients presented less frequently ex-novo triad findings and had a reduced prevalence of