Heart Valvular Alterations in a Multicentre Italian Cohort of SSC Patients


AB1065

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Background: systemic sclerosis (SSc) in a chronic autoimmune disease characterized by endothelial dysfunction, diffuse microangiopathy, and fibrosis of skin and visceral organs. Typical cardiac involvement may includes microvascular ischemia, contraction band necrosis, and patchy fibrosis, leading mainly to arrhythmias and conduction defects, diastolic dysfunction, or right ventricular failure (secondary to pulmonary arterial hypertension) [1]. Valvular disease are poorly described and generally not considered a typical sign of SSc [2-4].

Objectives: we aimed to describe valvular alterations in a multicentre cohort of SSC patients.

Methods: we consecutively recruited 118 SSC patients (M/F: 14/104, mean age 56.7±12.4 years, median disease duration 10 years, limited/diffuse skin subsets: 95/23, anti-centromere/anti-Sc170/others autoantibodies: 35/37/46) in 3 Rheumatology Centres in Sicily, Italy, from January to December 2019. Considering the cardiovascular risk factors, 40 (34%) patients were smokers, 7 (6%) diabetics, 12 (10%) showed hypercholesterolemia, 38 (32%) arterial hypertension, while none was obese. Transthoracic echocardiogram was carried out in all patients during their follow-up.

Results: valvular abnormalities were as follow: mitral valve: insufficiency 85 (72%) cases - mild in 77/85, stenosis 2 (2%) - mild in 25/28, sclerosis/ticken- ing 36 (30%), and calcification 9 (8%) patients; aortic valve: insufficiency 28 (24%), stenosis 4 (3%), sclerosis 29 (25%), and calcification 7 (6%) patients; tricuspid valve: insufficiency 91 (77) cases, no cases of stenosis, sclerosis 5 (4%), and calcification 1 (1%) patients; pulmonary valve: insufficiency in 13 (11%) patients.

As expected, tricuspid insufficiency (TI) was associated with pulmonary arterial hypertension (PAH) (moderate TI in 20% of patients with every TI and PAH vs. 4% of patients with TI without PAH, p=0.019).

Aortic sclerosis (AS) was associated with the presence of arthritis (AS in 35% of patients with arthritis vs. 16% of patients without, p=0.023).

No association was found with age, gender, disease duration, skin subset, autoantibodies, capillaryoscopic patterns, presence of digital ulcers, lung, renal, or digestive involvements.

Conclusion: in this multicentre SSC cohort study, we found that cardiac valve alterations are very common, even though generally not clinically relevant. The presence of PAH was associated with more severe TI. Finally, AS was associated with arthritis that could be considered sign of chronic inflammatory state, which is often linked with accelerated atherosclerosis and remodeling process of aortic valve [5].

References:

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Hospitalization in a Cohort of Patients with Idiopathic Inflammatory Myopathy: What is Happening in Argentina?

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Background: Inflammatory myopathies are rare diseases that affect multiple organs, with poor prognosis and high in-hospital mortality. [1-2] In Argentina there are few reported data regarding hospitalization and its outcomes in these patients.

Objectives: To analyze the characteristics of hospitalizations and the factors associated with poor outcome in adult patients with Idiopathic inflammatory myopathy (IMM).

Methods: Retrospective, analytical study. We included patients ≥ 18 years with IMM, according to Bohan and Peter and/or ACR / EULAR 2017 criteria, who were admitted in our hospital between 2003 and 2019 at least once. Sociodemographic and clinical data were recorded. We defined “unfavorable outcome” as the presence of one of the following events: death, mechanical respiratory assistance and/or critical care unit requirement. Continuous variables were compared by Student’s or Mann Whitney’s T test, and categorical variables by Chi² test or Fisher’s exact test. Binary logistic regression was performed to identify independent factors associated with an unfavorable outcome.

Results: 61 hospitalizations of 40 patients with IMM were evaluated; 67.3% of the patients were female (27/40), with a mean age of 52.5 years (SD± 13). The most frequent reason of admission was for diagnosis (44.3%) followed by disease activity (31.1%). In 78.7% of hospitalizations (48/61) the diagnosis was dermatomyositis. The median of hospitalization days was 14 (IQR 8-30). In 21 out of 61 hospitalizations (34.4%), an unfavorable outcome was observed, of which 17 (80.9%) ended in death. Respiratory muscle involvement (p = 0.01), thrombocytopenia (p < 0.001), treatment with intravenous methylprednisolone pulses (p = 0.032), Intravenous Immunoglobulin (IVlg) (p = 0.001), longer hospitalization (p = 0.001) and severe infections (p = 0.001) were associated with adverse outcomes.

In the multivariate analysis, serious infections (OR: 21.7; IC95 1.77 - 266; p = 0.016) and the requirement of Intravenous Immunoglobulin (IVlg) (VR: 5.45; IC95 1.4 - 214; p = 0.033) were found to be independently associated with an unfavorable outcome.

Conclusion: IMMs are diseases with high morbidity and mortality rate. In this cohort of hospitalized patients, we found a high percentage of unfavorable outcomes. Seriously ill patients received IVlg more frequently, and severe infections were associated with worse prognosis.

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AB0566
NAIFOLD EXTENT OF REDUCED CAPILLARY DENSITY IS ASSOCIATED WITH DIGITAL ULCERS AND WITH AN INCREASED RISK OF DIGITAL ULCERS IN SYSTEMIC SCLEROSIS

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Background: A growing evidence supports the role of microvasculopathy as a primary pathogenic event in systemic sclerosis (SSc). The most commonly used imaging technique to identify microangiopathy in SSc is high magnification videocapillaroscopy (VNC), and reduced capillary density and/or capillary loss, which is a typical feature of “sclerodema microangiopathy”, easily identified by VNC, has been associated with digital ulcers (DUs). Different approaches have been proposed to measure capillary density or capillary loss. Some of these were qualitative methods, others semi-quantitative, others only concerned a limited nailfold area, without ever evaluating the overall density, which is more suitable for quantitative estimate.

Objectives: To assess the association between the extent of different values of nailfold capillary density and the presence of DUs and to identify the risk of developing DUs, based on quantitative parameters.

Methods: The study involved 54 SSc selected patients (47 women and 7 men, mean age 59.5 years, 50 with limited and 4 with diffuse). The study population came from an ongoing database, that includes clinical and laboratory data of patients with definite SSc. A videocapillaroscope (VideoCap® 3.0, DS Medica, Milan, Italy) with a 200x optical probe was used. During examination, eight fingers (fingers 2–5 of each hand), 4 fields per finger, according to the standard literature were assessed. For each patient, a total of 32 images were collected, then classified as having either “normal”, “non-specific” or the “scleroderma pattern” (SP). Capillary density was defined as the number of capillaries/mm in the distal row, regardless of its shape and morphology. Vascular areas were defined by the absence of loops within a width/area extending over more than 500 microns. For each patient, the SP images were further graded with no/slight reduction of the capillary density (7–9 loops/mm) (NOR), with a well-defined reduction of digital ulcers (DUs). Different approaches have been proposed to measure capillary density or capillary loss. Some of these were qualitative methods, others semi-quantitative, others only concerned a limited nailfold area, without ever evaluating the overall density, which is more suitable for quantitative estimate.

Results: A total of 1728 images were analyzed. Patients with DUs were 16/54 (29.6%). All patients had a SP, but only five patients showed a SP along the entire nailfold. A comparison between patients with or without DUs showed a significant difference both for the overall extent of AA (p=0.032), and particularly for the overall extent of RED (p<0.001). No significant difference was found regarding the overall extent of the SP (p=0.085). Factor significantly associated with DUs in multivariate analysis was the overall extent of RED (p=0.0286). The ROC curve was very effective at discriminating the capillary feature able to distinguish patients with DUs from patients without DUs. The discriminatory power of the overall extent of RED was very good, with an AUC of 0.948 (95% CI 0.852 ± 0.990). Then, we calculated the cut-off values of the overall extent of RED for presence/absence of DUs with the highest discriminatory power of the overall extent of RED was very good, with an AUC of 0.948 (95% CI 0.852 ± 0.990). Then, we calculated the cut-off values (Y ourden index of 0.825) was >68.7 (sensitivity 92.31 %; specificity 90.24 %) with a LR+ of 9.46.

Conclusion: Our data strongly support that the capillary density between 4 and 6 loops/mm is the best capillaroscopic quantitative measure associated with DUs and able to discriminate the probability of having DUs. If all SSc-specific antibodies and/or other laboratory/clinical parameters are not yet available, the overall capillary density can allow physicians to assess SSc patients easily, regarding DUs and risk for developing DUs.

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AB0567
CHARACTERIZATION OF PATIENTS WITH IDIOPATHIC INFLAMMATORY MYOPATHY AND MYOCARDIAL INVOLVEMENT: A MONO-CENTRIC EXPERIENCE.

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Background: Idiopathic inflammatory myopathies (IIM) are immune-mediated disorders of the skeletal muscle, with dermatomyositis (DM), polymyositis (PM), inclusion body myositis (IBM) and immune-mediated necrotizing myopathy (IMNM) representing major subtypes. Beyond skeletal muscle, other organs may be affected and myocardial involvement may lead to severe life-threatening complications. The exact prevalence of myocardial involvement among IIM patients and its impact on other disease characteristics remain unclear.

Objectives: To investigate the prevalence of myocarditis in patients affected by IIM in and to determine whether the presence and extent of myocardial involvement identify a distinct disease phenotype.

Methods: 42 longitudinally followed IIM patients were routinely screened for myocardial involvement during a median [IQR] follow-up time of 4.2 [2.8-5.5] years. Patients with secondary causes of myocardial dysfunction were not included. Patients were considered to have myocarditis in case of: i) abnormal elevation of both circulating troponin T and troponin I, ii) signs of myocardial inflammation or necrosis/fibrosis at cardiac MRI, or iii) positive myocardial tissue histology. Demographic, clinical and serologic features of patients with myocarditis were compared to those with no sign of myocardial involvement. Moreover, we determined whether the extent of myocardial involvement based on troponin levels predicts skeletal muscle disease severity.

Results: 57% (24 of 42) of patients had myocarditis. The frequency of myocardial dysfunction was similar among patients with DM, PM or IBM and IMNM and was not related to autoantibody positivity. Myocarditis was not associated with sex or ethnicity. Patients with or without myocarditis were similar in terms of age at disease onset and extra-muscular manifestations including dysphagia, dysphagia, arthralgias or arthritis, Raynaud phenomenon or interstitial lung disease. Independent of the IIM subtype, the prevalence of perimysial macrophages at skeletal muscle biopsy seems to protect from myocarditis development (p=0.04). Patients with myocarditis had higher median [IQR] levels of aldolase (10.9 [7.8–15.8] vs. 5.6 [4.9–8.6], p=0.014) and creatine kinase (1785 [966–5852] vs. 685 [168–2255], p=0.04) compared to patients with no myocardial dysfunction. Among patients with myocarditis, levels of troponin I negatively correlated with manual muscle testing 8 (MMT8) score (r=-1, p=0.01), strength in biceps (r=-0.95, p=0.014) and wrist extenders (r=-0.95, p=0.014) at last visit. Troponin T and troponin I titers were similar among patients with different IIM subtypes. C-reactive protein (p>0.04) but not erythrocyte sedimentation rate was found to predict myocardial involvement.

Conclusion: Our findings suggest that myocarditis is a frequent occurrence among patients with IIM and should be routinely ruled out. A more severe skeletal muscle disease is associated with an increased likelihood of myocarditis development, presumably due to higher systemic disease activity or inefficient disease control. The extent of myocardial damage faithfully reflects the severity of skeletal muscle dysfunction.

References:

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AB0568
BASELINE EUROPEAN PATIENT DEMOGRAPHICS AND DISEASE CHARACTERISTICS IN A PHASE 3 STUDY OF SAFETY AND EFFICACY OF LENABASUM, A CB2 AGONIST, IN DIFFUSE CUTANEOUS SYSTEMIC SCLEROSIS

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Background: We previously presented on the baseline characteristics of a large cohort of diffuse cutaneous systemic sclerosis (dcSSc) patients enrolled in a Phase 3 trial of lenabasum, a selective cannabinoid receptor type 2 (CB2) agonist. Lenabasum, was safe and well-tolerated in a prior Phase 2