A COHORT STUDY OF MACROVASCULAR INVOLVEMENT IN PATIENTS WITH SYSTEMIC SCLEROSIS

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Background: It is well known that systemic sclerosis (SSc) affects microvessels, but data on macrovascular involvement are still lacking or debatable.

Objectives: Aim of this study was to estimate the prevalence of atherosclerotic plaques and their possible determinants in a large cohort of SSc patients.

Methods: One-hundred and four outpatients with SSc were enrolled. Data about disease characteristics and cardiovascular risk factors (diabetes, hypertension, dyslipidemia, smoke) were collected and patients underwent a full ecocolorDoppler ultrasonography of arteries of the neck and lower limbs (LL).

Results: Mean age of our cohort was 62±13 years and 17 (16.3%) patients were male. Seventy patients (67.3%) had a limited subset and the mean disease duration was 12±6.8 years. A previous history of digital ulcers was found in 27 cases (26.0%). Thirty-eight patients (36.5%) were on ongoing or previous treatment with immunosuppressive drugs.

Regarding cardiovascular risk factors, 22 (21.2%) had hypertension, 7 (6.7%) diabetes, 52 (50%) dyslipidemia and 45 (43.3%) were active or past smokers. Fifty-seven (54.2%) patients had plaques at carotids, 1 (1%) at LL and 37 (35.9%) at UL. Prevalences were higher in older patients, as expected.

Patients with carotid plaques were older (p=0.001), with hypertension (p=0.057), a limited disease subset (p=0.005), a more severe disease accordingly to Medsger severity score (0.048), worse renal function (p=0.012), higher glucose blood levels (p=0.001), homocysteinemia (p=0.006) and ESR (p=0.004) and less often on immunosuppressors (p=0.048) but more often on steroids (p=0.050).

Patients with LL plaques were older (p=0.001), male (p=0.003), treated with statins (p=0.056), with a worse renal function (p=0.001), higher glucose blood levels (p=0.038) and homocysteinemia (p=0.006).

In multivariate analysis with all variables with a p=0.010 in univariate, patients with carotid plaques were found to be older (p=0.003) and with a limited disease subset (p=0.012; OR 5.6, CI95% 1.5 to 21.5). These data were confirmed even after correcting also for other well-known risk factors for atherosclerosis. No variables were found to be statistically different between patients with and without LL plaques.

Conclusions: In this study we performed one of the most complete evaluation of macrovascular involvement in one of the most numerous cohort of SSc patients present in literature. The prevalence of carotid and LL plaques did not seem to be higher in SSc patients as compared to what reported in healthy subjects. Intriguing is that patients with limited disease have an increased risk of having carotid plaques even after correcting for possible confounders.

Disclosure of Interest: None declared


ORGAN INVOLVEMENT AND ILD PROGRESSION IN SCLERODERMA PATIENTS WITH ANTI-TOPOISOMERASE I SPECIFICITY AND LIMITED CUTANEOUS FORM

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Background: The great variability of clinical manifestations in systemic sclerosis (SSc) yields the need of defining prevalence and progression of organ involvement in the different subsets of the disease. Despite the classical association between anti-topoisomerase-I (SC170) positivity with diffuse cutaneous SSc (dSSc), and anti-centromere (ACA) with limited cutaneous SSc (lSSc), a population of patients with antiSC170 and ISSc has been described.1 Intertitial lung disease (ILD) is nowadays the leading cause of mortality in SSc; its progression can range from a self-limiting form to a rapidly progressive lung involvement, despite immunosuppressive treatment.

Objectives: The aim of our study was to evaluate the prevalence of different organ involvement in three groups of patients: ACA-ISSc, SC170-dSSc e SC170-lSSc. Our second endpoint was to investigate possible differences in ILD onset and progression between SC170-ISSc and SC170-dSSc patients.

Methods: Consecutive 260 patients attending the Rheumatology Unit of Padova University were included and retrospectively evaluated. Clinical, biochemical and functional parameters concerning pulmonary, cardiac and articular involvement were collected in all patients, with an average follow-up time of 15 years. As for lung involvement, spirometric indices (FVC e DLCO) and HRCT at ILD onset and at last follow up were considered (median ILD duration 8 years2,3).

Time between SC170 onset and ILD first evidence was defined as “scleroderma free ILD”. ILD grading was determined according to Kazerooni score. ILD progression was defined as either an HRTC score worsening of at least 2 points, or as a significant progression of spirometric indices (10% and 15% for FVC and DLCO respectively).

Results: 150 patients with ACA-ISSc, 58 with SC170-dSSc and 52 with SC170-lSSc were included in the study. SC170-ISSc patients presented more often with pulmonary and articular involvement with respect to ACA-ISSc patients (50% vs 6.9% and 42.9% vs 23.6% respectively), and less often compared to SC170-dSSc. In SC170-ISSc, cardiac and gastrointestinal involvement, BEV and digital ulcers were less prevalent with respect to SC170-dSSc (p=0.05). SC170-ISSc patient had a longer “scleroderma free ILD” compared to SC170-dSSc. At ILD onset pulmonary function was worse in SC170-dSSc group than in SC170-ISSc group (p=0.02 p=0.009 for FVC e DLCO respectively), even in the absence of a significant difference between HRTC scores (12.73±5.25 vs 10.25±4.93, p=0.284).

Total ILD progression was significantly higher in SC170-dSSc (p=0.0001).

Conclusions: Our study shows that SC170-ISSc patients appeared to have a different prevalence of targeted organ involvement and ILD progression with respect to both SC170-dSSc and ACA-ISSc. Since the correct classification of SSc patients is extremely important in view of a tailored treatment, our data suggest that it could be worth to identify patients with SC170 and limited cutaneous form as a different and specific subset.

REFERENCES:

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ASSESSMENT OF PERSISTENT ORGAN DAMAGE ACCORDING TO IMACS (INTERNATIONAL MYOSITIS ASSESSMENT AND CLINICAL STUDIES) MYOSITIS DAMAGE INDEX IN 92 PATIENTS WITH IDIOPATHIC INFLAMMATORY MYOSITIS

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Background: Number of work regarding long term organ damage caused by idiopathic inflammatory myopathies (IIl) and risk factors associated with organ damage have been undrstandted and never reported from Turkey; recently a new tool as developed fort that purpose.

Objectives: In this study we aimed to evaluate by long term organ damage and risk factors associated with these prospectively in IIM patients.

Methods: IIM patients (n=110) who has been followed up for at least six month by our clinic and fulfilling Bohan and Peter criteria were recruited. Demographic data, clinical and serological features, treatment and final clinical tatus was recorded. IMACS Myositis Damage Index (MDI) was determined twice in 92 patients (71% female) at the time of diagnosis from the records and at the at last clinical visit prospectively.

Results: Mean age of the 92 patients during the diagnosis was 46±14.7. Out of 92 patients 69% had dermatomyositis, 23% had polymyositis, 8% had necrotizing