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FRI0348 USE OF INTRAVENOUS IMMUNOGLOBULIN THERAPY IN PATIENTS WITH SYSTEMIC SCLEROSIS: A SPANISH MULTICENTER EXPERIENCE

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Objectives: To describe the efficacy of intravenous immunoglobulin (IVIG) therapy in different organic conditions of Systemic Sclerosis (SSc).

Methods: Retrospective multicenter observational study that enrolled patients with SSc treated with IVIG. We collected epidemiological data, SSc complications, treatments and functional tests. Regarding IVIG treatment we recorded the reason to use, number of cycles, and the clinical efficacy at the end of the last IVIG cycle were collected at investigator's discretion. For the comparison of variables, the Chi-square, Fisher's F test and the T test was used.

Results: 41 patients (83% women) were recruited, with a mean age of 58 ± 18.2 years old. The age of diagnosis was 48 ± 8 years old. The diffuse cutaneous SSc was the most frequent in the sample (61%) and 24/41 had Overlap syndrome (49% myositis). Regarding the different SSc complications in treated patients the most frequent were digestive involvement, arthritis and interstitial lung disease (78%, 73%, and 63% respectively). 12% of patients developed cancer and 25% died during the follow-up. The most frequent indication IVIG was myositis (51%) followed by cutaneous (17%). The mean of cycles was 10. 37% of patients had previously undergone treatment with biological therapy, being RTX the most used therapy. 80% of patients had a history of use with corticosteroid therapy. When evaluating the degree of skin involvement (mRSS) patients showed a significant improvement of -2.89 ± 5.2 (p < 0.03) at the end of the follow-up. Indeed we observed better results in the groups of patients with myositis. No differences were observed in the % FVC or % DLCO outcomes during the follow-up. However, when we compared patients with or without overlap syndrome we found differences in %FVC values at the beginning of the study that were not present at the end of the follow-up. Patients with antiScl70 seemed to have less response to IVIG therapy.

Conclusion: Our results suggest that IVIG can be useful for the management of some conditions in specific profiles of patients with SSc.

Prospective studies and randomized clinical trials are necessary to establish the efficacy and safety of IVIG to treat the different conditions of SSc.

	IVIG N=41(%)	Difuse n=25 (%)	Limited n=16(%)
Women	34(83)	21(84)	13(81)
Age	58 ± 18		
Overlap	24(59)	11(44)	13(81)
Clinical manifestations			
Joint	30(73)	22(88)	8(50)
Muscular	30(73)	16(64)	14(88)
Reflux GE	32(78)	20(95)	12(75)
Interstitial lung disease	26(63)	16(64)	10(63)
Pulmonary hypertension	5(12)	2(8)	3(19)

	myositis	Overlap	AntiScl70	PM/ Scl
Initial mRss	16±12	13±11	29±9	15 ±10
Final mRss	12±12	7±7	32±7	13 ±15
Initial FVC	68±25	64±24	56±23	60 ±13
Final FVC	73±26	72±25	58±20	67 ±18
Initial DLCO	56±16	56±17	55±16	49 ±13
Final DLCO	68±25	71±24	45±12	69 ±18

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FRI0349 RELATIONSHIPS BETWEEN BODY COMPOSITION AND NAILFOLD VIDEOCAPILLAROSCOPY PATTERNS IN SYSTEMIC SCLEROSIS PATIENTS

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Background: Systemic sclerosis (SSc) is a chronic autoimmune disease, characterized by microvascular damage and progressive fibrosis. Among clinical complications, abnormal body composition and sarcopenia have been reported in SSc patients(1,2). Nailfold videocapillaroscopy (NVC) is a safe diagnostic tool to assess microvascular progressive damage and may predict severe organ involvement in SSc(3).

Objectives: The aim of this study was to evaluate body composition in SSc patients and assess possible differences between patients with specific NVC patterns of microangiopathy.

Methods: 38 patients (6 men and 32 women) fulfilling ACR 2013 criteria for SSc underwent NVC to assess their NVC patterns ("Early", "Active" and "Late" pattern)(4) and DXA to evaluate body composition(5). In particular we analyzed: total mass, total lean mass, total fat mass, bone

mineral content(BMC), and bone mineral density(BMD) in seven body areas(head, upper limbs, lower limbs, trunk, spine, ribs, pelvis). Sarcopenia was diagnosed in patients with reduced skeletal muscle index (RSMI) below 5.45 Kg/m² for females and 7.25 Kg/m² for males(6). Statistical analysis was performed by non parametric tests.

Results: The mean age of patients was 64±11 years, mean disease duration 19.2 ±7.6 years, mean Rodnan skin score(mRSS) 11.5±9.3, and mean RSMI 6.3±1.05 g/cm². All the patients showed a NVC "scleroderma pattern": in particular 15 patients showed the "Late" pattern, 15 patients the "Active" pattern and 8 patients the "Early" NVC pattern. The "Late" NVC pattern group comparing to "Early/Active" group showed significantly lower total mass(58287±8217 vs 67238±11437 gr, p=0.01), lean mass (35249±3646 vs 41230±7954 gr, p= 0.05), RSMI(5.8±0.92 vs 6.6±1.02 g/cm², p=0.02), BMC(1839±339 vs 2183±502 gr, p= 0.04), trunk BMD(0.70 ±0.12 vs 0.87±0.13 g/cm², p=0.05) and spine BMD(0.91±0.17 vs 1.08 ±0.18 g/cm², p=0.008). No statically significant difference between the two group was observed regarding total fat mass, total body BMD and BMD at upper limbs, lower limbs, head, ribs and pelvis. Interestingly, 24% of SSc patients were found affected by sarcopenia, and the most of sarcopenic patients showed the "Late" NVC pattern(67%). Comparing age, disease duration, mRSS between sarcopenic and non sarcopenic patients there was no difference between the groups, but sarcopenic patients presented a statistically significant lower BMI(p=0.02).

Conclusion: This study demonstrates in SSc patients a relationship between a more severe microvascular damage ("Late" SSc pattern) and the body composition, characterized by lower weight, total lean mass, bone mineral content and sarcopenia, without any significant variation in total fat mass. These clinical conditions seem not to be associated with severity of skin involvement and/or disease duration.

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FRI0350 THE FREQUENCY OF LOW MINERAL DENSITY, FALLS AND FRACTURES IN PATIENTS WITH SYSTEMIC SCLEROSIS

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Background: Systemic sclerosis (SSc) is a severe connective tissue disorder causing vascular, immune, and fibrotic changes in the skin and internal organs. Patients with SSc may have an increased risk of osteoporosis (OP) and fractures due to a chronic inflammatory state, latent malabsorption or malnutrition, immobilization, and use of corticosteroid therapy.

Objectives: to determine the frequency of low mineral density, falls, low energy fractures and 10-year probability of new fracture in patients with SSc.

Methods: 191 patients with SSc were enrolled in the study: 160 women (mean age 51±13 yrs), among them 107 postmenopausal, and 31 men (mean age 53±14 yrs). Bone mineral density (BMD) was measured at lumbar spine (LS), femoral neck (FN) and total hip (TH) by dual energy X-ray absorptiometry (DXA, Hologic 4500A). BMD decreasing grade was determined in accordance to WHO criteria. All patients were interviewed on a special questionnaire and assessment of 10-year probability of new fracture by FRAX[®] was performed. Vitamin D level was measured in 104 patients.

Results: Low BMD was found in 68% women and 55% men: osteopenia in 30% and 32% and OP in 38% (21% in reproductive age and 50% in postmenopausal) and 23%, respectively. BMD in LS, FN and TH associated with body mass index (r=0.3, p=0.030; r=0.41, p=0.0027; r=0.49, p=0.0002, respectively) and duration of postmenopausal period in women (r=-0.56, p=0.023; r=-0.66, p=0.006; r=-0.63, p=0.009, respectively). BMD of LS and FN correlated with age (r=-0.22, p=0.045; r=-0.23, p=0.016, respectively), duration of SSc (r=-0.32, p=0.037; r=-0.31, p=0.046, respectively), glucocorticoid cumulative dose for LS only (r=-0.31, p=0.024). Mean 25(OH)D level was 19.83±11.06 ng/ml. Normal vitamin D level had 9% of SSc patients. No correlation between BMD and 25(OH)D level was found.

46 (24%) patients reported falls in the year prior to the interview, among them in 2 patients the fall led to fracture. A total of 48 (25%) patients had low energy fractures in the past, among them 8 (4%) of women had two or more fractures. Frequency of low energy fractures was 35%, 8% and 25% among postmenopausal women, women of reproductive age and men, respectively. The mean age at which the fracture occurred was 55±11 yrs. in postmenopausal women, 30±12 yrs. in young women and 60±12 yrs. in men. The most frequent were the fractures of distal forearm and vertebrae: 13 (7%) and 26 (14%) patients, respectively. Five patients each (3%) had ankle and humerus neck fractures and 7 – other localizations. Nobody reported the fracture of the proximal femur. Patients with abnormal BMD (OP or osteopenia) had a risk of falls and low energy fractures more than 2 times higher than patients with normal BMD (OR 2.93 [95% CI 1.11; 8.01], p=0.016 and OR 2.58 [95% CI 1.04; 6.6], p=0.025, respectively). 10-year probability of any major osteoporotic fracture was 18.4 ± 9.6% in women and 9.7 ± 8.6% in men and of hip fracture - 3.5 ± 3.7% and 1.5 ± 3.9%, respectively. Among all patients, 55% of women and 4% of men had a high risk of subsequent fractures using the FRAX[®] algorithm.

Conclusion: Low BMD was diagnosed in 68% of women and 55% of men with SSc. The correlations between BMD and age, body mass index, the duration of postmenopause were revealed. Additionally, we found associations with risk factors related to the disease itself: disease duration and glucocorticoid cumulative dose. Reduced BMD was associated with an increased risk of falls and fractures. 55% of women and 4% of men had a high risk of further fractures.

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FRI0351 DISTINCTION BETWEEN TELANGIECTASIA SECONDARY TO CONNECTIVE TISSUE DISEASE AND CUTANEOUS COLLAGENOUS VASCULOPATHY

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Background: Cutaneous collagenous vasculopathy (CCV), a recently described, rare, superficial dermal microangiopathy of unknown etiology, shares clinical and histopathological features in common with cutaneous lupus erythematosus and dermatomyositis. Fewer than 50 cases have been reported in the literature. Concurrence of CCV and connective tissue diseases (CTD) may present a diagnostic quandary.

Objectives: To determine whether CCV cases at our center were associated with CTD and to identify distinguishing features between CCV and cutaneous manifestations of CTD.

Methods: The laboratory information system at a single academic health care center (2000-2018) was searched to identify cases with a diagnosis of CCV. All pathological material and clinical charts were reviewed. The demographic, clinical and pathological data were documented.

Results: We identified 6 cases of CCV (4 female, 2 male; median age 57 years). Clinically, a telangiectatic eruption was present in all patients, affecting the lower limbs and other sites (N= 5) and the abdomen and arms (N=1). All patients had been seen by a dermatologist and 3 by a rheumatologist. A concurrent CTD was present in 2 cases; Sjogren's syndrome (N=1) and undifferentiated connective tissue disease (N=1). In both, the clinical differential diagnosis included CCV and cutaneous involvement by the CTD. In all 6 cases, histopathological examination revealed a non-inflammatory vasculopathy, with PAS-positive, diastase resistant, hyaline thickening of ectatic superficial capillary walls, and loss of pericytes. These hallmarks of CCV excluded skin disease attributable to CTD in the two relevant cases.

Conclusion: Concurrence of CCV and CTD, though uncommon, presents a diagnostic challenge. The clinical distribution of the CCV eruption, favouring the lower limbs, and its non-inflammatory character histopathologically distinguish it from cutaneous involvement by a CTD.

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