AB0667 CLINICAL AND THERAPEUTIC DIFFERENCES BETWEEN POLYMYOSITIS AND DERMATOMYOSITIS IN A COLOMBIAN COHORT WITH IDIOPATHIC INFLAMMATORY MYOPATHY

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Background: The idiopathic inflammatory myopathies (IIM) are a group of immune-mediated systemic conditions characterized by chronic muscle inflammation, resulting in muscle weakness (1).

Objectives: To characterize the disease in a Colombian cohort with idiopathic inflammatory myopathy, assessing differences in its classification, cutaneous and systemic manifestations, laboratory results, and therapeutic approach, according to the type of myopathy.

Methods: A cross-sectional study was conducted in 112 patients, in whom sociodemographic, clinical and therapeutic characteristics were analyzed based on the type of myopathy. Statistical association was examined by means of Chi-square tests, Mann-Whitney test, and logistic regression analyses.

Results: From the 112 patients recruited, 59 had polymyositis (PM) and 53 had dermatomyositis (DM). The patients were classified with Peter & Bohan criteria as: "definite" diagnosis 67 (60%), "probable" 35 (31%) and "possible" 9 (10%). A high proportion of males were found in this cohort. Our most notable findings are listed in Table 1, noticing for this population a high rate of polyautoimmunity associated to PM (OR 3.81 95%IC 1,003–14,53) and an association between ANAs antibodies positivity and DM (OR 7.03 95%IC 2,16–22,9). Patients with PM presented higher values of CK, LD and transaminases. Also, according to the therapeutic approach, PM was positively associated with the use of azathioprine and immunoglobulins (OR 2,59 95%IC 1,18–5,69 and OR 3,21 95%IC 1,19–8,19, respectively), while chloroquine and hydroxychloroquine were mainly used in DM patients.

Table 1. Sociodemographic and clinical characteristics of Colombian patients with idiopathic inflammatory myopathy

	Polymyositis N=59		Dermatomyositis N=53		p-value
	N	%	N	%	
Female	41	69,5	35	66	0,69
Age (mean)		54,3		49,4	0,09
Clinical characteristics					
Symmetrical muscle weakness	59	100	51	96,2	0,13
Gotttron's papules	0	—	49	92,4	<0,0001
Heliotrope rash	0	—	35	66	<0,0001
Shawl sign/ V sign	0	—	29	54,7	<0,0001
Polyautoimmunity	11	18,6	3	5,6	0,03
Muscle enzymes in serum					
CK (median)	3825		1012		0,006
LD (median)	554		433		0,003
ALT (median)	87		46,3		0,01
AST (median)	72		38		0,005
Aldolase	8	13,5	5	9,4	0,43
Autoantibodies					
ANA (+)	27	45,7	38	71,7	0,0006
Anti-Jo1 (+)	2	3,4	2	3,8	0,74
EMG					
Myopathic changes	41	69,5	27	50,9	0,34
Biopsy-proven myopathy					
Positive	30	50,8	16	30,2	0,7

SES, socioeconomic status; CK, creatine phosphokinase; LD, lactate dehydrogenase; AST, aspartate transaminase; ALT, alanine transaminase; EMG, electromyogram.

Conclusions: In this Colombian sample, a high proportion of patients were classified as definite diagnosis, high frequency of male-gender compromise, low association with cancer, and low prevalence of articular, pulmonary and cardiac involvements were found.

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AB0668 CALCINOSIS CUTIS AND ITS ASSOCIATION OF SERUM LEVELS OF OSTEOCALCIN, OSTEOCALCIN, NITRIC OXIDE AND TGF- β IN SYSTEMIC SCLEROSIS

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Background: Systemic sclerosis (SSc) is characterized by fibrosis, autoimmunity and vasculopathy. ES is classified subtypes: diffuse (dSSc) and limited (lSSc). More than 35% develop calcinosis. Calcium, phosphorus, parathormone, vitamin D, TGF- β , nitric oxide and osteonectin and osteopontin involved in bone mineralization.

Objectives: The aim was to compare osteonectin (ON), osteopontin (OP), TGF- β , nitric oxide (NO), Parathormone (PTH), Vitamin D and minerals concentrations in ES with and without calcinosis.

Methods: Cross-sectional study in ES patients (ACR criteria). We quantified OP, ON, TGF- β , ON, Calcium, Phosphorus, PTH and vitamin D in serum by ELISA. We performed descriptive statistics, Student t, Pearson correlation (significance $p < 0.05$) in SPSSv21 program.

Results: We included 71 patients, age 52.94 (± 11.47); 28 (40%) with calcinosis (18 dSSc/10 lSSc), and 43 (60%) without calcinosis (13 dSSc/30 lSSc). Biochemical parameters between two groups. In the whole population the higher PCR had moderate positive correlation ($r = 0.41$, $p = 0.042$), and serum calcium level had a moderate negative correlation ($r = -0.47$, $p = 0.021$); ON increased in direct relation to OP ($r = 0.3$, $p = 0.014$) and the serum levels of VitD had lower indirect relation with the evolution time of SSc ($r = -0.28$, $P = 0.025$) and the ON increase in direct relation to serum creatinine ($r = 0.039$, $p = 0.006$).

Conclusions: Patients with a longer time evolution of SSc have less serum levels of VitD and those with higher inflammation (PCR) have a higher TGF- β than a potent inducer of fibrosis. PCR and TGF- β have a moderate direct correlation, PCR and Calcium have moderate indirect correlation, ON and OP have a low direct correlation, VitD and evolution of diseases (years) had a low indirect correlation and NO and creatinine had a very low direct correlation.

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AB0669 THE EULAR SYSTEMIC SCLEROSIS IMPACT OF DISEASE (SCLEROID) SCORE – A NEW PATIENT-REPORTED OUTCOME MEASURE FOR PATIENTS WITH SYSTEMIC SCLEROSIS – PRELIMINARY RESULTS FROM THE ONGOING VALIDATION STUDY

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Background: Patient reported outcome measures (PROM) are required as key outcomes in therapeutic trials in systemic sclerosis (SSc). Given the unmet need of a validated, comprehensive PROM in SSc, the Scleroid questionnaire was developed by a team of patients with SSc and medical experts in the field. This is intended as a brief, disease-specific, patient-derived, disease impact score for scientific and clinical use in SSc.

Objectives: To present a preliminary analysis from the ongoing Scleroid validation study.

Methods: This EULAR-endorsed project involves 11 European centers special-

Abstract AB0669 – Table 1

ScleroID weighting (out of a total of 100 points)	Raynaud phenomenon	Hand function	Upper gastrointestinal tract symptoms	Pain	Fatigue	Lower gastrointestinal tract symptoms	Limitations of life choices and activities
Median (IQR)	15 (20)	10 (15)	6 (10)	10 (10)	10 (10)	5 (10)	5 (10)

ized in SSc. Patients fulfilling the ACR/EULAR 2013 criteria were prospectively included since 05/16 in the ongoing observational cohort study. Patients filled in the ScleroID questionnaire (Figure 1), as well as selected comparators SHAQ, EQ5D, SF36. Additionally, they weighted the 10 dimensions of the ScleroID by distributing 100 points according to the perceived impact on their health. The final score calculation will be based on the ranking of the weights. The study includes a reliability arm (follow-up questionnaire 7–10 days from baseline), as well as a longitudinal arm, looking at sensitivity to change at follow-up visits after 6 and 12 months from baseline.

Results: As of 01/2017 the study cohort included 224 patients with valid baseline data, 44 also had a reliability visit and 6 a 6-months follow-up visit. 84.4% of patients were female, 54.4% had limited SSc, median age 58, and median disease duration 8 years. The highest preliminary median weights for ScleroID domains were for Raynaud, impaired hand function, fatigue and pain (Table 1). Except for pain, these dimensions were also scored most highly in the ScleroID questionnaire at baseline.

The EULAR Scleroderma Impact of Disease Score (ScleroID)

How much have the different aspects of systemic sclerosis affected you during the last week? Please mark your responses on the scale by choosing the appropriate number for each of the following dimensions:

Raynaud's phenomenon:
Circle the number that best describes the severity of your Raynaud's phenomenon during the last week:
None 0 1 2 3 4 5 6 7 8 9 10 Extreme

Hand function:
Circle the number that best describes your hand function limitations due to your systemic sclerosis during the last week:
No limitation 0 1 2 3 4 5 6 7 8 9 10 Extreme limitation

Upper gastrointestinal tract symptoms (e.g. swallowing difficulties, reflux, vomiting):
Circle the number that best describes the severity of your upper gastrointestinal tract symptoms due to your systemic sclerosis during the last week:
None 0 1 2 3 4 5 6 7 8 9 10 Extreme

Pain:
Circle the number that best describes the pain you felt due to your systemic sclerosis during the last week:
None 0 1 2 3 4 5 6 7 8 9 10 Extreme

Fatigue:
Circle the number that best describes the impact of overall fatigue due to your systemic sclerosis during the last week:
None 0 1 2 3 4 5 6 7 8 9 10 Extreme

Lower gastrointestinal tract symptoms (e.g. bloating, diarrhea, constipation, anal incontinence):
Circle the number that best describes the severity of lower gastrointestinal tract symptoms during the last week:
None 0 1 2 3 4 5 6 7 8 9 10 Extreme

Limitations of life choices and activities (e.g. social life, personal care, work):
Circle the number that best describes how severe the limitations of life choices and activities due to your systemic sclerosis were during the last week:
None 0 1 2 3 4 5 6 7 8 9 10 Extreme

Body mobility:
Circle the number that best describes how much your body mobility was affected due to your systemic sclerosis during the last week:
Not affected 0 1 2 3 4 5 6 7 8 9 10 Extremely affected

Breathlessness:
Circle the number that best describes how severe your breathlessness due to systemic sclerosis was during the last week:
None 0 1 2 3 4 5 6 7 8 9 10 Extreme

Digital ulcers:
Circle the number that best describes how much your digital ulcers affected you overall during the last week:
None 0 1 2 3 4 5 6 7 8 9 10 Extreme

Conclusions: The EULAR ScleroID score is a novel tool designed for use in clinical practice and clinical trials to display the disease impact of SSc. In this preliminary analysis, Raynaud syndrome, impaired hand function, and fatigue were the main patient reported drivers of disease impact, however, further recruitment and validation of this new instrument is ongoing.

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AB0670 CLINICAL AND LABORATORY MANIFESTATIONS OF RNP-POSITIVE PATIENTS WITH SYSTEMIC SCLEROSIS

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Background: Among patients, who meet the classification criteria for systemic sclerosis (SSc), there are the patients without specific SSc anti-nuclear antibodies and who are positive for antibodies to ribonucleoprotein (RNP). The clinical significance of the RNP antibodies in SSc is not clear.

Objectives: To investigate clinical and laboratory characteristics of SSc patients positive for antibodies to RNP.

Methods: The study included 52 patients (49 women and 3 men, mean age 44.2±15 years, median disease duration from the beginning of Raynaud's phenomenon 11.2±7 years) who met the classification criteria of SSc (ACR/EULAR 2013) and have high level (>200 U/ml) of antibodies to RNP-70 (determined by ELISA, range 0–25 U/ml). Clinical, laboratory, and immunologic data were collected on all patients.

Results: All the patients met the criteria for classification of SSc ACR/EULAR 2013 (table 1). Other prevalent clinical manifestations were gastroesophageal reflux in 63, 5% of patients, joints involvement in 58%, myositis in 30, 7%, serositis in 27.5% (including 23% of pericarditis). All patients were positive for antinuclear factor (Hep-2) and in some cases RF, anti-SS-A, anti-SS-B, anti-Sm and anti-dsDNA antibodies were identified. The increased levels of CRP and/or ESR were found in 56% of cases. Among the SSc patients, there were 38 (73%) who satisfied for the classification criteria for mixed connective tissue disease (MCTD) proposed by Kasukawa et al. (1987).

Table 1. Frequency of items of SSc classification criteria (ACR/EULAR 2013) in the group under study

Items	52 patients
Skin thickening of the fingers extending proximal to the metacarpophalangeal joints	9 (17,3%)
Skin thickening of the fingers:	41 (78,8%)
puffy fingers	29 (55,7%)
sclerodactyly	14 (27%)
Fingertip lesions	22 (42,3%)
Telangiectasia	25 (48%)
Abnormal nailfold capillaries	45 (86,5%)
Lung involvement:	37 (72%)
Pulmonary arterial hypertension	2 (4%)
Interstitial lung disease	35 (68%)
Raynaud's phenomenon	52 (100%)
SSc-related autoantibodies:	12 (23%)
ACA	4 (7,7%)
Anti-Scl-70	8 (15,4%)

Conclusions: In the group under study the limited form of SSc dominated, with high frequency of the different manifestations of vasculopathy, esophagus, joint and muscle disorders symptoms. A subtype of RNP-positive SSc patients often met the criteria for MCTD. This fact makes it reasonable to discuss the possibility of overlap-syndrome between these two diseases.

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AB0671 INCREASED BODY FAT BUT DECREASED LEAN BODY MASS AND BONE MINERAL DENSITY IN PATIENTS WITH IDIOPATHIC INFLAMMATORY MYOPATHIES ARE ASSOCIATED WITH DISEASE DURATION, INFLAMMATORY STATUS, SKELETAL MUSCLE INVOLVEMENT AND PHYSICAL ACTIVITY

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Background: Idiopathic inflammatory myopathies (IIM) are characterized by inflammation and atrophy of skeletal muscles, pulmonary and articular involvement, which limit the mobility/self-sufficiency of patients, and can have a negative impact on body composition.

Objectives: To assess body composition and physical activity of IIM patients and healthy controls (HC).

Methods: 54 patients with IIM (45 females/9 males; mean age 57.3; disease duration 5.8 years; polymyositis (PM,22)/dermatomyositis (DM,25)/necrotizing