AB0643 CLINICAL AND ANALYTICAL DESCRIPTION OF A DERMATOMYOSITIS SERIES OF PATIENTS

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Background: Dermatomyositis (DM) is an idiopathic inflammatory myopathy. The recent years has increased its knowledge thanks to best characterization of myositis-specific antibodies that correlate with different clinical phenotypes.with different clinical phenotypes.

Objectives: To describe the clinical and analytical features of a series of dermatomyositis: clinical debut, clinical manifestations as well as the treatments received and the evolution of the disease.

Methods: Patients diagnosed of dermatomyositis in a tertiary hospital between the years 1978-2018 according to the criteria of Bohan and Peter (1975) and according to Dalakas' classification criteria (2015). Clinical, analytical and immunological profile data were collected, as well as treatments received and the evolution of the disease.

Results: A total of 59 inflammatory myopathies diagnosed between the years 1985 and 2018 were included. 46 were dermatomyositis (78%), 9 polymyositis (15%) and 4 necrotizing myositis (7%). 69% were women and 22% were smokers. Clinic started at 54 ± 17 years. The initial manifestation was pulmonary in 26.7% followed by cutaneous manifestations (24.4%) and the muscular (22.2%), while a 17.8% started skin and muscular manifestations at the same time. 45.5% behaved like a myopathic DM while a 28% as amyopathic DM and antisynthetase syndrome respectively. 38% presented interstitial involvement being the most common non-specific interstitial pneumonitis (76%)followed by usual interstitial pneumonitis (17%). In these patients,

DLCO was decreased (mean of 58.9% and 13.2) as well as the FVC (average of 58%, 2.5L). 73.3% presented cutaneous involvement being the most common manifestations the Gottron papules (37.8%) and the heliotrope rash (35.6%), and up to 27% had cuticular affectacion. 64% had muscle involvement, afecting proximal and symmetrical. Neck flexors were affected in a 38% of patients while 20% had dysphagia.Only 3 patients presented dysphonia (7%) and 2 myocarditis

Analytical data

Muscular enzyimes	
CK	821 ± 1260U/mL
LDH	425 ± 300U/L
Aldolase	11.5 ± 11U/L
Acute phase reactants	
ESR	30 ± 23 mm
RCP	13 mg/L ± 25
Myositis specific antibodies	
AntiJo1	27%
MDA5	18%
SAE1/2	15%

Conclusion: 80% received corticotherapy at a dose of mg/kg/day and 20% required high doses of metilprednisolone due to muscular involvement or pulmonary. 18% immunoglobulins and 11% cyclophosphamide. As maintenance 80% received disease modifiying antirheumatic drug in addition to corticosteroid therapy in descending doses (azathioprine 22%, dolquine 15%, tacrolimus 13.3%, rituximab 11%) due to muscle (29.9%), cutaneous (24.4%) and pulmonary involvement (22.2%). As complications, 2 cases of the syndrome were registered hemophagocytic Virtually all patients they presented pulmonary hypertension (mean 34 ± 12 mmHg). 5 patients (11.1%) were diagnosed with neoplasia, two of them after the diagnosis of DM. The mortality was 24%. 3 patients died due to a rapidly progressive pneumonitis, another 2 due to alveolar hemorrhage and three of them due to complication of the neoplastic disease.

Dermatomyositis occurs in a variable way, with predominance of pulmonary, skin and muscle manifestations. They require corticotherapy and immunosuppressive treatment for maintenance, even so, mortality is high.

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AB0644 RESULTS OF A TRAINING COURSE FOR CALCULATION OF THE MODIFIED RODNAN SKIN SCORE IN SCLERODERMA

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Background: Scleroderma is a connective tissue disease that is characterized by fibrosis of the skin.The modified Rodnan Skin Score (mRSS) is a measure generally used to assess the skin thickness in patients with scleroderma.Data on the effectiveness of the mRSS training courses differ in the literature.

Objectives: The objective of our study was to evaluate the effectiveness of the mRSS training course in rheumatology fellows in the rheumatology departments.

Methods: The study included 6 fellows from the departments of rheumatology.Participants were given a 1-hour-long theoretical training, including dermal involvement, and mRSS assessment by 3 rheumatology experts experienced in scleroderma, which was followed by an applied training on 4 patients for one-hour.Participants scored two patients before and after training on a form, which included 17 domains with a total score-range between "0" and "51". Then using the SPSS15 software program, inter-rater reliability was assessed with intraclass correlation(**ICC**) analysis for both pre- and post-training mRSS.Fleiss' kappa was used to measure the degree of agreement according to 12 Rodnan score areas before and after the training.

Results: The ICC value for pre-training and post-training total Rodnan scores was 0.867 (95% CI-0.625-1.00, P=0.05), and 0.905(95% CI 0.045-1.00, P=0.02), respectively. Individual analysis of score areas showed that after the training there was an increase in degree of agreement in some of these areas, while there was no difference in one area, and it decreased in others (Table 1).

Table 1: The ICC value and percentage of agreement for pre-training and post-training total Rodnan scores

	Pre-training		Post-training	
Body area	Fleiss's Kappa (%95 CI)	Agreement, %	Fleiss's Kappa (%95 GA)	Agreement, %
Right fingers	0.38 (0.003, 0.73)	**53.3**	0.60 (-0.18, 1.00)	**70.0**
Left fingers	0.42 (0.16, 0.68)	56.6	0.29 (0.29, 0.29)	46.6
Right hand	0.02 (0.02, 0.02)	**26.6**	0.11 (-0.06, 0.29)	**33.3**
Left hand	0.16 (-0.11, 0.42)	***36.6***	0.16 (-0.11, 0.42)	***36.6***
Right forearm	0.29 (0.29, 0.29)	46.6	0.02 (0.02, 0.02)	26.6
Left forearm	0.29 (0.29, 0.29)	46.6	0.02 (0.02, 0.02)	26.6
Right upper arm	0.11 (-0.06, 0.29)	33.3	-0.22 (-0.11, 0.06)	23.3
Left upper arm	0.24 (0.16, 0.33)	43.3	0.02 (0.02, 0.02)	26.6
Face	0.24 (0.16, 0.33)	43.3	0.02 (0.02, 0.02)	26.6
Anterios chest	0.24 (0.16, 0.33)	43.3	0.16 (-0.11, 0.42)	36.6
Abdomen	0.16 (-0.11, 0.42)	36.6	0.11 (-0.06, 0.29)	33.3
Right thigh	0.02 (0.02, 0.02)	**26.6**	0.29 (0.29, 0.29)	**46.6**
Left thigh	0.02 (0.02, 0.02)	**26.6**	0.20 (-0.50, 0.90)	**40.0**
Right leg	0.38 (0.003, 0.73)	53.3	0.24 (0.16, 0.33)	43.3

Conclusion: Several studies have demonstrated the applicability, reliability, and validity of mRSS, a measure of dermal involvement, and that evaluation of the score requires experience, and an attentive learning process.In the literature, the inter-rater ICC values during previous training courses are reported to range between 0.378 and 0.92(1). These studies show differences in terms of the number and experience of participants,

number of patients, course-length, and repeated courses. Similar to the observation by Ionescu *et al*(2), the pre-training inter-observer ICC value was very high(0.867), and it increased to 0.905 after the training. Presence of an increase in some areas, and reduction in some others in individual analysis of areas after the training may suggests the need for repeated training. Our limitations were low number of participants and patients. We obtained very good inter-rater ICC values in the mRSS training course given to rheumatology fellows; however we may benefit from new studies for optimization of these conclusions by increasing the numbers of trainees, patients, and the lenght of courses.

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AB0645 CANNABINOIDS IN THE TREATMENT OF PAIN RELATED TO SYSTEMIC SCLEROSIS SKIN ULCERS: OUR EXPERIENCE

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Background: Skin ulcers (SU) represent one of the most frequent complications of Systemic Sclerosis (SSc), characterized by severe chronic pain and frequent complications. Pain related to SSc SU(SSU) remains yet an area of significant unmet need. Moreover, pain control is fundamental for the wound care procedures in SSc patients (pt), increasing treatment adherence and compliance to skin ulcers dressing changes. The pain relief provided by standard therapy (i.e. NSAIDs, tramadol) is often inadequate or dose limited by side effects. Opioids currently are the mainstay of SSU pain treatment but burdened by side effect profile and/or ineffective. Thus, novel analgesic strategies need to be investigated. Cannabidiol (CBD), one of many constituents of the Cannabis sativa, has received renewed interest in the treatment of numerous pathological conditions.
Objectives: Evaluate our experience to define the efficacy of CBD preparation in patients with SSU.
Methods: 25 SSc pt (F/M 22/3, mean age 52.3 ± 12.9-SD-years), referred to our Scleroderma Unit during 2018, were consecutively included. In all pt the disease was complicated by long-standing, painful SU resistant to opioids. Pain was classified as severe, according to WHO guidelines in all subjects. 25/25 pt carried out systemic (calcium-channel blockers, prostanoids and/or anti-ET receptors) and local (debridement and dressing) therapies. The CBD (10% oral administration oil) was used daily for the treatment of SSU-related pain. We performed both an oral (five drops bid) as local treatment (two drops in the site of SSU) during surgical debridement of SSU for a period of 5.9 ± 3.2 SD months. Patients have been provided with a diary to record the following symptoms daily: self-evaluation of pain at the same time in the evening, using a visual analog scale (VAS), use of other analgesics, eventual side effects. Health Assessment Questionnaire-Disability Index (HAQ-DI) was administrated baseline and at the end of treatment. Safety of CBD was evaluated by patient's records of side effects, while vital signs and laboratory parameter variations were monitored at each weekly medication.
Results: The local treatment with CBD produced a significant reduction of SSU-related pain. After 1 month of therapy, pain VAS decreased from 94.8 ± 8.72 SD to 54.7 ± 9.4 SD (P<0.0001), total hours of sleep increased from 2.56 ±1.28 SD to 5.67 ± 0.85 SD (P <0.000). Additional analgesic therapy was necessary in 12/25 (48%). After 2 months, further clinical improvement was observed: the pain VAS reduced to 40.9 ± 12.9 SD, the mean total hours of sleep per night was 6.10 ± 0.85 SD and the HAQ-DI decreased from 1.1 ± 0.67 SD (baseline) to 0.46 ± 0.46 SD at the last patients' evaluation, when complete healing of SSU and pain relief were obtained and CBD was discontinued. 20/25 (80%) pt registered a better compliance to the local wound management.
No reported significant side effects with CBD oil.
Conclusion: Our study suggests that the use of CBD as a local therapy is effective and safe in maintaining analgesia in patients with SSU; not secondarily it could be essential for an adequate healing of a local wound with consequent improvement of SSc patients' quality of life and compliance on local SSU management. Further larger-scale studies will be needed to finally demonstrate CBD efficacy and to monitor long-term effects.
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AB0646 HEALTHCARE UTILIZATION AMONG INCIDENT CASES OF SYSTEMIC SCLEROSIS: RESULTS FROM A POPULATION-BASED COHORT (1988–2016)

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Background: Systemic sclerosis (SSc) is an autoimmune disorder associated with multi-organ dysfunction including but not limited to vascular, cardiac and pulmonary involvement. Few studies have estimated the healthcare resource usage of patients with SSc.
Objectives: To compare healthcare utilization among incident cases of SSc vs age- and sex-matched comparators.
Methods: This study utilized a retrospective, population-based cohort of physician-diagnosed patients with SSc in a geographically well-defined area from Jan 1, 1988 to Dec 31, 2016. A 2:1 cohort of age- and sex-matched non-SSc subjects from the same population base was randomly selected for comparison. Inpatient and outpatient utilization data were obtained from the Rochester Epidemiology Project beginning 12 months prior to the SSc incidence/index date. Patients were followed until death, migration from Olmsted County, or December 31, 2017. A maximum of 5 years following the incidence/index date was used for analysis and the follow-up of each matched triple was further truncated at the shortest length of follow-up for any member, to ensure similar periods of observation for SSc cases and non-SSc comparators. Services were summarized as visit-days (number of days at least one service in the category was billed) to avoid overestimation of services provided. Utilization was compared between SSc and non-SSc cohorts using negative binomial models.
Results: The cohort included 69 incident SSc cases and 138 non-SSc comparators (mean age of 57 ± 16 years at diagnosis/index, 90% female for both cohorts; 87% [SSc] and 95% [non-SSc] Caucasian). Patients with SSc had the highest utilization of outpatient physician, laboratory and combined radiology visit-days during the year of the SSc diagnosis compared with the year prior to diagnosis or years 1-4 after diagnosis of SSc (Table). Patients with SSc had higher utilization of outpatient physician, laboratory and combined radiology visit-days annually for the year prior to diagnosis of SSc and for each of the first 5 years after diagnosis of Ssc compared to patients without SSc. Rate ratios comparing utilization in patients with and without SSc ranged from 1.8 to 3.0 for all comparisons.
Conclusion: A higher utilization of outpatient physician, laboratory and radiology visits was observed among patients with SSc compared to non-SSc subjects throughout 5 years of disease duration, indicating high and continued care needs in this patient population.

Table. Comparison of outpatient visits, labs and imaging visit-days in patients with and without incident SSc

Services	Time Interval (years)	Number of patients, SSc/Non-SSc	SSc Median visit-days (IQR)	Non-SSc Median visit-days (IQR)	Rate Ratio (95% CI)
Outpatient visits	-1-0	69/138	4 (1–9)	2 (0–6)	1.8 (1.2–2.7)
	0-1	69/138	7 (4–12)	3 (0–5)	2.4 (1.7–3.3)
	1-2	66/132	4 (2–11)	2 (0–4)	2.0 (1.4–2.9)
	2-3	54/108	4 (2–10)	3 (0-5)	1.9 (1.4–2.7)
	3-4	48/96	4 (2–10)	3 (1–5)	1.9 (1.3–2.6)
	4-5	43/86	5 (2–12)	3 (0–6)	2.1 (1.4–3.0)
Labs	-1-0	69/138	3 (0-8)	1 (0-3)	2.1 (1.4–3.3)
	0-1	69/138	4 (2-10)	2 (0–3)	3.0 (2.0–4.6)
	1-2	66/132	5 (1-8)	1 (0–3)	2.0 (1.3–3.2)
	2-3	54/108	3 (1-9)	1 (0–4)	2.6 (1.7–4.3)
	3-4	48/96	3 (1-10)	1 (0–3)	2.7 (1.7–4.1)
	4-5	43/86	3 (1-12)	1 (0–3)	2.9 (1.7–4.7)
Radiology	-1-0	69/138	2 (0–5)	0 (0–2)	2.1 (1.6–4.2)
	0-1	69/138	3 (1–6)	1 (0–3)	2.6 (1.8–3.9)
	1-2	66/132	2 (1–6)	0 (0–2)	1.9 (1.8–3.1)
	2-3	54/108	2 (1–4)	1 (0–2)	2.1 (1.3–3.1)
	3-4	48/96	2 (1–4)	1 (0–2)	2.0 (1.3–3.1)
	4-5	43/86	2 (1–7)	1 (0–2)	2.6 (1.6–4.2)

IQR: interquartile range; CI: confidence interval