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AB0676

BODY COMPOSITION IN MYOSITIS PATIENTS AND THE ASSOCIATION WITH DISEASE SPECIFIC FEATURES, PHYSICAL ACTIVITY AND PLASMA LEVELS OF INFLAMMATORY CYTOKINES

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Background: Skeletal muscle, pulmonary, and articular involvement in idiopathic inflammatory myopathies (IIM) 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) and the association with selected inflammatory cytokines in IIM.

Methods: 54 patients with IIM (45 females; mean age 57.7; disease duration 5.8 years; PM, (22)/DM, (25)/IMNM, (7)) and 54 age-/sex-matched HC (45 females, mean age 57.7) without rheumatic/tumor diseases were included. PM/DM patients fulfilled Bohan/Peter criteria for PM/DM. We assessed body composition (densitometry: iDXA Lunar, bioelectric impedance: BIA2000-M), physical activity (Human Activity Profile, HAP questionnaire), disease activity (MITAX and MYOACT activity score), muscle involvement (manual muscle test, MMT-8 and functional index, FI2) and plasma levels of 27 cytokines (commercial multiplex ELISA kit, Bio-Rad Laboratories). Data are presented as mean±SD.

Results: Compared to HC, patients with IIM had a trend towards significantly increased body fat% (BF%) as assessed by iDXA (39.9±7.1 vs. 42.4±7.1 %, p=0.077), but significantly decreased lean body mass (LBM) as assessed both by iDXA (45.6±8.1 vs. 40.6±7.2 kg, p=0.001) and BIA (52.6±8.8 vs. 48.7±9.0 kg, p=0.023), and increased extracellular mass/body cell mass (ECM/BCM) ratio (1.06±0.15 vs. 1.44±0.42, p<0.001), reflecting deteriorated nutritional status and worse muscle predispositions for physical activity. Compared to HC, IIM patients had significantly lower bone mineral density (BMD: 1.2±0.1 vs. 1.1±0.1 g/cm², p<0.001). Disease duration negatively correlated with BMD (r=-0.392, p=0.004) and LBM-BIA (r=-0.272, p=0.047). Disease activity assessed by both MITAX and MYOACT positively correlated with LBM-BIA (MITAX: r=0.294, p=0.031; MYOACT: r=0.335, p=0.013) and LBM-DXA (MITAX: r=0.341, p=0.012; MYOACT: r=0.368, p=0.007), similarly as with basal metabolic rate (BMR; MITAX: r=0.336, p=0.014; MYOACT: r=0.351, p=0.010), and fat free mass (FFM; MITAX: r=0.338, p=0.014; MYOACT: r=0.356, p=0.009). CRP was positively associated with BF% assessed both by DEXA (r=0.276, p=0.035) and BIA (r=0.306, p=0.025). Higher BF%-DEXA was associated with worse physical endurance (FI2: r=-0.311, p=0.026) and worse ability to perform physical activity (HAP: r=-0.292, p=0.032). MMT-8 score negatively correlated with ECM/BCM ratio (r=-0.385, p=0.006). Plasma levels of IL-1ra positively correlated with body mass index (BMI, r=0.359, p=0.009), BF% (r=0.363, p=0.009), and visceral fat (VF, r=0.409, p=0.003). Plasma levels of MCP-1 positively correlated with ECM/BCM ratio (r=0.387, p=0.005). Increased plasma levels of IL-10 were associated with decreased LBM (r=-0.473, p=0.014).

Conclusion: Compared to healthy age-/sex-matched individuals we found significant negative changes in body composition of our IIM patients, which are associated with their disease activity and duration, inflammatory status, skeletal muscle involvement, and physical activity. Plasma levels of several inflammatory cytokines were associated with alterations of body composition in IIM patients and highlight the potential role of inflammatory status on impaired body composition during the course of IIM.

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AB0677

TREATMENT OF IDIOPATHIC INFLAMMATORY MYOPATHIES – A SINGLE CENTRE EXPERIENCE

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Background: There is no consensus or recommendations for the treatment of idiopathic inflammatory myopathies (IIM).

Objectives: We explored how we treated incipient IIM patients in our secondary/tertiary rheumatology centre.

Methods: We retrospectively included cases with IIM diagnosed between January 2010 and June 2018, who were followed at least 6 months. The remission inducing treatment and the treatment at the last follow-up were recorded.

Results: During the 102-month period we identified 102 IIM cases (71.6% female, median (IQR) age 62.7 (52.2–72.1) years): 27.5% dermatomyositis, 22.5% anti-synthetase syndrome, 14.7% myositis in an overlap syndrome, 13.7% immune mediated necrotizing myopathy, 11.5% polymyositis, 6.9% cancer associated myositis, 2.9% inclusion body myositis (IBM), 1% unspecified myositis. 94 (92.2%) patients received glucocorticoids (GC), 18/94 (17.6%) as monotherapy. The remaining 81 (79.4%) were treated with additional immunomodulatory agents (Table 1), most commonly methotrexate. In addition to therapy presented in Table 1, 13 (12.7%) also received human immunoglobulins (IVIG), one (1%) received plasma exchange therapy.

Of the 8 (7.8%) cases who were not treated with GC 3 received methotrexate, 1 rituximab, and 4 no additional treatment (1 paraneoplastic polymyositis received chemotherapy, 2 IBM, 1 mild polymyositis in elderly, multimorbid patient).

In the first 6 months following diagnosis, 11 patients died (3 due to infection, 2 as a consequence of IIM, 1 due to haemorrhagic shock, 5 of unknown causes), and 5 were lost to follow-up. The remaining 86 patients were followed for a median (IQR) 37.7 (20.2–62.6) months. At last follow-up 49 (57%) patients were still receiving GC, 8 (9.3%) in monotherapy. 9 (10.5%) cases were off GC and other maintenance treatment. Maintenance treatment at last follow-up is presented in Table 2. Treatment was changed in 13 (15.1%) patients (due to progression of pulmonary involvement in 3, active myositis in 4, pulmonary involvement progression and active myositis in 1 and due to intolerance to medication in 6 cases). In addition to therapy presented in Table 2, 2 (2.3%) patients received IVIG (1 concurrently with rituximab and mycophenolic acid due to recalcitrant anti-synthetase syndrome and 1 due to IBM complicated with dysphagia).

Table 1. Induction treatment of idiopathic inflammatory myopathy

Type of treatment	N (%)
Cyclophosphamide; median dose (range)	23 (22.5); 6 (2–12) g
Methotrexate	43 (42.2)
monotherapy	30 (69.8)
+ cyclosporine A	8 (18.6)
+ cyclosporine A and rituximab	4 (9.3)
+ chloroquine	1 (2.3)
Rituximab monotherapy	4 (3.9)
Azathioprine	3 (2.9)
Mycophenolic acid	3 (2.9)
Cyclosporine A monotherapy	1 (1.0)

Table 2. Maintenance treatment of idiopathic inflammatory myopathy

Type of treatment	N (%)
Methotrexate monotherapy	35 (40.7)
+ cyclosporine A	20 (57.1)
+ rituximab	11 (18.6)
+ chloroquine	2 (5.7)
Azathioprine monotherapy	13 (15.1)
Rituximab monotherapy	3 (3.5)
Mycophenolic acid monotherapy	15 (17.4)
+ tacrolimus	13 (86.7)
+ rituximab	1 (6.7)
Cyclosporine A	1 (1.2)
Tocilizumab*	1 (1.2)

* Anaphylactic reaction to rituximab, received tocilizumab due to pulmonary fibrosis progression as part of anti-synthetase syndrome

Conclusion: GC, methotrexate and cyclophosphamide (in case of pulmonary involvement) were the cornerstone treatment in our cohort.

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AB0678

ANTIPROLIFERATIVE AND VASOACTIVE TREATMENT MODALITIES IN 457 CONSECUTIVE PATIENTS WITH SYSTEMIC SCLEROSIS FROM ACADEMIC CENTERS IN GREECE

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Background: The pathophysiology of Systemic Sclerosis (SSc) encompasses autoimmunity, microvascular damage and fibrosis of the skin and internal organs. Accordingly, the two mainstays of treatment comprise antiproliferative and vasoactive regimens. So far there are limited data regarding the actual use of these agents in daily clinical practice.

Objectives: To describe current treatment modalities based on standard practice decisions according to published recommendations and/or guidelines for SSc, as well as to determine to which extent different disease modifying agents, namely antiproliferative and vasoactive drugs, are administered to these patients.

Methods: Consecutive patients from 7 academic rheumatology departments across Greece who fulfilled the 1980 American College of Rheumatology criteria for classification of SSc and were examined between January 2016 and December 2018 at least once, were included in the study. Patients' medical records were analyzed and antiproliferative and vasoactive agents administered anytime during the disease course were recorded.

Results: A total of 457 patients (87% women, 51% diffuse SSc, 62% with pulmonary fibrosis, 55% with digital ulcers, mean±SD age at diagnosis 49.2±13.1 years, disease duration 10.4±8 years) were studied. Methotrexate was the most frequent antiproliferative agent ever administered (53% of patients, 55% of them with diffuse SSc) followed by cyclophosphamide (26%, 78% of them diffuse), mycophenolate (11%, 63% of them diffuse), azathioprine (10%, 64% of them diffuse), rituximab (9%, 84% of them diffuse) and tocilizumab (3%, 68% of them diffuse). Among vasoactive agents, endothelin receptor antagonists (ERA) were most frequently ever administered in 39% of patients (64% with diffuse SSc) followed by iloprost in 7% (55% diffuse) and 5 phosphodiesterase (5PDE) inhibitors in 5.5% of patients (62% diffuse). Among all antiproliferative/vasoactive agents used, the greatest retention rate at last follow-up visit since initiation had ERA (82%) and 5PDE inhibitors (86%). Of note, 20% of SSc patients (90% women, 70% limited SSc, age at diagnosis 50.4±13.2

years, disease duration 8.4±6.4 years) had never been treated with either antiproliferative or vasoactive agents, whereas 41% had ever received only antiproliferative and 8% only vasoactive therapy. Moreover, 1/5 of patients with pulmonary fibrosis had never received antiproliferative treatment, whereas 1/3 of patients with digital ulcers had never received vasoactive drugs (Table).

Conclusion: About 20% of contemporary SSc patients, including patients with pulmonary fibrosis and digital ulcers, have never received antiproliferative or vasoactive therapy, possibly reflecting the spectrum of benign disease and/or a subgroup of under-treated patients.

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Table 1. Treatment modalities in consecutive SSc patients

	Combined antiproliferative and vasoactive agents	Only antiproliferative agents	Only vasoactive agents	Without antiproliferative and vasoactive agents
All patients n=457	31%	41%	8%	20%
Diffuse SSc n=233	43%	39%	7%	11%
Limited SSc n=224	17%	45%	11%	27%
Pts with Pulmonary fibrosis n=285	38%	40%	8%	14%
Pts with ulcers and/or pulmonary hypertension n=251	52%	23%	13%	12%

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AB0679

SKIN ULCERS IN SYSTEMIC SCLEROSIS: 10 YEARS' FOLLOW-UP OF 211 PATIENTS FROM A MONOCENTRIC COHORT IN THE NORTH EAST OF ITALY

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Background: Microvascular involvement plays a pivotal role in the pathogenesis of systemic sclerosis (SSc). Among vasculopathic manifestations, skin ulcers (SU) stand out for their high prevalence.

Objectives: The aim of this study was to evaluate the prevalence and the subtype of SU, the association with clinical phenotype and pharmacological approach in the cohort of scleroderma patients attending our tertiary referral Centre.

Methods: We retrospectively evaluated clinical records of patients affected with SSc according to ACR/EULAR 2013 criteria, attending the Rheumatology Unit of Padova University. Patients with SSc *sine scleroderma*, overlap syndrome and with a follow-up lower than 24 months were excluded. We evaluated the epidemiological and clinico-serological data, including autoantibody pattern, laboratory data (such as haemoglobin, ESR, CRP), organ involvement and pharmacological treatments. Prevalence of SU, type of occurrence (isolated, recurrent and persistent episodes) and site (digital and localized at the lower limbs, between knees and ankle-feet) were recorded. Statistical analysis was performed by SPSS v. 24.

Results: 211 patients were included in the study: 187 female and 24 male, aged 60.8 ±12.4 years, 147 (70%) with limited cutaneous and 64 (30%) with diffuse form. During the follow-up period of 120 months (50-216), 105 (50%) patients experienced at least one episode of SU: 74 experienced digital ulcers, 11 presented only SU at lower limbs and 20 both kinds of ulcers. Demographic, laboratoristic and clinical features of our cohort are shown in Table 1. At the multivariate analysis, diffuse cutaneous form, younger age at diagnosis, telangiectasias and joint involvement were independently associated to SU. Among 105 patients who experienced at least one episode of SU, 69 (66%) had recurrent/persistent ulcers. The latter group needed a combination of drugs, including: calcium-channel blockers (90%), low-dose aspirin (75%), prostanoids (85%) and/or endothelin receptor antagonists (58%); 27% requested opioids for analgesia. Concerning SU at lower limbs, 19/31 patients (61%) had macrovascular involvement and 14/31 (45%) persistent ulcers; 6/31 (19%) underwent autologous skin grafting.

Conclusion: In our cohort SU occur more frequently in SSc diffuse cutaneous form and are associated with visceral and articular involvement.