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

Stylianos Panopoulos¹, Maria Tektonidou¹, Katerina Chatzidionysiou¹, Alexandros Drosos², Stamatis-Nick Liossis³, Theodoros Dimitroulas⁴, Lazaros Sakkas⁵, Dimitrios Boumpas¹, Prodromos Sidiropoulos⁶, George Bertsis⁶, Paraskevi V. Voulgari², Dimitrios Daoussis³, Konstantinos Thomas¹, Georgios Georgiopoulos¹, Georgios Vosvotekas⁴, Dimitrios Vassilopoulos¹, Petros Sfrikakis¹. ¹Joint Rheumatology Program, National and Kapodistrian University of Athens Medical School, Athens, Greece; ²University of Ioannina, Medical School, Ioannina, Greece; ³University of Patras, Medical School, Patra, Greece; ⁴Aristotle University of Thessaloniki, Medical School, Thessaloniki, Greece; ⁵University of Thessaly, Medical School, Larissa, Greece; ⁶University of Crete, Medical School, Crete, Greece

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

Pamela Polito, Elisabetta Zanatta, Mara Felicetti, Maria Favaro, Mariagrazia Lorenzin, Roberta Ramonda, Franco Cozzi, Andrea Doria. Rheumatology Unit, Department of Medicine DIMED, University of Padova, Padova, Italy

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.

Digital ulcers tend to recur during the disease course requiring several drugs, while SU at lower limbs tend to be more persistent.

Table 1. Demographic, clinical and laboratory features of the whole cohort and of the 2 groups (patients with and without SSc).

Patients, n (%)	Total 211 (100)	SSc- 106 (50)	SSc+ 105 (50)	P value	OR (CI 95%)
Demographic characteristics					
Age at Raynaud's phenomenon, mean (SD) years	42.9 (14.2)	40.9 (13.4)	45.0 (14.4)	0.007*	
Age at SSc diagnosis, mean (SD) years	48.3 (13.3)	50.7 (12.4)	45.8 (13.7)	0.000*	
Diagnostic latency, mean (SD) years	1 (0-3)	1 (0-3)	1 (0-3)	0.496*	
Female, n (%)	187 (89)	98 (93)	89 (85)	0.079*	
Cutaneous subset, n (%)					
Diffuse	64 (30)	21 (20)	43 (41)		
Limited	147 (70)	85 (80)	62 (59)	0.001*	2.8 (1.5-5.2)
Autoantibodies, n (%)					
ACA	88 (42)	48 (45)	40 (39)		
Anti-Scl70	79 (38)	32 (30)	47 (45)		
Anti-RNAPolIII	11 (5)	7 (7)	4 (4)		
ANA without specificity	32 (15)	19 (18)	13 (12)	0.139*	
Laboratory exams, n (%)					
Anemia (Hb<12.5 g/dL)	43 (20)	12 (11)	31 (30)	<0.001*	3.7 (1.8-7.7)
CRP<6 mg/L	30 (14)	13 (13)	17 (16)	0.031*	2.1 (1.0-4.4)
ESR<20 mm/h	40 (20)	11 (11)	29 (28)	0.001*	3.4 (1.6-7.3)
Main disease manifestations, n (%)					
Lung involvement	70 (34)	25 (24)	45 (44)	0.002*	2.5 (1.4-4.5)
Heart involvement	33 (16)	9 (9)	24 (23)	0.004*	3.2 (1.4-7.4)
GI involvement	160 (76)	82 (78)	78 (75)	0.817*	
Renal involvement	8 (4)	3 (3)	5 (5)	0.496*	
Articular involvement	83 (40)	30 (29)	53 (51)	0.001*	2.5 (1.4-4.5)
Telangiectasia	101 (48)	34 (32)	67 (64)	<0.001*	3.0 (1.7-5.5)
Puffy fingers	40 (19)	41 (40)	37 (35)	0.027*	0.5 (0.3-0.9)
Proximal myopathy	81 (41)	11 (11)	70 (67)	<0.001*	10.1 (8.0-12.0)
Calcinosis	37 (24)	8 (8)	29 (28)	0.001*	4.3 (1.8-10.2)

SSc- patients with history of skin ulcers, SSc+ patients without history of skin ulcers, SD: standard deviation, OR: odds ratio, CRP: C reactive protein, ESR: erythrocyte sedimentation rate.

Hb: haemoglobin, G-I: gastro-intestinal.

* T test, * Pearson test, * Fisher test, * Mann-Whitney test.

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AB0680

NAIFOLD VIDEOCAPILLAROSCOPY PATTERN CORRELATES WITH ORGAN INVOLVEMENT AND DISEASE OUTCOME IN SYSTEMIC SCLEROSIS: A SINGLE-CENTER OBSERVATIONAL STUDY

Argyro Repa, George Bertisias, Nikolaos Kougkas, Nestor Avgustidis, Maria Terizaki, Paraskevi Kytonidou, Prodromos Sidiropoulos. *University Hospital of Heraklion, Department of Rheumatology, Clinical Immunology and Allergy, Heraklion, Greece*

Background: Systemic sclerosis (SSc) is characterized by systemic autoimmunity, vasculopathy and fibrosis. Nailfold Videocapillaroscopy (NVC) is a safe method to detect and to analyze morphological microvascular abnormalities in SSc patients. There is circumstantial evidence to suggest an association between NVC abnormalities and disease severity, particularly vascular manifestations.¹

Objectives: To evaluate the association of NVC patterns with demographic, clinical, laboratory features and disease progression in the SSc cohort of University Hospital of Heraklion.

Methods: Retrospective study (2011-2018) of SSc patients who underwent NVC. NVC findings were classified into three distinct patterns namely early, active and late according to *Cutolo et al*². NVC patterns were correlated with the clinical characteristics of the patients. We also correlated baseline NVC patterns with the progression of the disease at 3 years.

Results: 62 patients [(88.8% women, mean age of diagnosis 49.8 years, range 18-82)] were included. Compared to early/active pattern, patients with late pattern were more likely to have diffuse SSc (dSSc) (p=0.006), Interstitial Lung Disease (ILD) (80.0% vs 38.1% p=0.002) and esophageal involvement (80.0% vs 47.6% p=0.0034). There was a trend for increased frequency of Pulmonary Arterial Hypertension (PAH), digital ulcers (DU) and calcinosis in patients with late pattern (p=0.079, p=1.103 and p=0.079 respectively). Patients with active NVC pattern were more likely to have arthritis (early/active/late: 36.4%/75.9%/55.0%, p=0.054). A total 42 patients had NVC data at the time of SSc diagnosis. At 3 years there were no significant differences regarding disease progression, although patients with late NVC pattern had more hospitalizations due to infections (33.3% vs 9.1%, p=0.191), higher death rate (16.7% vs. 8.3%, p=0.543), need to use cyclophosphamide (33.3% vs 17.4%, p=0.391) and/or bosentan (40.0% vs 22.7%, p=0.426).

Conclusion: In this single-center SSc cohort, NVC pattern was found to correlate with organ involvement especially ILD, esophageal involvement

and dSSc. Serious infections and treatment intensification were also more frequent among patients with late NVC pattern. NVC may be a useful prognostic marker for SSc patients.

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AB0681

SCLERODERMIC HAND: SUBCLINICAL JOINT INVOLVEMENT?

Laura Belloli, Cinzia Casu, **Mariaeva Romano**, Elisa Verduci, Maria Di Cicco, Emanuela Schito, Valeria Campanella, Michel Chevallard, Marina Muscarà, Davide Antonio Filippini, Oscar Massimiliano Epis. *Grande Ospedale Metropolitano Niguarda, Milan, Italy*

Background: Systemic sclerosis (SSc) is a chronic autoimmune connective tissue disorder of not fully understood aetiology characterized by vascular and fibrotic abnormalities in the skin and internal organs. Musculoskeletal involvement, mainly affecting hands and feet, is frequent in SSc and represents a major cause of disability. Ultrasonography is a non-invasive, cost-effective imaging modality which is able to detect the presence of joint involvement in SSc.

Methods: 61 consecutive patients (60 women) were enrolled. Each patient has completed a dichotomous survey to evaluate the presence of joint symptoms (presence of pain, swelling, morning stiffness in the last 3 months). Subsequently a rheumatologist - blinded to clinical data and survey results - performed the ultrasound examination of hands and wrists to evaluate the presence of effusion, synovial hypertrophy, power-Doppler signal, erosions and tenosynovitis defined according to the OMERACT definitions. The ultrasound examination was performed using the Esaote MyLab70 with a multifrequency linear probe (6-18 MHz).

Results: Of the 61 patients analyzed, 29 showed joint involvement on the ultrasound examination (47.54%), in particular, 37.93% had tendon involvement and 72.41% joint involvement (Table 1). 31 patients did not report joint symptoms (50.82%) but 13 (41.93%) still had ultrasound alterations (Table 2).

Table 1. Musculoskeletal involvement in SSc

	n. pazienti (%)
Joint effusion	14/29 (48.28%)
Synovial hypertrophy	18/29 (62.07%)
Erosions	2/29 (0.69%)
Tenosynovitis	11/29 (37.93%)
Power-Doppler signal	9/29 (31.03%)

Table 2. Musculoskeletal involvement in asymptomatic patients

	n. pazienti (%)
Joint effusion	3/13 (23.08%)
Synovial hypertrophy	8/13 (61.54%)
Erosions	0/13 (0%)
Tenosynovitis	5/13 (38.46%)
Power-Doppler signal	3/13 (23.08%)

Conclusion: Musculoskeletal involvement is frequent in SSc and can be asymptomatic in more than 20% of patients. The ultrasound examination can make it possible to detect articular and periarticular involvement at an early stage of the disease before the onset of disability.

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