

Methods: We selected the NVCs of 273 patients affected by SSc (according to 2013 ACR criteria) who presented one of the validated NVC scleroderma patterns (81 with "Early" pattern, 84 with "Active" pattern, 92 with "Late" pattern, 16 with "scleroderma-like" pattern) (2). Among the 273 SSc patient with an established NVC scleroderma-pattern, a number of 54 subjects who had previously NVC analyses performed before the development of the scleroderma-pattern were enrolled. Time of evolution was calculated, and a detailed pilot study of capillaroscopic characteristics was random executed on 10 of those patients. The analysis included the number and the limbs diameters (arterial, venous, and apical) of capillaries with a diameter over 30 µm, together with the total number of capillaries and microhemorrhages, in 16 images per subject.

Results: All the 54 patients (100%) showed enlarged capillaries with an average diameter over 30 µm in their previous NVC. At the follow up, thirty-one patients (57%) developed an "Early" scleroderma pattern in the following 3 years, 6 patients (11%) developed an "Active" pattern in 4 years, 3 patients (6%) evolved in "Late" NVC pattern in 5 years, whereas 14 patients (26%) developed a "scleroderma-like pattern" in 4 years. The average time of evolution in a scleroderma-pattern was 4 years. The detailed pilot morphological analysis conducted on 10 patients revealed an average total number of capillaries of 8.6/mm at last non-specific NVC analysis; among these 2.66 (31%) capillaries showed a diameter over 30 µm. The mean value diameter of the most dilated capillary was 35.74 µm (arterial limb 33.34 µm, apical limb 43.94 µm, venous limb 30 µm); the mean value for microhemorrhages was 0.6/mm. The mean number of capillaries reduced from 8.6±0.8 to 6.9±2.2/mm (p=0.01) during follow-up (4 years).

Conclusion: Present detailed pilot study demonstrates that SSc patients showed a significant increase of nailfold capillary diameter over 30 µm before development of a validated NVC scleroderma-pattern at follow up. Even if this is an independent predictor for development of SSc as previously demonstrated, the data could help in intercepting patients with RP at higher risk of evolution in a validated SSc NVC pattern.

REFERENCE

[1] Trombetta AC, et al. *J Rheumatol*. 2016;43:599-606. 2. Cutolo M, et al. *J Rheumatol* 2000;27:155-60.

Disclosure of Interests: None declared

DOI: 10.1136/annrheumdis-2019-eular.6691

SAT0294 ANALYSIS OF POLIAUTOIMMUNITY IN THE DIFFERENT SUBSETS OF SCLERODERMA

Daniel Sánchez-Cano¹, Norberto Ortega¹, Jose Luis Callejas-Rubio¹, Carles Tolosa², Alfredo Guillén del Castillo³, Dolores Colunga Argüelles⁴, Luis Sáez-Comet⁵, Manuel Rubio-Rivas⁶, Ana Argibay⁷, Vicent Fonollosa-Pla³, Carmen Pilar Simeón-Aznar³, On Behalf of Rescle Investigators, Autoimmune Diseases Study Group (Geas)⁸. ¹Hospital Universitario San Cecilio, Unit of Systemic Autoimmune Diseases, Granada, Spain; ²Hospital Parc Taulí, Internal Medicine, Sabadell, Spain; ³Hospital Vall D Hebron, Unit of Autoimmune Diseases, Barcelona, Spain; ⁴Hospital Universitario Central De Asturias, Internal Medicine, Oviedo, Spain; ⁵Hospital Universitario Miguel Servet, Internal Medicine, Zaragoza, Spain; ⁶Hospital Universitario de Bellvitge-IDIBELL, Unit of Autoimmune Diseases, Barcelona, Spain; ⁷Complejo Hospitalario Universitario de Vigo, Unit of Systemic Autoimmune Diseases and Thrombosis, Vigo, Spain; ⁸Sociedad Española de Medicina Interna (SEMI), Madrid, Spain

Background: Coexistence of different connective tissue diseases based on their common autoimmune base is a feasible circumstance, which is known as poliautoimmunity (PAI).

Objectives: Evaluation of the occurrence of PAI in systemic sclerosis (SSc) according to subtypes in the patients included in the Spanish SSc Registry (RESCLE). Causes of death were also analyzed.

Methods: A nationwide, cross-sectional study was carried out. All participating centers had obtained local ethics committee approval.

Results: PAI was present in 46% out of 1911 patients, of whom, 33% had more than one association. Most of these patients were women

Table 1. Prevalence of PAI according to subsets.

	Isolated SSc	PAI	P-VALUE
lcSSc	588 (57%)	554 (63%)	0.008
dcSSc	210 (20%)	188 (21%)	0.611
ssSSc	121 (12%)	85 (9.7%)	0.160
earlySSc	28 (2.7%)	16 (1.8%)	0.222
preSSc	85 (8.2%)	36 (4.1%)	<0.001

(93%, p<0.001), with no significant differences regarding first manifestation: Raynaud's phenomenon, puffy hands, arthralgia or skin sclerosis. Isolated SSc was more frequent in all subsets (table 1). Sjogren's syndrome was by far the most frequent associated disorder, with a RR of 1376 (table 2). Causes of death (SSc-related or not) did not significantly differ whether PAI was present or not.

Table 2. Prevalence of the different associated autoimmune disorders.

	TOTAL	lcSSc	dcSSc	sineSSc	earlySSc	preSSc
Sjögren's syndrome	475 (25%)	317 (28%)	106 (27%)	34 (17%)	6 (14%)	12 (9.9%)
Thyroid autoimmune disorder	268 (14%)	173 (15%)	36 (9.0%)	35 (17%)	8 (18%)	16 (13%)
Primary biliary colangitis	88 (4.6%)	61 (5.3%)	5 (1.3%)	17 (8.3%)	0 (0.00%)	5 (4.1%)
Sclerosing colangitis	6 (0.31%)	4 (0.35%)	0 (0.00%)	0 (0.00%)	0 (0.00%)	2 (1.7%)
Autoimmune hepatitis	20 (1.0%)	17 (1.5%)	2 (0.50%)	0 (0.00%)	0 (0.00%)	1 (0.83%)
Systemic lupus erythematosus	11 (0.58%)	7 (0.61%)	1 (0.25%)	3 (1.5%)	0 (0.00%)	0 (0.00%)
Antiphospholipid syndrome	16 (0.84%)	8 (0.70%)	5 (1.3%)	2 (0.97%)	1 (2.3%)	0 (0.00%)
Inflammatory myopathy	99 (5.2%)	45 (3.9%)	47 (12%)	6 (2.9%)	1 (2.3%)	0 (0.00%)
Rheumatoid arthritis	6 (0.31%)	4 (0.35%)	2 (0.50%)	0 (0.00%)	0 (0.00%)	0 (0.00%)

Conclusion: A rather higher prevalence than reported was observed, although distribution of the associated disorders was similar. No remarkable differences were found regarding SSc subsets, either PAI prevalence, initial manifestations or causes of death.

Disclosure of Interests: None declared

DOI: 10.1136/annrheumdis-2019-eular.4870

SAT0295 ABSTRACT WITHDRAWN

SAT0296 FAST TRACK ALGORITHM: HOW TO DIFFERENTIATE A SCLERODERMA PATTERN FROM A NON-SCLERODERMA PATTERN

Vanessa Smith^{1,2,3}, Amber Vanhaecke^{1,2}, Miguel Guerra⁴, Rossella De Angelis⁵, Ellen Deschepper⁶, Christopher Denton⁷, Oliver Distler⁸, Ivan Foeldvar⁹, Eric Hachulla¹⁰, Francesca Ingegnoli^{11,12}, Ulf Müller-Ladner¹³, Yves Piette², Valeria Riccieri¹⁴, Barbara Ruardo¹⁵, Alberto Sulli¹⁵, Jacob M. van Laar¹⁶, Ariane Herrick^{17,18}, Maurizio Cutolo¹⁵. ¹Ghent University, Department of Internal Medicine, Ghent, Belgium; ²Ghent University Hospital, Department of Rheumatology, Ghent, Belgium; ³VIB Inflammation Research Center (IRC), Unit for Molecular Immunology and Inflammation, Ghent, Belgium; ⁴Centro Hospitalar Vila Nova de Gaia/Espinho, Vil Nova de Gaia, Rheumatology Department, Vil Nova de Gaia, Portugal; ⁵Carlo Urbani Hospital, Polytechnic University of Marche, Department of Clinical and Molecular Sciences, Rheumatology Unit, Jesi, Italy; ⁶Ghent University, Department of Biostatistics, Ghent, Belgium; ⁷University College London, Royal Free Hospital, Department of Rheumatology, London, United Kingdom; ⁸University Hospital Zurich, Department of Rheumatology, Zurich, Switzerland; ⁹Centre for Pediatric and Adolescent Rheumatology, Hamburg, Germany; ¹⁰Univ. Lille, CHU Lille, Département de Médecine Interne et Immunologie Clinique, Centre de Référence des Maladies Systémiques et Auto-Immunes Rares du Nord-Ouest (CERAINO), LIRIC, INSERM, Lille, France; ¹¹University of Milan, Department of Clinical Sciences and Community Health, Milan, Italy; ¹²ASST G. Pini, Division of Rheumatology, Milan, Italy; ¹³Justus-Liebig University of Giessen, Kerckhoff Klinik, Department of Rheumatology and Clinical Immunology, Bad Nauheim, Germany; ¹⁴Sapienza University of Rome, Department of Internal Medicine and Medical Specialties, Rome, Italy; ¹⁵IRCCS San Martino Polyclinic Hospital, University Of Genoa, Research Laboratory And Academic Division Of Clinical Rheumatology, Department Of Internal Medicine, Genoa, Italy; ¹⁶University Medical Center Utrecht, Department of Rheumatology and Clinical Immunology, Utrecht, Netherlands; ¹⁷University of Manchester, Salford Royal Hospital NHS Foundation Trust, Division of Musculoskeletal and Dermatological Sciences, Manchester, United Kingdom; ¹⁸NIHR Manchester Musculoskeletal Biomedical Research Centre, Central Manchester NHS Foundation Trust, Manchester Academic Health Science Centre, Manchester, United Kingdom

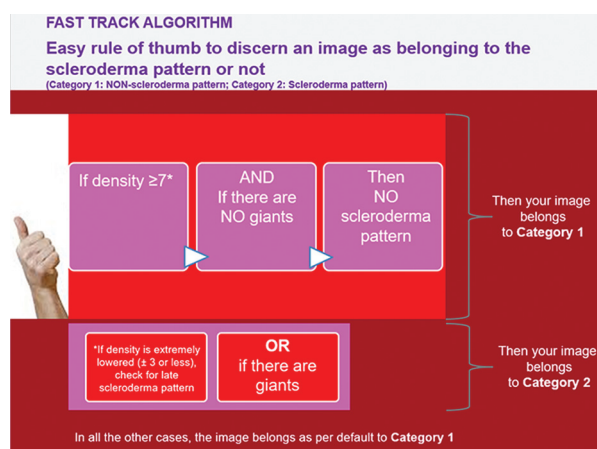
Background: The European League Against Rheumatism Study Group on Microcirculation in Rheumatic Diseases is a non-profit international network of expert centres. Its main research focus is to investigate the

morphology and function of the microcirculation with different non-invasive techniques such as nailfold videocapillaroscopy (NVC). NVC is of paramount importance for the differential diagnosis of primary and secondary Raynaud's phenomenon [1], and is part of the 2013 ACR/EULAR classification criteria for systemic sclerosis. As there is a wide variety of non-scleroderma patterns, the categorisation of capillaroscopic images as non-scleroderma patterns may be a challenge to the capillaroscopist.

Objectives: This study was designed to propose a simple fast track algorithm to differentiate scleroderma patterns from non-scleroderma patterns and to assess its interobserver reliability.

Methods: During the 8th EULAR course on capillaroscopy held in Genoa, September 2018, a lecture on teaching the fast track algorithm to categorise an image as non-scleroderma (category 1) or scleroderma pattern (category 2) (see figure) was given to all attendees (from 43 different countries): 6 experts, 68 novices, 53 moderately experienced (< 5 years of experience with capillaroscopy) and 14 experienced physicians (> 5 years of experience with capillaroscopy). Immediately after training, an examination was performed on 30 images. Classification of the images was defined by the presenter (VS) as the gold standard. Interobserver reliability was assessed by the calculation of kappa coefficients and proportion of agreement versus the gold standard.

Results: The light kappa was 1 for the independent experts (n=6) and 0.92 for the attendees (n=135). When comparing with the gold standard, an equal mean kappa of 0.96 (95%CI 0.95 – 0.98) was found for both independent experts and attendees. Analyses according to the reported level of experience on capillaroscopy revealed a mean index of reliability of 0.98 (95%CI 0.96 – 0.99) for novices, 0.96 (95%CI 0.93 – 0.99) for moderately experienced raters and 0.93 (95%CI 0.85 – 1.01) for experienced raters.



Conclusion: For the first time a fast track algorithm has been developed that was trainable within an hour to non-experienced capillaroscopists and has an excellent reliability to discern a non-scleroderma from a scleroderma pattern by medical doctors with varying levels of expertise in capillaroscopy.

REFERENCE

[1] Koenig M, et al. *Arthritis Rheum.* 2008;58(12):3902-12.

Disclosure of Interests: Vanessa Smith: None declared, Amber Vanhaecke: None declared, Miguel Guerra: None declared, Rossella De Angelis: None declared, Ellen Deschepper: None declared, Christopher Denton Grant/research support from: GlaxoSmithKline, Inventiva, CSF Behring, Consultant for: Roche-Genentech, Actelion, GlaxoSmithKline, Sanofi Aventis, Inventiva, CSL Behring, Boehringer Ingelheim, Bayer, Oliver Distler Grant/research support from: Prof. Distler received research funding from Actelion, Bayer, Boehringer Ingelheim and Mitsubishi Tanabe to investigate potential treatments of scleroderma and its complications, Consultant for: Prof. Distler has/had consultancy relationship within the last 3 years with Actelion, AnaMar, Bayer, Boehringer Ingelheim, ChemoAb, espeRare foundation, Genentech/Roche, GSK, Inventiva, Italfarmaco, iQvia, Lilly, medac, MedImmune, Mitsubishi Tanabe Pharma, Pharmacia, Novartis, Pfizer, Sanofi, Serodapharm and UCB in the area of

potential treatments of scleroderma and its complications. In addition, he had/had consultancy relationship within the last 3 years with A. Menarini, Amgen, Abbvie, GSK, Meppha, MSD, Pfizer and UCB in the field of arthritides and related disorders, Ivan Foldvari Consultant for: Chugai, Novartis, Eric Hachulla Consultant for: Received consulting fees or other remuneration from Actelion, GSK, Pfizer, and Bayer, Francesca Ingegnoli: None declared, Ulf Müller-Ladner Grant/research support from: Projekt supported by an unrestricted educational grant from Celgene GmbH., Yves Piette: None declared, Valeria Ricciari: None declared, Barbara Ruaro: None declared, Alberto Sulli: None declared, Jacob M. van Laar Grant/research support from: Genentech, Consultant for: F. Hoffmann-La Roche, Ariane Herrick: None declared, Maurizio Cutolo: None declared
DOI: 10.1136/annrheumdis-2019-eular.819

SAT0297

DISTINCT CLINICAL PROFILES OF CHINESE AND SWEDISH IDIOPATHIC INFLAMMATORY MYOPATHY PATIENTS WITH ANTI-MELANOMA DIFFERENTIATION ASSOCIATED GENE 5 (MDA5) ANTIBODY

Ho SO¹, Tak Lung Victor Wong¹, Maryam Dastmalchi², Valerie Leclair², Fabricio Espinosa-Ortega², Ingrid E. Lundberg². ¹Kwong Wah Hospital, Kowloon, Hong Kong (SAR); ²Karolinska Institutet, Solna, Sweden

Background: Anti-melanoma differentiation-associated gene 5 antibody (anti-MDA5 Ab) is one of the most clinically important myositis specific antibodies. It has been found to be associated with severe cutaneous and pulmonary complications. Nonetheless, there appears to be geographical variations in the prevalence and phenotypic presentation of the antibody [1]. We conducted the first comparative study of two ethnogeographically different idiopathic inflammatory myopathy (IIM) cohorts focusing on anti-MDA5 Ab, using standardized classification criteria, antibody detection methods and clinical features assessment.

Objectives: The objectives of the study were to compare the prevalence of the anti-MDA5 Ab and its associated clinical features between Chinese IIM patients in Hong Kong and Caucasian patients in Sweden.

Methods: This multicenter retrospective cohort study was conducted on IIM patients followed up in the rheumatology clinic or admitted to the rheumatology wards of the participating hospitals in Hong Kong and Sweden with anti-MDA5 Ab tested between September 2015 and August 2018. The diagnosis of IIM was based on the 2017 European League Against Rheumatism/American College of Rheumatology Classification Criteria with definite or probable cases being included. Demographics and clinical features were collected by reviewing the medical records. A commercial line blot immunoassay kit (EUROIMMUN) was used to detect the anti-MDA5 autoantibodies.

Results: Altogether, 409 patients with IIM (Sweden: 206, Hong Kong: 203) were recruited. The Swedish patients were generally older (mean age 58.1 vs 51.5 years, $p < 0.001$) and more of them were male (41.4% vs 23.2%, $p < 0.001$). Subgroups of patients were: dermatomyositis 42.9%, polymyositis 39.3%, inclusion body myositis 15.7% and clinically amyopathic dermatomyositis 2.1% in the Swedish cohort, and 39.4%, 39.4%, 0% and 21.2% respectively in the Chinese cohort. Anti-MDA5 Ab was found in 11.8% of the Chinese and 5.83% of the Swedish patients ($p = 0.032$). The Hong Kong Chinese patients were found to have significantly more ILD (52.7% vs 34.7%) and RPILD (9.41% vs 3.65%), while the Swedish patients had more dysphagia (55.3% vs 29.7%) and cardiac involvement (14.0% vs 5.3%). Anti-MDA5 Ab was associated with male gender, younger age at diagnosis, amyopathy, refractory rash, ILD and RPILD in the Swedish patients. The same associations were found in the Chinese patients except the young age at diagnosis. Besides, in the Chinese cohort, the antibody was also positively associated with skin ulceration, vasculitis and hoarseness, but negatively associated with cancer, dysphagia and other connective tissue diseases. When comparing only patients with anti-MDA5 Ab, Chinese patients were significantly older at diagnosis (mean age 47.9 vs 36.4 years, $p = 0.001$), more likely to be amyopathic (79.2% vs 18.2%, $p = 0.002$) and had more vasculitic skin changes (65.2% vs 0%, $p < 0.001$) than the Swedish patients. They also had a tendency towards developing more skin ulcers (45.8% vs 18.2%, $p = 0.15$) and less dysphagia (8.33% vs 36.4%, $p = 0.063$).

Conclusion: The distinct clinical profiles identified in the Chinese and Swedish IIM patients with anti-MDA5 Ab could depend on different genetic or environmental factors, and may enhance our knowledge about