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SAT0323

MYOSITIS-SPECIFIC AND MYOSITIS-ASSOCIATED AUTOANTIBODIES IN A LARGE INDIAN COHORT OF INFLAMMATORY MYOSITIS REVEAL NOVEL CLINICO-PHENOTYPIC PATTERNS

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Background: Idiopathic Inflammatory Myositis (IIM) are heterogenous, with distinct autoantibodies reflecting upon possible clinical evolution and outcomes. Ethnicity has major influence on both antibody prevalence patterns as well as phenotypic behaviours linked to them.

Objectives: Thus we sought prevalence and co-existence of myositis specific autoantibodies (MSAs) and myositis associated autoantibodies (MAAs) and associated clinical characteristics in a large cohort of patients with IIM.

Methods: Adult patients with a physician diagnosis of IIM as per ACR/EULAR classification criteria were investigated for the presence of MSAs/MAAs by Line immunoassay (G4, Euro-Immune, Lubeck, Germany). Anti-Nuclear Antibody (ANA) was tested by Immunofluorescence assay (IFA), and patterns in various antibody subsets explored. Prevalence and associations of different antibodies were assessed in disease subsets and clinical phenotypes.

Results: MSA and MAAs were tested in 250 IIM patients (F:M 3.8:1) of median age 37 (25-47) and disease duration 6 (3-17) years. Dermatomyositis (DM) was seen in most patients 83 (33.2%) followed by overlap myositis (OM), juvenile DM, Anti-synthetase syndrome (ASS), polymyositis (PM), and cancer associated myositis (CAM). MSAs/MAAs were found in 148 (59.2%) of patients, of which 95 (64.2%) had an MSA and 53 (35.8%) had MAAs (Fig. 1A). 93 (62.8%) of autoantibody positive patients were positive for a single antibody, and only 2 (0.8%) of total had more than one MSA (Table 1).

Table 1. Multiple antibodies positive upon testing for MSA and MAA by the LIA

AutoAntigen	Auto-Ab (n)	None	Jo-1	PL-7	PL-12	MDAS	SAE	MI2	NXP2	TIF1y	SRP	Ku	Pm/ScI	Ro52
Jo-1**	21	3			1								1	17
PL-7**	4	1											1	1
PL-12**	7	0	1										1	5
MDAS*	14	8								1*				1
SAE*	5	3						1*						1
MI2	19	17					1							1
NXP2	9	8										1		2
TIF1y	12	9				1								4
SRP**	5	0											1	1
Pm/ScI	12	11		1										3
Ro52	72	33	17	1	5	5	1	1		2	4	3		
Ku	10	6												3

Note: ** PL-7 co-exists with Ku + Pm/ScI, **PL-12 co-exists with Pm/ScI + Ro52, **SRP co-exists with Pm/ScI + Ro52

The most frequently detected MSA was anti-Jo-1 (8%), with a further 9 specificities each found in 0.5–7.0% of patients. Amongst the autoantibody positive patients, 21% (n=53) had isolated MAA positivity, anti-Ro52 (33, 62.3%) being the most common, followed by anti-Pm/ScI (11, 20.8%) and anti-Ku (9, 17.0%) (Fig. 1B).

Figure 1. A. MSA and MAA in Indian cohort of myositis **B.** ANA patterns in myositis **C.** MSA in ANA negative IIM

On ANA, 76.0% (172 of 226) were positive, with speckled being the most common pattern (37%, Fig. 1C). Of those ANA negative (n=54), 61% had either MSA or MAA (Fig 1D). 18 (54.6%) had autoantibodies associated with cytoplasmic patterns suggesting that cytoplasmic ANA may be underreported.

Clinical presentation akin to DM was seen with all MSA except anti-SRP. PM group was heterogenous, and included ASS, OM and necrotizing phenotype (Fig. 2A). On occasion, anti-SRP, anti-Mi-2 and anti-MDA5 presented with clinical phenotype of ASS. (Fig 2A,C). Patients with ARS or anti-SAE were often clinically amyopathic (Figure 2 B,C)

Figure 2. A. Phenotypic associated with various antibody subsets **B,C and D.** MSA/MAA in muscle weakness, rash and ILD phenotype. **E.** Unique feature of eye-lid edema in some patients with MDA-5 positive myositis

ARS were associated with mechanic's hand (p<0.0001,OR 7.6), ILD (p<0.0001,OR 4.4), and arthritis (p=.002,OR 2.6) though there was no difference between Jo-1 and non-Jo-1 ASS. Anti-MDA-5 associated with fever (p=0.003,OR 12) and weight loss (p=0.008,OR 10.2) and unique phenotype of eye-lid edema in some adults (Figure 2E) and arthritis in children (p=0.01, OR 11.5). Anti-TIF-1y associated with alopecia (p=0.007,OR 5.9) and malignancy (p= <0.0001,OR 34) in adults but not children.

Conclusion: Myositis autoantibodies are seen in two-thirds IIM and identify distinct clinical subsets as well as unique phenotypes. MSA/MAA are positive in two-thirds of those negative on ANA, adding diagnostic value. MSAs are nearly always mutually exclusive and thus useful as biomarkers for diagnosis.

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SAT0324

SEXUAL HEALTH IN WOMEN AND MEN WITH SYSTEMIC SCLEROSIS: A CROSS-SECTIONAL STUDY

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Background: Systemic sclerosis (SSc) is a chronic multisystem disease that may affect all aspects of life including sexual function.

Objectives: To assess sexual function, pelvic floor function in 80 women and 14 men with SSc compared to age-/sex-matched healthy controls (HC) and to analyze the potential impact of clinical features on sexual health.

Methods: In total 80 women (mean age: 49, disease duration: 6.1 years, lcSSc/dcSSc: 53/27, mRSS: 9.3, ESSG activity index: 2.1) and 14 men with SSc (mean age: 51.6, disease duration: 3.1 years, lcSSc/dcSSc: 6/8, mRSS: 12.9, ESSG activity index: 2.2) who fulfilled the ACR/EULAR 2013 criteria, filled in 12 well-established and validated questionnaires assessing sexual function, pelvic floor function, quality of life, fatigue, physical activity and depression. Full names of questionnaires are listed in Table 1. These questionnaires were also completed by 80 healthy women and 14 healthy men with identical age. Data are presented as mean±SEM.

Table 1

Clinical parameter	Correlated parameters	r	p-value
ESSG diseases activity index	FSFI Arousal domain	-0.300	p<0.0001
	IIIEF EF	-0.597	p=0.0240
CRP	IIIEF Organic Function	-0.668	p=0.0081
mRSS	MSHQ Sexual Desire	0.555	p=0.0613
FIS	BISF-W	-0.410	p=0.0002
MAF	FSFI	-0.399	p=0.0002
BDI-II	BISF-W	-0.381	p=0.0002
	IIIEF EF	-0.801	p=0.0005
HAP	SQoL-F	0.383	p=0.0006
	BISF-W	0.456	p<0.0001
	IIIEF EF	0.639	p=0.0062
SHAQ	FSFI	-0.409	p<0.0001
	BISF-W	-0.451	p=0.0002

Name of Questionnaires/Acronyms: FSFI: Female Sexual Function Index; BISF-W: Brief Index of Sexual Function for Women; SQoL-F/M: Sexual Quality of Life Questionnaire-Female/Male; PISQ-12: Pelvic Organ Prolapse/Urinary Incontinence Sexual Questionnaire short form; PFIQ7: Pelvic Floor Distress Inventory Questionnaire; IIIEF: International Index of Erectile Function; MSHQ: Male Sexual Health Questionnaire; FIS: Fatigue Impact Scale; MAF: Multidimensional Assessment of Fatigue; BDI-II: Beck's Depression Inventory II; HAP: Human Activity Profile; SHAQ: Scleroderma Health Assessment Questionnaire; ESSG: European Scleroderma Study Group; CRP: C-reactive protein; mRSS: modified Rodnan skin score

Results: Compared to HC, women with SSc had significantly higher prevalence and greater severity of sexual dysfunction [FSFI total score (SSc: 16.4±1.3, HC: 25.6±1.2, p<0.0001) as well as in all subscales (p<0.0001 for all), BISF-W total score (SSc: 17.2±2.3, HC: 32.0±1.9, p<0.0001)], dysfunction of pelvic floor [(PISQ-12 (SSc: 3.6±0.7, HC: 8.9±0.6, p<0.0001), PFIQ7 (SSc: 33.4±5.6, HC: 6.8±1.4, p<0.0001)], and worse sexual quality of life [SQoL-F (SSc: 55.3±3.3, HC: 82.1±2.1, p<0.0001)]. Men with SSc also reported more severe sexual dysfunction: [IIIEF - Erectile function (EF) (SSc: 15.4±3.0, HC: 26.2±2.1, p=0.004), IIIEF - Organic function (SSc: 6.4±1.2, HC: 9.0±0.6, p=0.045), IIIEF - Intercourse satisfaction (SSc: 9.7±1.8, HC: 15.7±1.3, p=0.008), MSHQ - Erectile Function (SSc: 9.0±1.2, HC: 12.9±0.9, p=0.005), MSHQ - Satisfaction (SSc: 19.2±2.1, HC: 26.8±0.9, p=0.001)] and worse sexual quality of life [SQoL-M (SSc: 68.5±7.4, HC: 86.7±6.2, p=0.023)]. According to the IIIEF classification, 71 % of SSc men reported mild to severe erectile dysfunction. No significant differences were found in pelvic floor function. Significant associations with major clinical parameters are presented in Table 1.

Conclusion: Both women and men with SSc reported significantly impaired sexual function compared to HC with identical age. Worse scores in SSc were associated with disease activity, increased systemic inflammation, health status, physical activity, fatigue and depression.

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SAT0325

STELLATE BLOCKADE COMBINED TO ILOPROST AS SUPPORTIVE TREATMENT OPTION IMPROVES PAIN AND ISCHAEMIC SYMPTOMS IN PATIENTS WITH SYSTEMIC SCLEROSIS

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Background: Peripheral ischaemia is a common symptom in systemic sclerosis (SSc) patients with risk of development of digital ulcers (DU). For its treatment, intravenous iloprost is the most effective option. Accompanying pain symptoms worsen the ischaemic symptoms, so a combination with anaesthetic procedures may improve ischaemic status and the subjective sensation of raynaud and pain. The aim of this study was to observe the impact of a combined treatment of iloprost with stellate blockade (ILOST) in improvement of ischaemic symptoms compared to iloprost treatment only (ILO).

Objectives: To evaluate efficacy of the ILOST treatment on changes in vascularisation and sensation of patients with SSc and indication for vasodilative treatment with Iloprost.

Methods: Twenty SSc-patients with indication for ILO-treatment (prophylactic or due to digital ulcerations (DU)) will be included in a prospective observational study. Patients will be offered to combine ILO with stellate blockade (ILOST). Beside documentation of disease activity characteristics (mRSS, number of DU, capillary microscopy at baseline, after ILO-treatment and at week 12), patients are assessed using fluorescence-optical imaging (FOI) as innovative method for illustration of changes in microvascularisation and patient reported outcomes (DASH, VAS) at week 12.

Results: This interim analysis includes the result of the first 11 patients treated. Mean baseline characteristics (age and gender) are well balanced. Iloprost treatment was initiated due to prophylactic treatment to avoid DU in all patients. 100% of the patients in the ILOST-group were diagnosed as limited SSc compared to 60% in the ILO-group (diffuse type with 40%). All patients showed abnormalities in capillary microscopy (ILOST group: 83,3% late pattern 16,7% active pattern; ILO group: 80% late pattern, 20% early pattern). mRSS was low in both groups with 1.8, the disease duration in mean 15.3 years in the ILOST-group compared to 13.2 in the ILO-group, respectively. In both groups, no new DU occurred in the 12-week follow-up. Improvement in VAS pain was reported in 83% of the patients in the ILOST group compared to 60% in the ILO group. DASH improved with a mean of 5.5 points in the ILOST group compared to 3 points in the ILO group. FOI was compared individual at both arms in the ILOST group only. The arm with stellate blockade showed a pronounced increase of FOI signals of 5% in mean whereas the opposite site showed a decrease of the signal shortly after ILO treatment indicating a pronounced increase of vascularisation in the ILOST treated body site.

Conclusion: A new treatment approach to improve acute ischaemic symptoms was tested by combining stellate blockade to iloprost treatment. No new DU occurred up to 12 weeks after treatment in all patients of both groups indicating the relevance of iloprost as effective vascular dilatative therapy in SSc. The additional intervention was well tolerated and asked to repeat. Subjective sensation on pain of the hands as well as DASH was improved in the combined group. FOI showed a relevant increase in vascularisation in the blockade arm compared to the opposite site in which signals decreased indicating a stronger effect of the combined treatment for improvement of vascularisation.

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SAT0326

SYSTEMIC SCLEROSIS WITHOUT ANTINUCLEAR ANTIBODIES: A MULTI-CENTER STUDY OF EUSTAR COHORT IN CHINA

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Background: The presence of circulating antinuclear antibodies (ANAs) is a hallmark of immune dysregulation and malfunction in patients with systemic sclerosis (SSc) [1]. A variety of ANAs [2], including anti-centromere antibody, anti-topoisomerase I antibody, and anti-RNA polymerase III antibody, are associated with unique sets of disease manifestations and widely used in routine clinical practice for diagnosis, clinical subgrouping, risk stratification and prediction of future organ involvements and prognosis in SSc patients [3,4].

Objectives: This study aimed to investigate the clinical features of SSc patients with negative ANAs in a European League Against Rheumatism Scleroderma Trials and Research Group (EUSTAR) and Chinese Rheumatism Data Center (CRDC) multi-center cohort in China.

Methods: Patients were prospectively recruited between April 2008 and June 2019 based on the EUSTAR database and CRDC multi-center cohort from 154 clinical centers nationwide, all of whom fulfilled the 2013 ACR/EULAR classification criteria for systemic sclerosis. Antinuclear antibody testing result was intensively collected. Demographic, clinical, and laboratory data were compared between ANA-positive SSc patients and those with negative ANAs. T-test and chi-square analysis were performed in the comparisons.