Conclusion: Using a SLR and modified nominal technique, we have developed a preliminary pSScHI consensus-based definition and started a validation process for it to be used in clinical research and clinical practice.

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AB0557

CO-EXISTENCE OF SYSTEMIC SCLEROSIS HALLMARK AUTOANTIBODIES ASSOCIATES WITH DISTINCT CLINICAL PHENOTYPE

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Background: Systemic sclerosis (SSc) is typically manifests with distinct SSc-specific antibodies (SSc-Abs): anti-topoisomerase I (ATA), anti-centromere (ACA), anti-RNA polymerase III (ARA), anti-U3RNP (u3RNP), anti-U1RNP (U1RNP), anti-PmScl (PmScl), anti-Ku (Ku) and anti-Th/T0 (Th/T0), each being characterised by different clinical features and prognosis. The presence of >1 SSc-Abs is rare with minimum data about these patients' clinical phenotype.

Objectives: To describe and compare the clinical features of SSc patients with >1 SSc-Abs

Methods: The autoantibody profiles of 2799 SSc patients from February 2001 to June 2017 were retrospectively reviewed. Patients with >1 SSc-Abs were identified. Clinical features were collected and compared to historical cohorts of SSc patients with single SSc-Abs positivity. Patients were excluded if treated prior to their immunology test with rituximab, iv immunoglobulins or stem cell transplantation. Statistical analysis was performed using Fisher exact test.

Results: 72 patients (2.6%) with >1 SSc-Abs were identified. Full clinical data were available for 63 patients. 60 patients (2.1%) had double Ab positivity and 3 patients had triple Ab positivity (0.1%). 13 Ab combinations were present. U1RNP and ATA was the most frequent combination (35%), patients were significantly younger (51.38 years) than both U1RNP (58.64 years, $p=0.050$) and ATA (62.03 years, $p=0.002$) patients and more commonly of diffuse subset (dcSSc) ($p=0.001$ and $p=0.041$ respectively). Compared to ATA patients overlap features were more frequent (43% vs 15%, $p=0.004$) including inflammatory arthritis ($p=0.025$) and myositis ($p=0.013$) (Table 1). U1RNP and ACA had a significantly higher prevalence of pulmonary arterial hypertension compared to U1RNP (young) and ACA ($p=0.022$) patients, and compared to ACA patients they were younger (57.88 vs 68.75, $p=0.015$) with a higher incidence of myositis ($p=0.001$). U1RNP and ARA patients were more frequently dcSSc subtype compared to U1RNP patients (75% vs 21%, $p=0.040$). U1RNP and PmScl patients had a higher prevalence of myositis compared to U1RNP patients ($p=0.006$). ATA and ACA patients behaved similarly to ATA patients with a significantly higher prevalence of lung fibrosis ($p=0.006$) and myositis ($p=0.041$) compared to ACA. ACA and PmScl (7%) had higher prevalence of myositis compared to ACA patients ($p=0.04$).

Table 1. Frequency of clinical features in some of the double antibody group combinations, compared to our cohort of patients with only one of the SSc specific antibody. Significant p values (<0.05) highlighted in bold. ILD (interstitial lung disease), PAH (pulmonary arterial hypertension), SRC (scleroderma renal crisis).

Double antibody group	Comparator group	Clinical features											
		Arthritis		ILD		PAH		SRC		Cardiac		Myositis	
		n	p	n	p	n	p	n	p	n	p	n	p
U1RNP and ATA	U1RNP	6 (29%)		15 (68%)		1 (9%)		0		0		4 (39%)	
	ATA	14 (18%)	1	38 (50%)	0.136	12 (16%)	0.287	1 (1%)	1	2 (3%)	1	12 (16%)	0.744
U1RNP and ACA	U1RNP	2 (10%)		2 (10%)		4 (50%)		0		0		1 (17%)	
	ACA	14 (18%)	1	38 (50%)	0.267	12 (16%)	0.039	1 (1%)	1	2 (3%)	1	12 (16%)	0.148
U1RNP and ARA	U1RNP	23 (30%)	0.087	43 (11%)	0.233	56 (15%)	0.022	6 (2%)	1	10 (3%)	1	3 (1%)	0.001
	ARA	0		2 (50%)		0		1 (25%)		1 (25%)		0	
U1RNP and PmScl	U1RNP	14 (18%)	0.311	38 (50%)	1	12 (16%)	1	1 (1%)	0.098	2 (3%)	0.144	12 (16%)	1
	PmScl	4 (13%)	1	64 (45%)	1	15 (10%)	1	34 (24%)	1	4 (3%)	0.131	5 (13%)	
ATA and ACA	ATA	1 (13%)		2 (66%)		12 (16%)	1	1 (1%)		2 (3%)		3 (100%)	
	ACA	14 (18%)	1	38 (50%)	1	12 (16%)	1	1 (1%)	1	2 (3%)	1	12 (16%)	0.006
ACA and PmScl	ACA	7 (13%)	1	27 (50%)	1	3 (6%)	1	3 (6%)	1	1 (2%)	1	21 (39%)	0.009
	PmScl	0		1 (25%)		0		0		0		1 (25%)	
ACA and U1RNP	ACA	28 (10%)	1	213 (79%)	1	12 (4%)	1	17 (6%)	1	20 (7%)	1	10 (4%)	0.533
	U1RNP	23 (30%)	1	43 (11%)	0.006	56 (15%)	1	6 (2%)	1	10 (3%)	1	3 (1%)	0.041
U1RNP and PmScl	U1RNP	1 (25%)		1 (25%)		0		0		0		1 (25%)	
	PmScl	23 (30%)	0.227	43 (11%)	0.385	56 (15%)	1	6 (2%)	1	10 (3%)	1	3 (1%)	0.04
ACA and U1RNP	ACA	7 (13%)	0.457	22 (50%)	0.612	3 (6%)	1	3 (6%)	1	1 (2%)	1	21 (39%)	1
	U1RNP	0		1 (50%)		0		0		0		1 (50%)	
U1RNP and PmScl	U1RNP	23 (30%)	1	43 (11%)	0.216	56 (15%)	1	6 (2%)	1	10 (3%)	1	3 (1%)	1
	PmScl	3 (6%)	1	12 (24%)	0.456	13 (26%)	1	5 (10%)	1	6 (12%)	1	9 (18%)	1

Conclusion: Coexistence of hallmark autoantibodies is exceedingly rare in SSc patients. When combined, both SSc-Abs have the potential to synergistically interact and modify the clinical phenotype.

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AB0558

RISK FACTORS FOR THE DEVELOPMENT OF BREAST CANCER IN SYSTEMIC SCLEROSIS

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