Abstract AB0781 – Table 1. Characteristics of 16 patients with Scleroderma renal crisis

Pt.No	Age(y)	Sex	SRC duration(y)	Prednisone before SRC	Ssc form	Pulmonary fibrosis	Cardiac involvement	PAH	MAHA	BP (mmHg)	Ser (μmol/L)	ACE inhibitor	Outcome
1	62	F	0.25	10mg/d	dcSSc	+	+	+	+	170/100	206	None	Recovered
2	20	F	10	None	dcSSc	+	+	+	+	168/120	299	Irbesartan	died
3	61	F	1	30mg/d	dcSSc	+	+	+	+	170/100	153	Losartan	Recovered
4	60	M	1	30mg/d	dcSSc	+	+	+	+	175/128	511	Captopril	CKD
5	65	M	5	None	dcSSc	+	+	+	+	198/97	174	Captopril	Recovered
6	64	M	5	5mg god	dcSSc	+	+	+	+	190/90	504	Captopril	CKD
7	62	F	1	10mg/d	dcSSc	+	+	+	+	130/102	420	Captopril	died
8	64	F	10	None	dcSSc	+	+	+	+	160/110	259	None	CKD
9	58	M	2	15mg/d	dcSSc	+	+	+	+	189/122	149	Captopril	Recovered
10	24	M	0.5	30mg/d	dcSSc	+	+	+	+	200/120	956	Captopril	Dialysis
11	64	M	8	None	lcSSc	+	+	+	+	194/102	400	Captopril	CKD
12	45	F	15	10mg/d	dcSSc	+	+	+	+	200/120	248	Captopril	died
13	61	M	2	15mg/d	dcSSc	+	+	+	+	110/70	969	Captopril	Dialysis
14	61	F	1	15mg/d	lcSSc	+	+	+	+	193/110	587	None	CKD
15	58	F	1	None	dcSSc	+	+	+	+	150/86	473	None	died
16	46	F	3	35mg/d	dcSSc	+	+	+	+	180/100	392	Captopril	died

SRC: Scleroderma renal crisis, SSc: systemic sclerosis, dcSSc: diffuse cutaneous SSc, lcSSc: limited cutaneous SSc, PAH: pulmonary arterial hypertension, MAHA: microangiopathic haemolytic anaemia, CKD: chronic kidney disease

Conclusions: Medium-to-high doses of glucocorticoids (prednisone ≥ 7.5 mg/d) were associated with an increased risk of SRC. Cardiac involvement was common and associated with high mortality in SRC patients.

Disclosure of Interest: None declared

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AB0782 SYSTEMIC SCLEROSIS PATIENTS WITH CONCOMITANT PSORIASIS: A PROOF-OF-CONCEPT PILOT STUDY

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Background: Psoriasis and Systemic Sclerosis (SSc) are chronic inflammatory diseases characterised by a systemic immunological response which is mainly driven by activated T helper (Th) Th1/Th17 lymphocytes.^{1,2} Further, genome-wide association studies (GWAS) in SSc have demonstrated an association at PSORS1C1, the same gene linked to psoriasis susceptibility³

Objectives: Evaluate the statistical significance of a clinical correlation between Scleroderma and Psoriasis

Methods: From April 2014 to April 2017, we enrolled 180 consecutive patients with a diagnosis of SSc fulfilling 2013 ACR/EULAR classification criteria. Patients with Localised Scleroderma (Morphea) had to have a diagnostic skin biopsy before being included. Patients satisfying VEDOSS criteria⁴ were also included. Patients with psoriasis had a dermatologist-proven diagnosis.

Results: 11/180 (6.1%, 95% IC:0.03–2.9) with scleroderma had a dermatologist-proven diagnosis of Psoriasis, and 23 patients (12.7%) had a family history of Psoriasis in a first-degree relative. For all the psoriatic patients except one, BMI was calculated; 50% had a BMI of >20 (average: 23.7). A metabolic syndrome was present in 3 out of 11 (27.2%), and in ten patients there were data available to calculate the presence of average moderate cardiovascular (CV) risk (11.5%). Finally, 3/11 patients (27.2%) had Psoriatic Arthritis (CASPAR classification criteria). In all 11 patients, Psoriasis was defined as mild (PASI ≤ 10).⁵ The average value of modified Rodnan Skin Score (mRSS) was curiously low (0.5), [limited cutaneous (lcSSc) 5 patients, Morphea 3 patients, Very Early Systemic Sclerosis (VEDOSS) 2 patients, and diffuse cutaneous disease (dcSSc) 1 patient. Anticentromere antibodies (ACA) were detected in 5/11 patients; antinuclear antibodies, with negative extractable nuclear antigens in 4/11; Anti-topoisomerase I were found in two cases.

Conclusions: Psoriasis was significantly associated with SSc ($p=0.014$), with a 2-fold higher frequency than that observed in general population (3%). Psoriasis +SSc, represent a further increased risk of CV disease. Th17 seems to play a crucial role in the pathogenesis of both diseases.

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AB0783 CLINICAL CHARACTERISTICS OF A COHORT OF PATIENTS WITH ANTI-JO1 ANTIBODIES

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Background: Anti-synthetase syndrome (ASSD) is characterised by myositis associated with anti-synthetase antibodies, fever, arthritis, Raynaud's phenomenon, "mechanic's hands" and diffuse interstitial lung disease (ILD); 80% of patients present incomplete forms. The most frequently detected anti-synthetase antibody is anti-Jo1 (anti-histidyl-tRNA synthetase), which usually leads to a greater lung involvement. In some cases of myositis, the simultaneous presence of anti-Jo1 and anti-Ro52 antibodies has been described, and it is debated whether the presence of both antibodies is accompanied by more severe pulmonary involvement.

Objectives: To compare the clinical manifestations of the positive anti-Jo1 patients, with and without associated anti-Ro52 antibodies.

Methods: This is a retrospective observational study of anti-Jo1 positive patients confirmed in the Immunology Laboratory, between 2009 and 2018. Two techniques were used to identify the anti-Jo1 and anti-Ro52 antibodies: Western blot (EUROLINE Myositis Profile 3 (IgG)) and Fluoroenzimimmunoassay (EliA, Thermofisher).

Results: In this study, 22 patients with anti-Jo1 antibodies were included, 16 (72.7%) women and 6 (27.2%) males. There was no association with FR or ACPA. The mean age was 57.3 years (SD:14.6) and the mean time of follow-up was 4.01 years (SD: 2.1). Of all patients, 5 had ASSD, the rest expressed the clinical picture of incomplete form. The most frequently clinical manifestations associated with anti-Jo1 were: muscular involvement ($n=18$, 81.8%), ILD ($n=13$, 59.1%), arthritis ($n=9$, 40.9%), Raynaud's phenomenon ($n=4$, 18.2%). No patient developed lesions of mechanic's hands. The majority of patients with muscular involvement (83.3%), had histological confirmation of inflammatory idiopathic myopathy (Dermato or Polymyositis). The most frequently found X-ray pattern (HRCT) in patients with ILD was Non-specific interstitial pneumonia (NSIP) ($n=9$, 69.2%), followed by 2 Respiratory bronchiolitis interstitial lung disease (RB-ILD) and 2 Idiopathic interstitial pneumonia (IIP). Nine patients presented ILD and Myositis concomitantly (40.9%), more than half had fibrosing pulmonary involvement. 50% ($n=11$) also had criteria for other autoimmune diseases (2 rheumatoid arthritis, 4 polymyositis, 4 dermatomyositis, and 1 overlap). Half of the patients with anti-Jo1 presented also anti-Ro52, which 27.3% exclusively developed myopathy, 18.2% ILD and both entities 54.5%. Half of the ILD presented a radiological fibrosing radiological pattern. During follow-up, 7 patients (31.8%) died, 2 of metastatic cancer (one renal and one ovarian) and 5 of complications of the ILD, of which 80% were anti-Jo1/anti-Ro52 "doublet" positives.

Abstract AB0783 – Table 1

	Age years (SD)	Follow-up years (SD)	Arthritis n(%)	Raynaud n(%)	Myositis n(%)	ILD n (%)	ILD/ Myositis n(%)	Exitus n(%)
Anti-Jo1 (n=22)	57,3 (14,6)	4,0 (2,1)	9 (40,9)	4 (18,2)	18 ^(81,8)	13 ^(59,1)	9 (40,9)	7 (31,8)
"Doublet"	59,2 (15,3)	3,8 (2,0)	6 (54,5)	3 (27,3)	9 (81,8)	8 (72,7)	6 (54,5)	4 (36,4)
Anti-Jo1/ Anti-Ro52 (n=11)								

Conclusions: The ILD is a frequent manifestation and the main cause of death in the patients with anti-Jo1 of our cohort. The presence of anti-Jo1/anti-Ro52 "doublet" seems to lead to a worse prognosis in these patients.

Disclosure of Interest: None declared

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