Microangiopathy evolution score was 5.19 (2.04), VAS for DU was 75.52 (16.17), VAS for Raynaud was 87.43 (14.16), HAQ was 1.62 (0.55). 5 patients received Bosentan less then 16 month, so they were excluded from the statistical analysis. 6 month month evolution revealed significant decrease in the number of DU (p<0.01), the VAS for DU (p<0.01), the VAS for Raynaud (p=0.03) and the HAQ (p=0.04), but not of the microangiopathy evolution score. No significant difference was noticed of the above mentioned parameters at the next follow-up evaluations. Regarding Bosentan safety: 6 patients died during the follow up (3 cases of severe pulmonary arterial hypertension, 1 scleroderma renal crisis, 1 heart failure, 1 post vascular surgery). Bosentan was stopped due to lack of efficacy in 2 case and due to side effects in 3 cases: 2 elevated liver enzymes, 1 severe trombocytopenia and 1 dyspepsia agravation.

12 patients had a follow up after a 6 months Bosentan stop. We did not notice any significant increase in the number of DU, the VAS for DU or Raynaud, the capillary microcircemia and the number of UVs on the skin.

Conclusions: We noted a significant decrease in the number of DU, patients perception of Raynaud and DU after 6 months of treatment and the effect was maintained in the 3 years follow-up, even 6 months after Bosentan was stopped. In this long-term follow-up no new unidentified adverse reactions were found, except for the unexpected severe trombocytopenia. The present study is limited due to the small sample size, to the observational nature and should be viewed as descriptive. Questions rise about drug costs (6 months or long term treatment), but it also has to be emphasised that most of these lesions were chronic and non-responsive to previous treatments.

REFERENCES:

Disclosure of Interest: None declared
DOI: 10.1136/annrheumdis-2018-eular.5875

AB0780 PROLONGED PROTON PUMP INHIBITOR EXPOSURE IS ASSOCIATED WITH DEVELOPMENT OF CALCINOSIS IN SYSTEMIC SCLEROSIS

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Background: Long-term use of proton pump inhibitors (PPI) has been associated with some safety concerns, including potential vascular calcification. In a previous retrospective analysis, we noted a possible association between PPI use and calcinosis in scleroderma (SSc).

Objectives: To perform a retrospective case series analysis of the characteristics, management and outcomes of SRC in Chinese SSc patients.

Methods: SSc patients hospitalised at Sun Yat-Sen Memorial Hospital from January 1992 to December 2017 were recruited. Clinical data were collected. SRC was defined as new onset, with blood pressure (BP) >140/90 mmHg or a >30 mmHg rise in BP from baseline, rising serum creatinine (Scr) levels and/or oligoanuria. Data were showed as mean ±standard deviation.

Results: There were 749 SSc patients recruited and 16 patients (2.1%) of them were hospitalised for SRC. Among these 16 patients, 56% were females, age was 54.6±13.6 years, mean duration from SSc onset to SRC occurred was 4 years (3.3 months to 10 years).

Conclusions: All 16 patients manifested progressive renal failure, with Scr levels increase to 969 μmol/L. Ten patients manifested new onset hypertension, with systolic BP 175±21 mmHg and diastolic BP 108±13 mmHg. Five patients who had a history of well-controlled hypertension manifested accelerated increase in BP 178 ±17/108±7 mmHg. One patient was normotensive, but manifested rapidly progressive oliguric renal failure with Scr increase to 969 μmol/L, massive proteinuria and hemolytic anemia.

AB0781 GLUCOCORTICOID DOSE AND CARDIAC INVOLVEMENT MIGHT BE POTENTIAL RISK FACTORS FOR SCLERODERMA RENAL CRISIS

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Background: Scleroderma renal crisis (SRC) is a rare but life-threatening complication of systemic sclerosis (SSc). SRC remains a major risk factor for mortality in SSc. It is important to identify potential risk factors for SRC, and avoid developing overt SRC.

Objectives: To perform a retrospective case series analysis of the characteristics, management and outcomes of SRC in Chinese SSc patients.

Methods: SSc patients hospitalised at Sun Yat-Sen Memorial Hospital from January 1992 to December 2017 were recruited. Clinical data were collected. SRC was defined as new onset, with blood pressure (BP) >140/90 mmHg or a >30 mmHg rise in BP from baseline, rising serum creatinine (Scr) levels and/or oligoanuria. Data were showed as mean ±standard deviation.

Results: There were 749 SSc patients recruited and 16 patients (2.1%) of them were hospitalised for SRC. Among these 16 patients, 56% were females, age was 54.6±13.6 years, mean duration from SSc onset to SRC occurred was 4 years (3.3 months to 10 years).

Conclusions: All 16 patients manifested progressive renal failure, with Scr levels increase to 969 μmol/L. Ten patients manifested new onset hypertension, with systolic BP 175±21 mmHg and diastolic BP 108±13 mmHg. Five patients who had a history of well-controlled hypertension manifested accelerated increase in BP 178 ±17/108±7 mmHg. One patient was normotensive, but manifested rapidly progressive oliguric renal failure with Scr increase to 969 μmol/L, massive proteinuria and hemolytic anemia.

Twelve patients (75%) had pulmonary fibrosis, 11 patients (68.8%) had cardiac involvement, 6 patients had pulmonary arterial hypertension (PAH) and 6 patients had gastrointestinal dysfunction. Cardiac involvement was common, manifested pericarditis, myocardial damage and heart failure (n=7, 43.8%, respectively). All 5 dead patients were accompanied by cardiac involvement.

Eleven patients had Raynaud’s phenomenon, 8 patients had digital ulcers, 5 patients had arthritis and 2 patients had oliguria. Thirteen patients (81%) manifested anaemia, 8 patients (50%) manifested thrombocytopenia, and 8 patients (50%) manifested microangiopathic haemolytic anaemia (MAHA).

Eleven patients (68.8%) received ACE inhibitor treatment. Fifteen patients were treated with glucocorticoid and 12 patients with immunosuppressant (Cyclophosphamide n=10, Azathiohpine n=2). After treatment, renal recovered in 4 patients (25%), kidney function improved and developed to chronic kidney disease (CKD) without dialysis in 5 patients (31%), 2 patients required permanent dialysis (13%). Five patients (31%) died.

**RESULTS:**

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 with a diagnosis ofSSc fullfilling 2013ACR/EULAR classification criteria and associated with an increased risk of SRC. Cardiac involvement was common and associated with high mortality in SRC patients.

**Disclosure of Interest:** None declared


**Abstract AB0783 – 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 with 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 with a diagnosis ofSSc fullfilling 2013ACR/EULAR classification criteria and associated with an increased risk of SRC. Cardiac involvement was common and associated with high mortality in SRC patients.

**Disclosure of Interest:** None declared


**Abstract 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 intestinal lung disease (ILD); 80% of patients present incomplete forms. The most frequently detected anti-synthetase antibody is anti-Jo1 (anti-histidyl-RNA 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 (IgG3) and Fluoroenzimunnoasyssay (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=9,4.00%9.1), 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 (RBILD) 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.

**Disclosure of Interest:** None declared


**Abstract AB0783 – Table 1**

<table>
<thead>
<tr>
<th>Age (years)</th>
<th>Follow-up (years)</th>
<th>Arthritis n(%)</th>
<th>Raynaud n(%)</th>
<th>Myositis n(%)</th>
<th>ILD n(%)</th>
<th>ILD/ Myositis n(%)</th>
<th>Exitus n(%)</th>
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</thead>
<tbody>
<tr>
<td>Anti-Jo1</td>
<td>57.3 (14.6)</td>
<td>4.0</td>
<td>9 (40.9)</td>
<td>4 (18.2)</td>
<td>18 (31.8)</td>
<td>16 (29.1)</td>
<td>9 (40.9)</td>
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<td></td>
<td>Anti-Ro52</td>
<td></td>
<td></td>
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</tr>
<tr>
<td>(n=22)</td>
<td>(2.1)</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>(31.8)</td>
</tr>
<tr>
<td>&quot;Doublet&quot;</td>
<td>59.2 (3.8)</td>
<td>6 (54.5)</td>
<td>3 (27.3)</td>
<td>9 (81.8)</td>
<td>8 (65.4)</td>
<td></td>
<td>4</td>
</tr>
<tr>
<td>Anti-Jo1</td>
<td>(15.3)</td>
<td>(2.0)</td>
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<td></td>
<td></td>
<td>(72.7)</td>
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<tr>
<td>Anti-Ro52</td>
<td>(36.4)</td>
<td></td>
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<td></td>
<td></td>
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</tr>
<tr>
<td>(n=11)</td>
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</table>

**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