FRI0471

PREDICTION OF PROGRESSION OF INTERSTITIAL LUNG DISEASE IN PATIENTS WITH SYSTEMIC SCLEROSIS: THE SPAR MODEL

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Background: The natural disease course of interstitial lung disease associated with systemic sclerosis (SSc-ILD) is highly heterogeneous. Currently, no data are available to distinguish a progressive disease course from a stable course when mild interstitial lung disease (ILD) is diagnosed in patients with systemic sclerosis (SSc).

Objectives: This study aimed to identify predictive clinical characteristics and establish a prediction model for the progression of mild ILD at 1 year follow-up in SSc patients.

Methods: Patients with SSc from two independent prospective cohorts were included in this observational study. All patients fulfilled the ACR^{EULAR 2013} criteria, had mild ILD at baseline diagnosed by HRCT (ILD extent <20% lung involvement on HRCT), available baseline and follow-up pulmonary function tests, at least one annual follow-up visit, and no concomitant pulmonary hypertension or airflow obstruction. ILD progression was defined as a relative decrease in FVC % $\geq$ 15%, or FVC% $\geq$ 10% combined with DLCO% $\geq$ 15% at 1 year follow-up. Candidate predictors for multivariate logistic regression were selected by expert opinion based on previous studies and clinical significance. Multiple imputation was used to address missing data. A prediction model for ILD progression was established in the derivation cohort and validated in the multinational validation cohort.

Results: A total of 25/98 and 25/117 SSc patients showed ILD progression in the derivation cohort and the validation cohort, respectively. Lower SpO₂ after six-minute walk test (6MWT) and arthritis ever were identified as independent predictors for ILD progression in the derivation, validation and pooled cohorts (figure 1). The optimal cut-off value for SpO₂ after 6MWT for ILD progression was determined as 94% by ROC curve analysis. In a simplified model, the presence of both SPO₂ after 6MWT $\leq$ 94% and Arthritis ever were set to 1, giving a SPAR score ranging from 0 to 2. The derived SPAR model increased the prediction rate for ILD progression from 7.4% (scoring 0) to 91.7% (scoring 2) with an AUC [95% CI] of 0.83 [0.73 to 0.93] in the derivation cohort, and a similar AUC [95% CI] of 0.82 [0.70 to 0.94] in the validation cohort.

Abstract FRI0471 – Figure 1. Multivariate logistic regression models in the derivation cohort, the validation cohort and the pooled cohort (both original and multiply imputed datasets, adjusted for age, gender, Anti-Sci-70 positive, ACA positive in the pooled cohort)

Conclusions: The evidence-based SPAR prediction model developed in our study might be helpful for the risk stratification of patients with mild SSc-ILD in clinical practice and cohort enrichment for future clinical trial design.

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TOWARDS A MULTIDIMENSIONAL PATIENT REPORTED OUTCOME MEASURES ASSESSMENT: DEVELOPMENT AND VALIDATION OF A QUESTIONNAIRE FOR PATIENTS WITH SYSTEMIC SCLEROSIS/ SCLERODERMA

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Background: Systemic sclerosis is a chronic multisystem autoimmune disorder associated with high morbidity and mortality rates. A multidisciplinary approach is necessary due to the complexity of the disease and its associated multi-organ affection. It is important to understand and monitor the impact of systemic sclerosis on the patients, provide them with high quality of care and to endorse the ownership of their disease process as early as possible to prepare them for the management this life-long illness.

Objectives: To assess the validity, reliability of a specific multidimensional patient self-reported questionnaire that can assess construct outcome measures of patients with systemic sclerosis/scleroderma.

Methods: The questionnaire was developed by integrating information obtained from patients suffering from systemic sclerosis as well as scleroderma based on the Rasch model. The questionnaire includes assessment of functional disability, quality of life, 0–10 numeric visual analogue scale (VAS) to rate the severity of the musculoskeletal pain, difficulty in breathing, gastrointestinal symptoms (e.g. swallowing difficulty/reflux/bloating/Faecal soilage/diarrhoea/constipation), Raynaud's phenomenon, fingers ulcers as well as the global assessment of the disease impact on the patient's life. In addition, the questionnaire includes 2 mannequins, one for self-reported body pains and the other one for self-reported skin tightness. Also there is a review of the possible comorbidities for the patient to tick in front of whatever he/she developed in the past month; as well as patient motivation. The questionnaire was completed by 52 consecutive patients with systemic sclerosis³¹ and scleroderma.²¹

Results: The multidimensional PROMs questionnaire was reliable as demonstrated by a high-standardised alpha (0.894–0.993). The questionnaire items correlated significantly ($p<0.01$) with clinical parameters of disease activity. Patient reported tender spots and skin tightness correlated significantly with the physician's as well as Rodnan skin scores (correlation coefficient 0.848 and 0.821 respectively). Changes in functional disability, quality of life and motivation scores showed significant variation ($p<0.01$) with diseases activity status. The PROMs questionnaire showed also a high degree of comprehensibility (9.3).

Conclusions: The developed PROMs questionnaire is a reliable and valid instrument for assessment of patients suffering from systemic sclerosis/scleroderma. Being short, rapid and comprehensive, this adds more to its applicability. The data support the value of completion of the simple 2 pages patient questionnaire, which provides a quantitative written documented record by the patient, at each visit to the rheumatologist.

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FRI0473

MPO-ANCA POSITIVITY IS RELATED TO INTERSTITIAL LUNG DISEASE IN SYSTEMIC SCLEROSIS

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Background: Recently, there were several reports that patients with systemic sclerosis (SSc) showed acute renal failure with anti-neutrophil cytoplasmic

Case	1	2	3	4	5	6	7	8
Sex	Female	Female	Female	Female	Female	Female	Female	Female
Age (years old)	73	62	34	64	64	59	58	75
Disease Duration (years)	6	30	9	3	17	26	6	25
Disease Type	Diffuse	Diffuse	Diffuse	Diffuse	Diffuse	Limited	Limited	Diffuse
Autoantibody	ARA	ACA	ATA	ATA	ATA	ATA	ND	ND
Digital Ulcer	-	+	-	-	-	-	-	+
Interstitial Lung Disease	+	+	+	+	+	+	+	+
CKD (eGFR<60 mL/min/1.73 m2)	+	-	-	-	-	-	-	-
MPO-ANCA titer (IU/mL)	14.2	10.0	63.8	38.7	9.1	11.5	20.1	13.9
PSL administration	+	+	-	+	+	+	+	-
PSL Dosage (mg/day)	1	5	-	10	10	2	6	-
Previous History of IVCY	+	-	-	+	+	-	+	-
Follow-up Period (months)	39	39	5	39	38	38	39	23
Newly Onset of AAV	-	-	+	-	-	-	-	-
Newly Onset of Acute Renal Dysfunction	+	-	+	-	-	-	-	-

ARA: anti RNA polymerase 3 antibody, ATA: anti topoisomerase 1 antibody, ACA: anti centromere antibody, ND: not detected, CKD: chronic kidney disease, PSL: prednisolone, IVCY: intravenous cyclophosphamide pulse therapy, ACEi: angiotensin converting enzyme inhibitor, ARB: angiotensin receptor blocker

antibody (ANCA) positivity. On the other hand, it is also reported that 7%–13% of patients with SSc revealed myeloperoxidase-ANCA (MPO-ANCA) positivity without vasculitis manifestation in 1990s, but their clinical characteristics were unclear. It is also unknown whether ANCA positivity leads to AAV or not in patients with SSc. It is important for physicians to clarify the characteristics of SSc patients with ANCA positivity, and answer the question whether they will shift ANCA-associated vasculitis (AAV).

Objectives: To assess the prevalence of ANCA positive patients with SSc, and clarify the characteristics of these patients.

Methods: We enrolled the 333 consecutive patients with SSc who visited our clinic during October 2014 to September 2015, all of who were checked MPO-ANCA using fluorescent-enzyme immune-assay. Clinical manifestation and laboratory data were obtained from medical chart. The data were assessed by chi-square analysis and Welch's t test.

Results: Two patients were diagnosed AAV before October 2014. Eight patients (2.4%) revealed MPO-ANCA positivity without vasculitis manifestation. All of MPO-ANCA positive patients were female, and mean age and disease duration were 61.1 years old and 17.2 years, respectively, and there's no statistically significant differences comparing MPO-ANCA negative patients. As a result of evaluating clinical manifestations, we found that patients with MPO-ANCA positivity more frequently had interstitial lung disease than patients without MPO-ANCA positivity (87.5% vs. 36.7%, $p < 0.01$). The clinical characteristics of 8 patients were shown in table 1. Only one patient out of 8 patients with MPO-ANCA positivity newly diagnosed AAV during mean of 33 months follow-up period.

Conclusions: The prevalence of MPO-ANCA positivity in SSc patients were lower than previous reports. MPO-ANCA positivity may be related to interstitial lung disease in SSc. MPO-ANCA positive patient may occasionally reveal AAV in the future, and careful observation are needed.

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Vasculitis

FRI0474

CLASSIFICATION OF SKIN INVOLVEMENT IN LEVAMISOLE-ADULTERATED COCAINE INDUCED VASCULOPATHY

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Background: Up to 88% of cocaine is tainted with levamisole, an anthelmintic withdrawn from the market due to toxicity. Since 2010 levamisole-adulterated cocaine induced vasculopathy (LACIV) patients, characterised by retiform purpura, ear necrosis, multisystemic compromise and positivity for multiple autoantibodies, have been reported. Knowing the pattern and the severity of skin involvement is essential in the approach of these patients.

Objectives: To describe the cutaneous manifestations of patients with LACIV and to propose a classification of skin involvement.

Methods: We describe the skin compromise of 30 patients with LACIV evaluated between December 2010 and May 2017. Based on this series and the review of the literature, we propose a classification according to the distribution and severity of the lesions.

Results: All patients were mestizo, median age of 31 (IQR 27–38), male:female ratio 5:1, time from symptoms to diagnosis 12 months (IQR 6–24). The most frequent clinical manifestations were skin lesions: ear necrosis (73%) and retiform purpura (83%) affecting the extensor part of the limbs, buttocks, face, and abdomen; sparing the scalp, palms and soles. Retiform purpura was classified in four grades according to distribution and severity (image). Skin biopsies revealed leukocytoclastic vasculitis (24%), pseudo-vasculitis (19%), thrombotic vasculopathy with leukocytoclastic vasculitis (19%), thrombotic vasculopathy with pseudo-vasculitis (19%), and pyoderma gangrenosum with vasculopathy (5%).

Image: LACIV retiform purpura classification. A. Grade 1: livedo reticularis or racemosa with incipient purpura (individual lesions ≤ 1 cm). **B. Grade 2:** More extended purpuric lesions which sometimes coalesce (individual lesions > 1 cm).

C. Grade 3: Purpuric lesions with haemorrhagic blisters. **D. Grade 4:** Deep purpuric lesions with associated ulceration.

Conclusions: Given the higher consumption of cocaine and its contamination with levamisole, the report of LACIV patients is increasing. A classification of the skin involvement in LACIV is proposed, according to the frequency of affection and the stratification of purpuric lesions in four degrees of severity. Cutaneous involvement is one of the pillars for the diagnosis and properly treatment, therefore a detailed description of distribution and characteristics of the lesions are fundamental for these patients care.

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