THU0430

DESCRIPTION AND PROGNOSIS FACTORS OF SYSTEMIC SCLEROSIS-ASSOCIATED INTERSTITIAL LUNG DISEASE OUTCOME ON SERIAL HRCT

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Background: Interstitial lung disease (ILD) is a leading cause of morbidity and mortality in systemic sclerosis (SSc). While factors associated with the presence of ILD in SSc (SSc-ILD) are identified, those associated with ILD outcome are still debated and studies assessing the evolution of SSc-ILD on HRCT are scarce. Yet, it is important to identify patients at risk of SSc-ILD worsening because those patients are thought to benefit the most from immunosuppressants.

Objectives: Thus, the aims of our study were: to describe the evolution of HRCT extension and patterns of SSc-ILD, to identify baseline prognosis factors of ILD outcome on serial HRCT and to investigate whether the evolution of pulmonary function tests (PFTs) parameters correlated with the evolution on HRCT.

Methods: We included 58 SSc patients with HRCT proven ILD, with at least two available HRCT, and collected clinical, biological data and PFT at baseline. We collected all HRCT and PFTs available during follow-up. We modeled PFTs and HRCT evolution using linear mixed model with random coefficients.

Results: Mean ILD extension at baseline was 32.3%±28.7%. During a mean follow-up of 5.3±4.9 years, we found a significant mean progression of ILD extension of 0.92%±0.36% per year (p=0.018). Male sex, anti-topoisomerase 1 antibodies, diffuse cutaneous SSc were associated with faster progression of ILD extension. Limited ILD according to Goh *et al.* staging system, and a coarseness score at zero (meaning 100% of ground glass opacification) were associated with a faster progression of ILD extension. We also found a significant decline of DLCO, FVC and TLC during follow-up. There was a significant correlation between the progression of ILD extension on HRCT and the decline of DLCO, but not with the evolution of FVC.

Conclusions: Male patients, patients with diffuse SSc/antitopoisomerase 1, patients with less severe and less extensive ILD at baseline were more likely to experience a faster progression of ILD extension on serial HRCT. To our knowledge, this is the first study that clearly highlighted the diffuse form of SSc/presence of antitopoisomerase 1 as a worsening factor of SSc-ILD on HRCT. FVC might not be the best mirror of ILD progression while DLCO significantly correlated with change in ILD extension. Our study helps to define the profile of patients who are going to experience a progression of ILD on HRCT during follow up.

Disclosure of Interest: None declared

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THU0431

ARE EXTREMITY TELANGIECTASES RELATED TO SEVERE DISEASE IN SYSTEMIC SCLEROSIS ?

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Background: The number and morphology of telangiectases (T) have been studied in terms of severity and organ involvement in systemic sclerosis (SSc). T are located more frequently on face and trunk than extremities.

Objectives: We aimed to evaluate the impact of the localisation of T on different skin areas in addition to number on disease severity of SSc.

Methods: SSc patients fulfilling ACR/EULAR classification criteria (2013) who had the manifestation T were included. The number of T were calculated by using telangiectasia score (TS) (Shah A. *et al*) and localisation was classified according to presence of T on extremities or not. Simultaneously; early, active and late scleroderma patterns (Cutolo *et al.*) were determined qualitatively and capillary number (CN) was calculated per linear mm at distal row quantitatively by using nail fold video-capillaroscopy (NVC) in all patients.

Results: In 113 (106 female) SSc patients with T; the mean age was 52±12, the duration of follow-up 57±62 months, Raynaud and non-Raynaud symptom 10 ±8 and 7±7 years. Limited cutaneous form was found to be in 77 (%68), ANA positivity in 102 (%90) ve anti-Scl70 positivity in 33 (%29) patients. In SSc patients with TS score ≥6 or extremity T; the duration of non-Raynaud symptom was found to be longer (p=0.010 or 0.009), MRSS and activity scores were higher (p=0.004 or 0.012 and p=0.010 or 0.009) and severity scores of general, peripheral vascular involvement and skin were higher (p=0.022 or 0.014, p=0.030 or 0.025 and p=0.006 or 0.02), digital ulcers and flexion contractures were more frequent (p=0.008 or 0.035 and p=0.027 or 0.032), late NVC pattern was more frequent and CN was lower (p=0.001 or 0.003 and p=0.001 or 0.007). When patients were classified in 3 groups according to TS and presence of extremity T, differences in terms of organ involvement, disease activity and severity scores and NVC findings were summarised in table 1.

Abstract THU0431 – Table 1. The scores of disease activity, severity and capillaroscopy in SSc patients grouped according to TS and localisation of T.

	All patients N=113	TS ≤ 6 Extremity T (-) N=77	TS ≤ 6 Extremity T (+) N=11	TS > 6 Extremity T (+) N=25	P*
Duration of non-Raynaud's (year)	6±7	6±6	8±5	10±9	P=0,008
MRSS	7,7±6,5	6±6	9±6	11±9	P=0,013
Disease activity score (Valentini)	1,4±1,2	0,7±0,9	1,2±1,0	1,6±1,4	P=0,015
Disease severity score (Medosger)	5,6±3,7	5,3±4,1	5,9±2,6	6,4±2,7	P=0,024
severity-general	0,6±0,7	0,5±0,6	0,7±0,7	1±0,8	P=0,050
severity-PVI	1,5±0,8	1,4±0,8	1,6±0,5	1,8±0,8	P=0,023
severity-skin	1,1±0,5	1±0,5	1,2±0,4	1,3±0,6	P=0,012
Clinical findings					
digital ulcer	52(%46)	29(%38)	7(%64)	16(%64)	P=0,033
Flex contr.	16(%14)	7(%9)	2(%18)	7(%28)	NS
renal crisis	1(%1)	0(%0)	1(%9)	0(%0)	P=0,010
Low DLCO	55(%49)	33(%44)	7(%70)	15(%60)	NS
Fibrosis	48(%43)	28(%38)	7(%70)	13(%52)	P=0,028
high PAB	20(%18)	10(%13)	2(%20)	8(%32)	P=0,034
IS receivers	64(%57)	39(%54)	8(%80)	17(%68)	NS
ERA receivers	30(%27)	15(%21)	3(%33)	12(%50)	P=0,028
NVC					
Normal	7(%6)	6(%9)	0(%0)	0(%0)	NS
Early	23 (%20)	19(%25)	2(%18)	2(%8)	NS
Active	22(%19)	19(%25)	1(%9)	2(%8)	NS
Late	62(%54)	33(%43)	9(%82)	21(%84)	P<0,001
Capillary number	5,8±2,0	6,4±2,1	5±2	4,9±1,8	P<0,001

NS=not significant, MRSS=modified Rodnan skin score, PVI=peripheral involvement, IS=Immunosuppressives, ERA=endothelin receptor antagonists, *Kruskal-Wallis H/Chi-Square tests

Conclusions: Disease duration was shown to be long, disease activity and severity were high and NVC findings were severe in patients with high scores of TS and extremity T. In patients with lower TS the presence of T on extremities was found to be related to severe disease. The number and localisation of T was emphasised as they are easy to evaluate in clinical practice and may be useful in determining severe patients with SSc.

Disclosure of Interest: None declared

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THU0432

PERICARDIAL EFFUSION IS AN INDEPENDENT FACTOR PREDICTIVE OF SCLERODERMA RENAL CRISIS

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Background: Scleroderma renal crisis (SRC) adversely affects renal and patient survival in systemic sclerosis (SSc).^{1, 2} The survival rate of SRC has been

improved dramatically by angiotensin-converting enzyme inhibitor therapy, but SRC still has a poor prognosis.³ Factors predictive of SRC include early diffuse skin involvement, rapid skin thickening, anti-RNA polymerase (RNAP) III antibodies, arthralgia/synovitis, and high glucocorticoid dosage.⁴ Although classical data have implicated pericardial effusion as another predictive factor of SRC,⁵ its role in SRC has not been well established.

Objectives: To clarify the clinical impact of pericardial effusion as a predictor of SRC.

Methods: Ninety-five patients diagnosed with SSc at our hospital between January 2003 and December 2017 were enrolled in the study. They were divided into a pericardial effusion group (n=21) and non-pericardial effusion group (n=74), and their clinical features retrospectively compared. Cox-regression analysis was performed to identify factors predictive of SRC.

Results: The patients comprised 14 men and 81 women with an average age of 57.4 years (range, 14 to 82) and the mean observation period was 65 months (range, 1 to 125). Pericardial effusion was detected in 21 of 95 (22.1%) cases. In the pericardial effusion group, SRC, modified Rodnan's total skin thickness score (mRSS), C-reactive protein, maximum glucocorticoid dose, proteinuria, finger apex ulcers, and interstitial pneumonia were significantly more prevalent compared to the non-pericardial effusion group. Cox regression analysis indicated that pericardial effusion (hazard ratio; HR 11.8 [95% CI 1.6–84.6], p=0.014) and anti-RNAP III antibodies (HR 11.1 [95% CI 2.0–59.6], p=0.005) were independent risk factors for SRC, while mRSS (HR 1.0 [95% CI 0.9–1.1], p=0.12), finger apex ulcers (HR 0.57 [95% CI 0.073–4.2], p=0.57), max glucocorticoid dose (HR 1.0 [95% CI 0.9–1.0, p=0.89]), and interstitial pneumonia (HR 0.9 [95% CI 0.2–3.7], p=0.96) were not. In the Kaplan-Meier method, SRC was significantly increased in the pericardial effusion group compared to non-pericardial effusion group (p<0.0001 by log rank test).

Conclusions: Pericardial effusion is another independent factor predictive of SRC in addition to anti-RNAP III antibodies.

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Vasculitis

THU0433 THE PREVALENCE OF SPONDYLOARTHRITIS IN PATIENTS WITH TAKAYASU ARTERITIS

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Background: Takayasu arteritis (TA) is characterised by inflammation of large arteries causing stenosis, occlusion, dilatation and/or aneurysm of affected vessels. TA is most commonly seen in younger women between 20–30 ages. Etiopathogenesis of TA is largely unknown although evidence suggest complex interplay between environmental and genetic factors such as HLA (human leukocyte antigen) groups. The coexistence of TA with spondyloarthropathies (SPA) has been reported in limited case series, raising hypotheses about shared pathogenetic mechanisms.

Objectives: To determine prevalence of spondyloarthropathy in patients with TA.

Methods: Detailed clinical and demographic features of TA patients were recorded and all were screened for the presence of SPA following recommendations of (ASAS). Patients were questioned for inflammatory back pain, enthesitis, uveitis, inflammatory bowel disease, peripheral arthritis, and investigated accordingly with HLA-B27, plain X-rays of pelvis and sacroiliac magnetic resonance imaging. Radiographic spondyloarthritis was reported in case of bilateral grade ≥2 or unilateral grade ≥3 sacroiliitis.

Results: There were 65 patients (61 female, 4 male) in the cohort. Mean age was 43±13 years and age at the diagnosis of TA was 35±13 years. Inflammatory bowel disease, psoriasis and psoriatic arthritis were observed in four, three and one patients. Chronic axial pain was reported by 26 (40%) patients but inflammatory back pain was evident in 13 (20%) patients. Chronic arthritis was observed in 4 patients. HLA-B27 was positive in three patients. Six patients were diagnosed as radiographic SPA and 3 were diagnosed as non-radiographic SPA. In sum nine patients were diagnosed as SPA (14.2%).

Conclusions: Our study demonstrated that SPA is common in patients with Takayasu arteritis suggesting shared pathogenetic mechanisms.

Disclosure of Interest: None declared

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THU0434 PREDICTORS OF LONG-TERM GLUCOCORTICOID THERAPY IN POLYMYALGIA RHEUMATICA: DISCONTINUATION IS MORE COMMON FOR PATIENTS TREATED WITH AMINOBISPHOSPHONATES

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Background: Glucocorticoids (GCs) are the cornerstone of polymyalgia rheumatica (PMR) therapy. Although guidelines for PMR generally recommend tapering GCs after 12–24 months, most patients are unable to discontinue GCs within the recommended time-frame. However, glucocorticoid-related adverse events can occur in up to 85% of treated cases. Patients treated with GCs should receive aminobisphosphonates (N-BPs) for the prevention of GCs-induced osteoporosis.

Objectives: In this retrospective observational study, we aimed to establish: 1) the proportion of patients with PMR who do not discontinue GCs, and 2) whether the use of N-BPs may be associated with a discontinuation of GCs.

Methods: Data were collected from electronic medical records of Rheumatology Unit at Azienda Ospedaliera Universitaria Integrata (AOUI) Verona, Italy. Patients were eligible for inclusion if they fulfilled the 2012 EULAR/ACR classification criteria for PMR. The following exclusion criteria were applied: a history of large vessel vasculitis and other diagnoses that could explain the symptoms. The main outcome was the long-term use of GCs, defined as a patient still receiving active treatment with GCs at the last evaluation available. Putative predictors included age, sex, dosage of prednisone, inflammatory markers (ESR and CRP), haemoglobin, peripheral joint involvement, use of DMARDs, number of relapses, osteoporosis and use of N-BPs. Univariable and multivariable Cox regression analyses were used to examine the association between several predictors and the outcome.

Results: 385/467 patients screened were eligible (median age 72 years [IQR 66–78], 64% females). Peripheral joint involvement was detected in 29%; 22% received DMARDs. The initial prednisone dose (median daily dose 20 mg^{13–25}) was correlated with age, haemoglobin, CRP and ESR. More than 60% of patients were treated with N-BPs, of whom only 26% were diagnosed with osteoporosis. The median follow up time was 38 months [IQR 9–57]. Disease relapse occurred in 307/467 patients (80%). GCs were discontinued in 47% after a median time of 20 months [IQR 14–27], but were restarted in 39%. At the last evaluation, 276 patients (72%) were still receiving active treatment with GCs (median daily dose 5 mg [IQR 0–8]). Multiple Cox regression analysis showed that older age (HR 1.02, 95% CI 1.00–1.04, p=0.006) and higher CRP at baseline (HR 1.24, 95% CI 1.10–1.40, p=0.001) were associated with the long-term use of GCs, whereas significant predictors of a shorter treatment duration were the use of N-BPs (HR 0.66, 95% CI 0.50–0.88, p=0.004; figure 1) and a higher initial prednisone dose (HR 0.98, 95% CI 0.96–0.99, p=0.002).

Abstract THU0434 – Figure 1. Risk function of long-term GC use for the use of antiresorptive medications.

Adjusted HR 0.66 (95% CI: 0.50, 0.88), p=0.004.