THU0430 DESCRIPTION AND PROGNOSIS FACTORS OF SYSTEMIC SCLEROSIS-ASSOCIATED INTERSTITIAL LUNG DISEASE OUTCOME ON SERIAL HRCT

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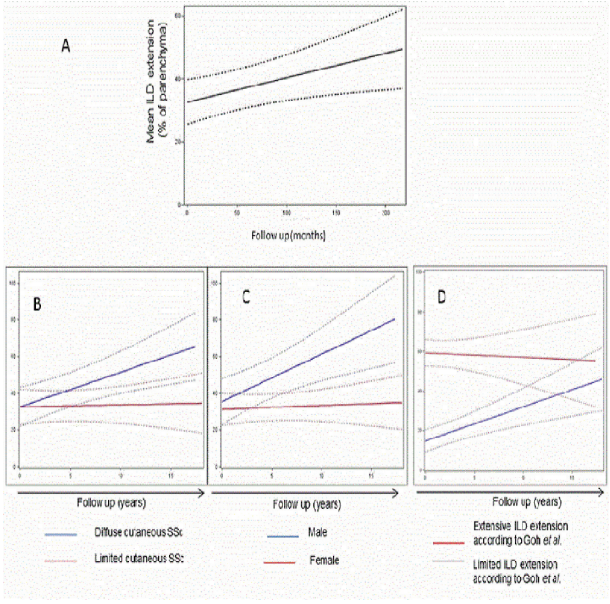
Background: Interstitial lung disease (ILD) is a leading cause of morbidity and mortality in systemic sclerosis (SSc). While factors associated with the presence of ILD in SSc (SSc-ILD) are identified, those associated with ILD outcome are still debated and studies assessing the evolution of SSc-ILD 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 modelized 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 experiment 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
DOI: 10.1136/annrheumdis-2018-eular.5493

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 sytemic 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; ealy, 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=0010 or 0,009), MRSS and activity scores were higher (p=0004 or 0012 and p=0010 or 0,009) and severity scores of general, peripheral vascular involvement and skin were higher (p=0022 or 0,014, p=0030 or 0025 and p=0006 or 0,02), digital ulcers and flexion contractures were more frequent (p=0008 or 0035 and p=0027 or 0,032), late NVC pattern was more frequent and CN was lower (p=0001 or 0003 and p=0001 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 (Medsger)		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:Immunusuppressives, 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
DOI: 10.1136/annrheumdis-2018-eular.3996

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 scleroderma (SSc)[1, 2] The survival rate of SRC has been

Ann Rheum Dis: first published as 10.1136/annrheumdis-2018-eular.3996 on 12 June 2018. Downloaded from http://ard.bmj.com/ on February 26, 2022 by guest. Protected by copyright.