Scleroderma, myositis and related syndromes

AB0401

CAN DUAL-ENERGY CT LUNG PERFUSION DETECT ABNORMALITIES AT THE LEVEL OF LUNG CIRCULATION IN SYSTEMIC SCLEROSIS (SSC)? PRELIMINARY EXPERIENCE IN 101 PATIENTS

V. Koether^{1,2}, A. Dupont³, J. Labreuche⁴, P. Felloni³, T. Perez³, P. Degroote⁵, E. Hachulla^{1,2,6}, J. Remy³, M. Remy-Jardin³, D. Launay^{1,2,6}. ¹Lille, CHU Lille, Service de Médecine Interne et Immunologie Clinique, Centre de référence des maladies autoimmunes systémiques rares du Nord et Nord-Ouest de France (CeRAINO), Lille, France; ²Lille, Université de Lille, U1286 - INFINITE - Institute for Translational Research in Inflammation, Lille, France; ³Lille, From the Department of Thoracic Imaging, Hôpital Calmette, Lille, France; ⁴Lille, Université de Lille, CHU Lille, ULR 2694-METRICS: Évaluation des technologies de santé et des pratiques médicales, France (J.L.), Lille, France; ⁵Lille, Université de Lille, CHU Lille, Département de Cardiologie, Institut National de la Santé et de la Recherche Médicale, Lille, France; ⁶Lille, Inserm-U1286, Lille, France

Background: Systemic sclerosis (SSc) is an autoimmune disorder that is characterized by a interplay of vascular abnormalities, immune system activation and an uncontrolled fibrotic response associated with interstitial lung disease affecting about 40% of patients. Identification of ILD relies on high-resolution CT that identify features suggestive of the histologic patterns of SSc(1). CT is used to determine pattern and extent of ILD and participates in the prediction of ILD progression(2).

All group of pulmonary hypertension (PH) may occur with an overall prevalence reported in up to one fifth of patient. Whereas extensive SSc-ILD can be responsible for PH, PH can also be seen as a consequence of myocardial abnormalities or as primarily affecting small pulmonary arteries and classified as pulmonary arterial hypertension.

Dual-energy CT introduction offers perspectives in the evaluation of SSc-related pulmonary manifestations. While these are not strictly perfusion images, they have been reported as adequate surrogate markers of lung perfusion (3). In the field of PH, detection of perfusion defects highly concordant with V/Q scintigraphic findings has been reported in the diagnostic approach of CTEPH but also in the differential diagnosis between PAH and peripheral forms of CTEPH (4).

Objectives: To investigate lung perfusion abnormalities in patients with SSc.

Methods: The study population included 101 patients who underwent dual-energy CT (DECT) angiography in the follow-up of SSc. CT examinations were obtained on a 3rd-generation dual-source CT system with reconstruction of morphologic and perfusion images. All patients underwent pulmonary function tests within two months of the follow-up CT scan. Fifteen patients had right heart catheterization-proven PH.

Results: Our population included patients without SSc lung involvement (Group 1; n=37), patients with SSc-related ILD (Group 2; n=56) of variable extent (Group 2a: ≤10%: n=17; Group 2b: between 11-50%: n=31; Group 2c: >50%: n=8) and patients with PVOD/PCH (Group 3; n=8). Lung perfusion was abnormal in 8 patients in G 1 (21.6%), 14 patients in G 2 (25%) and 7 patients in G 3 (87.5%). Perfusion changes were mainly composed of bilateral perfusion defects, including patchy, PE-type perfusion defects and areas of hypoperfusion of variable size. In G 1 and G 2a (n=54): (a) patients with abnormal lung perfusion (n=14) had a significantly higher proportion of NYHA III/IV scores of dyspnea (p=0.031), a shorter mean walking distance at the 6MWT (p=0.042) and a trend towards lower mean DLCO% (p=0.055) when compared to patients with normal lung perfusion (n=40); (b) a negative albeit weak correlation was found between the iodine concentration in both lungs and the DLCO% (r=-0.27; p=0.059) whereas no correlation was found with PAPs (r=0.16; p=0.29) and walking distance during the 6MWT (r=-0.029; p=0.84).

Conclusion: DECT lung perfusion provides complementary information to HRCT scans, depicting perfusion changes in SSc patients with normal or minimally infiltrated lung parenchyma.

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AB0402

SELF-ASSESSMENT OF SCLERODERMA SKIN THICKNESS: DEVELOPMENT AND VALIDATION OF THE PASTUL QUESTIONNAIRE

J. Spierings^{1,2}, V. Ong¹, C. Denton¹. ¹Royal Free and University College Medical School, University College London, Division of Medicine, Department of Inflammation, Centre for Rheumatology and Connective Tissue Diseases, London, United Kingdom; ²University Medical Centre Utrecht, Rheumatology and Clinical Immunology, Utrecht, Netherlands

Background: Evaluation of skin is central to both clinical practice and trials in systemic sclerosis (SSc). This is generally done with the modified Rodnan Skin Score (mRSS). Remote consultations are now widely implemented in response to the COVID-19 pandemic, which has inevitably limited evaluation of skin. To monitor skin during this pandemic and to further explore ways to assess skin, we developed the PASTUL (Patient self-Assessment of Skin Thickness in Upper Limb) questionnaire.

Objectives: This study evaluated feasibility and validity of PASTUL in SSc.

Methods: The PASTUL questionnaire uses a simple self-assessed grading of skin as normal, mild, moderate, or severely thickened at eight sites of upper limb corresponding to mRSS. Assessed grades were converted to an integer scale [0, 1, 2, 3]. Detailed instructions for patients were provided. Scleroderma Skin PRO (SSPRO) and Scleroderma Health Assessment Disability Index (SHAQ-DI) were also completed. For comparison, physician assessed mRSS was performed in a subgroup of patients. Construct validity was evaluated by examining the correlation between PASTUL, mRSS, SSPRO and SHAQ-DI using Pearson's correlation coefficient. Content validity was evaluated by scoring relevance, clarity and practical difficulty. Test-retest reliability was estimated using intraclass correlation coefficient (ICC).

Results: 130 patients were invited of which 104 (80%) completed the questionnaires. The mRSS was undertaken in 78 patients (n=42, 54% limited cutaneous SSc (lcSSc)). The PASTUL was completed by patients (86%) or by a partner/friend (14%). Mean PASTUL score was 11 (SD 7), mean HAQ-DI 1.41 (SD 0.77) and mean SSPRO 48.3 (SD 27.0). PASTUL strongly correlated with total SSPRO and SSPRO subdomain physical limitations (r=0.60 and 0.62, respectively) (Figure 1A). Correlations between PASTUL and mRSS and mRSS upper limbs were moderate (r=0.56 and 0.58, respectively) (Figure 1B). An overview of all correlations is provided in Table 1. Correlation between PASTUL and mRSS was stronger in lcSSc compared to diffuse cutaneous SSc patients (r=0.53 vs 0.43) and when assessed by a partner/friend compared to patients themselves (r=0.90 vs 0.54). The PASTUL demonstrated excellent test-retest reliability (ICC of 0.93, p<0.001) and good content validity.

Table 1. Correlations between PASTUL, mRSS, SHAQ-DI and SSPRO.

Outcome measure	Pearson's correlation coefficient	P-value
mRSS	0.56	<0.001
mRSS upper limbs	0.58	<0.001
SHAQ-DI	0.38	<0.001
SSPRO VAS scores		
- VAS pain	0.28	0.004
- VAS GI	0.01	0.333
- VAS breathing	0.17	0.168
- VAS RP	0.16	0.114
- VAS DU	0.26	0.008
- VAS Limitations	0.32	0.001
SSPRO	0.60	<0.001
SSPRO subdomains		
- Physical effects	0.59	<0.001
- Physical limitations	0.62	<0.001
- Emotional effects	0.48	<0.001
- Social effects	0.42	<0.001

DU, digital ulcers; GI, gastrointestinal; mRSS, modified Rodnan Skin Score; PASTUL, Patient self-Assessment of Skin Thickness in Upper Limb; RP, Raynaud's Phenomenon; SD, standard deviation; SHAQ-DI, Scleroderma Health Assessment Questionnaire Disability Index; SSPRO, Scleroderma Skin Patient-Reported Outcome; VAS, visual analogue score

Conclusion: The significant correlation of PASTUL scores with total SSPRO and physical limitation scores and moderate correlation with mRSS support the potential of PASTUL for remote evaluation of skin thickness in virtual clinical settings. Future studies may explore sensitivity to change and utility in clinical trials.

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