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

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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

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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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Figure 1. Correlations between PASTUL and SSPRO (A) and mRSS (B)

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AB0403

ORBITAL MYOSITIS: AN UNCOMMON OPHTHALMIC PRESENTATION IN RHEUMATIC DISEASES - CASE SERIES AND SYSTEMIC REVIEW OF CASE REPORTS

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Background: Ophthalmic conditions are common manifestations in patients with rheumatic diseases (RDs), orbital myositis (OM) remain rare. Only 1 case each for scleroderma and undifferentiated connective tissue disease (UCTD) reported. **Objectives:** Report frequency of Rheumatic Diseases with OM (RDs-OM), diagnosis and treatments.

Methods: Patients database were obtained from trust electronic record and literature review of case reports were performed.

Results: 4 out of 7 patients in our clinic with RDs-OM. Both Scleroderma-myositis (SCM) patients were positive for anti-PM/SCL antibody. All received glucocorticosteroid (GCS) and Mycophenolate as steroid-sparing or rescue therapy with good tolerance and outcome. One idiopathic OM has residual muscle paresis.

Table 1. Case series of patients with Orbital Myositis

	Idiopathic OM (n=3)	Rheumatology Diseases (n=4)	p-value
Age (years)	48 (27-72)	42.75 (21-60)	P= 0.8571
Gender – Female (Male)	2 (1)	4(0)	
Median Period Of Follow Up (months)	24.60 (7-37)	47.25(1-120)	
Underlying RD			
I.UCTD	-	1	
II.Overlap Scleromyositis	-	2	
III.Behcet's Disease	-	1	
Biopsy			
I.Pathological significant	1	1	
II.Normal	1	1	
III.Not done	1	2	

Total 93 cases with RDs-OM reviewed and the characteristic of patients represented in Figure 1.

Figure 1. Characteristics of orbital myositis in patients with rheumatic diseases. F: female, M: male, NS: not-specified, PrePD: onset before RD, AtD: onset with RD, PostPD: onset after RD, GCS 1st: Glucocorticoid as first line treatment, GCS res: Glucocorticoid responder, BxOM: biopsy of extra-ocular muscle, OtherTs: Other Tissue A: Median age at diagnosis, Sarc: sarcoidosis, SLE: Lupus, EGPA: Eosinophilic Granulomatous Polyangiitis, GPA: Granulomatous Polyangiitis, RA: rheumatoid Arthritis, BD: Behcet's Disease, RP: Relapsing Polychondritis, DM: Dermatomyositis, LoSSC: Localized Scleroderma, SPA: Spondyloarthropathy, ASOD: Adult Onset Still's Disease, RPF: Retroperitoneal Fibrosis, GCOM: Giant Cell OM, SJS: Sjogren Syndrome, DL: Discoid Lupus, KD: Kawasaki Disease, SAPHO: Synovitis, acne, pustulosis, hyperostosis, and osteitis, APLS: Antiphospholipid Syndrome.

OM although is rare but can present in most RDs. Our review showed OM commonest in patients with RA, GPA, EGPA and sarcoidosis. Only 1 each for SCM, APLS, UCTD, SAPHO and Kawasaki Disease.

Most RDs-OM patients are female. 89.2% OM diagnosed concurrently with primary RD diagnosis. Majority RA, BD and GPA developed OM along the course of RDs (83%-90%).

Except for routine RDs screening, thyroid eye disease, infections, malignancy (eg. lymphoma) and myasthenia gravis, atypical infective screenings include mycobacterium, fungal, *Borrelia* and spirochetes were excluded. Histopathological diagnosis is important diagnostic tool for IgG4 disease, neoplasm, sarcoidosis and vasculitis. 59% patients underwent tissue biopsy, 71% of these were sampled from EOM, some from involved organs. Orbital MRI or CT were the commonest diagnostic imaging, 5 reported had Ocular ultrasound as single or complementary imaging and 1 had PET-CT for exclusion of other diagnosis.

Only 40% GPA and 20% of EGPA patients were positive for ANCA either c-ANCA, p-ANCA and MPO which is similar in other literatures.

97.8% (n=91) patients received GCS with 79% of these patients reported improvement or resolution of ocular symptom with or without steroid-sparing agents. Immunosuppressants used are those not foreign among rheumatologists while biologics prescribed for refractory cases were Rituximab(2), Infliximab(4), Adalimumab(2) and Tocilizumab(1). Orbital decompression and orbital radiation were other measures. 13 patients had residual EOM paresis.

Conclusion: 1.RDs-OM is increasing recognized.

2.Most OM in RA, BD and GPA occurs along its primary RDs.

3.40% GPA and 20% of EGPA patients were ANCA positive.

4.GCS remain the primary treatment, three quarter patients responded.

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AB0404

EXTENDED MYOSITIS PANEL AND THE CLINICAL ASSOCIATION IN PATIENT WITH SUSPECTED INFLAMMATORY DISEASE, A RETROSPECTIVE STUDY

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Background: The idiopathic inflammatory myopathies (IIMs) are a heterogeneous group of conditions characterized by proximal muscle weakness. Autoantibodies are identified in more than 80% of patients with polymyositis (PM) or dermatomyositis (DM). Some are also found in other connective tissue diseases (CTD), while others are more specific to IIM. Thus, they are classified into two categories named myositis associated antibodies (MAA) and myositis specific antibodies (MSA). MSA have been reported as being 90% specific for IIM while MAA are found in up to 50% of myositis patients. ¹

Objectives: The aim of the study was to evaluate the myositis antibody prevalence and to assess the associations of these antibodies with clinical manifestations, final diagnosis, medication received and outcomes.

Methods: A retrospective chart review study was conducted at St. James's Hospital from 2015-2020. All positive myositis panels were obtained. The MAA evaluated were PMScl (100/75), U1snRNP, Ku and Ro52, while MSA were Mi2a, Mi2b, TIF1, MDA5, NXP2, SAE1, Jo-1, SRP, PL-7, PL-12, EJ and OJ.

Results: We identified 52 patients who were positive for one or more MSA/MAAs. The mean age was 58.9 years, the majority were female (65.3%). The most prevalent MAA was anti-Ro52 (29/52) followed by anti-PMScl 100/75 (7/52), anti-Ku and anti-U1RNP were seen in (3/52) each. The prevalence of MSA were (4/52) for anti-PL12 and (3/52) for anti-SAE1 followed by (2/52) for each of anti-Mi2b, anti-NXP2, anti-Jo, anti-SRP, anti-PL7, anti-EJ and anti-OJ while only (1/52) for anti-MDA5. The most common observed clinical phenotypes in our cohort were arthralgia (20/52), ILD (18/52) and cutaneous manifestations (15/52). Less than a quarter of the studied population had arthritis (8/52), myositis (7/52), raynauds (7/52) and malignancy (4/52). Various diagnosis were allocated for these patients, while only eight cases were diagnosed with dermatomyositis. Medication received and the final outcome for those with strong positive MSA were summarized in table1.

Conclusion: IIM was the final diagnosis in only 15% of positive myositis panels and the presence of antisynthetase antibodies didn't necessarily indicate the presence of antisynthetase syndrome thus signifying low specificity in our cohort.

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