DIFFERENTIATING THE DOMINANCE OF PULMONARY VASCULAR DISEASE OR INTERSTITIAL LUNG DISEASE ON HEMODYNAMIC ABNORMALITIES IN SYSTEMIC SCLEROSIS AND CLARIFYING EACH CHARACTERISTIC BY USING QUANTITATIVE EVALUATION OF CHEST CT

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Background: Group 1 and 3 pulmonary hypertension (PH) develop through different pathological mechanisms but have similar hemodynamic abnormalities. Systemic sclerosis (SSc) is associated with both pulmonary vascular disease (PVD) and interstitial lung disease (ILD), making it challenging to differentiate group 1 and 3 PH in those patients. A previous study using quantitative evaluation of chest computed tomography (CT) demonstrated that normal lung volume was inversely correlated with mean pulmonary arterial pressure (mPAP) in patients with group 3 PH (1).

Objectives: In this study, we aimed to assess the dominance of PVD or ILD in SSc patients by quantitative evaluation of chest CT and to evaluate each characteristic.

Methods: A total of 76 SSc patients who underwent right heart catheterization (RHC) were included. Chest CT was evaluated by using a software (Synapse Vincent Ver.3.0, Fujitlim) which quantified normal and total area of the lung. Then, we calculated abnormal area by drawing normal area from total area in the lung (%). Pulmonary function test (PFT) and serum biomarkers, such as KL-6 and LDH, were also evaluated. The dominance of PVD or ILD was defined as divergent or parallel change between the first and last assessments, respectively. In mPAP and abnormal area in the lung calculated using the software. Increase or decrease by over 10% in the last assessment compared to the first assessment was considered as a significant change in mPAP or abnormal area in the lung.

Results: The median (range) values of mPAP and abnormal area in the lung at baseline were 23 [9-65] mmHg and 30.2 [0-100] %, respectively. Of 37 SSc and PH patients, 18 were defined as having PVD dominance while 19 as ILD dominance. Abnormal area in the lung at baseline was greater in patients with ILD dominance compared to those with PVD dominance (39.1 [16.3-98.3] v.s. 14.0 [0-99] %, p=0.002), whereas mPAP was higher in patients with PVD dominance than those with ILD dominance (42.5 [23.0-65.0] v.s. 28.0 [16.0-42.0] mmHg, p=0.002). PFT parameters including forced vital capacity were not different between the two groups. The ratio of mPAP/KL-6 showed a great difference between the two groups with its significant elevation in patients with PVD dominance (p=0.007).

Conclusion: Quantitative evaluation of chest CT showed great efficiency in differentiating the dominance of PVD or ILD in patients with SSc and PH. In addition, the ratio of mPAP/KL-6 may be easily used as a parameter for dominance evaluation.

REFERENCES:

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ELIGIBILITY FOR ANTI-FIBROTIC TREATMENT WITH NINTEDANIB OF PATIENTS WITH SYSTEMIC SCLEROSIS RELATED INTERSTITIAL LUNG DISEASE

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Background: Pulmonary involvement is the main determinant of disease-related mortality in systemic sclerosis (SSc). The SENSCIS and INBUILD trials have documented the efficacy of the antifibrotic drug Nintedanib (NTN) in the treatment of SSc-related interstitial lung disease (SSc-ILD) and progressive forms of ILD. However, the use of this drug in patients with SSc-ILD in clinical practice are currently being defined.

Objectives: To evaluate the proportion of patients eligible for NTN treatment based on the enrolment criteria for the SENSCIS and INBUILD studies in a real-life cohort of SSc-ILD patients.

Methods: We considered consecutive patients with ILD extension on CT ≥10% at baseline or disease duration ≥5 years with at least one visit in the periods between September 2016 and December 2019. Data of the clinical visits were retrospectively evaluated. For each patient, we examined the visits during which a well-tolerated immunosuppressive therapy was modified because of progression of SSc-ILD and the most recent follow-up visit. Eligibility of patients for NTN was defined according to the inclusion criteria of the SENSCIS and INBUILD trials. Patients with more than 2 acral ulcers at the time of evaluation, history of digital amputation, pulmonary hypertension (functional class III-IV) and increased hemorrhagic or thrombotic risk were judged not eligible to NTN in trials.

Results: A total of 177 visits regarding 78 patients were examined (females 80.8%, diffuse skin disease 51.3%, anti-Scl70 antibodies positivity 55.7%, age 54.8±16.0 years, disease duration 4.0±2.4 years). Considering the visits in which a therapeutic change was given, 54 patients (54.5%) were eligible for NTN according to SENSCIS criteria and of these 31 (31.3%) also according to INBUILD criteria (Figure 1). In this group, 25 patients were treated with mycophenolate mofetil, 11 with azathioprine, 10 with cyclophosphamide, 7 with methotrexate and 8 with rituximab (2 in combination). At the latest available evaluation, 42 patients (62.8%) were eligible for NTN according to SENSCIS criteria and of these 12 (15.4%) also according to INBUILD criteria (Figure 1). In this group, 30 patients were in mycophenolate mofetil (6 in combination with biologic treatment), 5 in azathioprine, 1 in cyclophosphamide, 1 in methotrexate, 6 in rituximab, 2 in tocilizumab and 1 in pirenidone. Overall, the factors limiting NTN start according to the trial enrollment criteria would have been: uncompromised (19.2%) or too low (6.4%) DLco values, too low FVC (3.4%), severe acral disease with ulcers (16.9%), severe pulmonary arterial hypertension (6.2%), increased thrombotic or haemorrhagic risk (6.2%). In the scenario of eligibility, skin progression would be detectable in 43.4% of all visits.

Conclusion: Treatment that can modify the progression of SSc-ILD are currently limited. Based on our retrospective analysis, the use of NTN in accordance with current clinical evidence could be considered in a significant percentage of patients with SSc-ILD.

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REDUCED BONE MINERAL DENSITY IN PATIENTS WITH IDIOPATHIC INFLAMMATORY MYOPATHIES: A CASE CONTROL STUDY

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Background: Idiopathic Inflammatory Myopathies (IIMs) patients are at risk of bone mineral density (BMD) loss due to systemic inflammation, use of glucocorticoids (GCs) and disability. Cross sectional study showed 70% of IIMs patients had reduced BMD but whether they were at excessive risk compared to controls were unknown.

Objectives: To compare the prevalence of reduced BMD between IIMs patients, non-rheumatological controls, rheumatoid arthritis (RA) and systemic lupus erythematosus (SLE) patients and to determine the clinical determinants of BMD in IIMs patients.

Methods: This was a single centre retrospective case control study. BMD at lumbar spine L1-L4 and neck of femur (NOF) were assessed by dual-energy X-ray absorptiometry (DXA) scans. The prevalence of reduced BMD and osteoporosis in Chinese IIMs patients and age-and-sex-matched non-rheumatological controls were compared. The BMD of female IIMs were then compared to age matched female RA and SLE patients in the secondary analysis. Binary logistic regression was used for adjustment of confounders. The demographics and