The Relevance of High-Resolution Computed Tomographic Findings and Pulmonary Arterial Hypertension in Systemic Sclerosis-Associated Interstitial Lung Disease

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Objective: *To compare the high-resolution computed tomographic (HRCT) findings between systemic sclerosis-associated interstitial lung disease (SSc-ILD) with and without pulmonary arterial hypertension (PAH), as well as to correlate the calculated HRCT scores and the estimated systolic pulmonary artery pressure (sPAP).*
Material and Method: *The medical records of all SSc-ILD patients who presented at the Rheumatology Clinic, Chiang Mai University Hospital were retrospectively reviewed. Patients with the availability of echocardiography performed within six months of the corresponding HRCT were included. The extent of ground glass, lung fibrosis, and honeycombing were scored. The maximum diameter of the main pulmonary artery (MPAD) and ascending aortic diameter (AD) were measured. The PAH was defined by sPAP ≥45 mmHg.*
Results: *Fifty patients with SSc-ILD diagnosed with HRCT were included. Echocardiography identified 19 (38.0%) patients with PAH. The SSc-ILD with PAH had significantly higher mean (SD) lung fibrosis (9.9 [3.6] vs. 7.8 [3.5], p = 0.03), and CT-total scores (20.5 [6.9] vs. 14.9 [6.2], p<0.01) than those without PAH. In the total group, the CT-total score correlated positively with sPAP (r = 0.384, p<0.01). No significant correlation of MPAD or MPAD/AD with sPAP was found.*
Conclusion: *SSc-ILD with PAH had more severe lung fibrosis than those without PAH. The calculated total HRCT score may be useful to identify PAH in SSc-ILD.*

Keywords: *High resolution computed tomography (HRCT), Systemic sclerosis (SSc), Echocardiography, Interstitial lung disease (ILD), Pulmonary arterial hypertension (PAH)*

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Interstitial lung disease (ILD) and pulmonary arterial hypertension (PAH) are the most common pulmonary complications[1], and are now the leading causes of morbidity and mortality in systemic sclerosis (SSc)[2]. The prognosis of SSc associated PAH (SSc-PAH) is poor, independent of ILD[3,4].

Right-heart catheterization (RHC) serves as the gold standard test for the diagnosis of PAH in SSc. However, it is impractical as a screening test because the RHC is invasive, associated with complications, and costly. Currently, echocardiography is a practical measure for screening SSc patients with suspected clinical PAH, although not fully validated[5].

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High-resolution computed tomography (HRCT) of the chest is a sensitive measure of the extent and severity of lung involvement in systemic sclerosis associated interstitial lung disease (SSc-ILD). Currently, there are many HRCT scoring methods used for scoring the extent and severity of ILD on HRCT, but these are not yet fully validated as outcome measures of SSc-ILD[6-10].

The usefulness of HRCT-determined extent of fibrosis to identify PAH in ILD patients have been investigated, and the results were controversial. Zisman et al[10] demonstrated that the extent of pulmonary fibrosis on HRCT, main pulmonary artery diameter (MPAD) and the MPAD/ascending aortic diameter (AD) ratio did not differ between advanced idiopathic pulmonary fibrosis (IPF) patients with and without PAH determined by RHC. However, a recent study reported that the fibrotic score from HRCT, the MPAD and the MPAD/AD ratio correlated with the

peak pulmonary artery pressure determined by echocardiography in patients with SSc[11].

The aim of the present study was to compare the HRCT findings that consisted of the calculated fibrotic HRCT scores, the MPAD, and the MPAD/AD ratio between SSc-ILD with and without PAH. Additionally, we also determined the correlation of the calculated HRCT scores with the estimated systolic pulmonary arterial pressure (sPAP) obtained by echocardiography.

Material and Method

Patients

The study received ethics approval from the Research Ethics Committee of the Faculty of Medicine, Chiang Mai University for the chart review and data analysis, following the Ethics Principle of Declaration of Helsinki. The medical records of all patients with SSc who presented at the Rheumatology Clinic, Chiang Mai University Hospital, between March 2005 and 2010, diagnosed as ILD, confirmed by HRCT of the chest were retrospectively reviewed. If patients had multiple HRCT results, the first available HRCT results were used for the analysis. The rheumatologists who took care of SSc patients in the rheumatology clinic decided on the need for HRCT of the chest to confirm the diagnosis of ILD, as well as the need for echocardiography to identify PAH.

Patients were selected for the present study if they fulfilled all of the following criteria: (1) diagnosis of SSc according to the criteria of the American College of Rheumatology[12], (2) 18 years and older, (3) availability of results of HRCT, and (4) had echocardiography performed within six months of the corresponding HRCT. The SSc patients were classified as having limited cutaneous SSc (LcSSc) or diffused cutaneous SSc (DcSSc), according to LeRoy's and Medsger's[13] classification criteria. Patients were excluded if they had an overlap syndrome or had ILD secondary to conditions other than SSc. Patients with inadequate TR regurgitant signal, left ventricular dysfunction (% of LVEF <50%), congenital heart disease, porto-pulmonary PAH, or HIV infection were also excluded. The medical records were reviewed for the demographic data, clinical characteristics, New York Heart Association Functional Classification (NYHA FC), SpO_2, current medications used, laboratory investigations, sPAP, and% of LVEF measured by echocardiography. The SpO_2 was measured through a digital probe oximeter.

We defined PAH as an estimated systolic pulmonary artery pressure (sPAP) of ≥45 mmHg measured by echocardiogram. All echocardiograms including Doppler interrogations were obtained with sonography machines (Agilent Sonos® 4500, 5500 [Andover, MA, USA]). An experienced cardiologist (N.P.) reviewed all of the echocardiographic parameters blinded to clinical and laboratory data. The systolic pulmonary artery pressure was estimated[14] by using the summation between the estimated right atrial pressure (RA) and the calculated systolic transtricuspid gradient (ΔP). By using the maximum velocity (V) of the regurgitant jet, ΔP was derived by the modified Bernoulli equation ($\Delta P = 4V^2$). The RA pressure estimation was derived from the inferior vena cava diameter variation between end-expiration and sniff.

Chest HRCT scoring system

All HRCTs were obtained with 1 of 2 MDCT platforms (Somatom Definition, [Siemens, Forchheim, Germany] or Aquilion 16 [Toshiba, Tochigi-Ken, Japan]). The volumetric scan technique was performed with a high spatial resolution of 1-mm thickness and interval image reconstruction in the supine position with deep inspiration. Sampling the HRCT with 1-mm slice thickness during expiration was constructed with at least six levels to cover the whole thorax. To exclude ground glass opacity from dependent atelectasis, prone inspiratory HRCT was done to cover the suspected area.

All images were reviewed by thoracic radiologist with 17 years of experience (J.E.), blinded to clinical and laboratory data. Soft copy DICOM images were retrieved from a picture archiving and communication system (PACS, CMUPACS; Faculty of Medicine, Chiang Mai, Thailand) and axial and multiplanar reformatted (MPR) images were reviewed with workstations (version 2.0.1; Panacea, Bangkok, Thailand). The extent of parenchymal abnormalities was classified into three categories: ground-glass opacity (GG), lung fibrosis (Fib), and honeycombing (HC). The following radiographic definitions were used: GG, hazy parenchymal opacity with preserved underlying bronchovascular structure without architectural distortion; Fib, interlobular septal thickening, intralobular lines, traction bronchiectasis, and traction bronchiolectasis; and HC, clustered air-filled cyst with a dense wall.

The calculated HRCT-determined fibrosis method used in the present study was derived from Patiwetwitoon et al[9], which scores the amount of

GG, Fib, and HC of each lung lobe (right upper, right middle, right lower, left upper, and left lower) by using a Likert scale (0 = absent; 1 = 1-25%; 2 = 26-50%; 3 = 51-75%; 4 = 76-100%) modified from the method of Kazerooni et al[8]. The total (t)-GG, t-Fib, and t-HC scores were computed by the summation of all of the GG, Fib, and HC scores from the five lung lobes, respectively, in which each of the total scores ranged from 0 to 20. The t-GG, t-Fib, and t-HC scores were aggregated to produce a total calculated HRCT score (CT-total) ranging from 0 to 60.

The MPAD was measured at its widest dimension on the supine full-chest sequence. At this same level, the widest AD was measured. The ratio of MPAD to AD (MPAD/AD) was calculated.

Statistical analysis

Data were expressed as a percentage or mean ± SD. The mean values of the HRCT variables between SSc-ILD patients with and without PAH were compared using the Student's t-test or Mann-Whitney U test. Comparison of categorical variables used the Chi-square test or Fisher's exact test, as appropriate. Spearman's correlation coefficients (r-value) were used to determine the correlations of the sPAP and the HRCT variables consisting of the calculated HRCT scores, the MPAD, and the MPAD/AD ratio. All data analyses were performed using SPSS software (version 17.0; Chicago, Illinois). All tests were 2-tailed, and $p<0.05$ was considered significant.

Results

Patient characteristics

Of the 60 SSc-ILD patients with available echocardiography performed within six months of the corresponding chest HRCT were initially enrolled, two were excluded because they had the overlap syndrome of SSc with rheumatoid arthritis (1 patient) and with systemic lupus erythematosus (1 patient). Seven cases were excluded because the echocardiographic images were suboptimal for assessing the transtricuspid regurgitant jet and one was excluded because of a left ventricular ejection fraction of <50% by echocardiography, which left a total of 50 SSc-ILD who were analyzed.

The demographic and clinical characteristics of SSc-ILD population studied (n = 50) were summarized in Table 1. Of the 50 SSc-ILD, 33 (66%) woman and 33 (66%) were classified as having DcSSc. The mean (SD) age was 54.8 (11.7) years and mean (SD) disease duration from non-Raynaud's phenomenon to undergoing the HRCT was 3.8 (4.2) years (range 0-15.7 years). The mean (SD) duration of time from echocardiogram study to the time of the HRCT chest evaluation was 0.4 (2.2) months (range ±6 months).

Nineteen (38%) of 50 SSc-ILD patients had sPAP ≥45 mmHg, which were classified as SSc-ILD with PAH. There was a significantly higher proportion of male gender in the SSc-ILD with PAH group than those without PAH (52.6% vs. 22.5%, $p = 0.029$). The SSc-ILD with PAH had more currently used azathioprine than those without PAH (15.8% vs. 0%, $p = 0.049$). There was higher proportion of current calcium channel blocker usage in SSc-ILD without PAH patients than those with PAH, (90.3% vs. 47.4%, $p = 0.002$). There were no significant differences among the SSc-ILD with and without PAH with respect to age, disease duration, disease subtypes (DcSSc or LcSSc), clinical symptoms (cough, dyspnea), NYHA FC, SpO_2, and basic laboratory investigations.

The antinuclear antibody (ANA) and the anti-topoisomerase (Scl-70) were tested in 42 (84%) and 33 (66%) of 50 SSc-ILD patients, respectively. The ANA test was positive in 42 (100%) of 42 patients and the anti-Scl-70 test was positive in 27 (81.8%) of 33 patients. Of the 42 patients who had ANA tested, no centromere pattern was found.

Comparisons of HRCT and echocardiographic findings between SSc-ILD patients with and without PAH

HRCT findings of the study population showed that lung fibrosis (Fib) was the most common lung abnormality, which was found in all 50 (100%) patients. The GG and HC were seen in 41 (82%) and 28 (56%) of 50 SSc-ILD patients, respectively. Regarding the regional distribution of abnormalities, the HRCT scan findings of Fib, GG, and HC were the most common in the lower lung zones in the absence of right or left predominance (data not shown).

As shown in Table 2, SSc-ILD with PAH patients had significantly higher mean (SD) t-Fib scores (9.9 [3.6] vs. 7.8 [3.5], $p = 0.03$) and the CT-total scores (20.5 [6.9] vs. 14.9 [6.2], $p<0.01$) than those without PAH, respectively. There were no significant differences between SSc-ILD patients with and without PAH with respect to t-GG, t-HC, the MPAD, and the MPAD/AD ratio.

Echocardiographic findings consisting of the percentage of LVEF, proportion of right ventricular dysfunction, and proportion of diastolic dysfunction

Table 1. Clinical characteristics of the study population

	Total (n = 50)	SSc-ILD-PAH (sPAP ≥45 mmHg) (n = 19)	SSc-ILD (sPAP <45 mmHg) (n = 31)	*p*-value*
Demographic				
Age at HRCT, mean (SD), year	54.8 (11.7)	57.2 (12.6)	53.3 (10.9)	0.257[a]
Gender, male/female, n	17/33	10/9	7/24	0.029
Disease duration since NRP, mean (SD), year	3.8 (4.2)	3.2 (4.3)	4.2 (4.2)	0.303[b]
SSc subtype, DcSSc/LcSSc, n	33/17	14/5	19/12	0.369
Clinical, n (%)				
Cough	25 (50.0)	12 (63.1)	13 (41.9)	0.145
Dyspnea	41 (82.0)	15 (78.9)	26 (83.8)	0.715
NYHA FC, n (%)				0.233
I	7 (14.0)	4 (21.1)	3 (9.7)	
II	27 (54.0)	7 (36.8)	20 (64.5)	
III	13 (26.0)	7 (36.8)	6 (19.4)	
IV	3 (6.0)	1 (5.3)	2 (6.4)	
% SpO_2, mean (SD)	92.4 (7.0)	90.1 (9.2)	93.8 (4.9)	0.120[a]
Current medications, n (%)				
Cyclophosphamide	11 (22.0)	2 (10.5)	9 (29.0)	0.170
Azathioprine	3 (6.0)	3 (15.8)	0 (0)	0.049
Prednisolone	17 (34.0)	6 (31.6)	11 (35.5)	0.777
Calcium channel blocker	37 (74.0)	9 (47.4)	28 (90.3)	0.002
Beraprost	4 (8.0)	2 (10.5)	2 (6.4)	0.629
Laboratory investigation, mean (SD)				
Hemoglobin, g/dL	11.6 (1.8)	11.9 (1.9)	11.3 (1.7)	0.242[a]
Creatinine, mg/dL	0.8 (0.3)	0.9 (0.4)	0.8 (0.1)	0.057[a]
Aspartate aminotransferase, U/L	35.8 (21.4)	38.8 (26.8)	33.9 (17.4)	0.555[b]
Alanine aminotransferase, U/L	25.1 (17.4)	26.6 (17.9)	24.1 (17.2)	0.545[b]

SSc = systemic sclerosis; ILD = interstitial lung disease; PAH = pulmonary arterial hypertension; sPAP = estimated systolic pulmonary artery pressure; HRCT = high-resolution computed tomography; NRP = non-Raynaud's phenomenon; DcSSc = diffused cutaneous SSc; LcSSc = limited cutaneous SSc; NYHA FC = New York Heart Association Functional Classification

* SSc-ILD-PAH vs. SSc-ILD-without PAH

[a] *p*-value from Student's t test, [b] *p*-value from Mann-Whitney U test

were comparable in both groups studied. As expected, the mean (SD) sPAP of the SSc-ILD with PAH were higher than the SSc-ILD without PAH (55.4 [10.6] vs. 32.6 [7.6], $p<0.01$).

Correlation between the calculated HRCT scores and sPAP

Table 3 summarizes the correlation between the calculated HRCT scores and sPAP. In the entire group of SSc-ILD studied (n = 50), the CT-total score showed a significant correlation with the sPAP ($r = 0.384$, $p<0.01$). However, no significant correlations of the t-GG, t-Fib, t-HC scores, the MPAD and the MPAD/AD ratio with the sPAP were observed. When dividing SSc-ILD to two subgroups including SSc-ILD with PAH (sPAP ≥45 mmHg, n = 19) and SSc-ILD without PAH (sPAP <45 mmHg, n = 31), there were no significant correlations of the t-GG, t-Fib, t-HC, CT-total scores, the MPAD and the MPAD/AD ratio with sPAP in both subgroups.

Discussion

PAH has been demonstrated as a major cause of mortality in SSc independent the presence of ILD[3,4]. Noninvasive approaches to the identification of PAH in SSc-ILD patients are needed. Recently, HRCT-determined fibrotic score, MPAD and MPAD/AD ratio has been used as a CT surrogate for pulmonary artery pressure on echocardiography in SSc patients[11]. In the present study, we found that SSc-ILD with PAH had more severe lung fibrosis (t-Fib and CT-total scores) than those without PAH. Further, the CT-total score does correlate with sPAP on echocardiography, which suggests that the

Table 2. HRCT and echocardiography findings of the study population

	Total (n = 50)	SSc-ILD-PAH (sPAP ≥45 mmHg) (n = 19)	SSc-ILD (sPAP <45 mmHg) (n = 31)	*p*-value*
The calculated HRCT scores, mean (SD)				
t-GG	5.6 (3.9)	6.5 (3.5)	5.1 (4.0)	0.168[b]
t-Fib	8.6 (3.6)	9.9 (3.6)	7.8 (3.5)	0.033[b]
t-HC	2.9 (3.8)	4.0 (4.8)	2.1 (2.9)	0.402[b]
CT-total	17.1 (6.9)	20.5 (6.9)	14.9 (6.2)	0.005[b]
The pulmonary artery diameter, mean (SD)				
MPAD, cm	3.0 (0.5)	3.1 (0.5)	2.9 (0.5)	0.181[a]
MPAD/AD ratio	0.9 (0.2)	0.9 (0.2)	0.9 (0.2)	0.739[a]
Echocardiography findings				
sPAP, mean (SD)	41.3 (14.2)	55.4 (10.6)	32.6 (7.6)	<0.001[a]
% LVEF, mean (SD)	68.0 (8.7)	67.9 (8.9)	68.1 (8.6)	0.977[a]
RV dysfunction, n (%)	6 (12.0)	4 (21.0)	2 (6.4)	0.184
Diastolic dysfunction, n (%)	4 (8.0)	2 (10.5)	2 (6.4)	0.618

t-GG = total ground glass score; t-Fib = total fibrosis score; t-HC = total honeycombing score; CT-total = a total CT score; MPAD = the maximum diameter of the main pulmonary artery; MPAD/AD ratio = the ratio of MPAD and the widest ascending aortic diameter (AD); sPAP = estimated systolic pulmonary artery pressure; % LVEF = % of left ventricular ejection fraction; RV = right ventricular

* SSc-ILD-PAH vs. SSc-ILD-without PAH

[a] *p*-value from Student's t test, [b] *p*-value from Mann-Whitney U test

Table 3. Spearman's correlation coefficients (r) between the calculated HRCT scores and sPAP of SSc-ILD

Variables	Correlation coefficient (r)					
	t-GG	t-Fib	t-HC	CT-total	MPAD	MPAD/AD
sPAP (total, n = 50)	0.159	0.264	0.149	0.384[a]	0.241	0.091

[a] $p<0.01$

cumulative amount of pulmonary fibrosis as measured by summation of the HRCT-determined fibrosis score (CT-total score) impacts the sPAP. However, no significant correlations of the MPAD and the MPAD/AD ratio with the sPAP were found, which suggests that the MPAD and the MPAD/AD ratio do not help in identifying PAH in our SSc-ILD patients.

Most of the present study population had DcSSc (66%) and the mean (SD) disease duration since non-Raynaud's phenomenon to undergoing the HRCT of chest to investigate ILD was 3.8 (4.2) years. The abnormal parenchymal findings on HRCT were Fib, GG, and HC, which were observed in 100%, 82%, and 56%, respectively. Thus, approximately half (56%) of our SSc-ILD patients had a pattern of usual interstitial pneumonitis (UIP), which revealed HC on HRCT. In the present study, 19 (38%) SSc-ILD patients were classified as having PAH determined by echocardiography.

Zisman et al[10] showed that the HRCT scores including Fib, GG, HC and total perfusion scores as well as their score weighed by relative lobar size and the maximum CT-Fib over all lobes did not differ between advanced IPF patients with and without pulmonary hypertension determined by RHC. However, the present study found that SSc-ILD with PAH determined by echocardiography had higher t-Fib and CT-total scores than those without PAH. Additionally, the previous study[10] reported that patients with advanced IPF did not have any correlations between HRCT-determined fibrotic scores and mean pulmonary artery pressure (mPAP) obtained by RHC. These inconsistent findings from the present study may be due to the differences of the population studied, HRCT scoring system, and the different measuring methods used for a diagnosis of pulmonary hypertension. Moreover, the extent of lung fibrosis in our population studied was not as severe as the study by Zisman et al[10].

The present study found significant correlation of the CT-total score with sPAP (r = 0.384, $p<0.01$) similar to the previous study by Pandey et al[11] which

observed the relationship between the fibrotic score on the HRCT and the peak pulmonary arterial pressure determined by echocardiography in the total group of SSc patients studied; although using different HRCT scoring methods. However, after dividing our SSc-ILD patients to two subgroups comprised of 31 ILD without PAH and 19 ILD with PAH, we did not find any correlation between the calculated HRCT scores and sPAP. We assume that the small sample size in each subgroup may explain this inconsistent finding.

The pathogenesis of PAH in a subset of SSc-ILD is not well described. Historically, PAH in SSc may be considered into one of two categories. The first group includes ILD patients who developed hypoxia-induced PAH secondary to severe lung fibrosis[15,16]. The second group, isolated PAH, includes SSc patients who later develop severe PAH, out of proportion to the severity of lung fibrosis for which pulmonary vasculopathy would be a major contributor to PAH[16,17]. Theoretically, in the hypoxia-induced PAH group, the degree of lung fibrosis should correlate with the degree of PAH. It seems logical that as pulmonary fibrosis progresses and the pulmonary vascular bed is reduced, and then pulmonary vascular resistance increases, and finally PAH develops. The present study is cross sectional study in design, although we found a significant correlation between the CT-total score and sPAP, we cannot conclude the cause and effect relationship between these findings.

The present study did not find any correlation of the MPAD and the MPAD/AD ratio with sPAP in SSc-ILD. Several previous studies of the association between the MPAD, the MPAD/AD ratio and pulmonary arterial pressure reported inconsistent findings[10,11,18]. Pandey et al[11], however, reported a significant correlation of the MPAD and the MPAD/AD ratio with peak pulmonary arterial pressure in 46 SSc patients with varying degrees of lung fibrosis of which the Fib, GG, and HC was found in 72%, 93%, and 17%, respectively. The present study sample had Fib, GG, and HC on the HRCT in 100%, 82%, and 56%, respectively. Therefore, our patients had more severe lung fibrosis on the HRCT than those in Pandey et al's[11]. Contrarily, Zisman et al[10] did not find any correlation between the MPAD or MPAD/AD ratio and mPAP in advanced IPF patients. The author suggested that the restrictive lung physiology in advanced IPF patients might result in a traction effect on pulmonary artery independent of the underlying pulmonary artery pressure. Devaraj et al[18] found that the MPAD significantly correlated with mPAP in the absence of lung fibrosis, whereas, no significant correlation was found in the presence of pulmonary fibrosis. Therefore, in view of the previous studies by Zisman et al[10] and Devaraj et al[18], this may be one of the explanations that a correlation of the MPAD and the MPAD/AD ratio with sPAP was not found in the present study population.

Certain limitations of the present study need to be acknowledged. This was a retrospective review of patients evaluated in a single center and the study population was small. The HRCT and echocardiogram were requested by the rheumatologists who took care of patients with SSc in the rheumatology clinic to confirm the diagnosis of ILD and to identify PAH based on individual consideration about ILD and PAH associated with SSc. Therefore, this could have created a bias regarding the patients included. The HRCTs were read by a single chest radiologist similar to some previous studies[10,11,19,20]. Several previous studies[8,21,22] showed good inter-observer accuracy in measuring MPAD, AD, and extent of pulmonary fibrosis. Therefore, we do not think that the results from a single radiologist would bias the findings in the present study. Only eight patients had pulmonary function test performed within six months from their corresponding HRCT because of the limited availability of the pulmonary function tests in our institution. Therefore, we could not determine the physiologic severity of lung function in most of the population studied.

Finally, the diagnosis of PAH in the present study was determined as having sPAP ≥45 mmHg obtained by echocardiography, whereas the gold standard for PAH diagnosis is right heart catheterization (RHC). Echocardiography has been found to have a lack of specificity since the measurements of pulmonary pressure may be influenced by many factors[5]; however, the present study used the cutpoint of sPAP ≥45 mmHg in which echocardiography has 97% specificity vs. RHC[23].

Conclusion

The authors found that SSc-ILD with PAH had higher t-Fib and the CT-total scores than SSc-ILD without PAH. The CT-total score showed significant positively correlation with sPAP in SSc-ILD. However, the MPAD and the MPAD/AD ratio did not show any relationship with sPAP. Therefore, the calculated total HRCT score for assessing severity and extent of ILD, but not pulmonary artery size may be useful for identifying PAH in SSc-ILD patients.

What is already known in this topic?

Pulmonary complications including ILD and PAH are the leading causes of mortality in patients with SSc. HRCT of the chest is a sensitive measure of the extent and severity of lung involvement in SSc-ILD, although there has been no fully validated scoring system used in general practice. The usefulness of HRCT-determined extent of fibrosis to identify PAH in patients with ILD had been investigated, and the results were inconclusive[10,11].

What this study adds?

The authors found that SSc-ILD patients with PAH had higher t-Fib and the CT-total scores than SSc-ILD without PAH. The CT-total score for assessing the severity of ILD, but not the pulmonary artery size was associated with estimated systolic pulmonary artery pressure determined by echocardiography in patients with SSc-ILD. Therefore, the CT-total score may be useful for identifying PAH in SSc-ILD patients.

Potential conflicts of interest

None.

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ความสัมพันธ์ระหว่างลักษณะความผิดปกติของภาพถ่ายทางทรวงอกด้วยคอมพิวเตอร์ความถี่สูง (HRCT) กับภาวะแรงดันหลอดเลือดแดงปอดสูง ในผู้ป่วยโรคหนังแข็งร่วมกับภาวะปอดเป็นพังผืด (SSc-ILD)

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วัตถุประสงค์: *เพื่อเปรียบเทียบลักษณะความผิดปกติของภาพถ่ายทางทรวงอกด้วย HRCT ในผู้ป่วยโรค SSc-ILD ระหว่างกลุ่มที่มีภาวะแรงดันหลอดเลือดแดงปอดสูง (PAH) และกลุ่มที่ไม่มีภาวะ PAH และเพื่อหาความสัมพันธ์ระหว่างค่าคำนวณผลอ่าน HRCT กับค่า estimated systolic pulmonary artery pressure (sPAP)*

วัสดุและวิธีการ: *การศึกษานี้เป็นการศึกษาย้อนหลัง ได้ทำการทบทวนเวชระเบียนของผู้ป่วย SSc-ILD ที่ได้รับการวินิจฉัยด้วย HRCT และมีผลการตรวจคลื่นเสียงสะท้อนหัวใจภายในระยะเวลา 6 เดือนของการตรวจ HRCT และเข้ารับการรักษาที่หน่วยโรคข้อและรูมาติสซั่ม โรงพยาบาลมหาราชนครเชียงใหม่ ได้ทำการประเมินค่าคะแนน ground glass, fibrosis และ honeycomb รวมทั้งได้วัดขนาดเส้นผ่านศูนย์กลางที่กว้างที่สุดของหลอดเลือดแดงปอด (MPAD) และหลอดเลือด ascending aorta (AD) จากภาพถ่าย HRCT ภาวะ PAH วินิจฉัยโดยใช้ค่า sPAP ⩾45 มิลลิเมตรปรอท*

ผลการศึกษา: *จากผู้ป่วย SSc-ILD จำนวน 50 ราย พบมีภาวะ PAH ร่วมด้วย 19 ราย (ร้อยละ 38) ผู้ป่วยที่มีภาวะ PAH ร่วมด้วยพบมีค่าเฉลี่ย (ค่าเบี่ยงเบนมาตรฐาน) คะแนนพังผืด (9.9 [3.6] vs. 7.8 [3.5], p = 0.03) และคะแนนรวมความผิดปกติของภาพถ่าย HRCT (CT-total score) (20.5 [6.9] vs. 14.9 [6.2], p<0.01) มากกว่ากลุ่มที่ไม่มีภาวะ PAH อย่างมีนัยสำคัญทางสถิติ พบค่า CT-total score มีความสัมพันธ์เชิงบวกอย่างมีนัยสำคัญทางสถิติกับค่า sPAP (r = 0.384, p<0.01) แต่ไม่พบความสัมพันธ์ระหว่าง MPAD หรือ สัดส่วนระหว่าง MPAD/AD กับ sPAP*

สรุป: *ผู้ป่วยโรค SSc-ILD ที่มีภาวะ PAH ร่วมด้วยพบมีค่าคะแนนประเมินพังผืดในปอดสูงกว่ากลุ่มที่ไม่มีภาวะ PAH ดังนั้นค่าคะแนน CT-total score อาจมีประโยชน์ในการช่วยวินิจฉัยภาวะ PAH ที่เกิดร่วมใน SSc-ILD*