

Progressive Systemic Sclerosis: Using High-Resolution CT to Detect and Characterize Interstitial Lung Disease in Siriraj Hospital

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ABSTRACT

Objective: To detect and characterize interstitial lung disease in patients with progressive systemic sclerosis in Siriraj Hospital using high-resolution computed tomography (HRCT) and to detect other associated findings in the chest.

Methods: Retrospective reviewed chest radiography and HRCT of twenty patients with progressive systemic sclerosis during January 2000- January 2005. The HRCT scans were reviewed by two radiologists for reticular opacities (interlobular septal thickening, traction bronchiectasis, honeycombing and bronchovascular interstitial thickening), nodular pattern (miliary, centrilobular and perilymphatic nodule), consolidation and ground-glass opacities. The associated findings in the chest such as esophageal dilatation, mediastinal lymphadenopathy and main pulmonary artery enlargement were also evaluated.

Results: Chest radiography detect interstitial lung disease in eighteen patients (90%). The findings are reticular opacities and fibrosis at both lower lobes. The HRCT shows interstitial lung disease in twenty patients (100%) with the greatest proportion of interlobular septal thickening (85%), followed by traction bronchiectasis (75%), honeycombing (40%) and ground-glass opacities (25%). The distribution of disease is peripheral and lower lung predominant. The associated findings: esophageal dilatation and mediastinal lymphadenopathy were presented in 55% of patients and main pulmonary artery enlargement was presented in 90% of patients.

Conclusion: HRCT is more sensitive than chest radiography for detecting and characterize interstitial lung disease in patients with progressive systemic sclerosis. Our study has evidences of interstitial lung disease in all patients with greatest proportion of interlobular septal thickening, followed by traction bronchiectasis, honeycombing and ground-glass opacities. Moreover HRCT can recognize cases with predominance of inflammatory process and direct clinician into more aggressive treatment, may be beneficial in preventing irreversible disease. HRCT was useful for detecting other associated findings in the chest such as esophageal dilatation, mediastinal lymphadenopathy and main pulmonary artery enlargement.

Keywords: High-resolution computed tomography (HRCT); interstitial lung disease; progressive systemic sclerosis

Siriraj Med J 2007; 59: 168-171

E-journal: <http://www.sirirajmedj.com>

Progressive systemic sclerosis (PSS) is a generalized disorder of connective tissue characterized by varying degrees of inflammation, fibrosis and vascular change in the skin and internal organs.¹⁻³ Pulmonary involvement is more common and more severe in systemic sclerosis than in other types of collagen vascular disease. The most common pulmonary manifestation is interstitial fibrosis, which occurs in approximately 80% of patients.⁴

Evidence of pulmonary disease has been described in chest radiography in 20% to 65% of patients affected by systemic sclerosis.⁵ HRCT is far more sensitive than chest radiography in detecting interstitial lung disease in patient

with scleroderma, detecting abnormalities in 90% of cases.^{6,7} Other associated findings in the chest on CT include mediastinal lymphadenopathy, pleural and pericardial effusion, esophageal dilatation, and pulmonary artery enlargement.^{5,8}

The aim of this study was to detect and characterize interstitial lung disease in patients with progressive systemic sclerosis in Siriraj Hospital using HRCT and to detect other associated findings in the chest.

MATERIALS AND METHODS

Patient population

Twenty patients with diagnosis of progressive systemic sclerosis at Siriraj Hospital between January 2000- January

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2005 who had medical record, chest radiography and HRCT scan were included in this study. Eleven of the patients were male and nine were female. The mean age was 51 years (age range: 32-73 years). All patients in our study had respiratory symptoms (dry cough and dyspnea) and underwent both chest radiography and HRCT scan.

HRCT

The HRCT was performed on a variety of scanner using 1 to 2 mm collimation at 10 mm intervals (n=5) or volumetrically on a multidetector CT scanner (n=15). The CT scans were performed at end inspiration with the patient supine and reconstructed using a high resolution edge-enhancing algorithm. The image were photographed at lung (level -600 to -700 HU, width 1,000-1,500 HU) and mediastinal windows (level 20-50 HU, width 300-500 HU).

Image evaluation

The chest radiography and HRCT scan were retrospectively reviewed by two radiologists and a decision was made by consensus. The radiologists evaluated the presence of reticular opacities (interlobular septal thickening, traction bronchiectasis, honeycombing and bronchovascular interstitial thickening), nodular pattern (miliary, centrilobular and perilymphatic nodule), consolidation and ground-glass opacities. The associated findings in the chest such as esophageal dilatation, mediastinal lymphadenopathy and main pulmonary artery enlargement were also evaluated.

Esophageal dilatation was diagnosed if the luminal diameter of esophagus at aortic arch level exceed 10 mm. in the coronal plane.

Mediastinal lymphadenopathy was diagnosed if their short axis diameter exceed 10 mm. All mediastinal nodes except hilar group were evaluated since no intravenous contrast medium was given.

Main pulmonary artery enlargement was diagnosed if diameter exceed 3 cm.

Statistic analysis

Interobserver agreement of characterize interstitial lung disease in patients with systemic sclerosis using HRCT



Fig 1. Chest radiography of a patient with systemic sclerosis shows reticular opacities and fibrosis at both lower lungs.

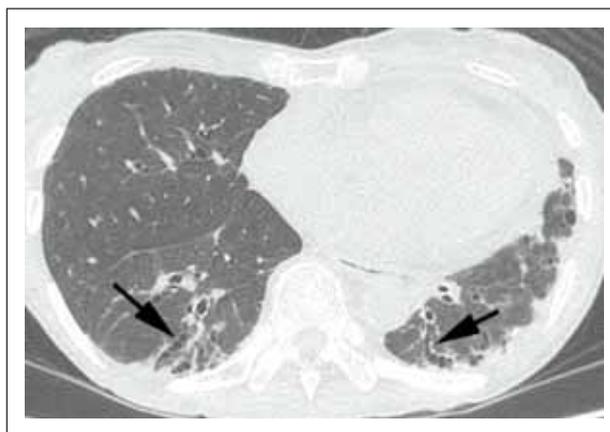


Fig 2. High-resolution CT scan shows evidence of traction bronchiectasis at both lower lungs.

was assessed by using Kappa statistics. Degree of observer agreement was graded as follows:

Kappa values of 0-0.20 slight agreement, 0.21-0.40 fair agreement, 0.41-0.60 moderate agreement, 0.61-0.80 substantial agreement, 0.81-1.00 almost perfect agreement.

Descriptive analysis was used mean and percentage to determine chest radiography abnormalities, HRCT findings and other associated findings.

RESULTS

The readers had substantial agreement in determining characterize interstitial lung disease in patients with progressive systemic sclerosis using HRCT (kappa = 0.73). Chest radiography abnormalities were detected in eighteen patients (90%). The findings are reticular opacities and fibrosis at both lower lobes (Fig 1). The HRCT has evidenced of interstitial lung disease in all patients (100%). HRCT findings included interlobular septal thickening in seventeen patients (85%) which predominate at peripheral (n=16) and lower lung (n=17), traction bronchiectasis in fifteen patients (75%) which predominate at peripheral (n=14) and lower lung (n=15) (Fig 2) and honeycombing in eight patients (40%) which all located at peripheral and lower lung (n=8) (Fig 3). Ground-glass opacities was found in five patients (25%) and all located at peripheral

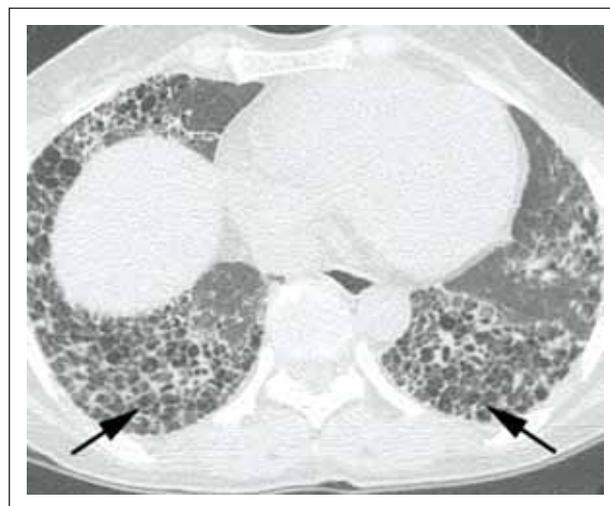


Fig 3. High-resolution CT scan shows interlobular septal thickening with honeycombing at bilateral lung bases, suggesting UIP pattern.



Fig 4. High-resolution CT scan shows predominant ground-glass opacities and superimposed interlobular septal thickening, no obvious honeycombing. The findings suggesting NSIP pattern.

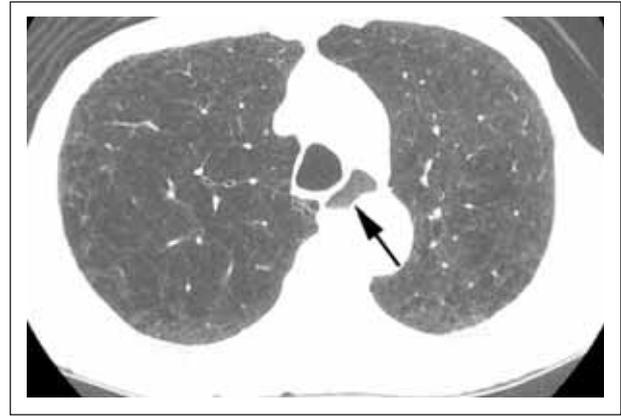


Fig 5. High-resolution CT scan with lung window setting shows dilatation of esophagus.

and lower lung (n=5) (Fig 4). The other HRCT findings included bronchovascular thickening in one patient, miliary nodule in one patient and consolidation in one patient (Table 1). The esophageal dilatation was presented in eleven patients (55%) with the coronal diameter ranging from 1.1-2.5 cm. (mean 1.57 cm.). Mediastinal lymphadenopathy were presented in eleven patients (55%) of patients and main pulmonary artery enlargement was presented in eighteen patients (90%).

DISCUSSION

Interstitial lung disease is common in patients with progressive systemic sclerosis and causes considerable morbidity and mortality.⁹ Evidence of pulmonary disease has been described in chest radiography in 20% to 65% of patients affected by systemic sclerosis.⁵ Our study has abnormal chest radiography findings in 90% of cases. The findings are reticular opacities and fibrosis, predominant at both lower lobes. A higher frequency of radiographic findings of reticular opacities and fibrosis in our study could be from more advanced stage of disease.

The HRCT shows evidence of interstitial pneumonitis and fibrosis in patients who have normal or questionable radiographic findings.^{5,6} Remy- Jardin et al reviewed the HRCT, pulmonary function test, and bronchoalveolar lavage (BAL) result of 53 patients who had progressive systemic sclerosis. Among the 32 patients who had abnormal HRCT finding, 26 (81%) had ground glass opacities and 19 (59%) had honeycombing. The abnormalities involve mainly

the lower lobes and have predominantly peripheral and posterior distribution.¹⁰ Schurawitzki et al studies 23 patients who had progressive systemic sclerosis. The HRCT findings in their patients include subpleural lines (74%), septum thickening or parenchymal band (43%) and honeycombing (43%). Parenchymal abnormalities had typical lower lung zone predominance.¹¹ Our study has evidenced of interstitial lung disease in all patients with greatest proportion of interlobular septal thickening (85%), followed by traction bronchiectasis (75%), honeycombing (40%) and ground-glass opacities (25%). The abnormalities have predominantly peripheral and lower lobe distribution.

The pattern of tomographic findings has a good correlation with histopathological findings, differentiating patterns with predominance of inflammatory process (ground-glass opacities) from predominantly fibrotic lesion (reticular opacities and honeycombing), with inflammatory patterns being associated with a superior response to treatment.¹² A current classification of the idiopathic interstitial pneumonias (According to ATS/ ERS International Multidisciplinary Consensus Classification of Idiopathic Interstitial Pneumonias), included four histopathologic subtypes: usual interstitial pneumonia (UIP), nonspecific interstitial pneumonia (NSIP), desquamative interstitial pneumonia (DIP) and acute interstitial pneumonia (AIP). Our study showed findings suggesting fibrosis (UIP) in the majority of cases (75%), while NSIP pattern suggesting inflammatory process were observed in 25% of cases. Although UIP pattern of lung parenchyma is a non-specific finding which can be found in many condition included: dust exposure (eg: asbestosis), drugs (eg: bleomycin), radiation and collagen vascular disease⁹ but other associated findings will

TABLE 1. HRCT findings and distribution of pulmonary abnormalities in scleroderma patients (n=20).

Abnormalities	Right lung		Left lung		Distribution		
	Upper	Mid	Lower	Upper	Lower	Central	Peripheral
Septal thickening	0	1 (5%)	16 (80%)	1 (5%)	17 (85%)	1 (5%)	16 (80%)
Traction bronchiectasis	0	2 (10%)	13 (65%)	0	15 (75%)	1 (5%)	14 (70%)
Honeycombing	0	0	8 (40%)	0	8 (40%)	0	8 (40%)
Bronchovascular thickening	0	1 (5%)	0	0	1 (5%)	1 (5%)	0
Miliary nodule	0	0	0	0	1 (5%)	0	1 (5%)
Centrilobular nodule	0	0	0	0	0	0	0
Perilymphatic nodule	1 (5%)	0	0	1 (5%)	0	0	1 (5%)
Consolidation	0	0	1 (5%)	0	0	0	1 (5%)
Ground-glass opacity	0	0	5 (25%)	5 (25%)	5 (25%)	0	5 (25%)

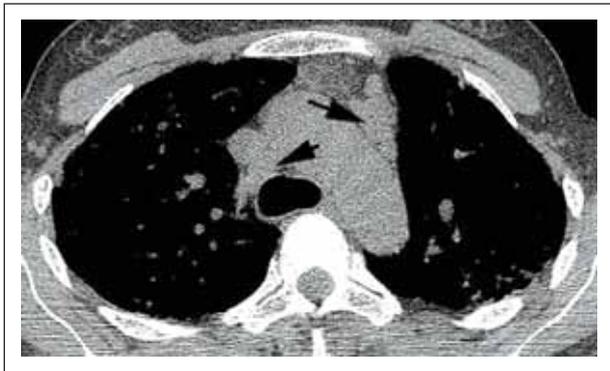


Fig 6. High-resolution CT scan with mediastinal window setting shows multiple mediastinal lymphadenopathy.



Fig 7. High-resolution CT scan shows enlargement of main pulmonary artery, measured about 3.7 cm.

narrowing the differential diagnosis.

Esophageal dilatation and mediastinal lymphadenopathy are frequent associated findings with pulmonary disease in progressive systemic sclerosis. Thoracic CT has been previously used to evaluate the esophagus and mediastinal lymphadenopathy in systemic sclerosis patients.

Bhalla et al demonstrated asymptomatic esophageal dilatation in 20/25 patients (80%) and mediastinal lymphadenopathy in 15/25 patients (60%) with systemic sclerosis.¹³ The author suggested that CT could be useful in detecting asymptomatic esophageal dilatation and could be used to narrow the differential diagnosis of patients who have diffuse interstitial lung disease. Our study has demonstrated esophageal dilatation in 55% and mediastinal lymphadenopathy in 55% of cases.

Pulmonary hypertension is a potentially fatal complication of scleroderma. The incidence of pulmonary hypertension has varied from 10 to 80%. On chest radiography it is characterized by an enlarged pulmonary artery and attenuation of the smaller vessels.¹⁴ Computed tomography is an accurate technique for measuring the size of the main pulmonary artery. Pulmonary diameter has been shown to generally correlate with pulmonary artery pressure. Enlargement of the pulmonary artery is predictive of pulmonary hypertension.¹⁵ Our study demonstrated main pulmonary artery enlargement in 90% of cases.

There were several limitations to our study. Firstly, it was retrospective with a small number of patients. Secondly, no histologic confirmation were obtained. Therefore we unable to correlate the HRCT findings with histopathology.

CONCLUSION

HRCT is more sensitive than chest radiography for detection and characterize findings of interstitial lung disease in patient with progressive systemic sclerosis. Our study has evidenced of interstitial lung disease in all patients with greatest proportion of interlobular septal thickening, followed by traction bronchiectasis, honeycombing and ground-glass opacities. Moreover HRCT can recognize those cases with predominance of inflammatory process

and direct the clinician into more aggressive treatment in these patient, may be beneficial in preventing irreversible disease.

HRCT was useful for detecting other associated findings such as esophageal dilatation, mediastinal lymphadenopathy and main pulmonary artery enlargement.

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