

Pulmonary Artery Hypertension Associated With Systemic Scleroderma

Correlation Among Lung Pulmonary Blood Volume, Lung Perfusion Single Photon Emission Computed Tomography, and Chest Computed Tomography Images

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A 68-year-old woman had been experiencing Raynaud symptom, palpitation of the heart, and short breath in walking stairs or slopes for 7 years. Recently, these symptoms had rapidly become worse. She admitted to our hospital. At that time, she was diagnosed with systemic scleroderma (limited scleroderma). Pulmonary hypertension was pointed out by echocardiography (estimated pulmonary artery pressure 35–43 mm[Hg]), and she was suspected as having pulmonary artery hypertension associated with systemic scleroderma.

At the admission, laboratory data showed as follows: plasma D-dimer 0.2 µg/mL, IgM 419.3 mg/dL, antinuclear antibody ×640, anticentromere antibody 140.6 index (+), and anti-Scl-70 antibody 2.7(–). Respiratory function test showed vital capacity 1.26 L, percentage of vital capacity 59.7%, percentage of forced expiratory volume in one second 80.8%, and percentage of carbon monoxide diffusing capacity 45.1%.

Pulmonary perfusion scintigraphy revealed partial perfusion defects in both lungs. Pulmonary ventilation scintigraphy showed no defects.

Chest computed tomography (CT) images showed that diffuse low- and high-attenuation areas (mosaic perfusion pattern) were found on both of the lungs. On pulmonary perfusion single photon emission computed tomography images, perfusion defects were correlated with mosaic perfusion pattern (low attenuation areas) on chest CT. On lung pulmonary blood volume images using dual energy CT (SOMATOM Definition, Siemens), the decrease of perfusion was correlated with mosaic perfusion pattern of chest CT and perfusion defects on pulmonary perfusion single photon emission computed tomography images (Figures 1 and 2). Lung pulmonary blood volume images well depicted abnormalities of pulmonary perfusion (reduced blood flow because of

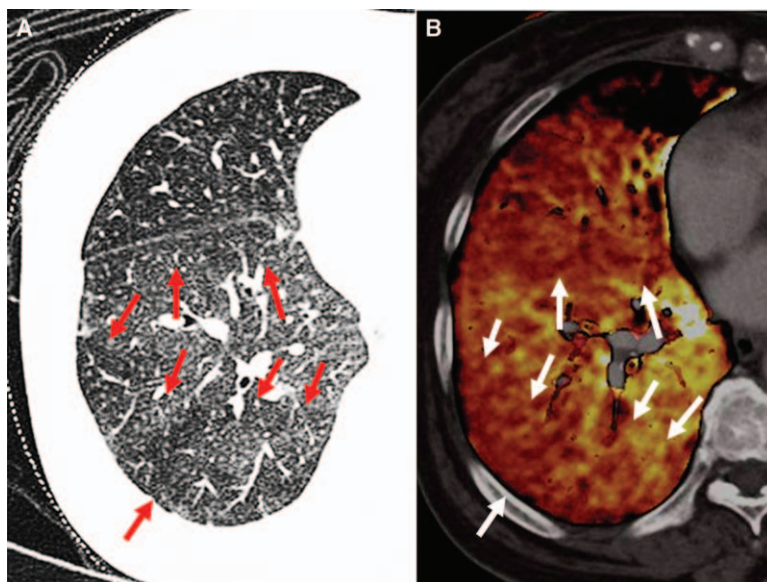


Figure 1. **A**, On chest computed tomography (CT), diffuse low- and high-attenuation areas (mosaic perfusion pattern) were found on both of the lungs (arrows). Low-attenuation areas suggest the decrease of perfusion. **B**, On lung pulmonary blood volume image using dual energy CT, the decrease of perfusion was correlated with mosaic perfusion pattern of chest CT (arrows).

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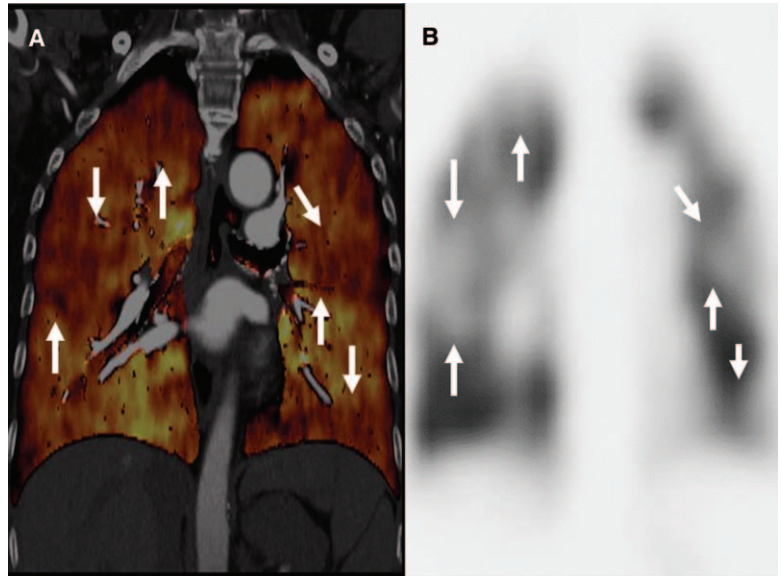
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Figure 2. **A** and **B**, On lung pulmonary blood volume image using dual energy computed tomography (**A**), the decrease of perfusion was correlated with perfusion defects on pulmonary perfusion single photon emission computed tomography images (**B**; arrows).



primary pulmonary hypertension associated with localized scleroderma).

Systemic scleroderma is classified into localized scleroderma and scleroderma diffuse types. Delayed pulmonary hypertension occurs in 50% of localized scleroderma.¹ In patients with systemic scleroderma, pulmonary hypertension can be caused by following a pulmonary artery thickening.²

For the diagnosis of pulmonary embolism, the use of lung pulmonary blood volume has been reported.³ However, to our knowledge, this is first report that the correlation among lung pulmonary blood volume, lung perfusion single photon emission computed tomography, and chest CT images in a patient with primary pulmonary hypertension associated with localized scleroderma.

Disclosures

None.

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