

Pulmonary hypertension and interstitial lung disease in systemic sclerosis

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Section: Chest imaging

Imaging Technique: CT

Imaging Technique: CT

Imaging Technique: CT

Imaging Technique: MR

Case Type: Clinical Cases

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Hilfiker

Patient: 60 years, male

Clinical History:

A 60 years old patient presented in the outpatient department with worsening of a exertional dyspnoe.

Imaging Findings:

A 60-year-old patient presented himself to the outpatient department with exertional dyspnea . His everyday activities were actually limited by worsening dyspnea. The patient had a known systemic sclerosis, which caused only moderate symptoms. Pulmonary function tests showed a slight restrictive ventilation disorder and a severe reduction of diffusion capacity. CT and HR-CT were performed. Multidetector computed tomography (MD-CT) was performed in spiral mode in addition to a high-resolution CT. Imaging showed all signs of pulmonary artery hypertension, including small peripheral pulmonary arteries in contrast to large central arteries, enlarged right ventricle and atrium, pericardial effusion, and horizontally displaced interventricular septum. Vena cava and hepatic veins were enlarged due to right-heart insufficiency. Several enlarged mediastinal lymph nodes were present (Figure 1a). Apical HR-CT images showed only minor pathologic changes: Faint, peripherally located inter- and intralobular septal thickening was present. The same changes were detected in the lower lobes to a much greater degree, causing traction bronchiectasis and ground-glass opacities. The esophagus was dilated and fluid-filled as a consequence of typical organ involvement.

Discussion:

Systemic sclerosis is characterized by induration of the skin, Reynaud's phenomenon (in 80-90%), and musculoskeletal and visceral manifestations. The current patient shows the typical signs of lung involvement in systemic sclerosis. Pulmonary involvement is present in 70-90% of cases (2) and is now the major cause of death (1). Fibrotic changes occur predominantly in the lower lobes and are typically peripherally located. HR-CT findings include 1. ground glass opacification, 2. fine reticular pattern through to honeycombing, 3. lines of various types (septal, subseptal long parenchymal lines) and 4. subpleural and intralobular micronodulation (1). The intralobular thickening may reach the point that it causes ground-glass opacity. Pneumonia is a recognized complication of systemic sclerosis. It may be due to aspiration in cases of severe esophageal dysmotility. However, most authors consider that aspiration does not play a significant role in pathogenesis pulmonary fibrosis. Historically, the fibrosing alveolitis associated with systemic sclerosis was considered the same as cryptogenic fibrosing alveolitis. However, the much better prognosis of fibrosing alveolitis in systemic sclerosis compared to cryptogenic pulmonary fibrosis

suggests a different pathogenesis (5). Exertional dyspnea is present in as much as 60% of patients with systemic sclerosis and is due to pulmonary hypertension (3). Pulmonary hypertension in systemic sclerosis, in contrast to pulmonary hypertension in other fibrosing lung diseases, is not strictly coupled to the grade of fibrosis. Both may develop independently. Its presence may be signaled by an isolated decrease in diffusing capacity. Pulmonary lymphadenopathy is present in 32-60% of cases (present case) (4). Findings in systemic sclerosis can be summarized as follows : 1. severe pulmonary hypertension (enlarged pulmonary artery, enlarged right ventricle, horizontal dislocation of the Septum interventriculare, pericardial effusion), 2. signs of basal / peripheral, slowly progressing pulmonary fibrosis, often presenting as ground glass attenuation and intralobular septal thickening, 3. lymphadenopathy in 32-60%, 4. signs of esophageal dysmotility including widening of the esophagus and secondary aspiration pneumonias.

Differential Diagnosis List: pulmonary hypertension and interstitial lung fibrosis in systemic sclerosis

Final Diagnosis: pulmonary hypertension and interstitial lung fibrosis in systemic sclerosis

References:

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AJR Am J Roentgenol 1996 Jul;167(1):101-4. (PMID: [8659350](#))

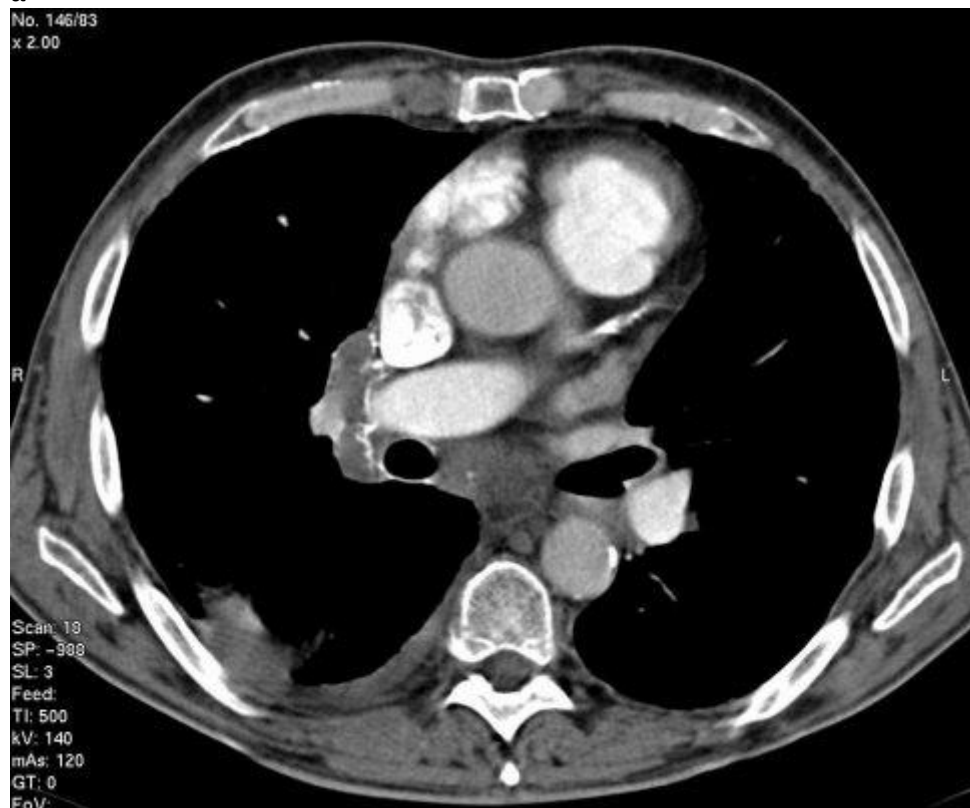
Papiris SA, Vlachoyiannopoulos PG, Maniati MA, Karakostas KX, Constantopoulos SH, Moutsopoulos HH.

Idiopathic pulmonary fibrosis and pulmonary fibrosis in diffuse systemic sclerosis: two fibroses with different prognoses.

Respiration 1997;64(1):81-5. (PMID: [9044481](#))

Figure 1

a



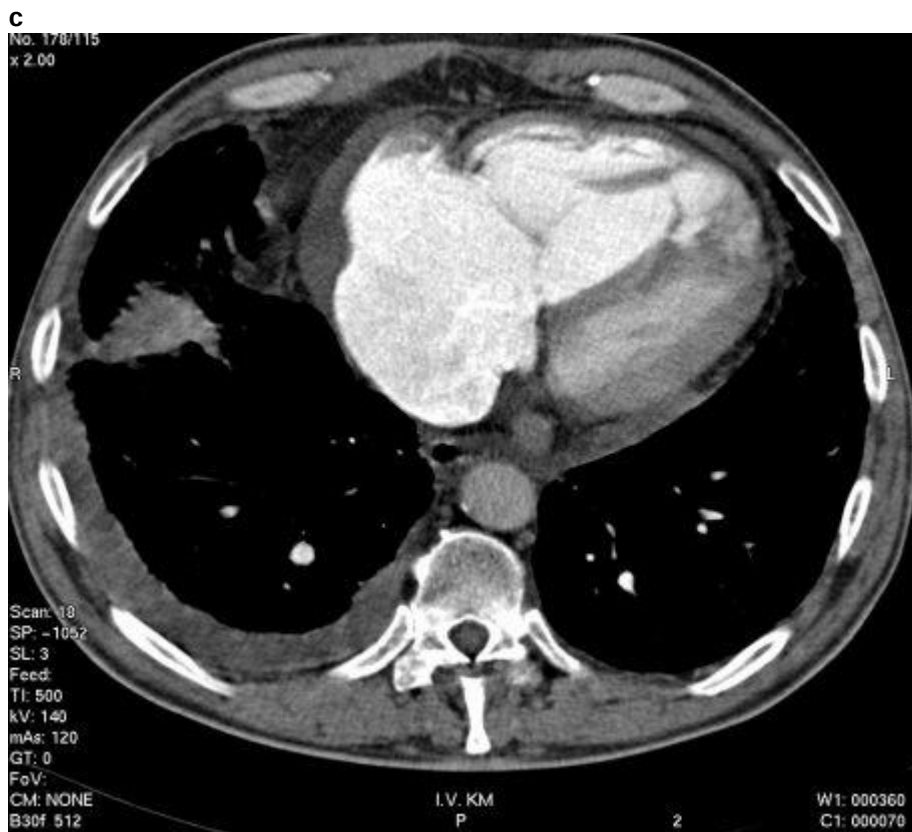
Description: Post-contrast CT image showing slightly enlarged right hilar and subcarinal lymph nodes.

Origin:

b

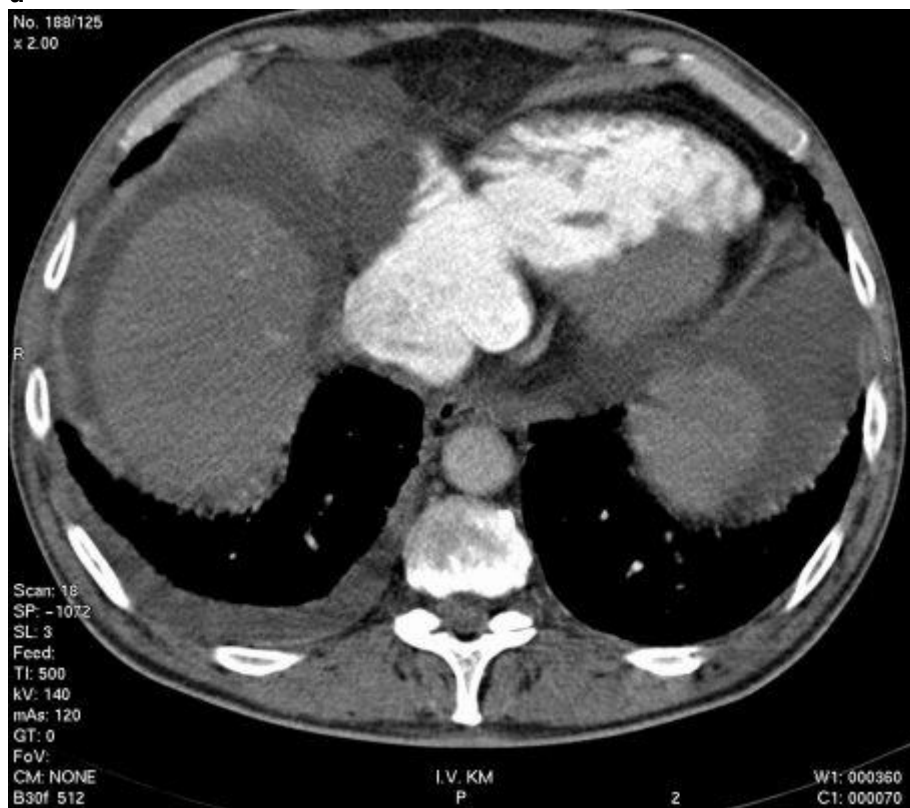


Description: Post-contrast CT image showing an enlarged pulmonary artery trunc as well as enlarged right and left pulmonary arteries **Origin:**



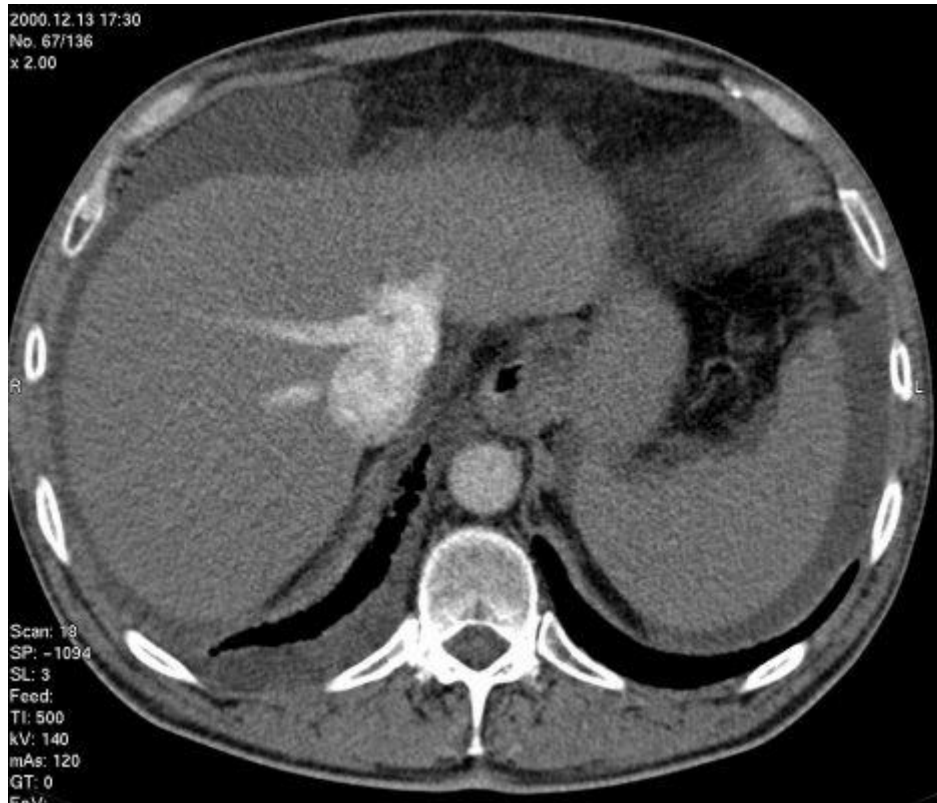
Description: Post-contrast CT image shows an widened right atrium and ventricle and a pericardial effusion. A pleural effusion is seen on the right side. **Origin:**

d



Description: Post-contrast CT image shows an enlarged right atrium and ventricle and a pericardial effusion. Note the horizontal displacement of the interventricular septum. **Origin:**

e



Description: Contrast media regurgitation into the enlarged hepatic veins occurs due to right heart insufficiency in cor pulmonale. Ascitis is present as a sign of chronic congestion. **Origin:**

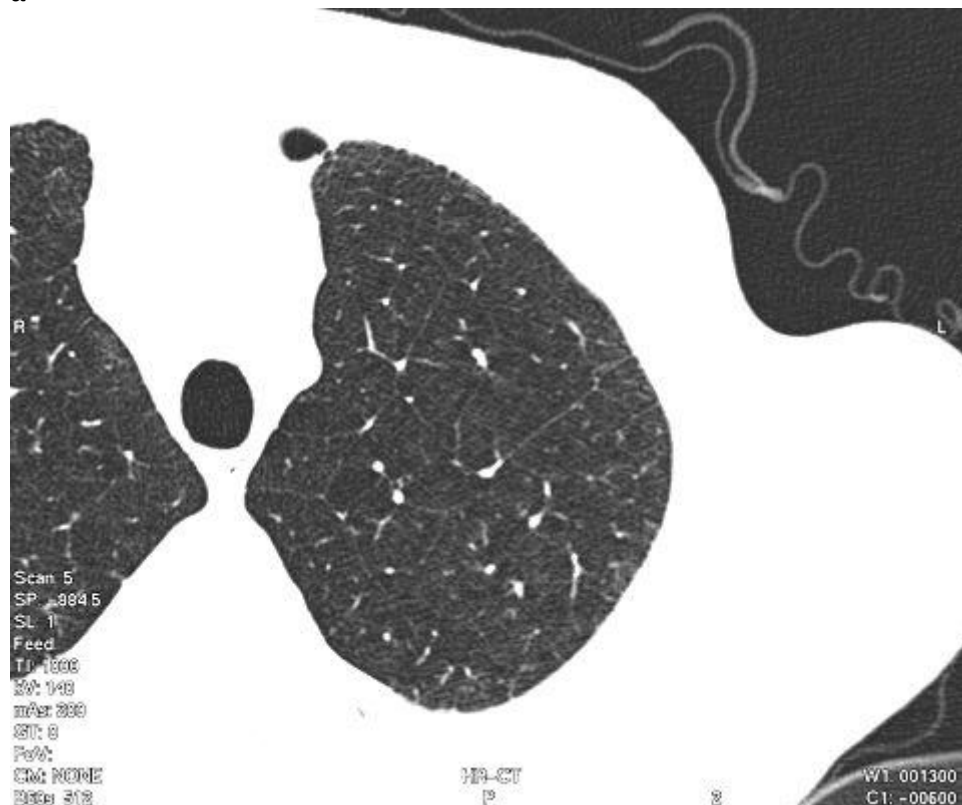
f



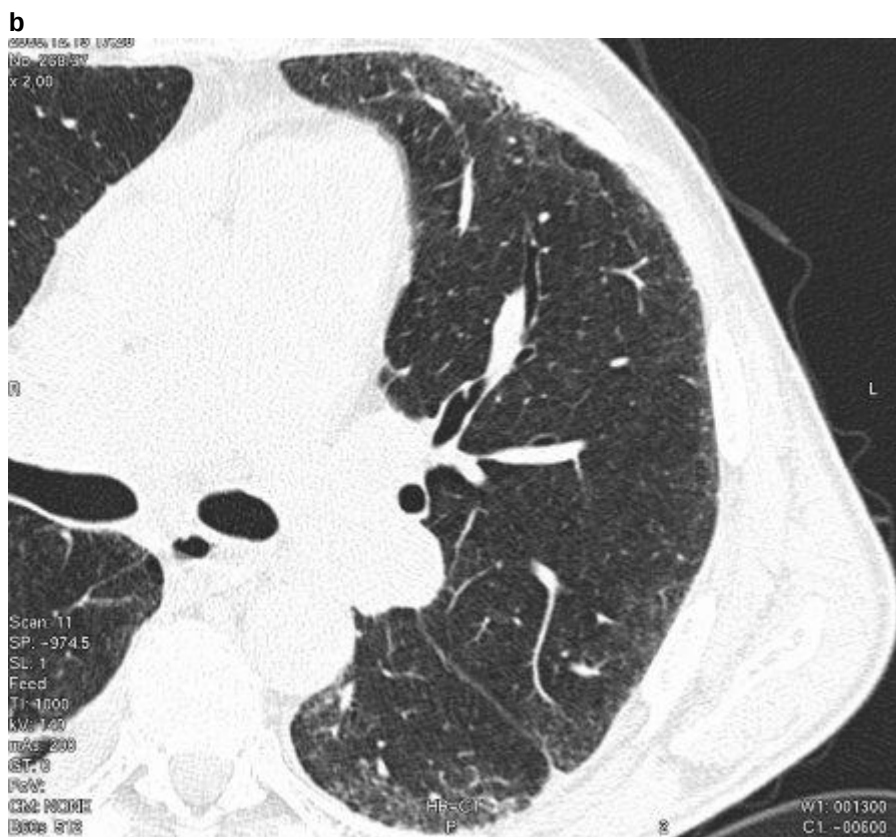
Description: Coronal multiplanar reconstruction shows the enlarged right pulmonary artery as well as the the enlarged right atrium. **Origin:**

Figure 2

a

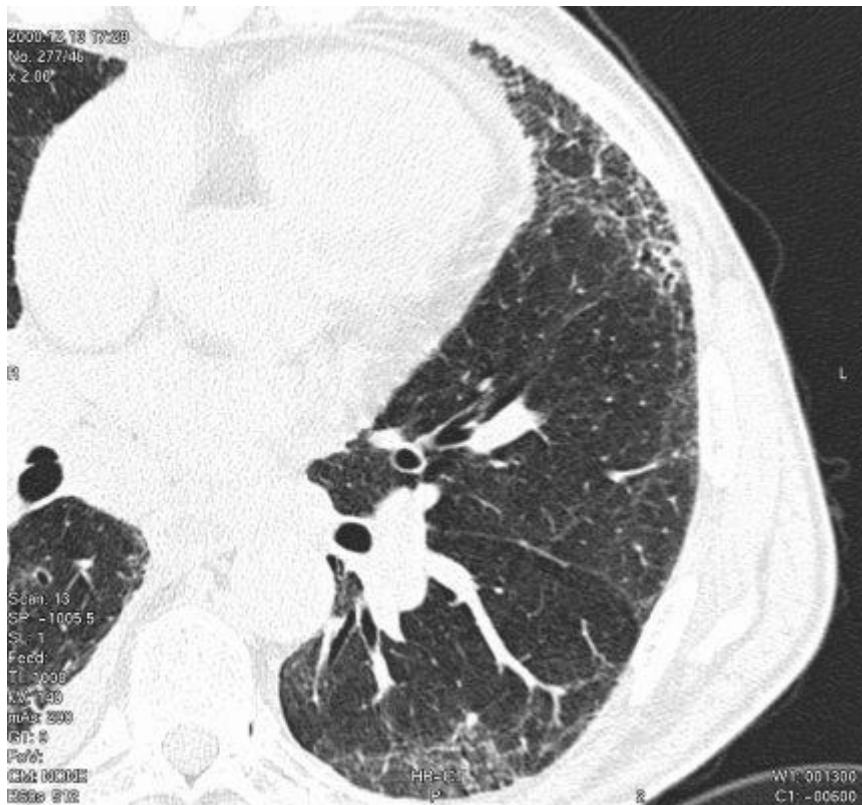


Description: HR-CT image of the left upper lobe showing only faint subpleural intralobular and interlobular interstitial thickening. **Origin:**



Description: HR-CT image of the left upper lobe/lingula and apical lower lobe at the level of the carina showing subpleural intralobular and interlobular interstitial thickening as well as areas of ground glass opacity. Several small subpleural cysts (honeycombing) are seen. **Origin:**

C



Description: HR-CT image at the level below the carina showing subpleural intralobular and interlobular interstitial thickening as well as areas of ground glass opacity. Several small subpleural cysts (honeycombing) are seen. **Origin:**

d



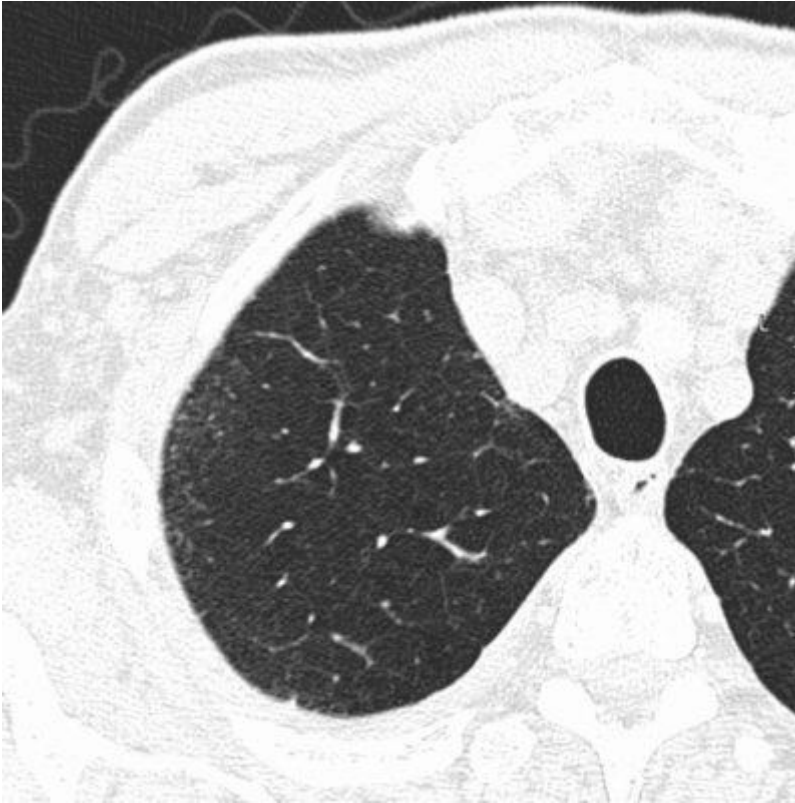
Description: HR-CT image showing subpleural fibrosis in the lower lobe and inferior lingula segment on the left. Note the traction bronchiectasis in the lingula. **Origin:**



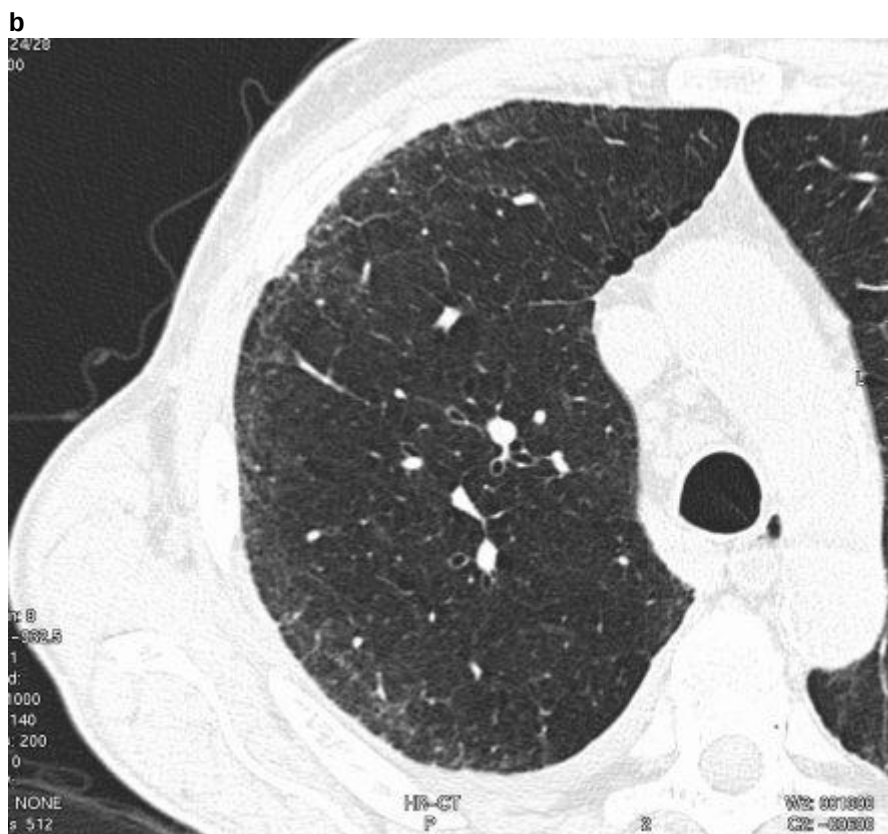
Description: HR-CT image at the base of the left lung showing extensive fibrosis with subpleural predominance. Traction bronchiectasis are seen. **Origin:**

Figure 3

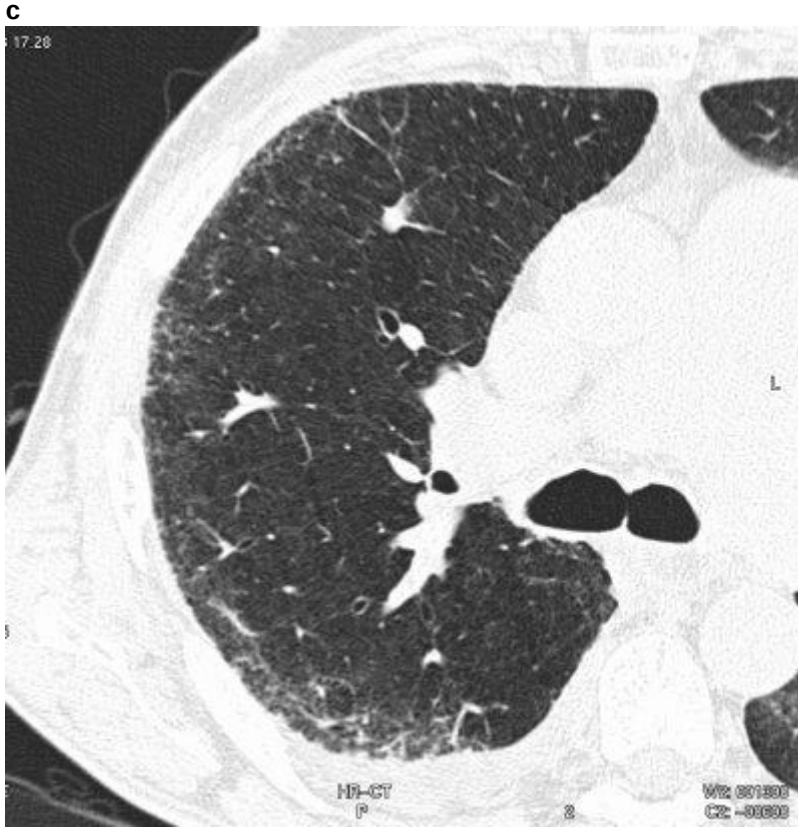
a



Description: HR-CT image showing only faint subpleural intralobular and interlobular interstitial thickening in the right upper lobe. **Origin:**

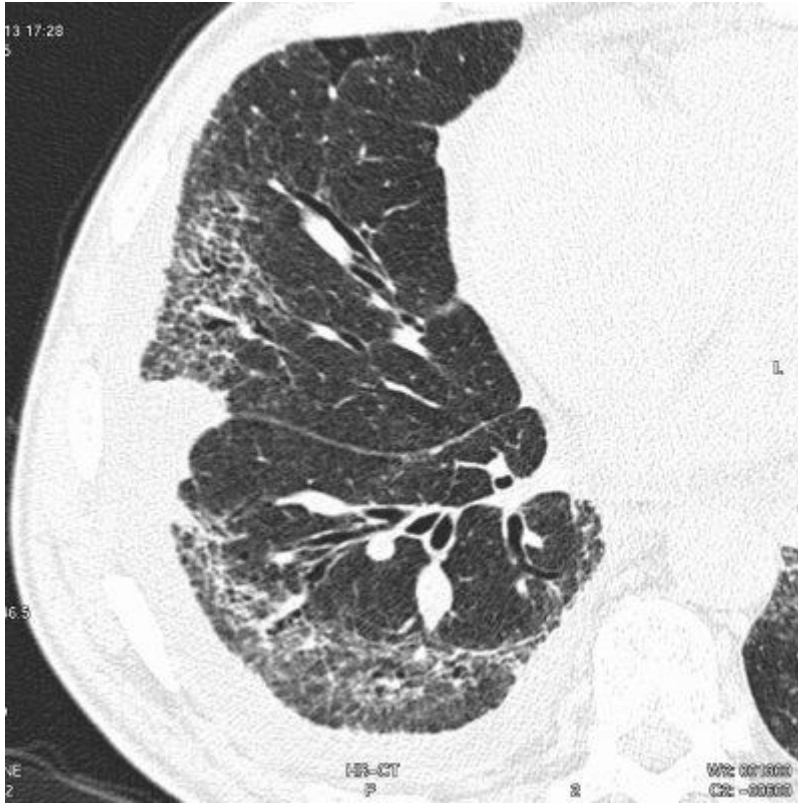


Description: HR-CT image showing increasing subpleural intralobular and interlobular interstitial thickening in a slice located more caudally than 3a. **Origin:**



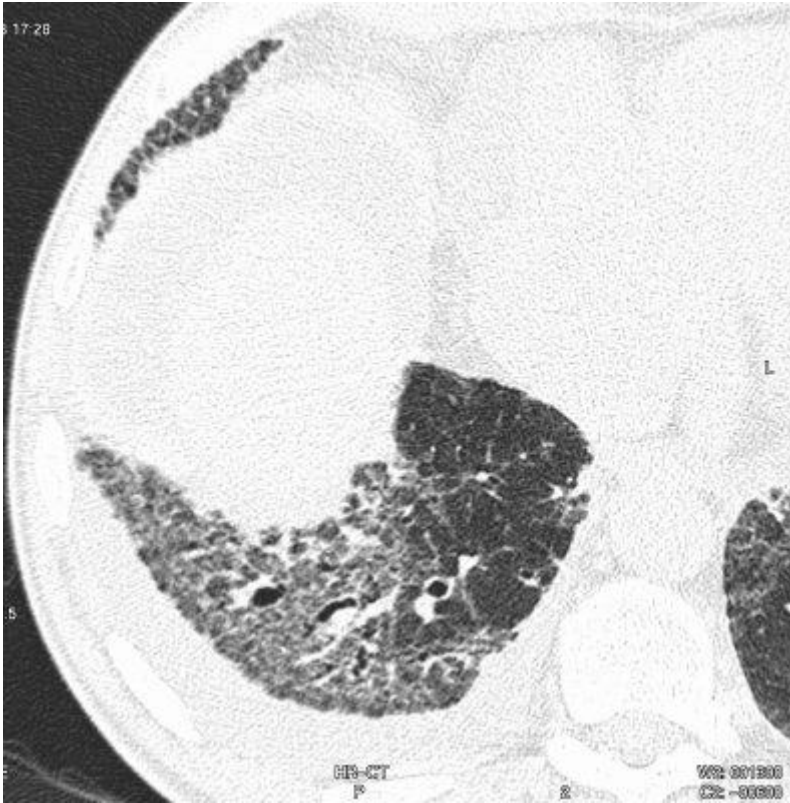
Description: HR-CT image of the left lung at the level of the carina showing intralobular and interlobular interstitial thickening as well as areas of ground glass opacity. **Origin:**

d



Description: HR-CT image of the lower lobe / middle lobe showing extensive intralobular interstitial thickening together with areas of ground glass opacity the lateral middle lobe and lower lobe with peripheral predominance. **Origin:**

e



Description: HR-CT image showing extensive fibrosis at the base of the right lung. Extensive intralobular thickening and bronchiectasis are seen. Only some small subpleural cysts (honeycombing) are detectable. **Origin:**