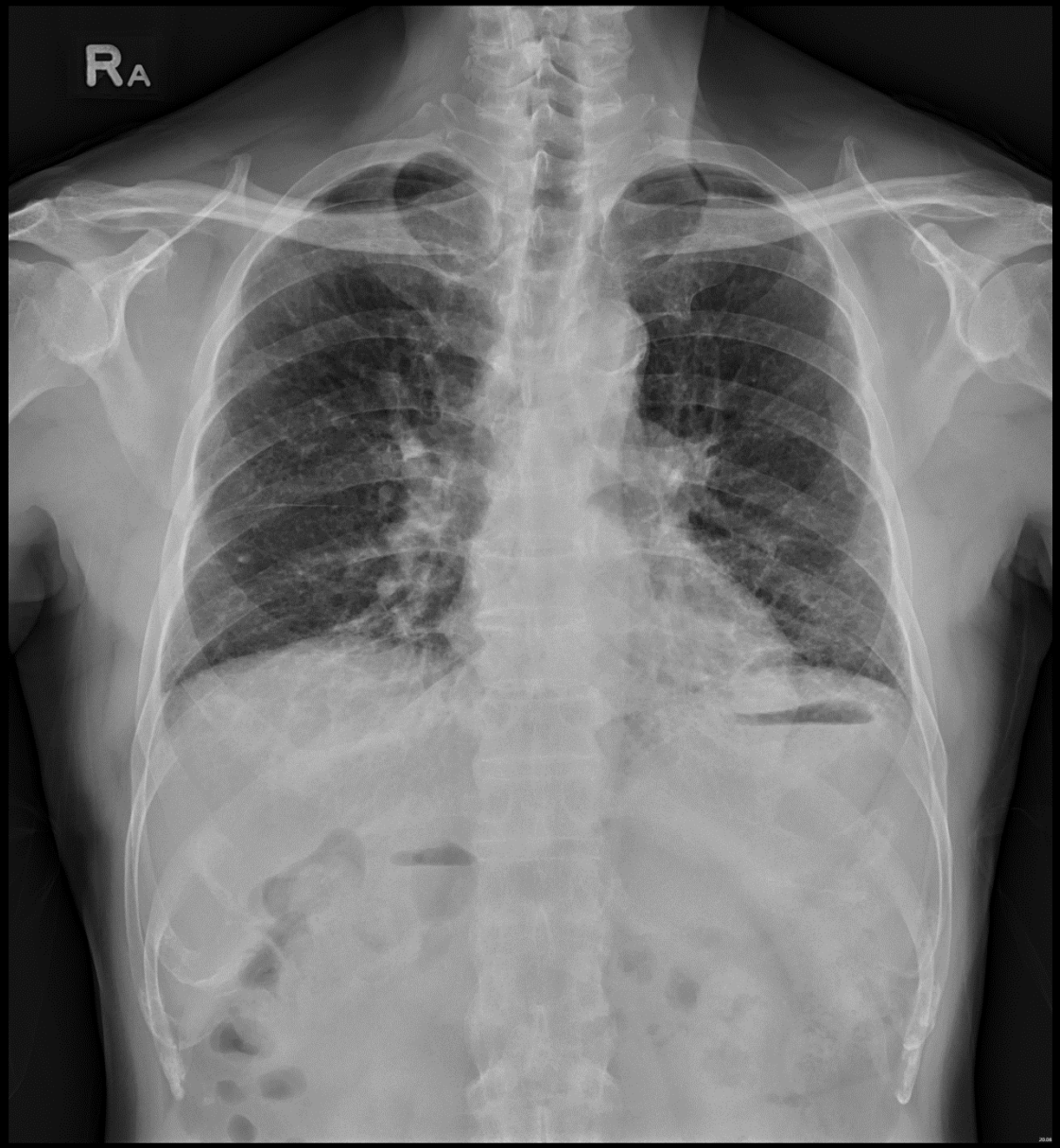
CASE 4

Case 4

- 69 y/o male
- PHx: Ex-smoker (+), HTN (+), GERD
- Occupation: Teacher
- S: Dypnea on exertion recently
- O: Bilateral lower lung dry rales

- 2020-06-01 CXR
- 2020-06-02 HRCT

2020-06-01 CXR



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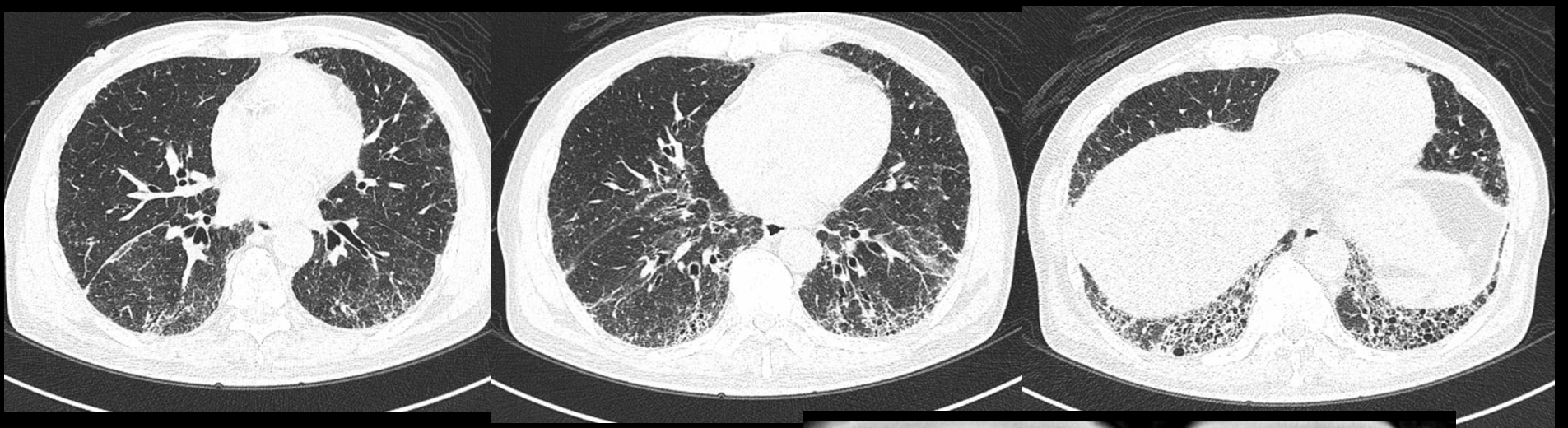
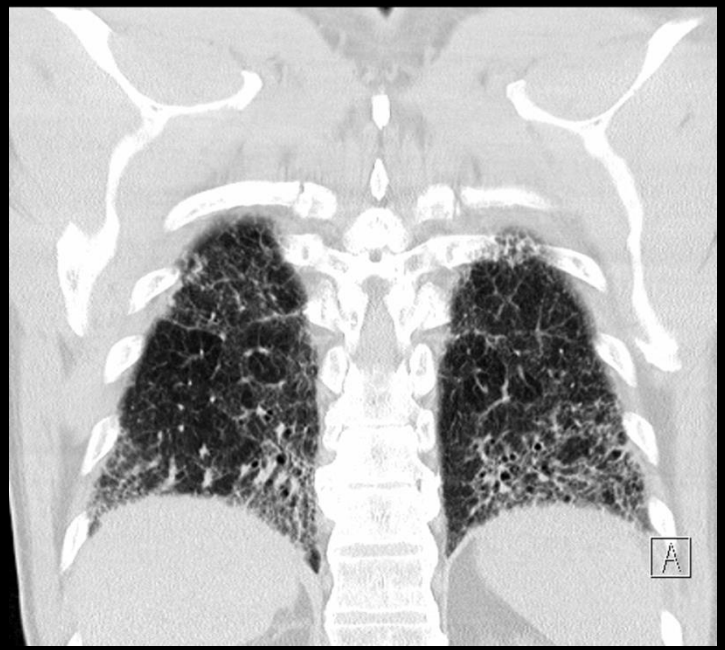
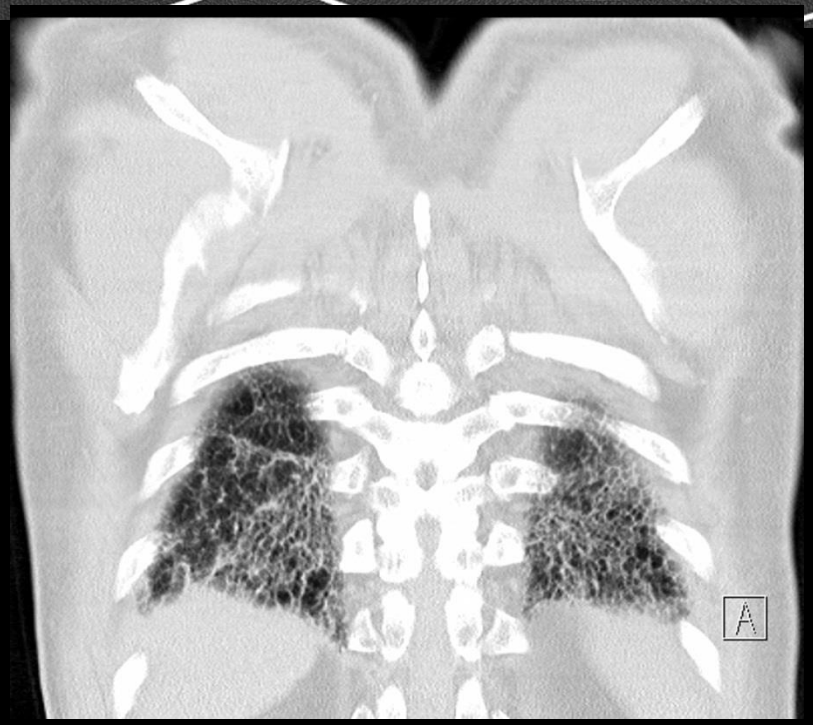






Differential diagnosis

- Usual interstitial pneumonia (UIP)
- Idiopathic pulmonary fibrosis (IPF)
- CTD-ILD
- Fibrosing hypersensitive pneumonitis

Diagnosis

- Scleroderma-related interstitial lung disease (SSc-ILD)

簽收日期	1090601	1090601
簽收時間	1550	1550
工作單號	1201334890	1201334891
試管編號	1201334890	1201334891
報告日期	1090601	1090603
報告時間	1734	1415
委外代檢機構		
ANA [抗細胞核抗體法]		Positive
Centromere [抗細胞核抗體法]		1:640(+)
IgG4(外送)	537.000 mg/dL [3.000-201.000]	

簽收日期	1091203	1091203	1091203
簽收時間	0954	0954	0954
工作單號	1201399279	1201399280	1201399280
試管編號	1201399279	1201399280	1201399280
報告日期	1091203	1091204	1091216
報告時間	1156	1441	1442
委外代檢機構		大安聯合檢驗所	
CRP (血液) [C 反應性蛋白試驗 － 免疫比濁法]	0.44 mg/dL [<0.5]		
IgG4(外送)		379.0 mg/dL [3.0-201.0]	
Anti-Scl-70 ab			Negative
Anti-CENP A ab			Strong Positive(3+)
Anti-CENP B ab			Strong Positive(3+)

CTD-ILD

RadioGraphics

Frequency of Pulmonary Manifestations of Collagen Vascular Diseases

Type of Collagen Vascular Disease	UIP	NSIP	COP	LIP	DAD	Hemorrhage	Airway Disease
Rheumatoid arthritis	+++	++	++	+	+	–	+++
Progressive systemic sclerosis	+	+++	+	–	+	–	–
Dermatomyositis/polymyositis	+	+++	+++	–	++	–	–
Sjögren syndrome	+	++	–	++	+	–	+
Mixed connective tissue disease	+	++	+	–	–	–	–
Systemic lupus erythematosus	+	++	+	+	++	+++	–

Note.—Symbols in columns indicate the frequency with which a feature or pattern of features occurs, with "+" indicating the lowest and "+++" the highest frequency, and with "–" indicating absence of the feature or pattern. COP = cryptogenic organizing pneumonia, DAD = diffuse alveolar damage.

Capobianco J, Grimberg A, Thompson BM, Antunes VB, Jasinowodolinski D, Meirelles GS. Thoracic manifestations of collagen vascular diseases. *Radiographics*. 2012;32(1):33-50. doi:10.1148/rg.321105058

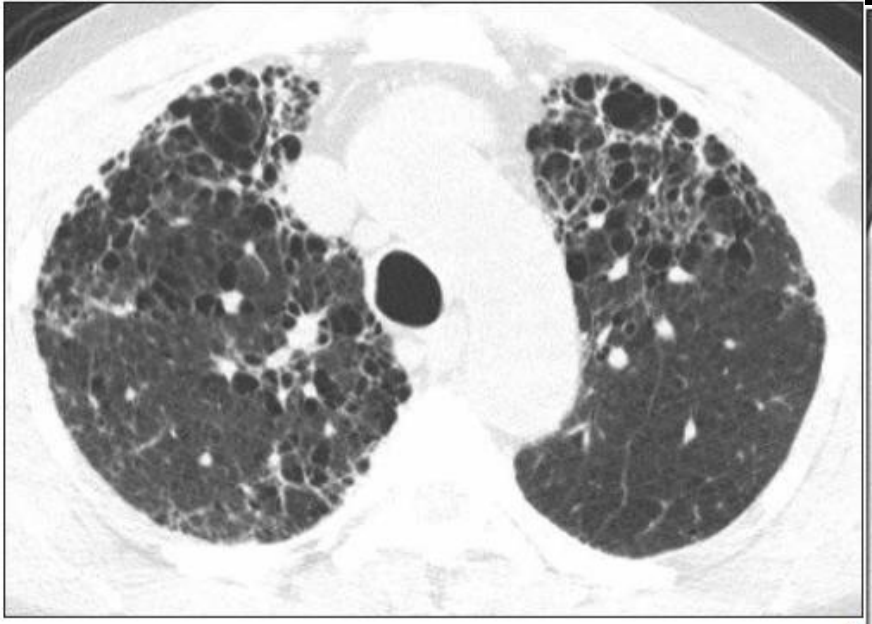
TABLE 3: Performance of Specific CT Signs in Differentiation of Connective Tissue Disease–Associated Interstitial Lung Disease (CTD-ILD) From Idiopathic Pulmonary Fibrosis (IPF) in Patients With Usual Interstitial Pneumonia CT Pattern

CT Sign	Percentage of Patients With IPF With CT Sign ($n = 133$)	Percentage of Patients With CTD-ILD With CT Sign ($n = 63$)	Sensitivity (%)	Specificity (%)	Positive Likelihood Ratio	Negative Likelihood Ratio	p
Anterior upper lobe	12.8 (17)	25.4 (16)	25.4	87.2	1.99	0.86	0.028[a]
Exuberant honeycombing	6.0 (8)	22.2 (14)	22.2	94.0	3.69	0.83	< 0.001[a]
Straight edge	6.0 (8)	25.4 (16)	25.4	94.0	4.22	0.79	< 0.001[a]
More than one sign	4.5 (6)	23.8 (15)	23.8	95.5	5.28	0.80	< 0.001[a]
Any CT sign	19.5 (26)	42.9 (27)	42.9	80.5	2.19	0.71	< 0.001

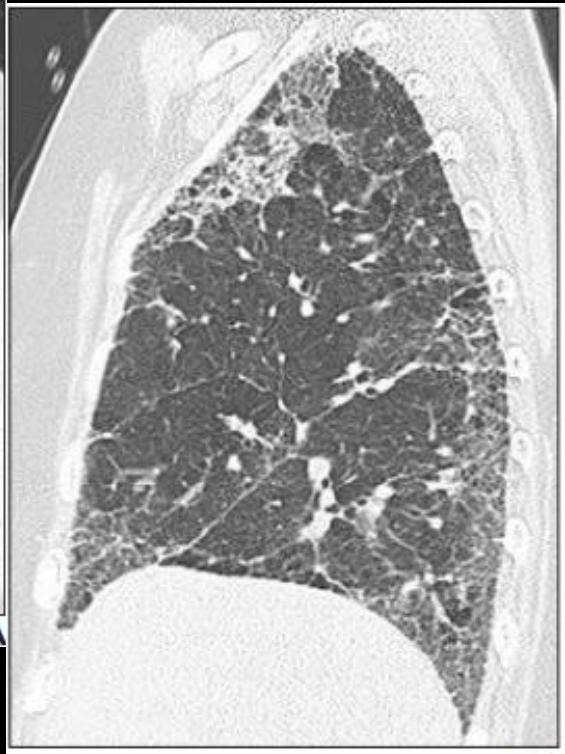
Note—Values in parentheses are number of subjects.
[a]Statistically significant.

- More common in CTD UIP than in IPF UIP
- Low sensitive of any single CT sign in detecting CTD UIP (22.2–25.4%), though specificity was high(87.2–94.0%)
- The highest specificity values: Exuberant honeycombing & Straight-edge signs

Chung JH, Cox CW, Montner SM, et al. CT Features of the Usual Interstitial Pneumonia Pattern: Differentiating Connective Tissu Disease-Associated Interstitial Lung Disease From Idiopathic Pulmonary Fibrosis. *AJR Am J Roentgenol*. 2018;210(2):307-313. doi:10.2214/AJR.17.18384

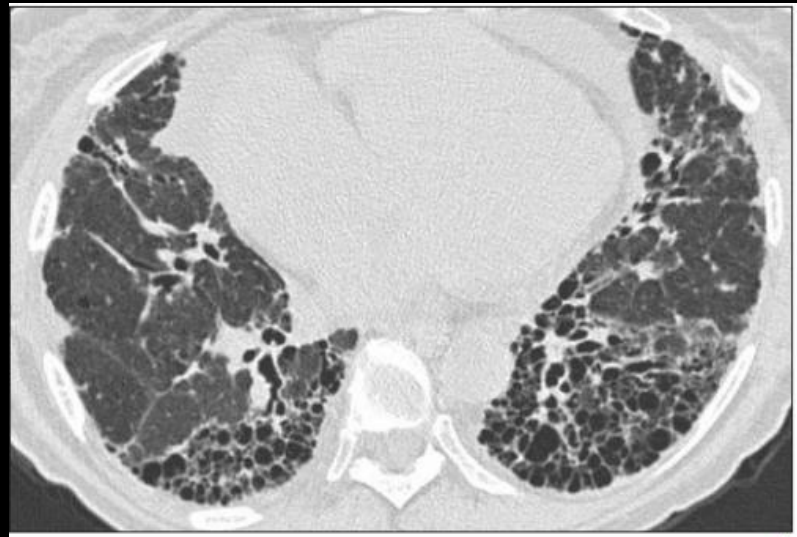


A

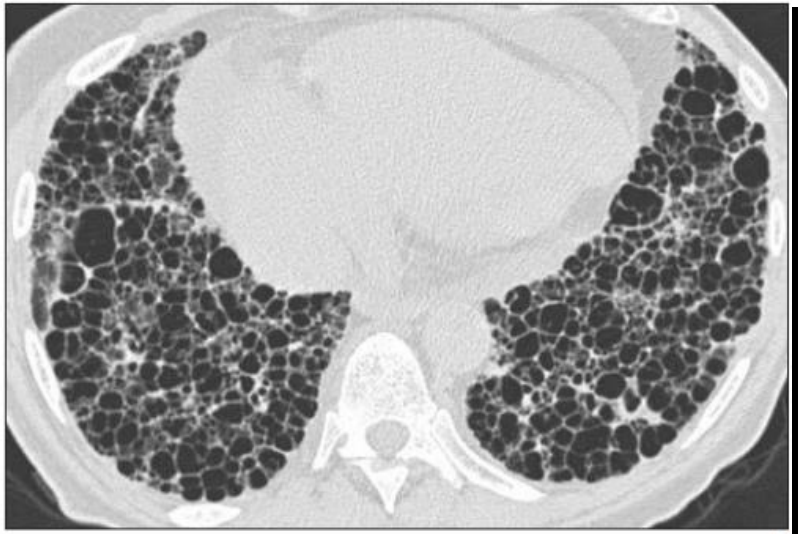


B

Fig. 1—65-year-old woman with connective tissue disease.
A and **B**, Axial (**A**) and sagittal (**B**) unenhanced chest CT images show substantial degree of reticulation, traction bronchiectasis and bronchiolectasis, and honeycombing within upper lobes, mostly concentrated in anterior aspect consistent with anterior upper lobe sign.



A



B

Fig. 4—57-year-old woman with known connective tissue disease.
A and **B**, Axial unenhanced chest CT images show peripheral- and basilar-predominant pulmonary fibrosis pattern characterized primarily by florid honeycombing consistent with exuberant honeycombing sign.

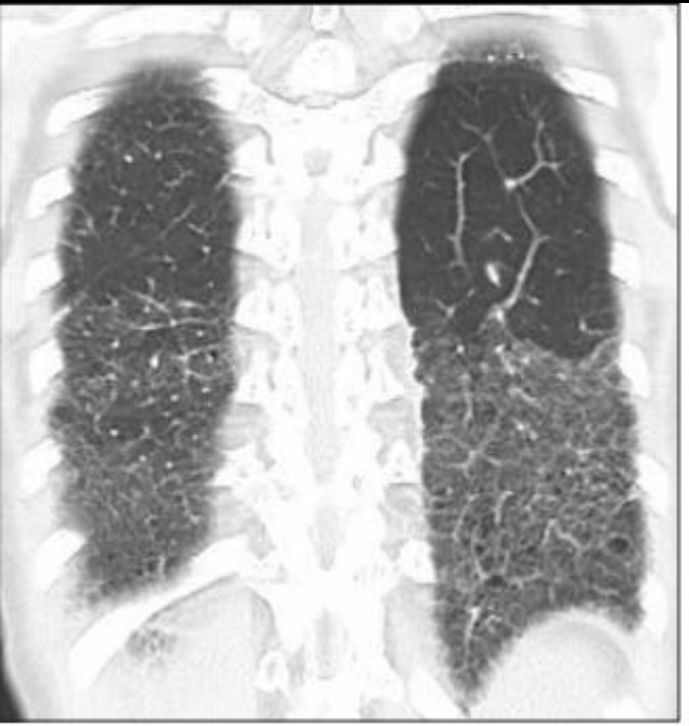


Fig. 5—49-year-old woman with connective tissue disease. Coronal unenhanced chest CT image shows basilar-preponderant pulmonary fibrosis characterized by ground-glass opacity and reticulation and traction bronchiolectasis. Along lateral aspect of lungs, fibrosis does not extend superiorly along chest wall but rather forms fairly straight interface between fibrosis and normal lung orthogonal to lateral chest wall surface, consistent with straight-edge sign.

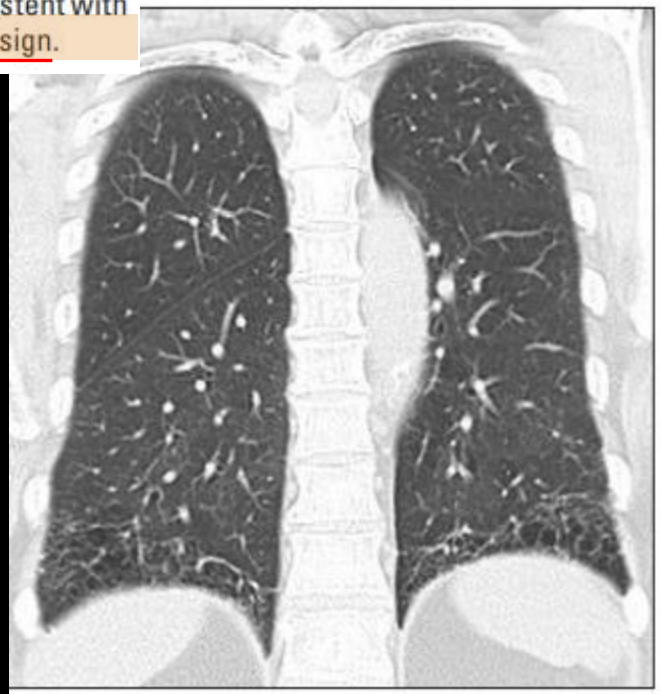


Fig. 6—31-year-old man with connective tissue disease. Coronal unenhanced chest CT image shows basilar-preponderant pulmonary fibrosis characterized mainly by large degree of honeycombing (exuberant-honeycombing sign). Along lateral aspect of lungs, fibrosis does not extend superiorly along chest wall but rather forms fairly straight interface between fibrosis and normal lung orthogonal to lateral chest wall surface, consistent with straight-edge sign.