Connective Tissue Disease–related Thoracic Disease

Yutaka Tsuchiya, MD[a,b,*], Aryeh Fischer, MD[c], Joshua J. Solomon, MD[d], David A. Lynch, MB[a]

KEYWORDS

• Airways disease • Connective tissue disease • Interstitial lung disease
• Nonspecific interstitial pneumonia • Usual interstitial pneumonia

KEY POINTS

- Understanding the prevalence of each entity and the characteristic imaging patterns of each connective tissue disease (CTD) manifestation helps to make the correct diagnosis for CTD-related thoracic disease.
- Drug-induced toxicity, pulmonary infection, and malignancy are frequently seen in patients with CTD. These complications should be excluded when thoracic involvement newly occurs.
- Innovative approaches for evaluating severity and therapeutic effect for patients with CTD-associated thoracic disease have been under development.

INTRODUCTION

Connective tissue disease (CTD) is a group of systemic disorders characterized by autoimmunity and autoimmune-mediated organ damage. It frequently targets the lungs and other thoracic manifestations. CTD-related thoracic disease comprises features directly related to the CTD (including interstitial lung diseases [ILDs], airway diseases, vascular diseases, lymphoproliferative disease, and pleural diseases) and indirect complications (including infections, drug toxicity, and malignancy).

Thoracic disease in CTD has several unique features (**Box 1**):

- It is often asymptomatic, discovered as a consequence of routine radiologic evaluation of a patient with collagen vascular disease.
- In contrast, thoracic disease may be the first manifestation of a CTD, which may develop up to 5 years after initial presentation with CTD.[1–4]
- Although CTD may involve a single lung compartment (pulmonary interstitium, airways, vessels, or pleura), it is common to have involvement of several compartments (eg, interstitial abnormality with pulmonary hypertension [PH] or pleural effusion).
- Although overlap occurs, each CTD has a characteristic pattern of pulmonary involvement, and the prevalence of each thoracic complication entity varies according to the specific CTD (**Table 1**).
- Clues to the diagnosis may be apparent on chest radiograph or computed tomography (CT). For example, a dilated esophagus

[a] Department of Radiology, National Jewish Health, 1400 Jackson Street, Denver, CO 80206, USA; [b] Department of Respiratory Medicine, Showa University Fujigaoka Hospital, 1-30 Fujigaoka, Yokohama 227-8501, Japan; [c] Department of Rheumatology, National Jewish Health, 1400 Jackson Street, Denver, CO 80206, USA; [d] Department of Respiratory and Critical Care Medicine, National Jewish Health, 1400 Jackson Street, Denver, CO 80206, USA
* Corresponding author. Department of Radiology, National Jewish Health, 1400 Jackson Street, Denver, CO 80206.
E-mail address: caeser-salad.2corn-soups@nifty.com

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Box 1
Features of CTD-related thoracic disease

- Often asymptomatic
- May predate other manifestations
- Involves 1 or many lung compartments: interstitium, airway, vessels, pleura
- Type of involvement varies with specific type of CTD
- Clues to diagnosis may be apparent on chest radiograph or CT

should suggest scleroderma, and joint erosions suggest rheumatoid arthritis (RA).

Understanding the prevalence of each entity and the characteristic patterns of each CTD manifestation contributes to the correct diagnosis for CTD-related thoracic disease. This article discusses the characteristic radiologic findings of each CTD-related thoracic disease.

IMAGING TECHNIQUES

Chest CT is helpful not only for diagnosis but also for evaluation for progression and treatment in patients with CTD-related thoracic disease.

The use of thin sections (0.5–1.5 mm) is essential if spatial resolution and lung detail are to be optimized. Also, using a high-resolution CT (HRCT) algorithm is a critical element in performing HRCT. Prone imaging is helpful for identifying early lung fibrosis in the posterior lungs.[5] Multiplanar CT reconstructions may be helpful in identifying distribution of disease. Postexpiratory HRCT scans are helpful for showing significant air trapping in patients with suspected airways or obstructive lung diseases, such as bronchiolitis obliterans (BO) (**Fig. 1**).

Table 1
Relative frequency of CTD-related manifestations in different diseases

	SSc	RA	pSS	MCTD	PM/DM	SLE
Airways	–	++	++	+	–	+
ILD	+++	++	++	++	+++	+
Pleural	–	++	+	+	–	+++
Vascular	+++	–	+	++	+	+
DAH	–	–	–	–	–	++

Abbreviations: DAH, diffuse alveolar hemorrhage; MCTD, mixed CTD; PM/DM, polymyositis/dermatomyositis; pSS, primary Sjögren syndrome; RA, rheumatoid arthritis; SLE, systemic lupus erythematosus; SSc, scleroderma.

Data from Fischer A, du Bois R. Interstitial lung disease in connective tissue disorders. Lancet 2012;380(9842):689–98.

IMAGING PATTERNS

Most of the parenchymal manifestations of CTD are similar to those found in idiopathic interstitial pneumonias (IIPs),[6] and can be classified using the same system.[7] The most common pattern of interstitial fibrosis seen in CTD is nonspecific interstitial pneumonia (NSIP) (**Fig. 2**).[8] However, usual interstitial pneumonia (UIP) (**Fig. 3**), organizing pneumonia (OP) (**Fig. 4**), and lymphocytic interstitial pneumonia (LIP) (**Fig. 5**) may also be seen. In addition to interstitial pneumonia, there may be evidence of airways disease, including bronchiectasis, obliterative bronchiolitis, or follicular bronchiolitis (FB). Bronchiectasis, usually found in RA, is usually cylindrical and must be distinguished from the traction bronchiectasis found in lung fibrosis. Obliterative bronchiolitis is characterized by mosaic attenuation and air trapping (see **Fig. 1**). FB may be associated with tree-in-bud pattern. In addition, clinicians should seek evidence of pleural disease and PH. Enlargement of the central pulmonary arteries is common in

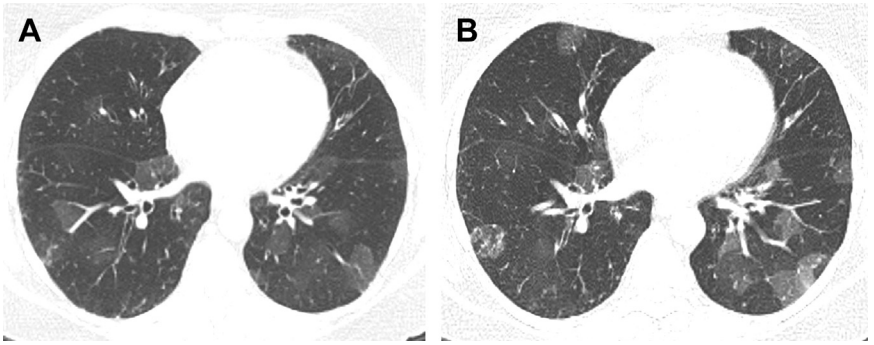

Fig. 1. BO in RA. (*A*) CT at full inspiration shows mosaic perfusion pattern as a result of BO. (*B*) End-expiratory CT shows marked lung inhomogeneity as a result of air trapping.