Imaging of Pulmonary Involvement in Rheumatic Disease

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KEYWORDS

• Computed tomography • Imaging • Connective tissue diseases
• Interstitial lung diseases

KEY POINTS

- There is a high degree of overlap between the pulmonary manifestations of the various connective tissue diseases (CTDs), particularly with respect to interstitial lung disease (ILD) patterns.
- Nonspecific interstitial pneumonia (NSIP) is the most common ILD pattern in all CTDs apart from rheumatoid arthritis, where usual interstitial pneumonia (UIP) is the most frequent pattern.
- Distinguishing between acute exacerbation, infection, drug toxicity, or pulmonary hemorrhage as the cause for acute deterioration in CTD-associated lung disease is impossible on high-resolution computed tomography (HRCT) appearances alone, but requires the integration of clinical and serologic data with the evolution of appearances on plain radiograph.
- A role for HRCT in staging disease and aiding prognostication has recently been shown, with traction bronchiectasis and extent of honeycombing both associated with increased mortality in CTD-ILD.

INTRODUCTION

The rheumatic diseases are a heterogeneous group of inflammatory disorders characterized principally by joint disease, but also, not infrequently, multiorgan dysfunction. Lung disease is common in connective tissue diseases (CTDs) and is an important

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cause of both morbidity and mortality. The CTDs (sometimes termed collagen vascular diseases) include rheumatoid arthritis (RA), systemic sclerosis (SSc), systemic lupus erythematosus (SLE), polymyositis/dermatomyositis (PM/DM), primary Sjögren syndrome (SS), mixed connective tissue disease (MCTD), and undifferentiated connective tissue disease (UCTD). Radiologic assessment has a definite role in the management of CTD-associated pulmonary disease. In this regard, plain chest radiography and high-resolution computed tomography (HRCT) are the principal tests. The indications for imaging will vary, but a typical scenario for the radiologist is to establish whether lung disease is present and, if so, to characterize its nature and extent. In cases in which a histospecific radiologic diagnosis of lung disease cannot be provided, HRCT may be the best guide to the optimal site for surgical biopsy. More recently, there has been interest in the utility of HRCT to assess longitudinal behavior and prognosis.

In the present article, a general discussion of radiologic features is followed by a description of the appearances in individual CTDs. We then consider the pulmonary complications of CTDs on imaging, the use of imaging in prognostication, and the role of multidisciplinary evaluation. Given the acknowledged limitations of plain chest radiography in characterizing patterns of diffuse lung disease in general[1,2] and in CTD-associated lung disorders,[3–5] the article focuses primarily on HRCT appearances. Pulmonary involvement also occurs in other rheumatic disorders, such as vasculitides and inflammatory disorders, including spondyloarthropathy, Behçet disease, and relapsing polychondritis, but the thoracic radiologic manifestations of these conditions are distinct from the CTDs listed previously and are outside the scope of this article.

GENERAL RADIOLOGIC AND PATHOLOGIC CONSIDERATIONS

The CTDs can affect the pulmonary and extrapulmonary components of the thorax to varying degrees. The main manifestations in the pulmonary interstitium, airspaces, airways, pulmonary vasculature, pleura, pericardium, heart, mediastinum, and thoracic musculature are given in **Table 1**. Among the thoracic manifestations of the CTDs, the interstitial diseases are perhaps the most intriguing and widely studied. In this regard, an important consideration is that almost all the patterns of idiopathic interstitial pneumonias (IIPs) (but, importantly, not their prevalences), are mirrored in CTD-related interstitial lung disease (ILD). The radiologic and histopathologic features of the IIPs have been well documented (**Table 2**).[6–8] Indeed, it may be argued that the true utility

Table 1
Thoracic manifestations of the connective tissue disorders

Compartment	Manifestation
Airways	Bronchial wall thickening, bronchiectasis, obliterative bronchiolitis
Lung parenchyma	Interstitial lung disease: interstitial pneumonias (see Table 2) Airspace disease: diffuse alveolar damage, pulmonary hemorrhage Necrobiotic nodules, infection, malignancy
Pulmonary vasculature	Acute/chronic pulmonary thromboembolism, pulmonary hypertension
Pleura/pericardium	Pleural/pericardial thickening, nodularity or effusion, pneumothorax
Mediastinum	Esophageal dilatation/dysfunction, enlarged mediastinal lymph nodes
Thoracic musculature	Muscle dysfunction leading to ventilatory impairment