

# Thoracic Manifestations of Systemic Autoimmune Diseases: Radiographic and High-Resolution CT Findings<sup>1</sup>

## CME FEATURE

See accompanying test at [http://www.rsna.org/education/rg\\_cme.html](http://www.rsna.org/education/rg_cme.html)

## LEARNING OBJECTIVES FOR TEST 5

After reading this article and taking the test, the reader will be able to:

- Identify the chest radiographic and CT findings in patients with systemic autoimmune diseases.
- Recognize the differences in imaging findings among the various systemic autoimmune diseases.
- Describe some of the clinical thoracic findings of the systemic autoimmune diseases.

Jennifer P. Mayberry, MD • Steven L. Primack, MD • Nestor L. Müller, MD, PhD

The systemic autoimmune diseases include collagen vascular diseases, the systemic vasculitides, Wegener granulomatosis, and Churg-Strauss syndrome. They can cause a variety of thoracic abnormalities that are influenced by the pathophysiologic characteristics of the underlying disease process. Although many of the abnormalities can be detected at chest radiography, high-resolution computed tomography (CT) has been shown to be superior in depicting parenchymal, airway, and pleural abnormalities. Thoracic manifestations of collagen vascular diseases include pleural disease, pulmonary fibrosis, diaphragm weakness, aspiration pneumonia, bronchiolitis obliterans organizing pneumonia, bronchiolitis obliterans, and bronchiectasis. Wegener granulomatosis may be associated with multiple nodules or masses with irregular margins that are frequently cavitated. Patients with Churg-Strauss syndrome often have consolidation or ground-glass attenuation at chest radiography and CT. Goodpasture syndrome is associated with extensive bilateral air-space consolidation.

**Index terms:** Arthritis, rheumatoid, 60.611 • Dermatomyositis, 60.614 • Lung, CT, 60.12118 • Lung, diseases, 60.61, 60.62, 60.691, 60.695 • Lupus erythematosus, 60.612 • Scleroderma, 60.613 • Spondylitis, 60.74 • Wegener granulomatosis, 60.622

**RadioGraphics 2000;** 20:1623-1635

<sup>1</sup>From the Department of Radiology, Oregon Health Sciences University, 3181 SW Sam Jackson Park Rd, Mail Code L340, Portland, OR 97201 (J.P.M., S.L.P.); and the Department of Radiology, University of British Columbia and Vancouver Hospital and Health Sciences Centre, Vancouver, British Columbia, Canada (N.L.M.). Presented as a scientific exhibit at the 1999 RSNA scientific assembly. Received February 21, 2000; revision requested March 31 and received May 3; accepted May 4. **Address correspondence** to S.L.P. (e-mail: [primacks@ohsu.edu](mailto:primacks@ohsu.edu)).

## Introduction

The autoimmune diseases comprise a group of immunologic disorders whose common denominator is the presence of an idiopathic systemic autoimmune process. These disorders include the collagen vascular diseases, the systemic vasculitides, Wegener granulomatosis, and Churg-Strauss syndrome. The systemic autoimmune diseases can cause a variety of pulmonary parenchymal, vascular, airway, and pleural abnormalities. The characteristic thoracic manifestations of these diseases are influenced by the underlying autoimmune process. Although many of the complications can be detected at chest radiography, high-resolution computed tomography (CT) has been shown to be superior to radiography in the assessment of the presence and extent of parenchymal, airway, and pleural abnormalities.

In this article, we illustrate the radiographic and high-resolution CT manifestations in the chest of systemic autoimmune diseases. Discussion of clinical symptoms and patient characteristics associated with each of these disorders is accompanied by description and illustration of the characteristic thoracic findings at radiography and high-resolution CT.

### Systemic Lupus Erythematosus

Systemic lupus erythematosus is an autoimmune disease of unknown pathogenesis characterized at histologic examination by deposition of autoantibodies and immune complexes that damage tissues and cells. The presentation is usually systemic and includes fatigue, malaise, anorexia, fever, and weight loss. The disease predominantly affects women (F:M, 10:1) aged 20–50 years.

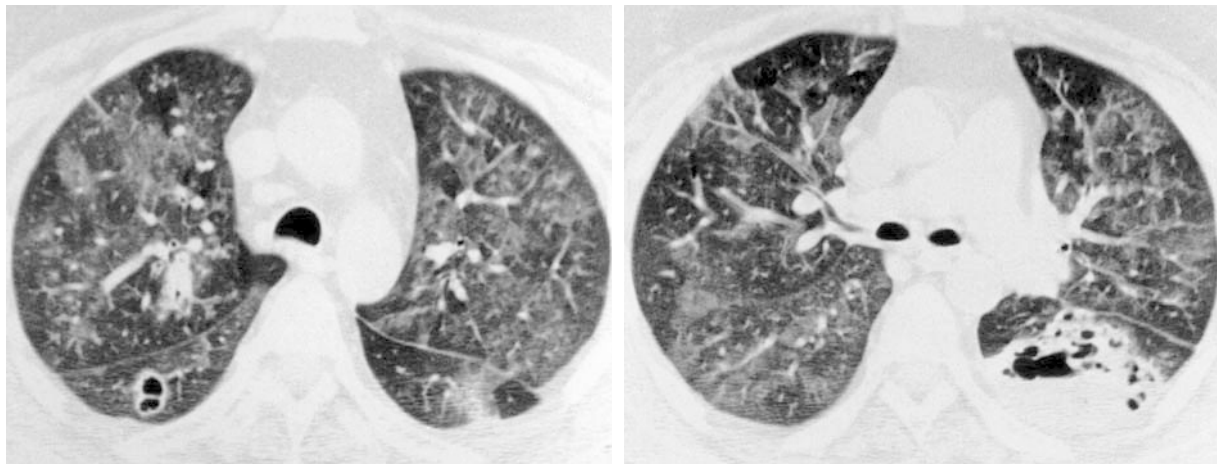
During the course of systemic lupus erythematosus, pleural disease develops in 50% of patients (1). The most common radiographic manifesta-



**Figure 1.** Systemic lupus erythematosus with pleural involvement. CT scan shows pleural effusion of the left lung, as well as pericardial effusion.

tion is unilateral or bilateral pleural effusion that frequently is associated with pericardial effusion (2) (Fig 1).

Pulmonary parenchymal abnormalities are also common. Parenchymal opacification may be caused by pneumonia, hemorrhage, acute lupus pneumonitis, or pulmonary edema, with pneumonia being the most common cause (1). Although in most patients pneumonia has a bacterial pathogenesis, in patients with systemic lupus erythematosus, opportunistic infections occur with increased frequency (Fig 2). Pulmonary hemorrhage is another, though less common, cause of air-space consolidation. Chest radiography usually shows extensive bilateral areas of air-space consolidation. The findings at CT consist of bilateral areas of consolidation and ground-glass attenuation. Acute lupus pneumonitis is a diagnosis of exclusion. The chest radiographic findings of acute lupus pneumonitis (which are similar to those of pneumonia and pulmonary



a.

b.

**Figure 2.** Systemic lupus erythematosus with an opportunistic infection. High-resolution CT scans show patchy ground-glass attenuation bilaterally and a cavitary nodule in the superior segment of the right lower lobe (**a**) and focal consolidation in the left lower lobe with multiple areas of cavitation (**b**). Results of bronchoscopic lavage and biopsy were positive for cytomegalovirus and aspergillosis.

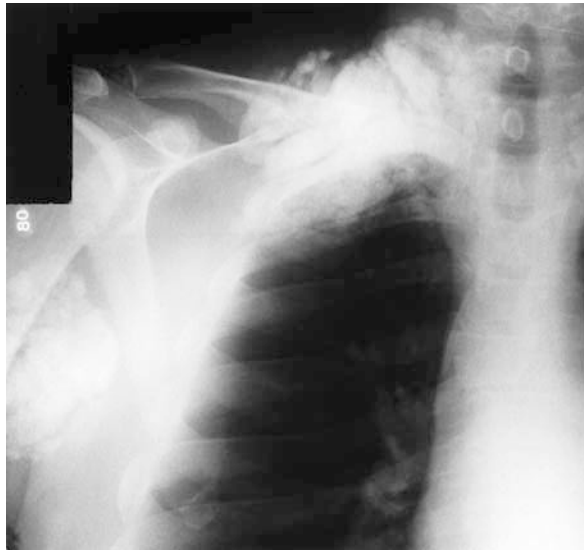


**Figure 3.** Systemic lupus erythematosus with lupus pneumonitis. Posteroanterior radiograph shows patchy bilateral areas of air-space consolidation.

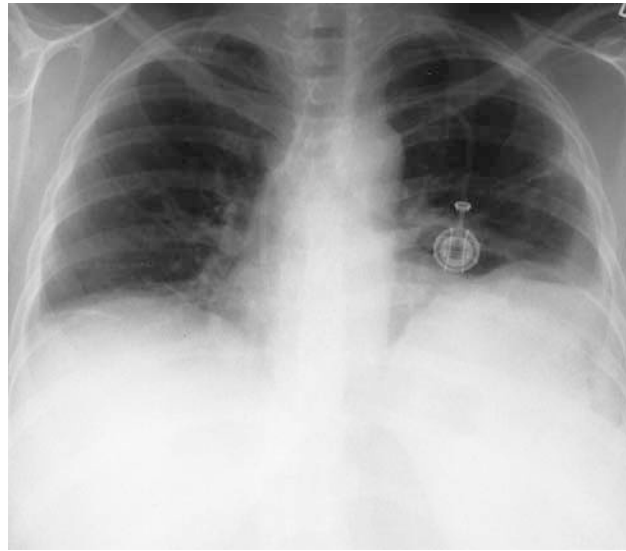
hemorrhage) usually consist of bilateral areas of consolidation (3) (Fig 3). On occasion, the consolidation is unilateral.

Pulmonary fibrosis is less common in systemic lupus erythematosus than in rheumatoid arthritis or scleroderma. High-resolution CT scans show pulmonary fibrosis much more frequently than do chest radiographs. In two recent studies in which high-resolution CT scans were evaluated, fibrosis was present in approximately 30% of cases (4,5). The fibrosis involved predominantly the lung periphery and lower lobes.

Other chest radiographic findings of systemic lupus erythematosus include loss of lung volume related to **diaphragmatic dysfunction**, pulmonary edema, musculoskeletal changes related to renal



4.



5.

**Figures 4, 5.** (4) Systemic lupus erythematosus with tumoral calcinosis. Posteroanterior radiograph of a patient with systemic lupus erythematosus and renal failure shows a large amorphous soft-tissue calcification projecting into the right supraclavicular region and right axilla. (5) Systemic lupus erythematosus with diaphragmatic dysfunction. Posteroanterior radiograph shows decreased lung volumes related to diaphragmatic dysfunction and bibasilar atelectasis.

failure, and bone changes related to corticosteroid therapy (Figs 4, 5).

### Rheumatoid Arthritis

Rheumatoid arthritis is an autoimmune disease of unknown pathogenesis that affects 1% of the population, with a 3:1 predilection for women between the ages of 20 and 50 years. The classic clinical manifestation is chronic symmetric polyarthritis due to a persistent inflammatory synovitis.

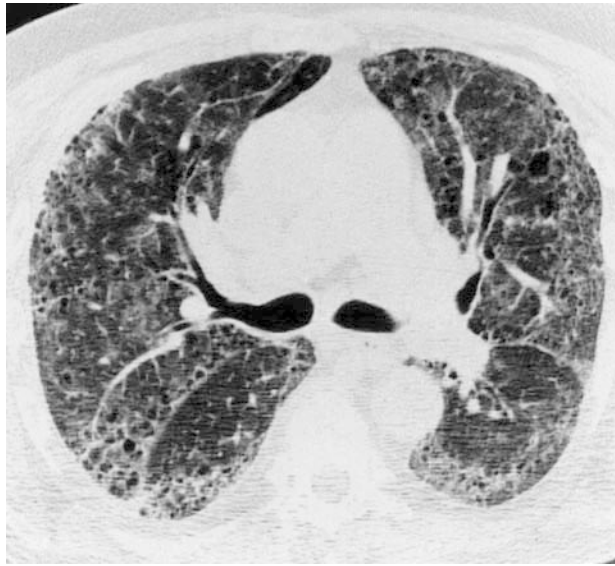
Thoracic involvement often develops in patients as their disease progresses. **Pleural disease is the most common thoracic manifestation and is seen much more frequently in men (6). Pleural thickening is the next most common finding and is seen more often than pleural effusion. Pleural effusions are usually unilateral and may be loculated (Fig 6). They usually occur late in the disease and are commonly associated with pericarditis and subcutaneous nodules.**

**Pulmonary fibrosis occurs in 2%–9% of patients with rheumatoid arthritis.** Chest radiographs typically show a reticular or reticulonodular pattern involving the lower lung zones (7,8). High-resolution CT scans show a reticular pattern and irregular interlobular septal thickening predominantly in the lung periphery and lower

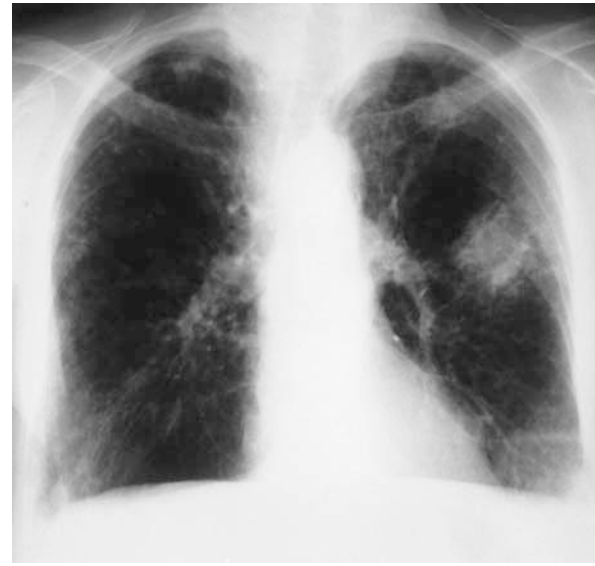


**Figure 6.** Rheumatoid arthritis with pleural effusion. CT scan shows a loculated left pleural effusion.

lung zones (Fig 7). Honeycombing and progressive volume loss develop as the disease progresses. **In rare cases, fibrosis may be limited to the upper lobes and contain areas of cavitation, characteristics that mimic those of tuberculosis.** Like any patient with pulmonary fibrosis, patients with pulmonary fibrosis associated with rheumatoid arthritis have an increased prevalence of lung cancer (9) (Fig 8).

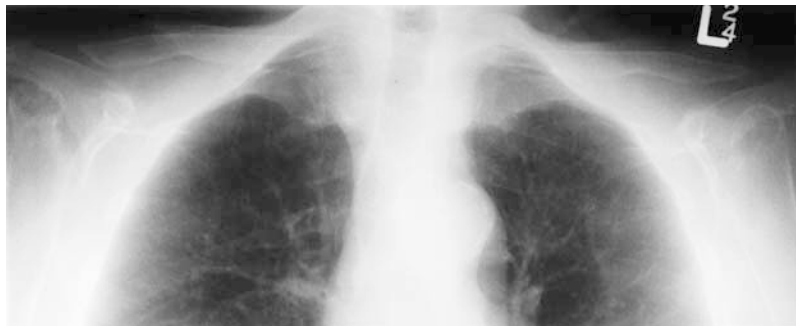


7.



8.

**Figures 7, 8.** Rheumatoid arthritis with pulmonary fibrosis. High-resolution CT scan shows honeycombing and reticulation predominantly in the lung periphery. **(8)** Rheumatoid arthritis with pulmonary fibrosis and lung cancer. Posteroanterior radiograph shows bilateral peripheral areas of coarse reticulation and an irregular lung mass in the left upper lobe. The diagnosis of lung cancer was proved at surgery.



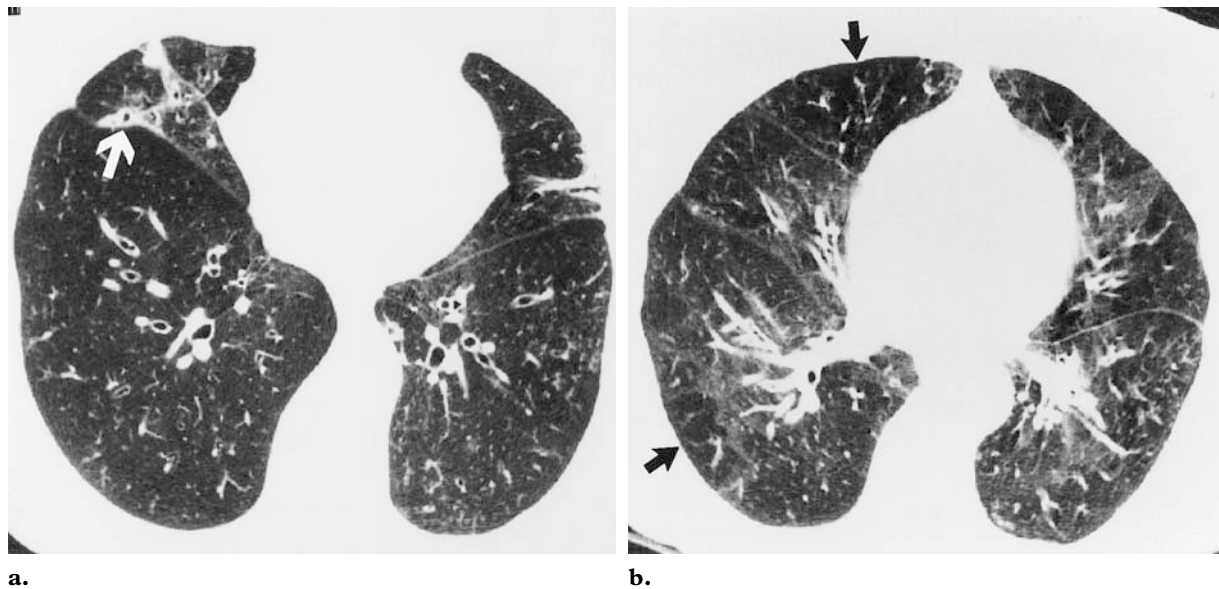
**Figure 9.** Rheumatoid arthritis with thoracic bone changes. Posteroanterior radiograph shows resorption of the distal clavicles and humeral heads.

Pulmonary nodules are uncommon in patients with rheumatoid arthritis and are usually associated with advanced disease and subcutaneous nodules. The nodules are pathologically identical to subcutaneous nodules. Usually multiple and well circumscribed, they often result in thick-walled cavities (10). Thoracic bone changes of rheumatoid arthritis include resorption of the distal part of the clavicles and erosive arthritis of the shoulders (Fig 9).

Patients with rheumatoid arthritis have an increased prevalence of airway diseases such as bronchiolitis obliterans and bronchiolitis obliterans organizing pneumonia. Obliterative bronchi-

olitis occurs with increased frequency in patients with rheumatoid arthritis regardless of whether they have received penicillamine or gold therapy. Findings at chest radiography are usually normal. High-resolution CT may show a characteristic mosaic pattern of attenuation and perfusion. Abnormal areas of lung tissue have decreased attenuation and vascularity due to redistribution of blood flow away from areas of abnormal ventilation. High-resolution CT performed at end-expiration is more sensitive than end-inspiratory scanning and shows areas of air trapping (11,12)





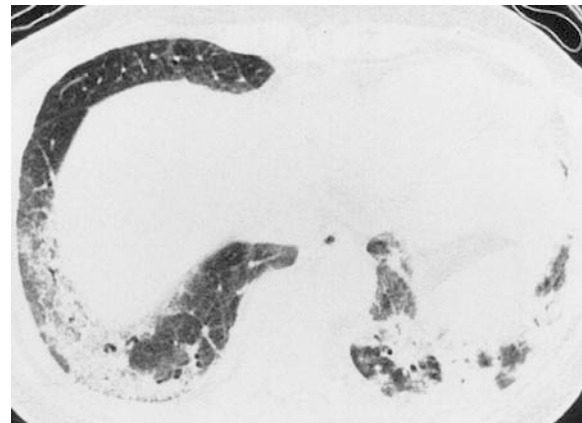
**Figure 10.** Rheumatoid arthritis with obliterative bronchiolitis in a patient who had undergone penicillamine therapy. **(a)** High-resolution CT scan obtained at end-inspiration shows cylindrical bronchiectasis in the right middle lobe (arrow). **(b)** High-resolution CT scan obtained at end-expiration shows patchy air trapping bilaterally, particularly in the right middle lobe and right lower lobe posteriorly (arrows).

(Fig 10). The predominant radiographic and high-resolution CT finding in patients with bronchiolitis obliterans organizing pneumonia is air-space consolidation that is usually bilateral and tends to have a patchy peripheral or peribronchial distribution.

Follicular bronchiolitis occurs with increased frequency in patients with rheumatoid arthritis. In a recent serial evaluation of high-resolution CT findings in 12 patients with follicular bronchiolitis, eight patients (66%) had rheumatoid arthritis (13). The major CT findings were small centrilobular nodules associated with patchy areas of ground-glass attenuation.

### Progressive Systemic Sclerosis

Progressive systemic sclerosis (scleroderma) is a connective-tissue disease of unknown pathogenesis that affects 30- to 50-year-old women three times as often as it affects men. This type of sclerosis is characterized by overproduction of collagen, which leads to fibrosis of the lungs, skin, vasculature, and visceral organs. Patients present with thickening and tightening of the skin; musculoskeletal manifestations; Raynaud phenomenon; and fibrosis of the lungs, kidneys, and gastrointestinal tract. Two-thirds of patients with progressive systemic sclerosis have clinical pulmonary symptoms, the most common of which are exertional dyspnea and dry, nonproductive cough.



**Figure 12.** Scleroderma with nonspecific interstitial pneumonitis. High-resolution CT scan obtained through the lung bases shows bibasilar areas of consolidation. Video-assisted thoracoscopic surgical biopsy showed nonspecific interstitial pneumonitis.

Pulmonary fibrosis is the most common radiographic finding, present in 20%–65% of patients (14,15). The fibrosis usually has a basilar predominance and appears initially as a fine reticular pattern that progresses to coarse reticulation and honeycombing (14) (Fig 11).

High-resolution CT may show evidence of fibrosis in patients with normal findings at radiography (15). In a prospective study of 23 patients with progressive systemic sclerosis, fibrosis was identified at chest radiography in 39% of patients and at high-resolution CT in 91% of patients (15). The predominant abnormalities at high-resolu-

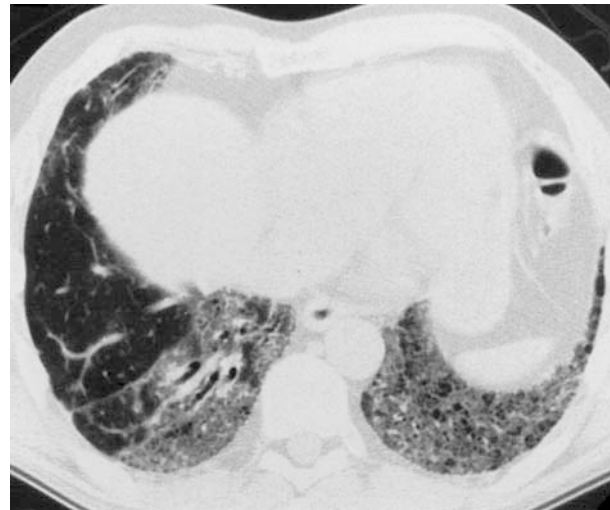


a.

**Figure 11.** Scleroderma with pulmonary fibrosis. **(a)** Posteroanterior radiograph shows low lung volumes and bibasilar, coarse reticular markings. **(b, c)** High-resolution CT scans show patchy areas of peripheral honeycombing **(b)** and diffuse honeycombing at the lung bases **(c)**.



b.

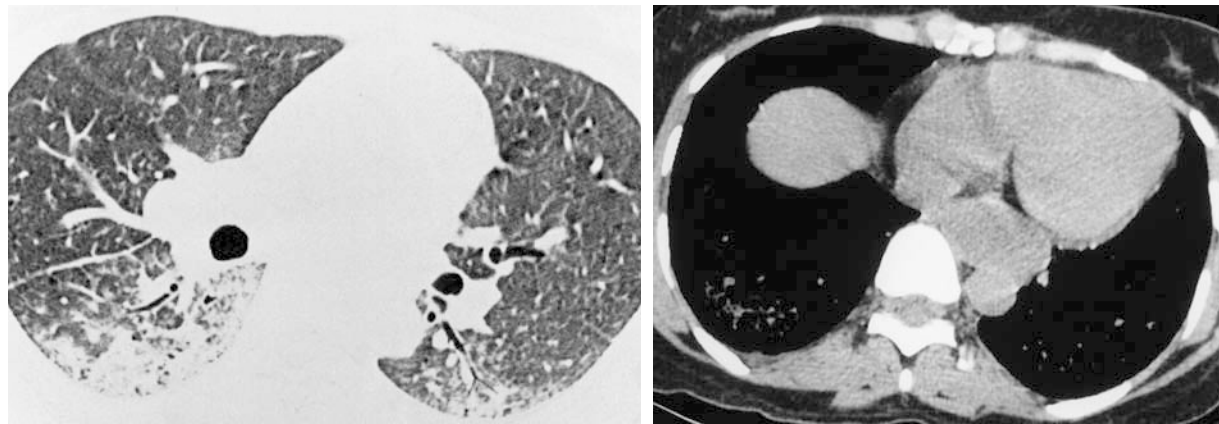


c.

tion CT consist of areas of ground-glass attenuation, poorly defined subpleural nodules, reticular pattern of attenuation, honeycombing, and traction bronchiectasis (15,16). As with rheumatoid disease and idiopathic pulmonary fibrosis, the abnormalities have a lower-lobe and peripheral predominance. The majority of patients with pulmonary fibrosis and progressive systemic sclerosis have a histologic pattern of usual interstitial pneumonia. However, many cases have a pattern of nonspecific interstitial pneumonitis (Fig 12). In a series of 64 patients with nonspecific interstitial pneumonitis, 10 (16%) had collagen vascular disease (17). The appearance of nonspecific interstitial pneumonitis at high-resolution CT is variable and nonspecific and consists of areas of ground-glass attenuation, consolidation, or a reticular

pattern (18). Patients with this disease have a much better prognosis than those with usual interstitial pneumonia.

There is an increased prevalence of lung cancer in patients with progressive systemic sclerosis, particularly in those with pulmonary fibrosis. Pleural disease is not a common manifestation, but when present, it is usually accompanied by parenchymal disease. The esophagus is usually found to be involved at clinical examination, and a dilated esophagus may be identified at chest radiography or CT. Aspiration pneumonia occurs with increased frequency owing to esophageal dysfunction (Fig 13).



**Figure 13.** Scleroderma with aspiration pneumonia. **(a)** High-resolution CT scan shows air-space consolidation in the superior segments of both lower lobes. **(b)** High-resolution CT scan (mediastinal windows) shows a dilated, fluid-filled esophagus.

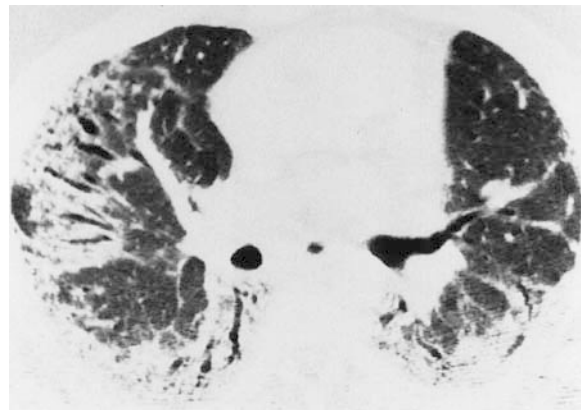
### Polymyositis and Dermatomyositis

Patients with polymyositis typically are first seen with progressive weakness of proximal striated muscles. Patients with dermatomyositis have skin changes in addition to muscle weakness. Women are affected more often than men, with a bimodal peak age of occurrence during childhood and middle adulthood. The most common radiographic finding is aspiration pneumonia secondary to pharyngeal muscle weakness. Involvement of the diaphragm leads to diaphragmatic elevation, reduced lung volumes, and basilar atelectasis.

Interstitial fibrosis occurs in 5%–30% of patients and appears as a fine reticular pattern that progresses to a coarse reticulonodular pattern and honeycombing (19) (Fig 14). The lung bases are most severely involved. Other parenchymal abnormalities include bronchiolitis obliterans organizing pneumonia and diffuse alveolar damage. The high-resolution CT findings of polymyositis and dermatomyositis have been recently described and consist predominantly of linear abnormalities and areas of ground-glass attenuation (20,21). Air-space consolidation is often also present, mainly in the middle and lower lung zones with a peribronchial and subpleural distribution. The consolidation is usually due to bronchiolitis obliterans organizing pneumonia (Fig 15).

### Sjögren Syndrome

Sjögren syndrome usually affects women (F:M, 9:1) in the fourth and fifth decades of life. Patients with Sjögren syndrome typically present with dry mouth and dry eyes. Histopathologic examination shows infiltration of exocrine glands

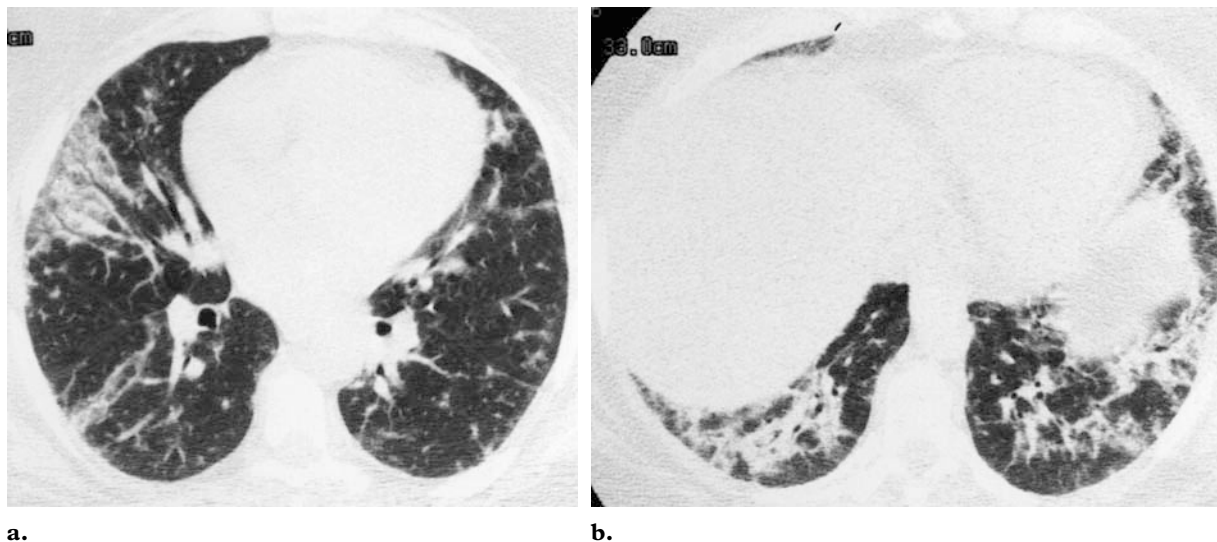


**Figure 14.** Polymyositis with pulmonary fibrosis (usual interstitial pneumonia). High-resolution CT scan shows peripheral architectural distortion and bilateral traction bronchiectasis.

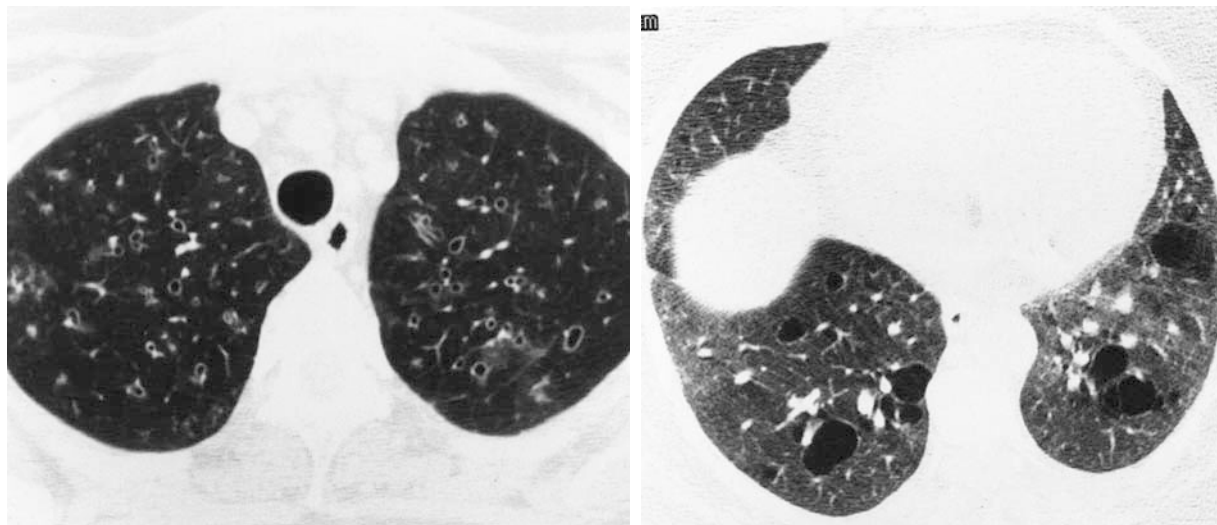
by immunoglobulin-producing lymphocytes. The salivary and lacrimal glands are most frequently involved, but extraglandular involvement occurs in 5%–10% of cases.

The most common radiographic finding associated with Sjögren syndrome is pulmonary fibrosis, seen in 10%–14% of cases. In an evaluation of high-resolution CT findings among 50 patients with Sjögren syndrome, the major abnormalities were bronchiectasis, bronchiolar inflammation, and increased parenchymal lines (22) (Fig 16). There is an increased prevalence of lymphocytic interstitial pneumonitis, which is seen radiographically as a reticulonodular pattern predominantly involving the lower lobes. A recent article described the high-resolution CT findings of lymphocytic interstitial pneumonia (23), with the most common findings being areas of ground-glass attenuation, thickening of bronchovascular bundles and interlobular septa, and cysts (Fig 17).





**Figure 15.** Dermatomyositis with bronchiolitis obliterans organizing pneumonia. High-resolution CT scans show a peripheral area of consolidation in the right middle lobe (a) and peribronchial consolidation in both lung bases (b).



**Figures 16, 17.** Sjögren syndrome with bronchiectasis. High-resolution CT scan shows extensive bilateral cylindrical bronchiectasis. (17) Sjögren syndrome with lymphocytic interstitial pneumonitis. High-resolution CT scan shows scattered bilateral thin-walled cysts.

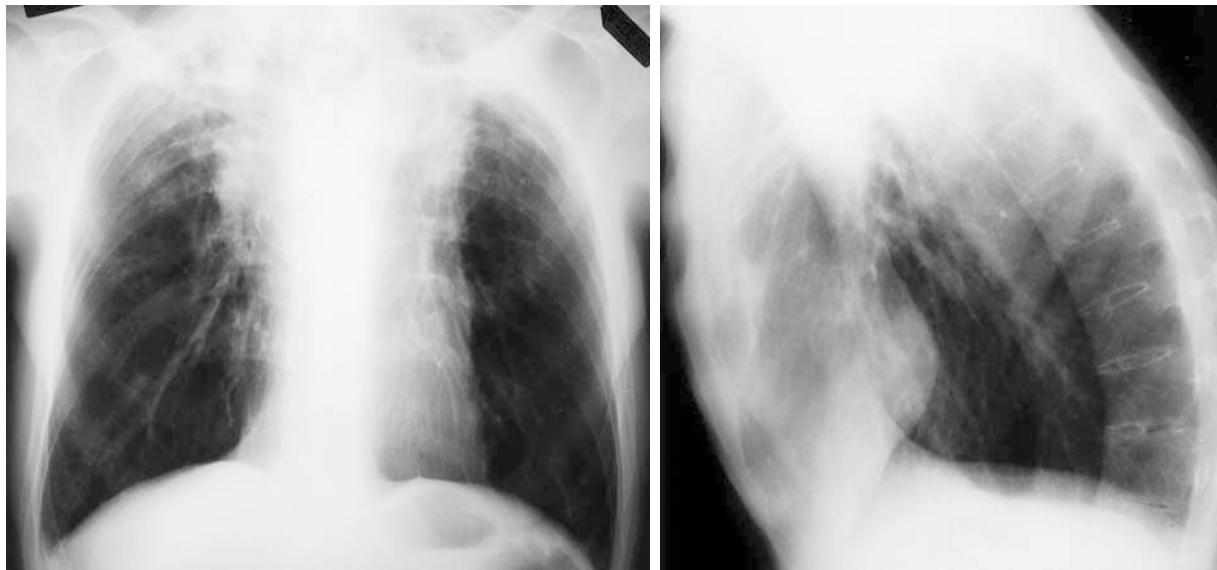
Patients with Sjögren syndrome also have an increased risk of lymphoma. Lymphoma should be suspected if a chest radiograph shows mediastinal lymphadenopathy or a pulmonary mass.

### Ankylosing Spondylitis

Ankylosing spondylitis is an autoimmune disease of unknown pathogenesis that primarily affects the axial skeleton. It has a male-to-female predominance of 3:1.

In a study of chest radiographic findings in 2,080 patients with ankylosing spondylitis, 26 (1.2%) had fibrosis in the upper lobes (24). The radiographic findings of ankylosing spondylitis consist of reticulonodular opacities in the lung

apices, which become confluent as the disease progresses (24). Common associated abnormalities include apical bullae and cavitation, potentially mimicking tuberculosis. The high-resolution CT findings in 26 patients with ankylosing spondylitis were recently described (25). The most common abnormalities are peripheral interstitial lung disease, bronchiectasis, paraseptal emphysema, and apical fibrosis (Fig 18). Radiographic changes of the spine, consisting of symmetric marginal syndesmophytes (“bamboo spine”), are usually evident when there is apical fibrosis. Restriction of the chest wall may result from fusion of the costovertebral joints.



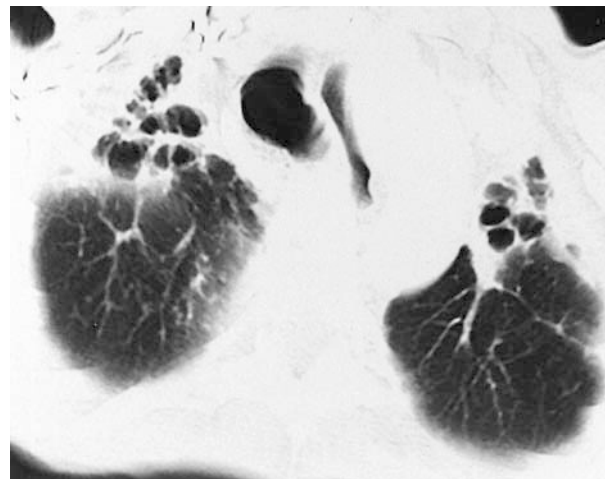
**Figure 18.** Ankylosing spondylitis. **(a)** Posteroanterior radiograph shows severe bilateral scarring in the upper lobes and volume loss. **(b)** Lateral radiograph shows flowing syndesmophytes of the thoracic spine. **(c)** CT scan shows severe volume loss of the lung apices with traction cystic bronchiectasis.

### Wegener Granulomatosis

Predominantly affecting male patients, Wegener granulomatosis is a systemic autoimmune disease characterized by granulomatous vasculitis of the upper and lower respiratory tracts, glomerulonephritis, and small-vessel vasculitis. The histopathologic hallmark of Wegener granulomatosis is a necrotizing vasculitis of small arteries and veins with granuloma formation.

Lung disease develops in most patients. The most common radiographic manifestation of Wegener granulomatosis is multiple nodules or irregularly margined masses with no zonal predominance (26,27). The nodules or masses are usually multiple but can be solitary in up to 25% of cases (27). Cavitation of the nodules occurs in approximately 50% of cases. The cavities usually have irregular, thick walls. With treatment, the nodules or cavities may resolve completely or result in a scar. At CT, the nodules typically have irregular margins and often have a peribronchovascular distribution (28,29) (Fig 19). Peripheral, wedge-shaped areas of consolidation representing infarcts may also be seen. Pleural effusions occur in less than 10% of cases. Mediastinal and hilar adenopathy are relatively uncommon.

Localized or diffuse areas of air-space consolidation may be present. These areas usually represent pulmonary hemorrhage, although pneumonia causes similar findings. Pulmonary hemorrhage can result in a focal area of dense



consolidation, patchy bilateral areas of consolidation, or diffuse air-space consolidation (27).

Involvement of the tracheal or bronchial walls usually consists of mucosal or submucosal granulomatous thickening. CT scans show smooth or nodular thickening of the tracheal or bronchial wall (Fig 20). The thickening may become severe, causing narrowing of the lumen and possible eventual calcification.

### Churg-Strauss Syndrome

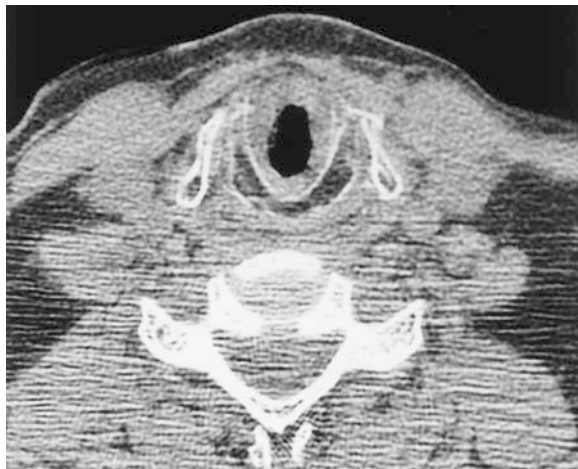
Churg-Strauss syndrome is an allergic angiitis and granulomatous necrotizing vasculitis that occur almost exclusively in patients with asthma. The syndrome is most common in patients aged 30–50 years and has no gender predilection. Patients are typically asthmatic and present with eosinophilia, fever, and allergic rhinitis. Findings of chest radiography are usually abnormal and most often consist of patchy, nonsegmental areas of consolidation with no zonal predominance (30) (Fig 21a).



**a.** **b.**  
**Figure 21.** Churg-Strauss syndrome. **(a)** Posteroanterior radiograph shows patchy bilateral areas of consolidation predominantly in the right lung. **(b)** High-resolution CT scan shows patchy bilateral ground-glass attenuation and consolidation of the right lower lobe.



**19a.** **19b.**  
**Figures 19, 20.** **(19)** Wegener granulomatosis. **(a)** Posteroanterior radiograph shows bilateral irregular nodules and a mass in the right lower lobe. **(b)** High-resolution CT scan shows multiple irregular nodules in a peribronchovascular distribution. **(20)** Wegener granulomatosis with tracheal involvement. CT scan shows diffuse thickening of the tracheal wall.



**20.**



Thoracic Manifestations of the Collagen Vascular Diseases						
	Systemic Lupus Erythematosus	Rheu- matoid Arthritis	Progressive Systemic Sclerosis	Polymyositis or Dermato- myositis	Sjögren Syndrome	Ankylosing Spondy- litis
Pleural disease	+++	+++	...	...	...	...
Pulmonary fibrosis	+	+++	+++	++	+	+
Diaphragm weakness	+++	...	...	+++	...	...
Aspiration pneumonia	...	...	+++	+++	...	...
Bronchiolitis obliterans organizing pneumonia	...	++	...	++	...	...
Bronchiolitis obliterans	...	++	...	...	...	...
Bronchiectasis	...	+	...	...	++	...
Apical fibrosis	...	...	...	...	...	+++

Note.— + = occasional occurrence, ++ = common occurrence, +++ = frequent occurrence or highly characteristic finding.

The areas of consolidation may have a peripheral distribution and are often transient. Nodules may occur, but cavitation is rare. Pleural effusions are present in approximately 30% of patients (30).

CT findings in 17 patients were recently described (31). The predominant findings were consolidation or ground-glass attenuation, seen in 10 (59%) patients (Fig 21b). Other, less common findings included pulmonary nodules, interlobular septal thickening, and bronchial wall thickening.

### Anti-Glomerular Basement Membrane Antibody Disease

Anti-glomerular basement membrane antibody disease (Goodpasture syndrome) is defined by a triad of diffuse pulmonary hemorrhage, glomerulonephritis, and circulating anti-glomerular basement membrane antibodies. The clinical pulmonary presentation usually consists of cough, mild shortness of breath, hemoptysis, and hypoxia in young male patients. Chest radiography typically shows extensive bilateral air-space consolidation (32) (Fig 22). The consolidation is usually symmetric, with a perihilar distribution and sparing of the lung apices. The consolidation usually resolves within 2–3 days and is replaced by a reticulonodular pattern and interlobular septal thickening. Findings at chest radiography may occasionally be normal despite the presence of diffuse pulmonary hemorrhage (32).



**Figure 22.** Anti-glomerular basement membrane antibody disease. Posteroanterior radiograph shows bilateral perihilar air-space consolidation.

### Conclusions

The systemic autoimmune diseases cause a variety of findings at chest radiography and high-resolution CT. Depending on the underlying autoimmune process, the pleura, pulmonary parenchyma, or airways may be predominantly affected. The Table summarizes the thoracic manifestations of the collagen vascular diseases. The remaining systemic autoimmune diseases—Wegener granulomatosis, Churg-Strauss syndrome, and Goodpasture syndrome—each cause distinct radiographic and CT findings.



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