**Imaging Manifestations of Collagen Vascular Disease**

*Steven Primack, MD*

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**THORACIC MANIFESTATIONS of COLLAGEN VASCULAR DISEASE**

Steven L. Primack, MD  
Department of Radiology  
Oregon Health and Science University

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**Objectives**

- Review spectrum of thoracic involvement in patients with collagen vascular disease
- Differences in thoracic manifestations of the various collagen vascular diseases

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**Collagen Vascular Disease**

- Systemic lupus erythematosus (SLE)
- Rheumatoid arthritis
- Progressive systemic sclerosis (scleroderma)
- Polymyositis / Dermatomyositis
- Sjögren’s syndrome
- Mixed connective tissue disease

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**SLE**

- 20-50 y/o women:men, 10:1
- Thoracic involvement in >50%
- Primary and secondary manifestations
- Pleural dz by far most common
- Infection relatively common
- ILD uncommon, < 5%

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**Primary Thoracic Manifestations**

- Pleuritis, pericarditis
- Acute lupus pneumonitis
- Pulmonary hemorrhage
- ILD: NSIP>>UIP, <5%, usually mild
- Diaphragm dysfunction
- PA HTN

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**Secondary Thoracic Manifestations**

- Pneumonia: community acquired, opportunistic
- Atelectasis
- Drug-induced lung disease
- Renal failure, pulmonary edema
**SLE**  
**Pleural Disease**

- Up to 50% during course of illness  
- Unilateral or bilateral effusions  
- Most effusions resolve, some residual thickening  
- Associated pericardial effusion  
- Ddx: parapneumonic, nephrotic syndrome, PE

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**SLE - Pleural Effusion**

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**SLE – Pleural and Pericardial Effusion**

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**SLE**  
**Acute Lupus Pneumonitis**

- Incidence 1 – 4%, usually established disease  
- DAD, necrosis, ± alveolar hemorrhage  
- Mimics pneumonia, infarction, hemorrhage  
- Patchy consolidation, usually bilateral  
- Diagnosis of exclusion
SLE
Pulmonary Hemorrhage
- Incidence <3%, usually established disease
- Rapid onset dyspnea, fever, hemoptysis
- Diffuse bilateral ground glass or consolidation
- Mortality 40-90%

SLE – Pulmonary Hemorrhage

Pneumocystis Pneumonia

Diaphragm Dysfunction
- Diaphragmatic weakness due to myopathy
- Incidence: ~20%
- Usually bilateral hemidiaphragm elevation
- “Shrinking lung” syndrome
Rheumatoid Arthritis

• 25 – 50 y/o, women:men, 3:1
• Thoracic disease more common in men
• Extraarticular involvement in ≈ 50%

Rheumatoid Arthritis
Thoracic Manifestations

• Pleural disease
• Bronchiectasis, BO (consider expiratory scan)
• UIP slightly more common than NSIP
• Organizing pneumonia
• Follicular bronchiolitis
• Necrobiotic nodules

Rheumatoid Arthritis
Pleural Disease

• Pleural effusion – usually unilateral, often loculated
• Pleural thickening very common, up to 50% at autopsy
• 4% before, 20% with onset of joint disease
• Associated pericarditis, subcutaneous nodules
### Nonspecific Interstitial Pneumonitis (NSIP)

- Fibrotic or cellular NSIP
- HRCT: ground-glass opacity, consolidation, reticulation, traction bronchiectasis
- Peripheral and lower lung zone
- May have subpleural sparing
- Honeycombing generally absent
RA – interval increase in GGO

RA – NSIP, Pneumocystis Pneumonia

RA – Necrobiotic Nodules
- Rare, <0.5% incidence on CXR
- Advanced RA and subcutaneous nodules
- Wax and wane with arthritis
- Cavitation common, irregular margins

Rheumatoid Arthritis: Organizing Pneumonia

RA – Follicular Bronchiolitis

Progressive Systemic Sclerosis (Scleroderma)
- Inflammatory, fibrotic changes of lungs, skin, vasculature, visceral organs
- Limited and diffuse forms
- 45 – 65 y/o women:men, 3:1 – 8:1
Scleroderma
Thoracic Manifestations

- Pulmonary fibrosis: 30 - 90% patients
  - NSIP >> UIP
- Esophageal dysmotility - dilated, air-filled esophagus, aspiration
- Follicular bronchiolitis
- PA HTN: ILD, direct vascular involvement
- Highest mortality of CVDs: PA HTN

33F with scleroderma

Scleroderma - NSIP

Scleroderma - UIP

Supine

NSIP

Prone

33F

SUNDAY
Polymyositis / Dermatomyositis

- Inflammatory myopathies of skeletal muscle
- Dermatomyositis – additional skin changes
- 40 – 50 y/o women:men, 2:1
- Progressive weakness of proximal muscles
- Hypoventilation, respiratory failure

Polymyositis/Dermatomyositis
Thoracic Manifestations

- Diaphragm and intercostal muscle weakness – elevated diaphragm, basilar atelectasis
- Pharyngeal muscle weakness – aspiration
- ILD: NSIP and OP most common, often coexist
  - UIP also occurs
  - ILD in 30 - 60%, most frequent if anti-Jo-1

Dermatomyositis 60m

Atelectasis v. ILD
Sjögren’s Syndrome

- Lymphocyte infiltration – salivary, lacrimal glands, extraglandular in 5 – 10%
- 30-50 y/o women:men, 9:1
- Primary or secondary (usually with RA)
- Lymphoma – as high as 44-fold increase

Thoracic manifestations

- Airway disease most common – bronchiectasis
- ILD - uncommon
- Lymphocytic interstitial pneumonitis (LIP)
- Follicular bronchiolitis
- Lymphoma – enlarged nodes, multifocal masses, consolidation, pleural effusion

Dermatomyositis – NSIP, OP
**Mixed Connective Tissue Disease**

- SLE, scleroderma, RA, PM/DM
- Pleural effusion
- ILD: NSIP > UIP
- PA HTN
- Diaphragmatic dysfunction
- Aspiration

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**Collagen Vascular Disease**

- NSIP/UIP: PSS, PM/DM, RA
  SLE, Sjögren’s
- Pleural: SLE, RA
- OP: PM/DM, RA
- BO: RA
- Bronchiectasis: RA, Sjögren’s
- Follicular bronchiolitis: RA, Sjögren’s

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**Collagen Vascular Disease**

- Aspiration: PM/DM, scleroderma
- Diaphragm: PM/DM, SLE
- Consolidation: pneumonia, OP
- Lung Cancer: increased with fibrosis
- LIP, lymphoma: Sjögren’s
- PA HTN: PSS and MCTD >> SLE
Suggested Reading