Interstitial Lung Disease in Collagen Vascular Diseases

Ami N. Rubinowitz, M.D
Associate Professor
Co-Chief of Thoracic Imaging
Dept. of Diagnostic Radiology
Yale University School of Medicine

Disclosures

• There are no commercial supporters of this workshop
• There are no conflicts of interest

Learning Objectives

• To recognize common CT patterns of lung injury in collagen vascular diseases.
• To highlight the salient features of lung injury in each CVD.
• To become familiar with ancillary imaging findings ("clues") that may suggest one CVD over another.

Collagen Vascular Diseases

Group of autoimmune disorders whose common denominator is damage to components of connective tissue at variety of sites in the body

Collagen Vascular Diseases

• RA
• Scleroderma (PSS)
• DM/PM
• SLE
• MCTD
• Sjogren’s

Thoracic Involvement of CVD’s

• Pt may be asymptomatic
• Often times is first manifestation of an underlying CVD
• If known CVD, no need for bx in most cases
• HRCT > CXR
Thoracic Involvement of CVD’s
- Parenchymal
- Pleural
- Airway
- Vascular
- Musculoskeletal

Collagen Vascular Diseases
- Rheumatoid Arthritis
- Scleroderma (PSS)
- DM/PM
- SLE
- MCTD
- Sjogren’s

RA--Pulmonary involvement
- 30-40% at HRCT will have pulmonary fibrosis
- *UIP (mc)
- NSIP
- OP

Collagen Vascular Diseases
- RA
- Scleroderma (PSS)
- DM/PM
- SLE
- MCTD
- Sjogren’s
Scleroderma--Thoracic involvement

- 40-100% of cases, Pulmonary fibrosis
- NSIP (mc), UIP, OP
- Pul HTN
- Dilated esophagus

Collagen Vascular Diseases

- RA
- Scleroderma (PSS)
- Dermatomyositis/Polymyositis
- SLE
- MCTD
- Sjogren’s

DM/PM--Pulmonary involvement

- 30% of cases, pul fibrosis: UIP, NSIP
- OP
- DAD
Collagen Vascular Diseases
- RA
- Scleroderma (PSS)
- DM/PM
- Systemic Lupus Erythematosus
- MCTD
- Sjogren’s

SLE
- Fibrotic ILD not common
- NSIP, UIP
- OP

SLE, NSIP

Collagen Vascular Diseases
- RA
- Scleroderma (PSS)
- DM/PM
- Systemic Lupus Erythematosus
- MCTD
- Sjogren’s

Mixed Connective Tissue Disorder
- Mixed features of: SLE, scleroderma and PM
- 20-80% have respiratory involvement
- NSIP, UIP, OP

Mixed Connective Tissue Disorder
- 28 yo male, MCTD
Collagen Vascular Diseases

- RA
- Scleroderma (PSS)
- DM/PM
- Systemic Lupus Erythematosus
- Mixed Connective Tissue Disorder
- Sjogren’s Syndrome

Sjogren’s Syndrome–Pulmonary Involvement

- Up to 30% have findings on CT
- 10-14% of cases, pul fibrosis (NSIP, UIP)
- OP, *LIP

What’s most common?

- UIP: RA
- NSIP: Scleroderma, DM/PM
- OP: DM/PM, MCTD
- LIP: Sjogren’s
- DAD: DM/PM, SLE
**Summary: ILD in CVD**

- May be the 1st manifestation of an underlying CVD
- Imp. to obtain HRCT in these patients
- Same patterns as see with the IIP’s
- Certain patterns more common with some CVD’s than with others

*RA, PSS, DM/PM*  *SLE, MCTD, Sjogren’s*

**References**

