A Great Imitator: 
Imaging the Musculoskeletal Sequelae of 
Systemic Lupus Erythematosus 

Daniel Driscoll, Harvard Medical School Year III 
Gillian Lieberman, MD
Agenda

1. Brief Introduction to Systemic Lupus Erythematosus

2. Index Patient Presentation
   • Basics of SLE Arthropathy Diagnosis, Imaging and Treatment

3. Imaging of additional musculoskeletal sequelae of SLE
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Introduction to Systemic Lupus Erythematosus

- SLE is a systemic autoimmune disease
- Epidemiology:
  - Females > Males
  - Over one million people with SLE in the US
- Pathogenesis:
  - There are two main mechanisms of SLE sequelae
    1. Reduced cell apoptosis
    2. Self-antibody generation

Image adapted from healthtap.com
Systemic Lupus Erythematosus Diagnosis Criteria

- SLE Diagnosis requires 4 of 11 of the following American College of Rheumatology Criteria

(Mnemonic: SOAP BRAIN MD)

- Serositis
- Oral ulcers
- Arthritis
- Photosensitivity
- Blood disorders
- Renal involvement
- Antinuclear antibodies
- Immunologic phenomena (eg, dsDNA; anti-Smith [Sm] antibodies)
- Neurologic disorder
- Malar rash
- Discoid rash
SLE is a Multisystem Disease

- **Cardiovascular SLE sequelae** (16-44% of patients)
  - Valve disease
  - Coronary artery disease
- **Gastrointestinal SLE sequelae** (20-40% of patients)
  - Pancreatitis
  - Colitis
- **Neurologic and Psychiatric SLE sequelae** (10-80% of patients)
  - Seizures
  - Psychosis
- **Respiratory SLE sequelae**
  - Serositis
- **Renal SLE sequelae**
  - Lupus nephritis
- **Musculoskeletal SLE sequelae** (90-95% of patients)
  - Joints
    - Lupus arthritis
    - Deforming Arthropathy
      - Mild Deforming Arthropathy
      - “Rhupus”
      - Jaccoud’s Arthropathy
  - Ligaments/Tendons
    - Tenosynovitis
  - Bones
    - AVN
    - Insufficiency Fracture
  - Muscles
    - Myositis
  - Fat
    - Panniculitis
SLE is a **Musculoskeletal Disease**

- **Musculoskeletal Sequelae** are found in 90-95% of SLE patients
- SLE affects different parts of the MSK system
  - **Joints**
    - Lupus arthritis
    - Deforming Arthropathy
      - Mild Deforming Arthropathy
      - “Rhupus”
      - Jaccoud’s Arthropathy
    - Tenosynovitis
  - **Ligaments/Tendons**
    - Tenosynovitis
  - **Bones**
    - Avascular necrosis (AVN)
  - **Muscles**
    - Myositis
  - **Fat**
    - Panniculitis

Images adapted from imgbuddy.com
There are two different types of SLE joint sequelae: (1) SLE Arthritis and (2) Deforming arthropathy

**SLE Arthritis:**
- **Common** in SLE patients (95% of patients)
- non-erosive, non-deforming arthritis
- moderately painful, symmetric and migratory

**Deforming Arthropathy:**
- **Less common** in SLE Patients
- Deforming but usually non-erosive process
- Several sub-classifications (see table on next slide for distinguishing factors):
  - Mild Deforming Arthropathy
  - Jaccoud’s Arthropathy
  - “Rhupus”
## SLE Joint Sequelae (2)

<table>
<thead>
<tr>
<th></th>
<th>SLE Arthritis</th>
<th>Mild Deforming Arthropathy</th>
<th>Jaccoud's Arthropathy</th>
<th>Rhupus</th>
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<tbody>
<tr>
<td>Erosions?</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>Yes</td>
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<tr>
<td>Deforming?</td>
<td>No</td>
<td>Yes, mild</td>
<td>Yes, severe</td>
<td>Yes</td>
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<tr>
<td>Percentage of SLE patients</td>
<td>~90%</td>
<td>~5%</td>
<td>10-35%</td>
<td>3-5%</td>
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<tr>
<td>Role for imaging?</td>
<td>Limited</td>
<td>Yes</td>
<td>Yes</td>
<td>Yes</td>
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</tbody>
</table>
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Our Patient:

• A 48 year old woman presents with one year history of pain and stiffness in digits 2, 3, 4 and 5 on her right hand.

• What is the appropriate imaging?
# ACR Appropriateness Criteria

**Clinical Condition:** Chronic Wrist Pain  
**Variant 1:** With or without prior injury. Best initial study.

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<th>Radiologic Procedure</th>
<th>Rating</th>
<th>Comments</th>
<th>RRL*</th>
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<td>MRI wrist without contrast</td>
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<td></td>
<td>O</td>
</tr>
<tr>
<td>MRI wrist without and with contrast</td>
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<td></td>
<td>O</td>
</tr>
<tr>
<td>MR arthrography wrist</td>
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</tr>
<tr>
<td>CT wrist without contrast</td>
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<td>CT wrist with contrast</td>
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<td>CT wrist without and with contrast</td>
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<td>CT arthrography wrist</td>
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<tr>
<td>US wrist</td>
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<td>O</td>
</tr>
<tr>
<td>Tc-99m bone scan wrist</td>
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<td>☀</td>
</tr>
</tbody>
</table>

*Relative Radiation Level

**Rating Scale:** 1,2,3 Usually not appropriate; 4,5,6 May be appropriate; 7,8,9 Usually appropriate
Hand Anatomy

Bones
- Distal Phalanges
- Intermediate Phalanges
- Proximal Phalanges
- Metacarpals
- Carpals

Joints
- Distal interphalangeal joints (DIP)
- Proximal interphalangeal joints (PIP)
- Metacarpophalangeal joints (MCP)
- Carpometacarpal joints (CMC)
- Carpal Ulnar Joints

Images adapted from Wikimedia Commons

Daniel Driscoll, HMS III
Gillian Lieberman, MD
Our Patient’s Radiographs (Right Hand)

Frontal  Lateral  Oblique
Our Patient’s Radiographic Findings

**Ulnar deviation and MCP subluxation**

**Boutonnière Deformity**

*NO erosions noted*

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Gillian Lieberman, MD

Images from PACS, BIDMC
Courtesy of Dr. Jim Wu
Initial Differential Diagnosis for our patient: Arthritis with Multiple Subluxations and/or Ulnar Deviation

- Rheumatoid Arthritis
- Ehlers-Danlos Syndrome
- Jaccoud’s arthritis (post-rheumatic fever)
- Juvenile chronic arthritis
- Systemic Lupus Erythematosus
- Mixed Connective Tissues
- Neuropathic arthropathy with or without destruction
- Other advanced arthritis
- Psoriatic Arthritis
More information about our patient:

- **Chief Complaint:** A 48 year old woman with long-standing history of SLE presents with one year history of inability to extend digits 2-5 on right hand.

- **HPI:**
  - Denies tingling or numbness
  - Patient is able to place joints in place manually

- **Physical Exam**
  - On exam, deformities are passively correctable

- **PMH and Labs:**
  - Positive ANA and anti-Sm antibody
  - Negative Anti-U1RNP Antibody
  - Negative Rheumatoid Factor and Anti-CCP Antibody
  - No history of rheumatic fever
Refined Differential Diagnosis for our patient

After learning more about our patient, we can dismiss nearly all potential diagnoses

- Systemic Lupus Erythematosus
- Rheumatoid Arthritis
- Ehlers-Danlos Syndrome
- Post-rheumatic fever Jaccoud’s arthritis
- Juvenile chronic arthritis
- Mixed Connective Tissue Disease
- Neuropathic arthropathy with or without destruction
- Psoriatic Arthritis
SLE Arthropathy Diagnosis Algorithm

Start HERE

MCP Finger deviation/subluxation

IMAGING: RADIOGRAPHY

Erosions?

Yes

No

IMAGING: Ultrasound or MRI

Erosions?

Yes

No

JAI > 5

JAI < or = 5

*JAI = Jaccoud Arthropathy Index

Diagnosis: "Rhupus"

Diagnosis: Jaccoud's Arthropathy

Diagnosis: Mild Deforming Arthropathy

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Adapted from Messuti et al. 2014
Classic Deformities of Jaccoud’s Arthropathy

“Z” Deformity

Ulnar Deviation

Boutonniere Deformity

Swan Neck Deformity

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Images from PACS, BIDMC and adapted from www.kleisertherapy.com
# Jaccoud Arthropathy Index (JAI)

(greater than 5 points is diagnostic for Jaccoud Arthropathy)

<table>
<thead>
<tr>
<th>Deformity</th>
<th>Affected Fingers</th>
<th>Points</th>
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</thead>
<tbody>
<tr>
<td>Ulnar Drift</td>
<td>&lt;=50%</td>
<td>2</td>
</tr>
<tr>
<td></td>
<td>&gt;50%</td>
<td>3</td>
</tr>
<tr>
<td>Swan Neck Deformities</td>
<td>&lt;=50%</td>
<td>2</td>
</tr>
<tr>
<td></td>
<td>&gt;50%</td>
<td>3</td>
</tr>
<tr>
<td>Boutonniere Deformities</td>
<td>&lt;=50%</td>
<td>1</td>
</tr>
<tr>
<td></td>
<td>&gt;50%</td>
<td>2</td>
</tr>
<tr>
<td>Z deformity</td>
<td>1</td>
<td>2</td>
</tr>
<tr>
<td></td>
<td>2</td>
<td>3</td>
</tr>
</tbody>
</table>

Adapted from Messuti et al. 2014
Which deforming arthropathy does our patient have?

1. Ulnar deviation of digits 2, 3, 4 and 5
2. No degenerative changes or erosions
3. JAI < 5

Diagnosis: Mild Deforming Arthropathy
Our patient received MCP joint arthroplasty in digits 2-5 as definitive treatment for her deforming arthropathy.
Companion Patient 1: Potential Rhupus Patient

MCP Joint subluxation
Subchondral erosions
Boutonnière Deformity

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Images from PACS, BIDMC
Courtesy of Dr. Jim Wu
Companion Patients 2 and 3

Ultrasound can be used to show tenosynovitis, joint erosion, or joint effusion in SLE patients.

Patient 2. SLE patient with tenosynovitis of third finger flexor tendon showing positive power Doppler signal.

Patient 3. SLE patient who is ANA positive with erosion and large effusion of the second MCP joint.
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Avascular Necrosis

Epidemiology: 13% of SLE patients have AVN

**Late AVN** lesions:
- detectable by Radiograph
- **Serpiginous sclerosis**

**Early AVN** lesions:
- detectable by MRI
- **Serpiginous sclerosis**

Myositis

- Definition: Inflammation of muscle tissue
- Epidemiology: 8% of SLE patients
- T2 MRI with fat suppression findings: linear areas of high signal intensity

Tenosynovitis

- **Definition:** Inflammation of synovium surrounding tendon
- **Epidemiology:** 10-44% of SLE patients
- **Ultrasound findings:** Fluid collection around the tendon and within its sheath
- **MRI findings:** Hyperintensity

Panniculitis

- **Definition:** Inflammation of subcutaneous fat tissue
- **Epidemiology:** <1% of SLE patients

**Imaging findings:**
- MRI STIR (post contrast)
  - hyperintensity
- MR T1 FS (post contrast)
  - hyperintensity

Daniel Driscoll, HMS III  
Gillian Lieberman, MD  
Images from PACS, BIDMC  
Courtesy of Dr. Jim Wu
Summary

1. SLE is an autoimmune disease with multiple potential effects on the body and musculoskeletal system

2. Radiographs and ultrasound, along with clinical history, are efficacious in the diagnosis of SLE deforming arthropathy

3. Additional musculoskeletal sequelae of SLE can be imaged with different modalities, with MRI being efficacious for detection of inflammation
Acknowledgements

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References


