Myositis and Your Lungs

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Myositis

Heterogeneous group of autoimmune syndromes characterized by chronic muscle weakness (muscle inflammation), other organ system involvement and a cause that is unknown.
Autoimmunity

- Immune response against *self*
  - loss of tolerance

- Unknown cause
  - susceptibility factors (genetic)
  - environmental triggers
    - e.g. infection

- Multiple diseases and "syndromes"
  - which sometimes run in families
# Autoimmune Diseases

<table>
<thead>
<tr>
<th>Disease</th>
<th>Target</th>
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<tbody>
<tr>
<td>Rheumatoid Arthritis</td>
<td>Joints (synovium)</td>
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<tr>
<td>Systemic Lupus Erythematousus</td>
<td>Skin, joints, kidneys</td>
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<tr>
<td>Scleroderma</td>
<td>Skin</td>
</tr>
<tr>
<td>Multiple Sclerosis</td>
<td>Nervous system</td>
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<tr>
<td>Myositis</td>
<td>Muscle</td>
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</tbody>
</table>

Nearly every AI disease has **multiple** targets!
Autoimmunity (cont’d)

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- Multiple diseases and “ syndromes”
  - which sometimes run in families

- Formation of autoantibodies
  - markers of autoimmunity

- Inflammatory in nature
Immune cells (lymphocytes) “attacking” normal muscle tissue in a patient with polymyositis
Systemic Features of Myositis

Musculoskeletal
- Weakness
- Muscle pain/ tenderness
- Muscle atrophy
- Arthralgias
- Arthritis

Cardiac
- Arrhythmias
- Congestive failure

Pulmonary
- Atelectasis from muscle weakness
- Aspiration pneumonia
- ILD

Gastrointestinal
- Dysphagia
- Reflux
- Dysmotility

General
- Fever
- Fatigue
- Weight loss
- Raynaud’s

Cutaneous
- Rashes
- Calcification

Slide, courtesy of Dr. Fred Miller
Systemic Targets of Myositis

- Skin
- Joint pain (arthritis)
- GI tract: difficulty swallowing
- **Lung**
  - Shortness of breath
  - Inflammation in lung tissue
  - Fibrosis (scar tissue)
  - Associated with markers in the blood called antibodies
Case Presentation

- 41 y.o. white male with hypertension, high cholesterol and allergies
- 3/20: swelling around eyes
- 3/27: joint pain (arthritis)
- 4/7: shortness of breath and fever
- 4/11: admitted to local hospital with abnormal chest x-ray and diagnosed with pneumonia
- 4/26: breathing worsens; poor response to antibiotics and transferred to UPMC
This patient did not have pneumonia or infection as the cause of his shortness of breath. He had interstitial lung disease (ILD) associated with his myositis.
How Common is ILD in Myositis?

- At least 30-40% of myositis patients have ILD
  - most commonly involved organ system in myositis

- There is no correlation between the extent and severity of muscle or skin disease and the development of ILD

The skin and muscle may be mild or even non-existent but the ILD may be severe
How Do We Diagnose and Follow ILD?

- Think about it!
- Chest x-rays
- High resolution CT scans
- Lung biopsy
- Pulmonary Function Tests (PFTs)
- Blood tests
  - Antibody markers
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The patient today had a marker in his blood that is associated with ILD in myositis patients.
Case Presentation

- Lung biopsy: inflammation
- Mild myositis on further testing
- Anti-Jo-1 autoantibody
- “Anti-synthetase syndrome”
“Anti-Synthetetase Syndrome”

- Acute onset of symptoms
- Lung symptoms may dominate the clinical picture in the form of ILD
- Also may have myositis, fever, arthritis, Raynaud phenomenon, “mechanic’s hands”
Mechanic’s Hands

13 days later
University of Pittsburgh Autoantibody Cohort

<table>
<thead>
<tr>
<th>Autoantibody</th>
<th>Number (% synthetases)</th>
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<tbody>
<tr>
<td>Jo-1</td>
<td>140 (60)</td>
</tr>
<tr>
<td>PL-12</td>
<td>36 (16)</td>
</tr>
<tr>
<td>PL-7</td>
<td>27 (12)</td>
</tr>
<tr>
<td>EJ</td>
<td>11 (5)</td>
</tr>
<tr>
<td>OJ</td>
<td>6 (3)</td>
</tr>
<tr>
<td>KS</td>
<td>9 (4)</td>
</tr>
<tr>
<td><strong>Total Synthetases</strong></td>
<td><strong>229</strong></td>
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Clinical Features of ILD in Myositis

- Shortness of breath with or without cough
- In about 1/3 of myositis patients, ILD precedes the muscle or skin manifestations
- Variable course:
  - Acute and dramatic or subacute
  - Chronic and more slowly progressive
  - Without symptoms (x-ray findings of mild fibrosis)
Other Causes of Breathing Problems in Myositis

- Inflammation in the breathing muscles
- Aspiration into the lungs because of swallowing problems
  - Pneumonia/infection
- Heart involvement
Treatment of ILD in Myositis Patients

- Steroids (prednisone) still the initial treatment
- Cyclophosphamide and azathioprine used early or in steroid-resistant cases with variable results
- CellCept is being increasingly used
- Cyclosporin A and tacrolimus, also used to prevent rejection of transplanted organs