Complications of Inflammatory Myopathy:
Lung Disease
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Case

- 41 y.o. white male with hypertension and hypercholesterolemia:

- **3/20/01**: periorbital edema

- **3/27/01**: acute polyarthritis treated with steroids

- **4/7/01**: dyspnea, fever

- **4/11/01**: admitted to outside hospital with abnormal chest radiograph and bilateral infiltrates

- **4/26/01**: worsening dyspnea; unresponsive to antibiotics and steroids and transferred to UPMC
- **ROS:** no Raynauds, mild joint pain, no dysphagia or weight loss

- **Exam** (Post bronchoscopy and BAL/biopsy):
  - dyspneic male with O2 saturation 90% (100% O2 mask/nasal cannula); otherwise normal VS and afebrile
  - erythematous rash but no heliotrope or Gottron’s sign
  - diffuse rales
  - no synovitis
  - normal muscle strength

- **Labs:** WBC=11.7; Hgb nl; renal normal; ANA negative; CPK=657

- **BAL/Biopsy:** organizing hyaline membranes; COP-like

- **EMG:** generalized myopathy
**Diagnosis:** Polymyositis

- anti-Jo-1 antibody returned later as positive
- Anti-synthetase syndrome

**Treatment:** pulse IV solumedrol; tacrolimus

**Course:**

- pneumomediastinum
- No intubation necessary
- off O2
- prednisone tapered and tacrolimus continued
Objectives

• Types of Lung Involvement

• Diagnosis of Lung Disease

• Management of Lung Disease
Lung Involvement

- multiple forms of lung involvement in myositis

- “Extrinsic”
  - weakness of respiratory muscles
  - aspiration (due to swallowing impairment)
  - opportunistic infection
  - congestive Heart Failure (rare)
  - pulmonary hypertension (rare)

- “Intrinsic”
  - Interstitial Lung Disease (ILD)
Lung Involvement

- at least 30% IIM patients have ILD
  - most commonly involved extramuscular organ system
- Anti-Jo-1 Ab found in 50–75% IIM pts with ILD
  - strong association of ILD with all anti-synthetase Ab in myositis
- lung disease may precede muscle involvement
- significant contribution to morbidity/mortality
  - 5 year survival with ILD ~70% (vs. ~ 85%)
  - likely depends on subtype of ILD

No correlation between extent/severity of muscle or skin disease and activity of ILD
Lung Involvement: Symptoms

- dyspnea with or without nonproductive cough
- no digital clubbing unlike idiopathic pulmonary fibrosis (IPF)
- pleuritis and pleural effusion are unusual (unlike SLE)
- variable presentation of ILD:
  - acute (ARDS) or subacute [Clawson, A&R, 1995]
  - chronic and more slowly progressive
  - asymptomatic (usually with basilar fibrosis)
Objectives

- Types of Lung Involvement
- Diagnosis of Lung Disease
- Management of Lung Disease
Diagnosis

1. Clinical suspicion

2. Imaging

3. Pulmonary Function Tests (PFTs)

4. Biopsy—rule out alternative processes
Diagnosis: Autoantibodies

Anti-synthetases

PL-7
PL-12
EJ
Jo-1

PM
SRP
TIF-1γ
DM
MJ
PM-Scl
SSc
U1RNP
Overlap
# Autoantibodies

<table>
<thead>
<tr>
<th>Antibody</th>
<th>Target</th>
<th>Subset</th>
<th>Phenotype</th>
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<tbody>
<tr>
<td>Mi-2</td>
<td>NuRD</td>
<td>DM</td>
<td>Shawl, V-neck, Gottron’s</td>
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<tr>
<td>CADM-140</td>
<td>MDA-5</td>
<td>DM</td>
<td>Amyopathic, ILD</td>
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<tr>
<td>SAE</td>
<td>SUMO</td>
<td>DM</td>
<td>ILD, dysphagia</td>
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<tr>
<td>MJ</td>
<td>NXP-2</td>
<td>JDM</td>
<td>Calcinsosis, Ulceration</td>
</tr>
<tr>
<td>p155/140</td>
<td>TIF1-g</td>
<td>DM, JDM</td>
<td>Severe skin, malignancy</td>
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<tr>
<td>SRP</td>
<td>72, 54 kDa</td>
<td>PM</td>
<td>Severe/refractory myositis</td>
</tr>
<tr>
<td>p200/100</td>
<td>HMGCR</td>
<td>IMNM</td>
<td>Necrotizing myopathy</td>
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<tr>
<td>Jo-1</td>
<td>ARS</td>
<td>PM/DM</td>
<td>Anti-synthetase syndrome</td>
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## Anti-synthetase Autoantibodies

<table>
<thead>
<tr>
<th>Antibody</th>
<th>Antigen (tRNA synthetase)</th>
<th>Prevalence in IIM (%)</th>
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<tbody>
<tr>
<td>Jo-1</td>
<td>histidyl</td>
<td>20-30</td>
</tr>
<tr>
<td>PL-7</td>
<td>threonyl</td>
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<tr>
<td>PL-12</td>
<td>alanyl</td>
<td>&lt;5</td>
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<tr>
<td>OJ</td>
<td>isoleucyl</td>
<td>&lt;5</td>
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<tr>
<td>EJ</td>
<td>glycyl</td>
<td>&lt;5</td>
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<tr>
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<td>asparaginyl</td>
<td>&lt;1</td>
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<tr>
<td>Tyr</td>
<td>tyrosyl</td>
<td>&lt;1</td>
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<tr>
<td>Ze</td>
<td>phenylalanyl</td>
<td>&lt;1</td>
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</tbody>
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Myositis--Autoantibodies