Myositis: It's Complicated

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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

Disease	Target
Rheumatoid Arthritis	Joints (synovium)
Systemic Lupus Erythematosus	Skin, joints, kidneys
Scleroderma	Skin
Multiple Sclerosis	Nervous system
Myositis	Muscle

Nearly every AI disease has **multiple** targets!

Autoimmunity (cont'd)

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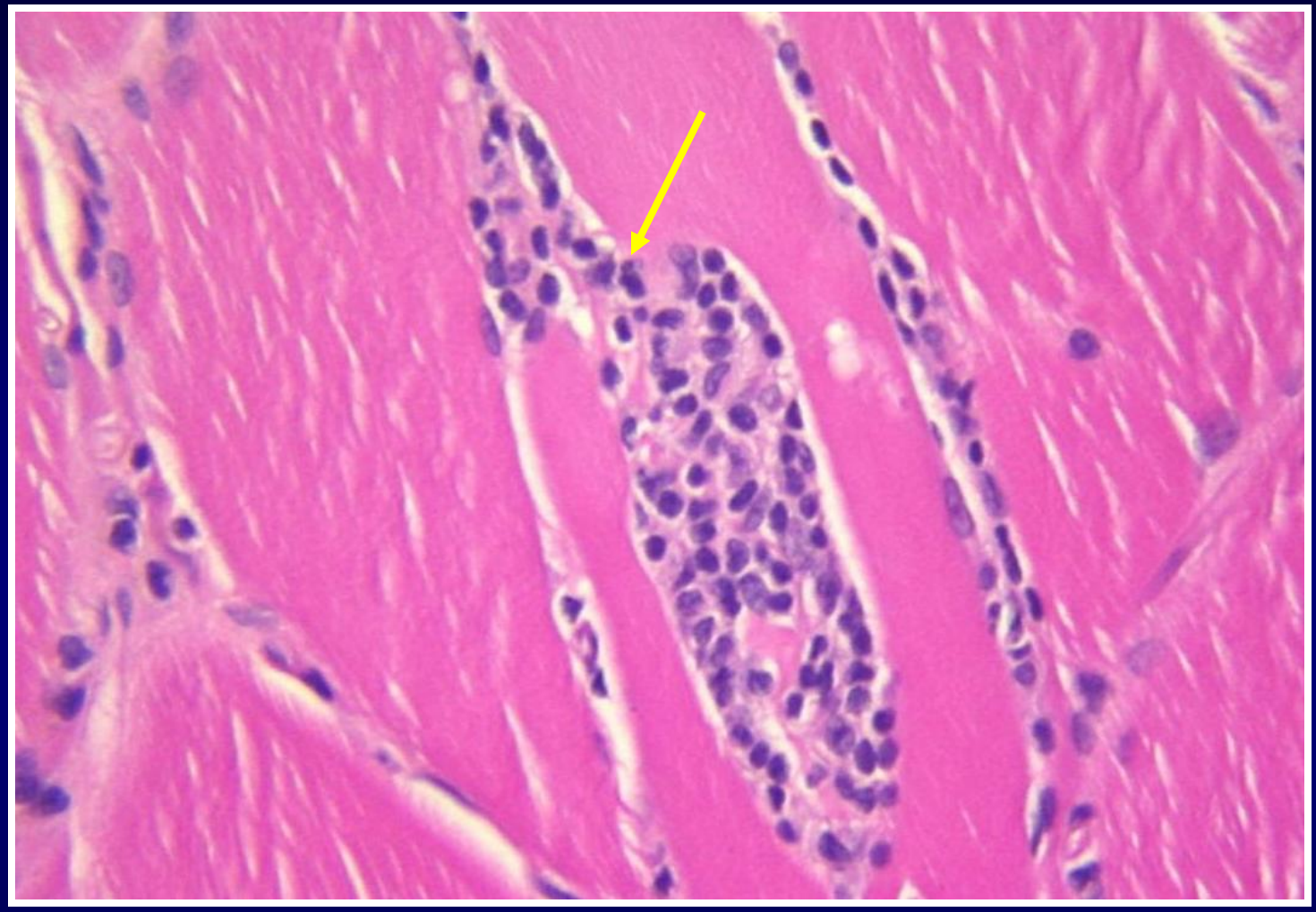
    - e.g. infection
- Multiple diseases and "syndromes"
  - which sometimes run in families
- Formation of autoantibodies
  - markers of autoimmunity
- Inflammatory in nature



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

Gastrointestinal

- Dysphagia
- Reflux
- Dysmotility

Cutaneous

- Rashes
- Calcification

Cardiac

- Arrhythmias
- Congestive failure

Pulmonary

- Atelectasis from muscle weakness
- Aspiration pneumonia
- ILD

General

- Fever
- Fatigue
- Weight loss
- Raynaud's

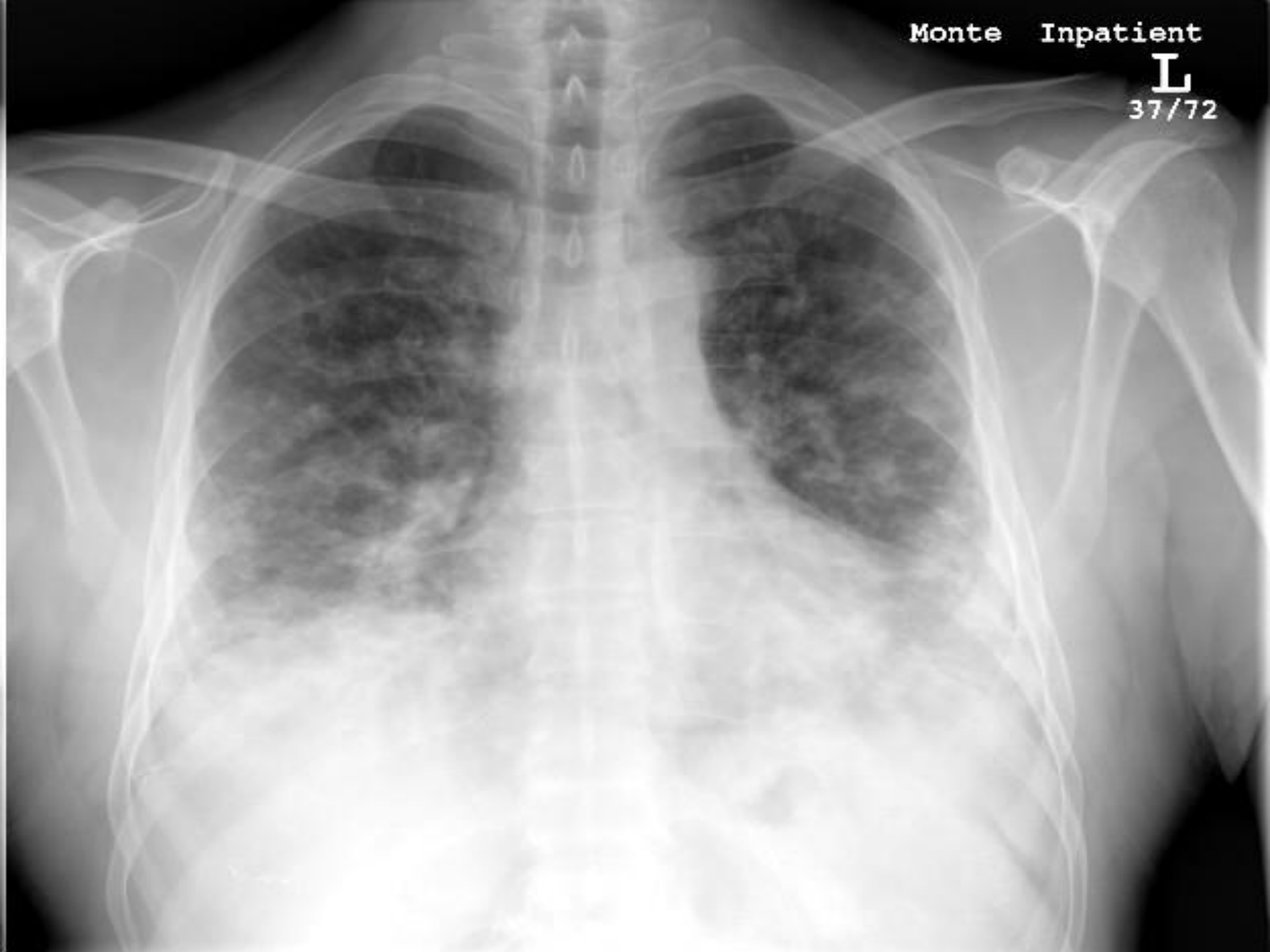
Slide, courtesy of Dr. Fred Miller

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


Monte Inpatient
L
37/72

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

Autoimmunity: Autoantibody Markers

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- Multiple diseases and "syndromes"
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- Formation of autoantibodies
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The patient today had a marker in his blood that is associated with ILD in myositis patients

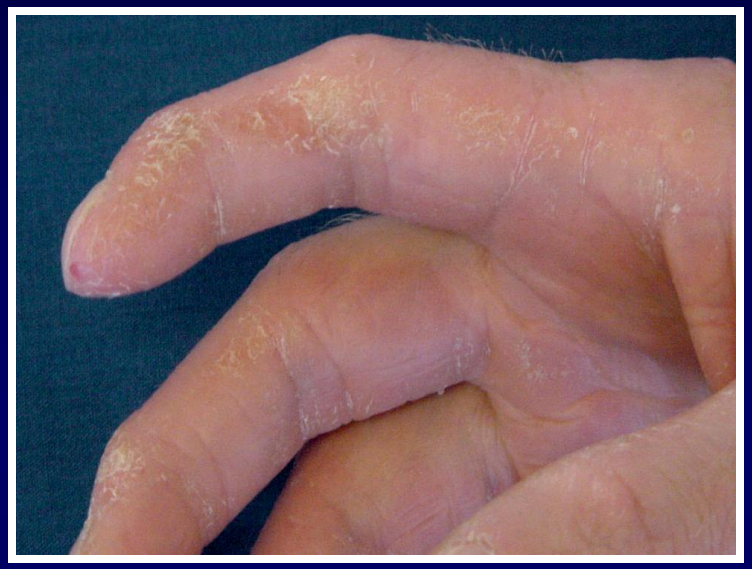
Case Presentation

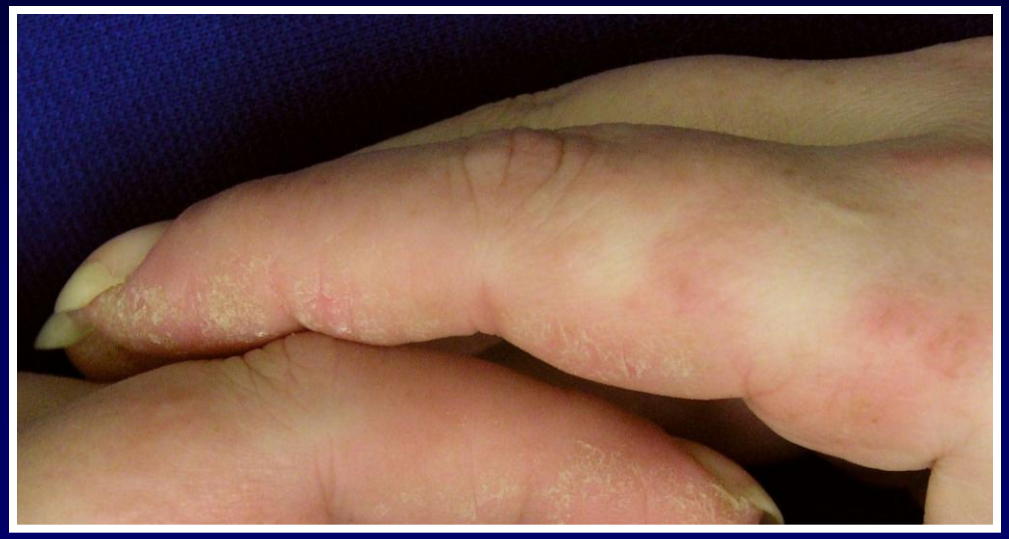
- Lung biopsy: inflammation
- Mild myositis on further testing
- Anti-Jo-1 autoantibody
- "Anti-synthetase syndrome"

"Anti-Synthetase Syndrome"

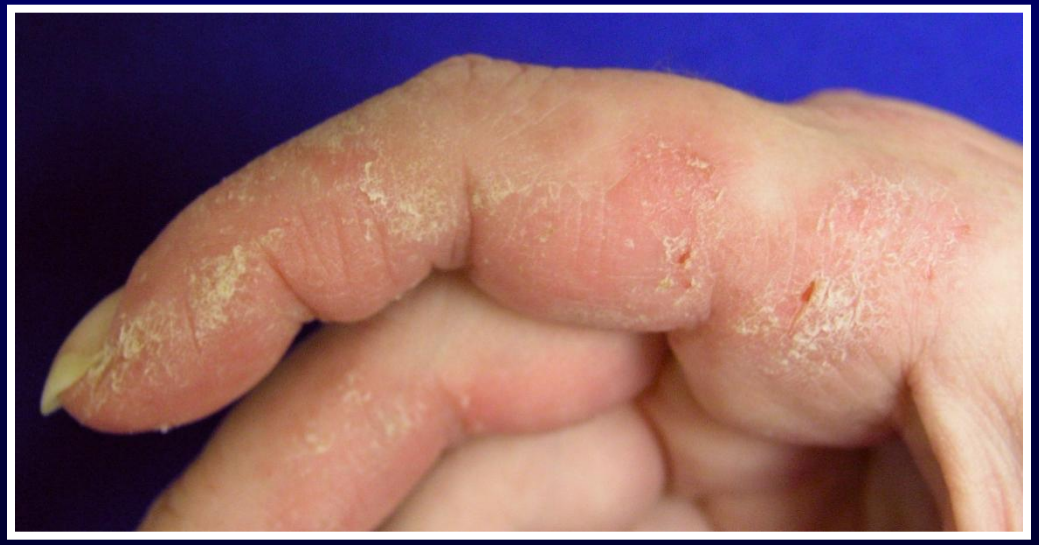
- Acute onset of symptoms
- Lung symptoms may dominate the clinical picture in the form of ILD
- Also may have myositis, fever, arthritis, Raynaud phenomenon, "mechanic's hands"

Mechanic's Hands





13 days later



University of Pittsburgh Autoantibody Cohort

Autoantibody	Number (% synthetases)
Jo-1	140 (60)
PL-12	36 (16)
PL-7	27 (12)
EJ	11 (5)
OJ	6 (3)
KS	9 (4)
Total Synthetases	**229**

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 (medications used to prevent rejection of transplanted organs)
- Maybe even some biologic agents like rituximab