Complications of Inflammatory Myopathy:

Lung Disease

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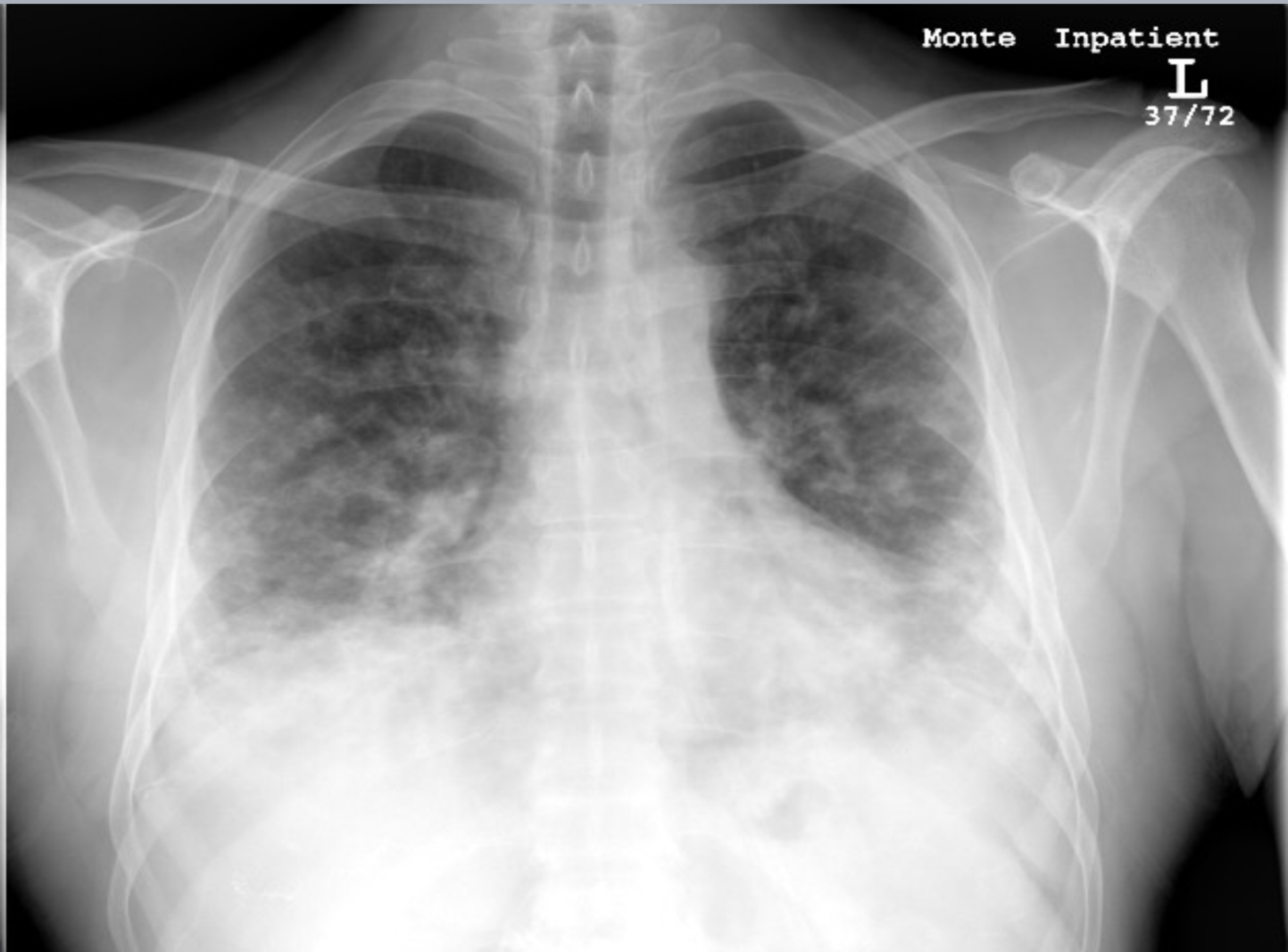
Case

- 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


Monte Inpatient
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- **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 hypertenison (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

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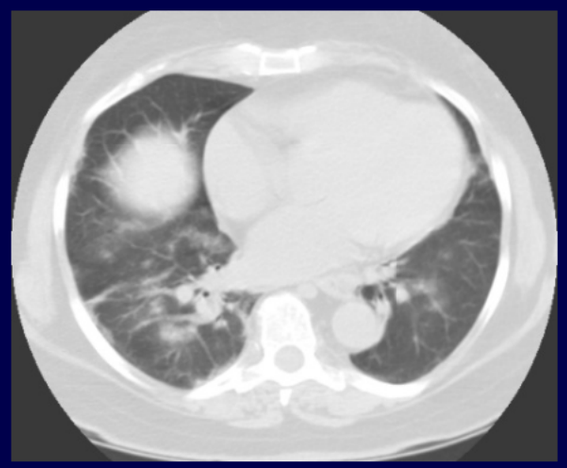
Diagnosis

1. Clinical suspicion

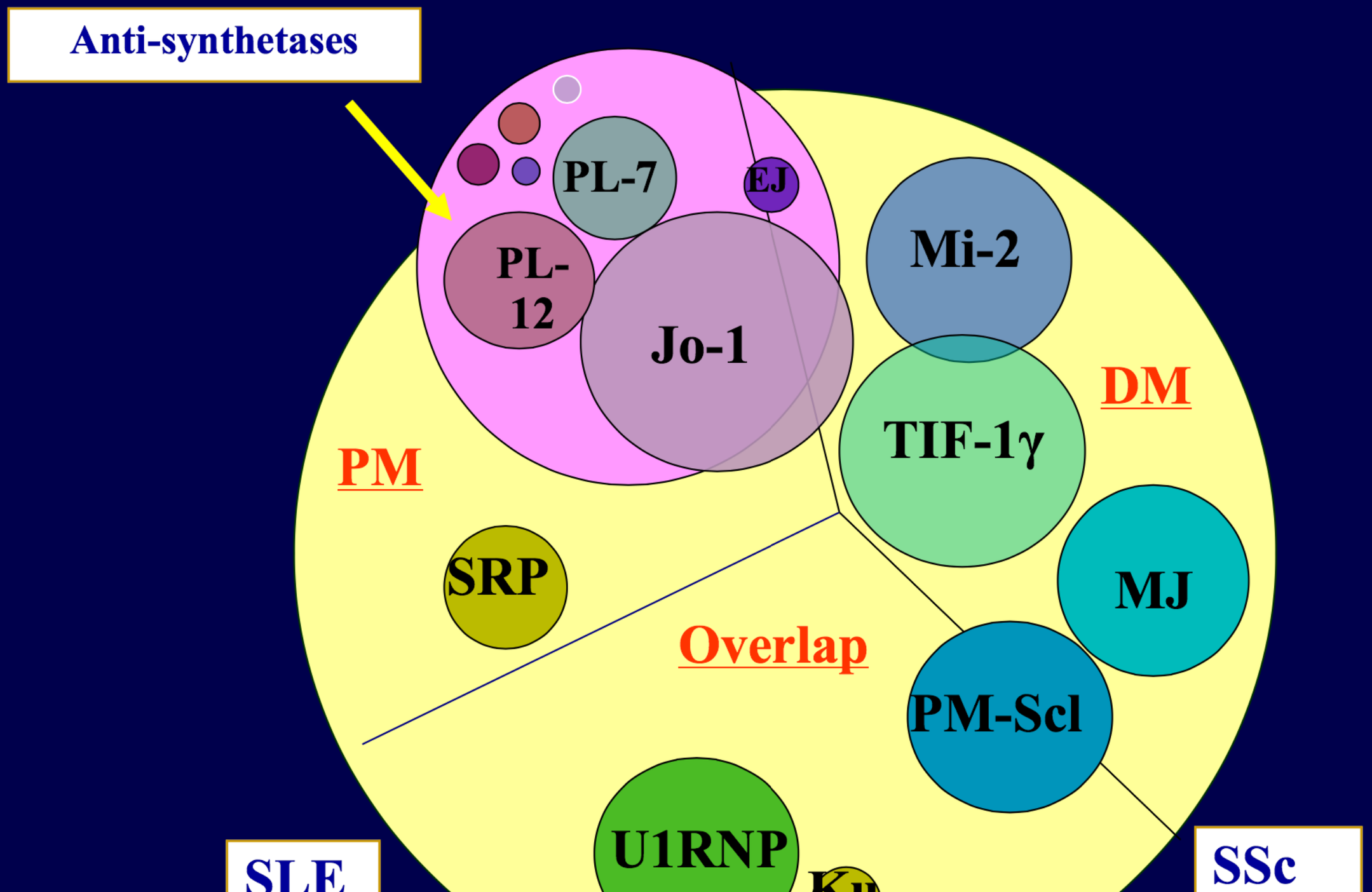


2. Imaging

3. Pulmonary Function Tests (PFTs)

4. Biopsy—rule out alternative processes

Diagnosis: Autoantibodies



Autoantibodies

Antibody	Target	Subset	Phenotype
Mi-2	NuRD	DM	Shawl, V-neck, Gottron's
CADM-140	MDA-5	DM	Amyopathic, ILD
SAE	SUMO	DM	ILD, dysphagia
MJ	NXP-2	JDM	Calcinosis, Ulceration
p155/140	TIF1-g	DM, JDM	Severe skin, malignancy
SRP	72, 54 kDa	PM	Severe/refractory myositis
p200/100	HMGCR	IMNM	Necrotizing myopathy
Jo-1	ARS	PM/DM	Anti-synthetase syndrome

Anti-synthetase Autoantibodies

Antibody	Antigen (tRNA synthetase)	Prevalence in IIM (%)
Jo-1	histidyl	20-30
PL-7	threonyl	<5
PL-12	alanyl	<5
OJ	isoleucyl	<5
EJ	glycyl	<5
KS	asparaginyl	<1
Tyr	tyrosyl	<1
Zo	phenylalanyl	<1

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Myositis--Autoantibodies