Inpatient Rheumatology Updates for the Hospitalist

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Disclosures

I have no relevant financial relationships to disclose.

Objectives

- Recognize rheumatologic emergencies presented in sample cases
- Understand the workup and treatment for sample cases
- Understand the role of rheumatology and other consultant teams in the management of case scenarios

Case 1

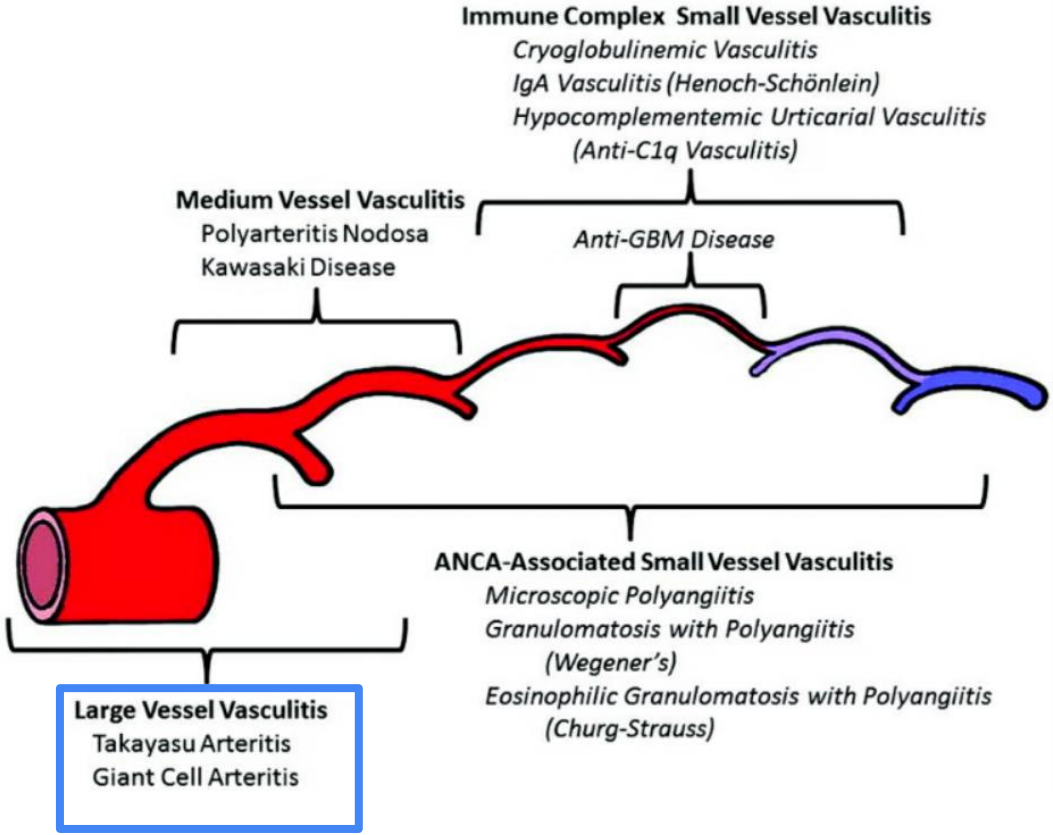
A 73-year-old man presents to the ED for an episode of vision loss in his right eye. He also reports right-sided headache over the last 2 weeks. He tried aleve for his headaches, but this did not help.

On examination, the right temporal artery pulse cannot be palpated.

Laboratory values are as follows: **C-reactive protein 11.5 *mg/dL*** (≤0.8), **Erythrocyte sedimentation rate 85 *mm/hr*** (0–15), **Leukocyte count 12,000 *cells/uL*** (4000–11,000), **Platelet count 550,000 *cells/uL*** (150,000–450,000)

What is the next best step in management?

A) Refer patient for a biopsy of the temporal artery
B) Order a temporal artery ultrasound + pulse dose steroids
C) Refer patient for a biopsy of the temporal artery + pulse dose steroids
D) Order a FDG-PET CT

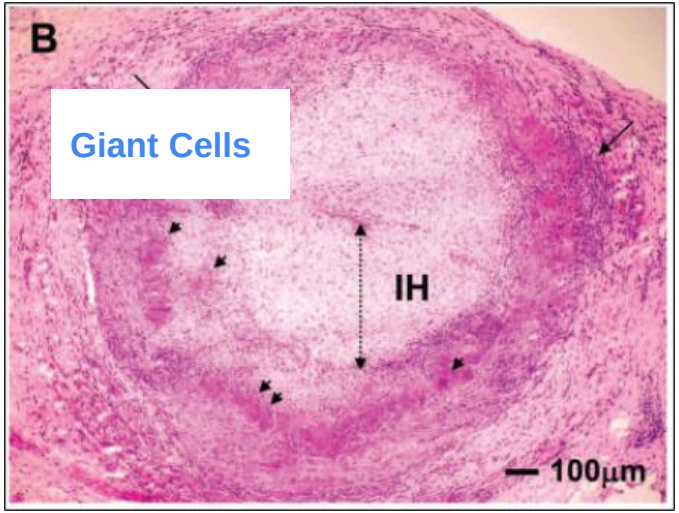


Jennette JC et al. *Arthritis Rheum.* 2013

Terrades-Garcia et al. *Rheumatology.* 2018

Giant Cell Arteritis:

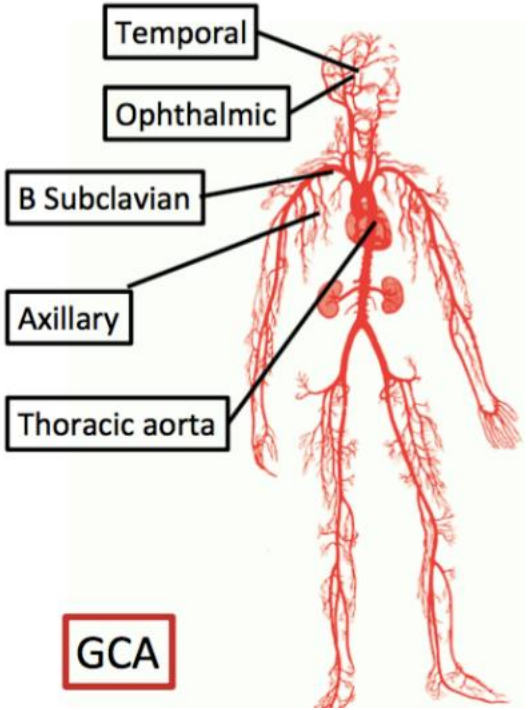
- A vasculitis of large- and medium-sized vessels due to potential involvement of aorta (especially thoracic aorta) and its branching vessels



When to Suspect GCA

Individuals > 50 with:

- Cranial symptoms
  - Headache (~70%) - **new, non-responsive to therapy**
  - Scalp tenderness
  - Jaw claudication (~50%)
  - Vision loss (up to 15%) and double vision (5%)
  - Stroke - especially in occipital lobe
- Extracranial symptoms (aorta and large vessels)
  - Constitutional symptoms, anemia, unexplained very high inflammatory markers
    - Fever of unknown origin in the elderly
  - Limb claudication/blood pressure discrepancies
  - Aortic aneurysms and aortic dissections
- Association with **polymyalgia rheumatica** (present in 40-60% of patients with GCA



Workup of GCA

- Complete Physical Exam:
  - Involve ophthalmology for vision symptoms - transfer if not available!
- Serology:
  - ESR/CRP are sensitive markers; 4% of patients have normal inflammatory markers
- ***Obtaining pathology is the current diagnostic gold standard!***
  - Always pursue a temporal artery biopsy > 1 cm (to limit "skip lesions") for cranial symptoms
    - General surgery, vascular surgery, oculoplastic surgery
  - Ideally within 2 weeks of starting steroids
  - Role for temporal artery ultrasound depending on local expertise
- **Large vessel imaging now is a standard part of workup:**
  - CTA/MRA neck/chest/abdomen/pelvis or FDG-PET
- Patient requires urgent inpatient rheumatology evaluation or outpatient visit scheduled visit on discharge - **soft handoff appreciated! :-)**

Management of GCA - American College of Rheumatology Guidelines

Clinical pearls

Pulse steroid (1,000 mg IV methylprednisolone x 3 days) is the standard of care for ocular involvement or critical cranial ischemia/stroke.

Case 2

A 42-ye on, reflux,
inflamm he ED for
non-ren

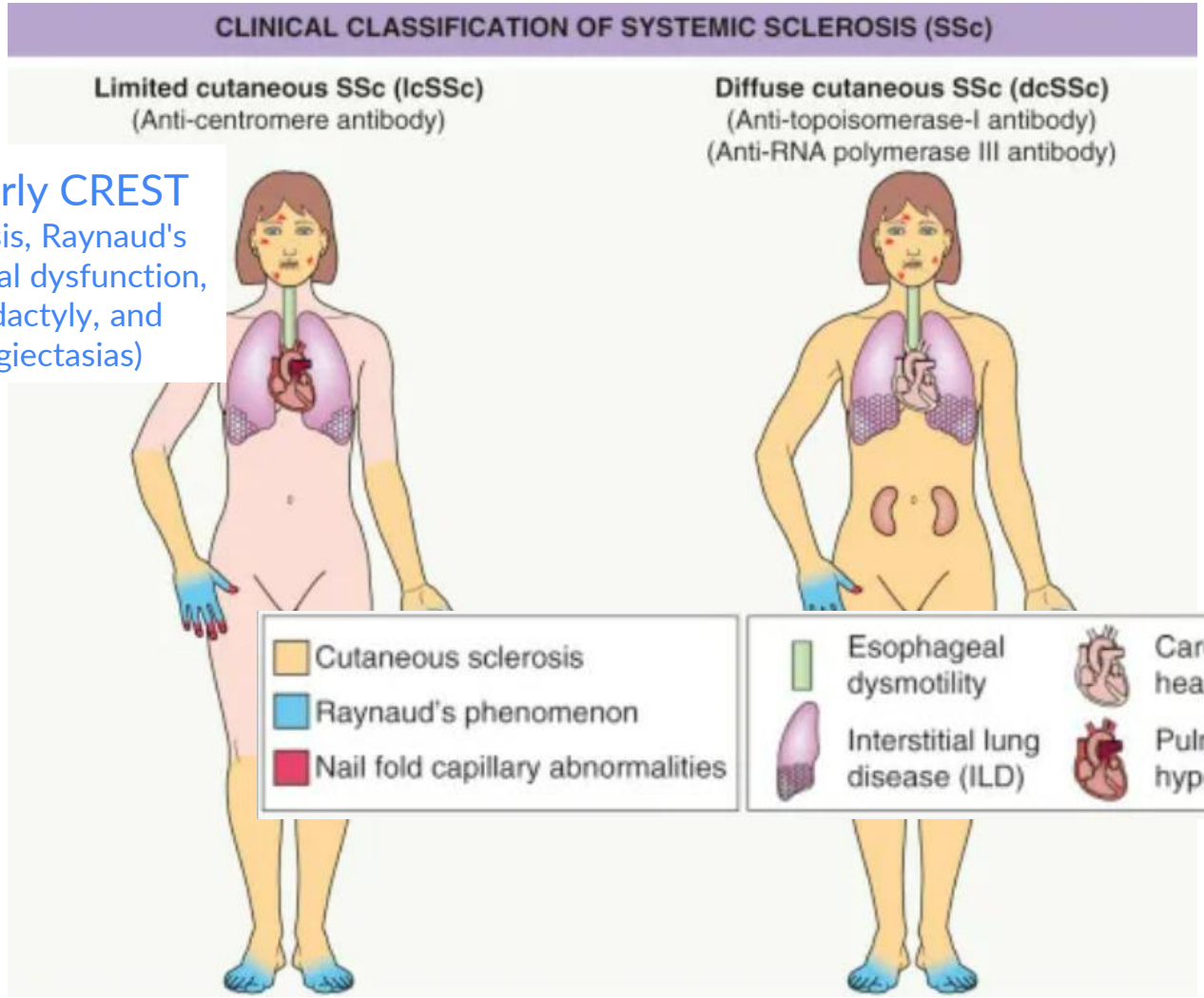
A scleroderma patient presents with critical digital ischemia!

What is

A) Start IV prostaglandins
B) Consult vascular surgery
C) Pain control
D) Request angiography of the left upper extremity

What is Systemic Scleroderma?

- “Systemic scleroderma” aka “scleroderma” aka “systemic sclerosis”
  - Chronic autoimmune condition characterized by widespread vascular dysfunction and progressive fibrosis of the skin and internal organs
  - Two main forms:
    - **LIMITED cutaneous systemic sclerosis (former CREST)**
    - **DIFFUSE cutaneous systemic sclerosis**



Formerly CREST
(calcinosis, Raynaud's esophageal dysfunction, sclerodactyly, and telangiectasias)

Karynne O Duncan, MD, https://plasticsurgerykey.com/systemic-sclerosis-scleroderma. 2019

Physical Exam Findings in Systemic Scleroderma

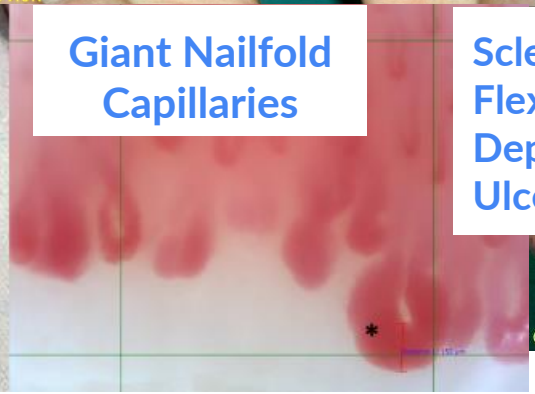
Puffy Fingers

Raynaud's

Calcinosis

Periungual Erythema

Giant Nailfold Capillaries



Sclerodactyly
Flexion Contractures
Depigmentation
Ulcers & pitting Scars

ACR Image Bank

Raynaud Phenomenon

- Expected in nearly all cases of scleroderma, a **precursor to critical digital ischemia**
- Reversible vasospasm leading to episodic color changes in response to cold or stress (white > blue > red upon re-warming)
- Associated paresthesia or numbness and pain
- Two forms:
  - Primary (idiopathic)
  - Secondary
    - Connective tissue disease, vascular disease, hyperviscosity, substances, vibration, endocrinopathies

Critical Digital Ischemia Is an Emergency

Courtesy of ACR VIRL

Schiopu et al., International J. of Rheumatology. 2010

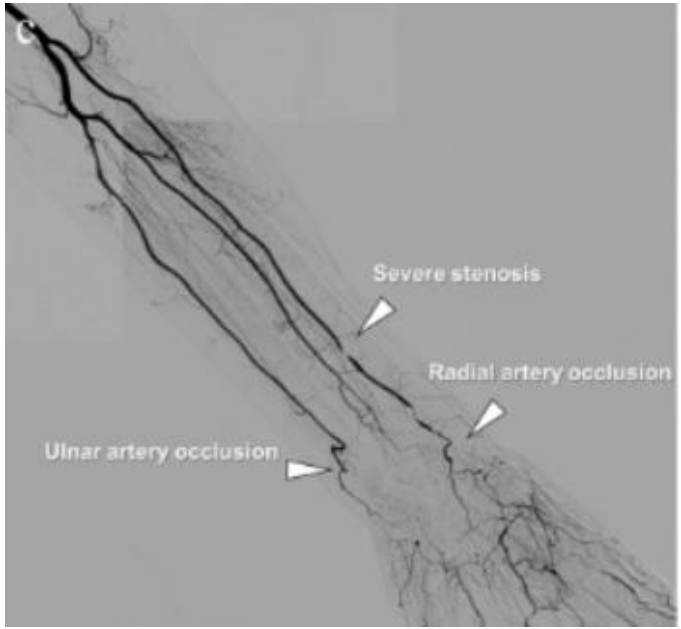
- **Time is tissue!**
- Critical ischemia is marked by:
  - Prolonged, refractory pain suggests impending tissue infarction
  - Color change due to infarction
  - Infarction progresses to gangrene

Management Approach to Critical Digital Ischemia in Scleroderma Patients: First Steps in All Patients

- 1. Stop working, rest, and admit to the hospital
- **2. Control pain - answer to case scenario!**
  - Analgesic medication, frequently need opioids
  - Digital block if needed (lidocaine)
- 3. Limit exacerbating factors (cold environment, stress, medications - amphetamines, certain B-blockers, chemotherapies, clonidine, cocaine)
- 4. Plan for vasodilative therapy (more details to come… consider ICU admission and obtaining a baseline echocardiogram)

Critical Digital Ischemia in Scleroderma Patients: Vascular Evaluation

- Doppler ultrasound
- Arteriogram (traditional angiogram) is gold standard
- MRA/CTA (limited use in distal circulation, but often requested before angiogram)



Shoji K et al. *Vasc Endovascular Surg.* 2022

Management Approach to Critical Digital Ischemia in Scleroderma Patients: Treatment

Clinical pearls

Pharmacologic treatment is required for critical digital ischemia in scleroderma patients. Surgical intervention typically is not indicated.

Scleroderma patients with critical digital ischemia should be urgently treated at a center with rheumatology that is able to provide IV prostaglandins.

Case 3

A 47-year-old woman with no significant past medical history presented to the ED with progressive dyspnea and a dry cough over the past 10 weeks. She has had no environmental exposures. On exam, she is afebrile, tachycardic, tachypneic and hypoxic to 70s on 3 L nasal cannula. She has bilateral inspiratory crackles, power was 5/5 strength in all extremities. Skin findings are shown.

CT chest showed extensive pneumomediastinum and subcutaneous emphysema and consolidations bilaterally.

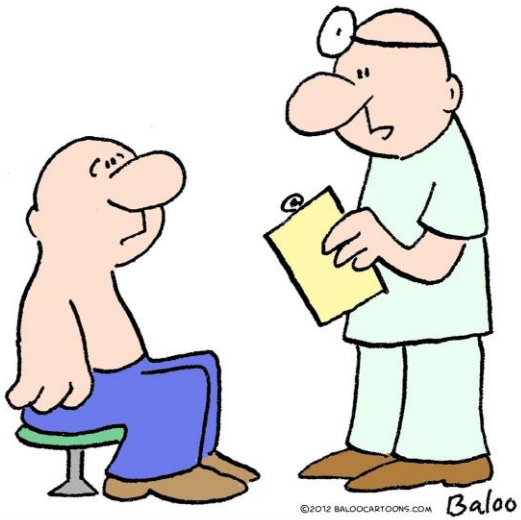
Which of the following is the most likely diagnosis?

A. Lupus-related interstitial lung disease
B. Anti-MDA-5 rapidly progressive lung disease
C. Hypersensitivity pneumonitis
D. Antisynthetase syndrome

Hands 6 weeks ago

Kurtzman et al. *Journal of American Academy of Dermatology*. 2018

What does this patient likely have?



"I'll give it to you straight — This disease is almost *impossible* to pronounce."

Anti-MDA5 (melanoma differentiation-associated protein-5) positive dermatomyositis

This entity is associated with a rapidly progressive interstitial lung disease with high mortality rates

Immune-Mediated Myopathies (IMMs) Introduced

- AKA idiopathic inflammatory myopathies
- Systemic autoimmune diseases characterized by chronic **muscle weakness**
- **Traditionally discussed as two main entities:**
  - **Polymyositis**
  - **Dermatomyositis**

Other features include: **dysphagia, rash, arthritis, Raynaud's, interstitial lung disease and less commonly myocarditis**

The antibody association informs clinical phenotype

Connolly CM. *Lancet Rheumatol.* 2024

Current Understanding of IMMs

Clinical pearl

Associated antibodies fit specific clinical phenotypes that predict outcomes.

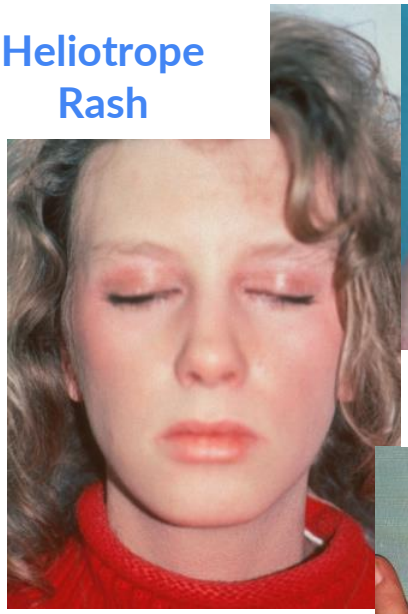
Anti-MDA5 positive dermatomyositis has predominant skin and lung manifestations

Int. Med. 2016

Overview of Diagnostic Approach to IMMs

- Clinical history and physical exam
  - Typically weeks to months of bilateral proximal muscle weakness (except inclusion body myositis) +/- dysphagia
- Serology
  - Including muscle enzymes and autoantibodies (more in upcoming slides)
- Muscle Imaging (MRI) or Electromyography (EMG)
- **High -resolution CT chest imaging - with any suspicion of lung disease**
- Skin biopsy or muscle biopsy rarely needed. Diagnostic detail not as specific as antibody profile!

Classic Dermatomyositis Rashes

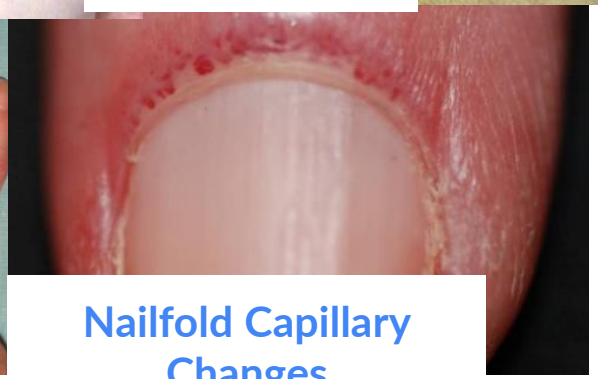


Heliotrope Rash

V-Sign

Holster Sign

Gottron Papules



Nailfold Capillary Changes

ACR Image Bank

Additional Specific Cutaneous Findings in Anti-MDA-5 Dermatomyositis

Clinical pearl

Anti-MDA5 dermatomyositis may be amyopathic! So, we must rely on other clues such as skin findings …

Serologic Workup for Inflammatory Myositis

- CBC/CMP
- Muscle enzymes: creatinine kinase, aldolase, lactate dehydrogenase
- Inflammatory markers: ESR, CRP
- Urinalysis
- Myositis panel (Oklahoma Medical Research Foundation - OMRF preferred)
- Anti-HMG-CoA Reductase (HMGCR) (separate order to evaluate for statin-induced myositis)
- Antinuclear Antibody testing (ANA) (to evaluate for overlap conditions)

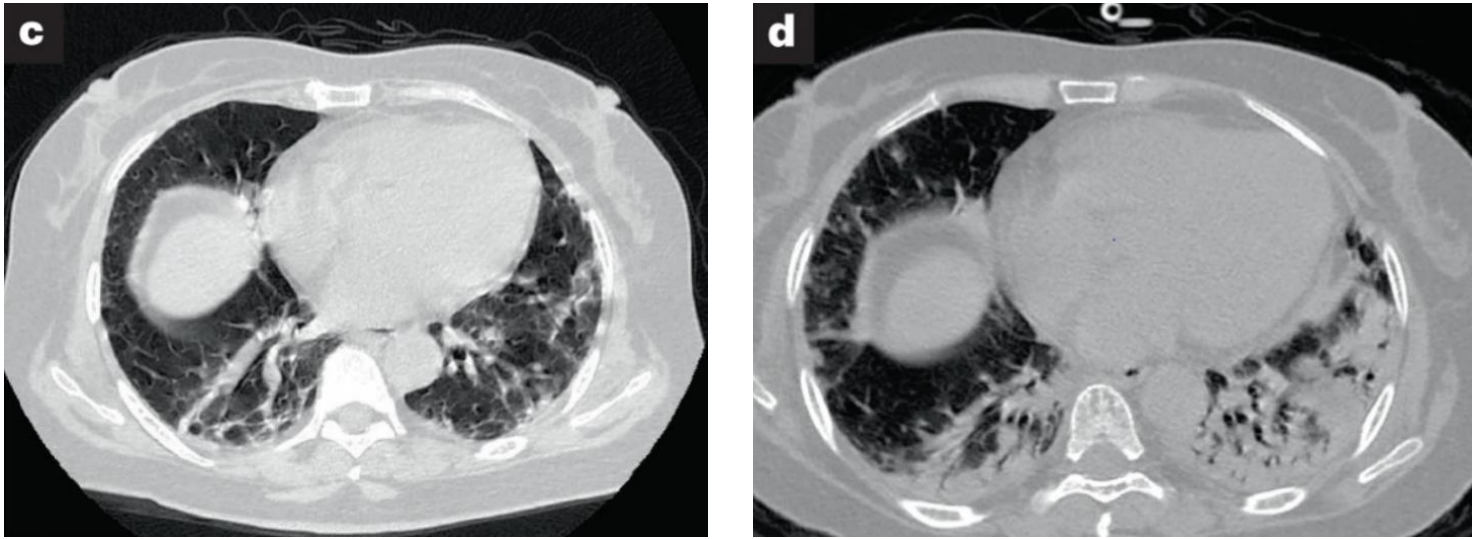
Many Causes of HyperCKemia

Clinical pearl

Consider myositis in the correct clinical context of subacute/chronic progressive muscle weakness.

- Viral illness

Rapidly-Progressive Lung Disease in Anti-MDA5 Positive Dermatomyositis



Typical radiological dynamic course: from subpleural opacities at the initial visit to wide consolidation after ten days

Lu X et al. *Nat Rev. Rheumatology.* 2024

Treatment of Rapidly-Progressive ILD

Clinical pearl

Rapidly-progressive interstitial lung disease seen in patient with Anti-MDA5 positive dermatomyositis require very aggressive immunotherapy and multidisciplinary care including rheumatology, critical care and pulmonology!

Mortality rates are high

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Thank You!