

Polymyalgia Rheumatica and Myositis: Oh My Aching Muscles!

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Dr. Rowland is a graduate of Rush Medical College in Chicago, Illinois, and completed residency at Advocate Illinois Masonic Medical Center and a fellowship at the University of Chicago. She is an associate medical editor for the AAFP's *FP Essentials* and serves on the editorial board for *Journal of Family Practice*. She enjoys teaching about topics that require research, synthesis, and empathy to understand. In addition, she enjoys teaching about evidence-based medicine topics. Dr. Rowland strives to make her lectures relevant to practice, thought provoking, and informative.

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Learning Objectives

1. Recognize early symptoms of PMR.
2. Differentiate PMR from other entities mimicking PMR.
3. Order appropriate test to differentiate PMR from other conditions.
4. Employ appropriate treatment strategies for PMR.

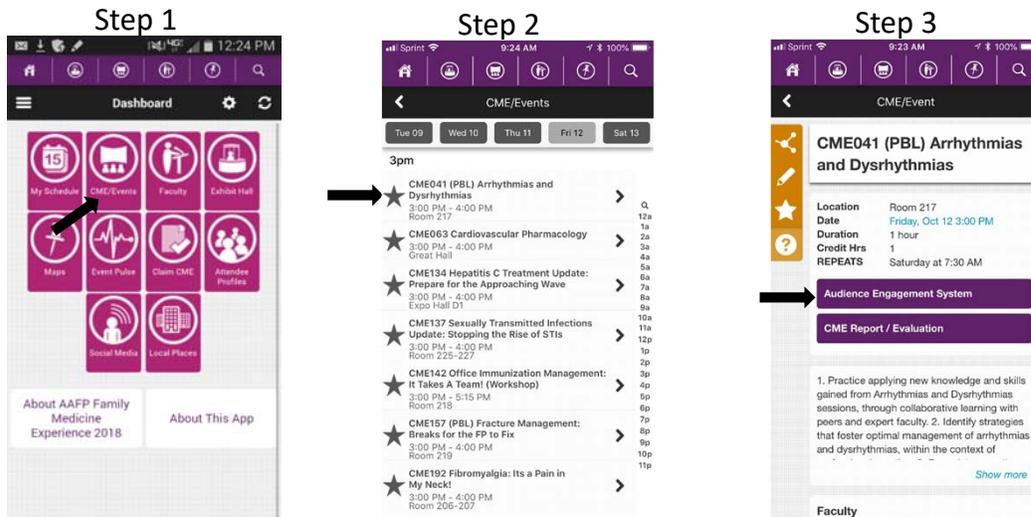
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Associated Session

- (PBL) Polymyalgia Rheumatica and Myositis: Oh My Aching Muscles!

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Audience Engagement System



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A textbook case

- A 72 year old woman with a history of hypertension presents with a 3-week history of worsening bilateral shoulder pain and bilateral hip pain, which she describes as constant and worse with activity. She notes her pain is present on awakening but that the worst symptom then is joint stiffness, which lasts at least an hour. She reports minimal relief with acetaminophen. ROS is positive for fatigue and malaise, although she continues to work three days a week (her usual schedule).

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AES Question

This patient has...

- A. Polymyalgia rheumatica
- B. Polymyositis
- C. Giant cell arteritis
- D. Statin-induced myopathy
- E. Dermatomyositis
- F. Fibromyalgia
- G. I don't know

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Polymyalgia rheumatica

- Chronic inflammatory condition in older adults
- Affects
 - Bursa
 - Synovia
 - Muscles
- Unclear etiology
 - Believed to be antigen-mediated
 - Autoimmune origin

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AES Question

In which decade of life does the prevalence of polymyalgia rheumatica peak?

- A. 30-40
- B. 40-50
- C. 50-60
- D. 60-70
- E. 70-80
- F. 80-90

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AES Question

Who is more likely to get PMR?

- A. Men
- B. Women

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Polymyalgia rheumatica

- Peaks between 70-80 years
- Uncommon (but not impossible!) below age 50, quite uncommon below age 40
- F:M=3:1
- More common in white people

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AES Question

What body part is most often affected in PMR?

- A. Neck
- B. Shoulders
- C. Hips
- D. Knees
- E. Ankles

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Polymyalgia rheumatica: symptoms

- Proximal large muscle groups and joints
 - Shoulders
 - Hips
 - Neck
- Usually bilateral
- Morning stiffness
- Symptoms worsen with movement
- Not improved with rest
- Fatigue or weight loss present
 - 30-50% of patients have these or other systemic symptoms
- Onset of symptoms is insidious
 - Subacute->chronic timeframe

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Polymyalgia rheumatica: physical exam

- Tenderness to palpation over affected muscles and joints
- Strength intact
- Synovitis
- Joint edema

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Differential diagnosis of PMR

- Dermatomyositis
- Drug-induced myalgia
- Rheumatoid arthritis
- Osteoarthritis
- Rhabdomyolysis
- Polymyositis
- Lupus
- Shoulder-specific pathology:
 - Adhesive capsulitis
 - Rotator cuff disease
 - Subacromial bursitis
 - Spondyloarthritis
- SEID
- Fibromyalgia

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Workup of patients with presumed PMR

- Workup targets accurate diagnosis and assessing for other things on the differential
- Also assesses for risk of long-term steroid use
- CBC
- CMP
- ESR
- TSH
- CK
- UA
- Rheumatoid factor \pm ANA
- SPEP/UPEP
- DEXA

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Polymyalgia rheumatica: diagnostic criteria from the British

- Age > 50
- Sx > 2 weeks
- Bilateral shoulder pain or pelvic girdle pain or both
- Morning stiffness \geq 45 minutes
- Acute-phase response (e.g.; elevated ESR)
- Less likely with:
 - Other inflammatory diseases
 - Drug-induced myalgias
 - Chronic pain conditions
 - Endocrine diseases
 - Neurologic conditions

Ref: Dasgupta B, Borg FA, Hassan N, et al.; BSR and BHPR Standards, Guidelines and Audit Working Group. BSR and BHPR guidelines for the management of polymyalgia rheumatica. *Rheumatology (Oxford)*. 2010;49(1):186–190.

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Treatment of PMR

- Steroids!
- Prednisone
 - May initially need a burst
 - Pt should improve by 70%+ within first 7 days of treatment
- Start at 15 mg daily
- Taper by 2.5mg every 3 weeks to 10 mg
- Then taper by 1 mg a month
- Slow taper prevents rebound symptoms
- Monitor symptoms at:
 - 1-3 weeks
 - 6 weeks
 - 3 months
 - 6 months
 - 9 months
 - 12 months
 - and PRN

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AES Question

Which of the following is true about giant cell arteritis?

- A. GCA is more common than PMR
- B. GCA affects younger patients than PMR
- C. Systemic symptoms are uncommon in GCA
- D. Headache is the most common symptom in GCA
- E. Blindness occurs in half of patients with GCA

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A textbook case

- A 72 year old woman with a history of hypertension and polymyalgia rheumatica presents with a 2 week history of headache. The headache is bitemporal, worse when she brushes her hair or wears her noise-canceling headphones. She has noted a low-grade fever (100.5F) for 5 days. She has no visual changes. She reports weight loss of about 4 pounds, although she also says that she has been on soft foods for about a week because her jaw hurts.

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AES Question

This patient has...

- A. Tension-type headache
- B. Polymyositis
- C. Giant cell arteritis
- D. TMJ synovitis
- E. Dermatomyositis
- F. Fibromyalgia
- G. I don't know

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Giant cell arteritis

- Inflammatory disorder
- Affects medium and large arteries
- Peaks between 70-80 years
- Virtually unheard of below age 50
- F:M=3:1
- More common in white people

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GCA and PMR

- PMR is a risk factor for GCA
 - As soon as the diagnosis of PMR is made, look for symptoms of GCA!
 - Approximately 10-20% of patients with PMR have or will have GCA
- 40-50% of people with GCA will have PMR

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Giant cell arteritis: symptoms

- New onset headache or new headache type
- Subacute course: symptoms arise and persist over weeks
- Jaw or tongue claudication
- Facial pain
- Systemic symptoms:
 - Fever
 - Weight loss
 - Fatigue
- Amaurosis fugax
- Diplopia
- Other visual changes

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Giant cell arteritis: atypical symptoms

- Cough
- FUO
- Extremity claudication
- Neuropathy
- TIA/CVA

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Giant cell arteritis: physical exam

- Head or scalp tenderness
 - Traditionally over the temples/temporal artery
- Loss of visual acuity
- Afferent pupillary defect
- Reduced ocular motility
- PE may be normal in patients with classic symptoms!

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Differential diagnosis of GCA

- TIA/CVA
- Vasculitis
- Malignancy
 - Primary
 - Secondary
- Ophthalmologic problems
 - Glaucoma
 - Optic neuritis
 - Retinal detachment
 - Central retinal artery/vein occlusion
- Headache syndromes
 - Migraine
 - Tension-type headache
 - ICH

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AES Question

What's the average ESR of a patient with GCA?

- A. 54
- B. 66
- C. 73
- D. 88
- E. 102

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AES Question

What percentage of people with biopsy-proven GCA had an ESR < 50?

- A. 0
- B. 3
- C. 5
- D. 7
- E. 9
- F. 11

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Workup of patients with GCA

- CBC
- BMP
- ESR
- CXR
- UA
- ANA
- Consider CT or MRI
- Ophtho eval
- If available, color Doppler ultrasound
 - May show arterial lesions
- Temporal artery biopsy

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ACR diagnostic criteria for GCA

Need 3 to make the diagnosis:

- Age \geq 50
- New headache or new type
- Temporal artery problem
 - Decreased pulsation
 - Tenderness to palpation
- ESR \geq 50
- + Temporal artery biopsy

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Treatment of GCA

- Steroids!
- Prednisone 40-60 mg *as soon as diagnosis is suspected*
 - Consider IV steroids if transient or evolving vision loss
 - Use 60 mg if jaw claudication or established visual loss
- Taper over months
- Offer aspirin if no contraindications to reduce risk of CVA

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Treatment of GCA and PMR: steroid precautions

- Consider a DEXA
- Offer bone protection
 - Calcium (1200 mg/day)
 - Vitamin D (800IU/day)
 - Bisphosphonates as needed
 - Weight bearing exercise
- Stomach protection
- Monitor blood pressure
- Monitor blood sugar

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A textbook case

- A 72 year old woman with a history of hypertension and hyperlipidemia presents with a 2 week history diffuse muscle pain. She says all her muscles ache, and she reports it is somewhat worse with movement. She comes in today because she notes she has become weak and has difficulty lifting her arms above her head. She has no relief with acetaminophen or ibuprofen. She continues to take her chlorthalidone and atorvastatin daily.

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AES Question

This patient has...

- A. Polymyalgia rheumatica
- B. Polymyositis
- C. Giant cell arteritis
- D. Statin-induced myopathy
- E. Dermatomyositis
- F. Fibromyalgia
- G. I don't know

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AES Question

Which statin is most likely to cause myalgias?

- A. Rosuvastatin
- B. Pravastatin
- C. Lovastatin
- D. Simvastatin

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Statins and muscles

- Myalgias: Muscle discomfort
- Myopathy: Muscle discomfort and weakness ± elevated CK level
- Myositis: Muscle inflammation, usually with pain
- Rhabdomyolysis: Muscle breakdown with renal failure or myoglobinuria

National Lipid Association Statin Muscle Safety Task Force

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Statins and muscles

- ~10% of patients report muscle pain with statins
 - 3-4% on placebo
- 0.5% have myopathy
- Rhabdomyolysis rates vary, but <1/10,000 patient/years

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Statins and muscles: risk factors

- Therapy choices
 - Combination with fibrates or niacin increases risk
 - Gemfibrozil>fenofibrate
 - More lipophilic statins (simva, lova) may be more likely to cause myopathy
 - Statins not metabolized by CYP3A4 (prava, rosuva) have reduced risk of drug-drug interactions, less likely to cause myopathy
- Patient factors
 - Comorbidities
 - Known or indolent muscular disease
 - Ethnic origin
 - Chinese>>European
 - Age
 - Thyroid disorders
 - Vitamin D deficiency?

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Statins and muscles: treatment

- For patients with CK levels WNL or <10x nl:
 - Once CK normalized, trial of
 - Pravastatin
 - Fluvastatin
 - Pitavastatin
 - Reduce dose or intensity
 - Evidence for CoQ10 mixed
- Patients with CK levels >10x nl:
 - Use caution when restarting
- Patients with statin-induced rhabdo:
 - Contraindication

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A textbook case

- 37 year old woman presents with 2 months of increasing pain and weakness in her shoulders, hips, and neck. She comes in today because she has difficulty getting out of a chair. Symptoms are present throughout the day and she doesn't notice any particular morning stiffness, although in the last year, her hands have turned white and painful when she gets cold. She also notes weight loss of about 4 pounds, although she says she has been on soft foods for a few weeks because she has trouble swallowing.

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AES question

This patient has...

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- B. Polymyositis
- C. Giant cell arteritis
- D. Statin-induced myopathy
- E. Dermatomyositis
- F. Fibromyalgia
- G. I don't know

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Polymyositis

- Inflammatory disorder of muscles
- Affects people <50
- African Americans affected more than other races/ethnicities
- Women>men
- Rare: 4-8 cases/100,000 people

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Polymyositis: symptoms and signs

- Muscle weakness
 - Typically symmetric
 - Large proximal groups
 - Neck often involved
 - Subacute course
 - Progressive
- ± Pain; when present, not prominent
- No sensory or extraocular findings
- May involve esophagus, pharyngeal muscles or diaphragm
- Cardiac or pulmonary involvement possible
- May be associated with Raynaud's and other CTD
- Normal DTRs on exam

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Polymyositis

- Diagnosed through
 - Symmetric proximal muscle weakness
 - Elevated CK
 - Abnormal EMG
 - Abnormal muscle biopsy
- If all present, diagnosis made, if 3 of 4, diagnosis likely

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Polymyositis

- Treatment
 - Steroids
 - Rituximab
 - Other immunomodulators

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Polymyositis: malignancy risk

- Associated with increased risk of malignancy
 - At the time of diagnosis and in the future
 - Estimates range from 3-40%
 - Most common types in polymyositis:
 - Adenocarcinoma
 - Lymphoma
 - Leukemia

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Polymyositis: malignancy risk

- Factors associated with malignancy:
 - Age >45
 - Male sex
 - Dysphagia
 - Cutaneous symptoms
 - Rapid onset of symptoms
 - Elevated ESR/CRP
- Reduced risk of malignancy:
 - Raynaud's
 - Anti-Jo antibody+
 - Arthritis

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A textbook case

- A 57 year old woman comes in with a complaint of rash on both hands and difficulty drying her hair for several months. She presents because both symptoms are worsening. The rash is reddish, on the backs of both hands, and mildly itchy. She has weakness and minimal tenderness in both shoulders, both hip flexors, and her neck, but denies any morning stiffness. A thorough review of systems is otherwise normal.

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AES Question

This patient has...

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- B. Polymyositis
- C. Giant cell arteritis
- D. Statin-induced myopathy
- E. Dermatomyositis
- F. Fibromyalgia
- G. I don't know

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Dermatomyositis

- Autoimmune muscle condition with characteristic skin findings
- Peaks in 50's
 - Can occur in children; average age of onset 5-15 years
- African Americans affected more than other races/ethnicities
- Women > men

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AES Question

Which of the following is pathognomonic for dermatomyositis?

- A. Gottron's papules
- B. Heliotrope rash
- C. Shawl sign
- D. Hairbrush sign

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Dermatomyositis symptoms

- *Dermato:*
 - Skin symptoms can be indicative
 - Heliotrope rash
 - Shawl sign
 - or pathognomic
 - Gottron's papules
- *Myositis:*
 - Proximal symmetric muscle weakness
 - Similar to polymyositis

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Dermatomyositis symptoms



Image credit:
<https://commons.wikimedia.org/wiki/File:Dermatomyositis.jpg>



Image credit: <https://pl.wikipedia.org/wiki/Plik:Dermatomyositis18.jpg>

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Dermatomyositis symptoms



Source: IMACS

Image credit: <https://commons.wikimedia.org/wiki/File:Dermatomyositis14.jpg>



Image credit: <https://www.maxpixel.net/Red-Heliotrope-Valerian-Flower-Perennial-Wild-413021>. CC0

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Dermatomyositis diagnosis

- Diagnosed through
 - Symmetric proximal muscle weakness
 - Elevated CK
 - Abnormal EMG
 - Abnormal muscle biopsy
 - Characteristic skin findings (Gottron's sign or heliotrope rash)
- DM diagnosed with rash + 3 of first 4; probably with rash + 2 of 4; possible with rash + 1 of 4

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Dermatomyositis treatment

- Steroids
- Rituximab
- Other immunomodulators

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Dermatomyositis and malignancy

- Approximately 15% of patients with DM have malignancy at the time of diagnosis
 - Pancreatic
 - Ovarian
 - Lung
 - Stomach
 - Colorectal
- Highest risk
 - Older age
 - Men
 - Shawl sign
 - Recurrent disease

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Recommended practice changes

- Diagnose PMR in a patient:
 - older than 50 years
 - with at least 2 weeks of bilateral shoulder or pelvic joint pain, elevated ESR, and morning stiffness.
 - (SOR C)
- Treat patients with polymyalgia rheumatica with low-dose oral corticosteroids.
 - (SOR B)
- Assess all patients with PMR for giant cell arteritis
 - (SOR C)

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Questions



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