Polymyalgia Rheumatica : A Diagnostic Dilemma

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Polymyalgia rheumatica(PMR)

- A clinical syndrome of proximal muscle pain in older patients .
- Often presents a diagnostic challenge because of

➢ the large differential diagnosis

➢ lack of definitive diagnostic criteria

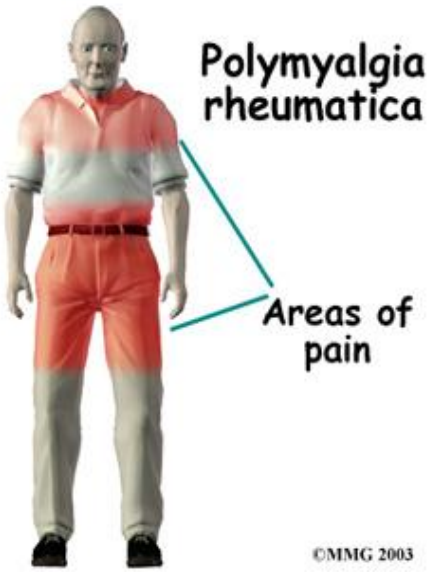
➢ relatively frequent "atypical" clinical findings, such as

peripheral synovitis

distal extremity pain

normal ESR and

mild weakness.



Epidemiology

- Almost exclusively a disease of adults over the age of 50 years.
- Prevalence increases progressively with advancing age.
- Peak incidence : 70 and 80 years of age .
- Female : Male = 3 : 1.
- Positive family history.
- Incidence : Highest in Scandinavian countries and in people of northern European descent . Lowest in Asian, African-American, and Latino populations.

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- PMR is two to three times more common than giant cell (temporal) arteritis (GCA).

- PMR can precede, accompany, or follow GCA.

(5 to 30 percent with PMR experience GCA whereas 50 percent of patients with GCA may have PMR).

Etiology

- Exact cause (or causes) of PMR : unknown.
- An autoimmune process : may play a role
- Associated with the HLA-DR4 haplotype.
- A high level of IL-6 is associated with increased disease activity.

Diagnostic criteria

Several diagnostic criteria for PMR exist. One set of diagnostic criteria is as follows ·····

- ✓ Age of onset 50 years or older
- ✓ Erythrocyte sedimentation rate ≥40 mm/h
- ✓ Pain persisting for ≥1 month and involving 2 of the following areas: neck, shoulders and pelvic girdle
- ✓ Absence of other diseases capable of causing the same musculoskeletal symptoms
- ✓ Morning stiffness lasting ≥1 hour
- ✓ Rapid response to prednisone (≤20 mg)

European League Against Rheumatism and the American College of Rheumatology published new provisional classification criteria 2012.

- PMR in patients aged 50 or older with bilateral shoulder aching and elevated inflammatory markers

Criteria	Points
Morning stiffness >45 minutes	2
Hip pain/limited range of motion	1
Absence of rheumatoid factor and/or anti-citrullinated protein antibody (anti-CCP)	2
Absence of peripheral joint pain	1

- A score of ≥4 points has a 68% sensitivity and 78% specificity for discriminating PMR from other comparison patients. There is also an additional ultrasound criteria (1 point if positive findings), which can add up to a score of ≥5 points that is associated with a 66% sensitivity and 81% specificity for PMR.

Investigations

(Joint guidelines from the EULAR and ACR)

- Rheumatoid factor and/or anti-cyclic citrullinated peptide antibodies (anti-CCP)
- C-reactive protein (CRP) and/or erythrocyte sedimentation rate (ESR)
-
- Complete blood cell count (CBC) with differential
- Blood glucose
- Serum creatinine
- Liver function tests
- Bone profile (including calcium and alkaline phosphatase)
- Dipstick urine analysis

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Additional studies to consider are as follows :

- Protein electrophoresis
- Thyroid-stimulating hormone (TSH)
- Creatinine kinase
- Vitamin D

If clinically indicated, tests such as the following may be considered to exclude alternative diagnoses :

- Antinuclear antibodies (ANA)
- Anti–cytoplasmic neutrophil antibodies (ANCA)
- Tuberculin test
- Chest radiograph

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- Synovial fluid analysis

In patients who have synovitis with effusions, synovial fluid analysis reveals

- signs of mild inflammation, including poor mucin clotting.

- Synovial fluid WBC counts range between 1,300-11,000 cells/µL (median 6,000 cells/µL), with 34% polymorphonuclear leukocytes (range 12-78%).

Imaging studies

- Radiographs reveal either normal joints or evidence of osteoarthritis.
- Magnetic resonance imaging (MRI) is not necessary for diagnosis, but MRI of the shoulder reveals subacromial, subdeltoid bursitis and glenohumeral joint synovitis in the vast majority of patients.
- Bursa ultrasonography may reveal an effusion within the shoulder bursae.

Differential diagnoses

Disease	Clinical feature	Investigation
Rheumatoid arthritis	▪In younger patients in their early fifties ▪ Symmetric polyarthritis of the small joints of the hands and feet, which is persistent and only partially responsive to low doses of prednisone.	Measurement of rheumatoid factor and antibodies to cyclic citrullinated peptide (anti-CCP) is mandatory
Multifocal local musculo-skeletal pain	•Symptoms and signs at the shoulders in PMR can be similar to those that result from subacromial/subdeltoid bursitis without PMR or from rotator cuff tendinitis •Morning stiffness is brief, •Constitutional symptoms are absent	Acute phase reactants are not elevated

Disease	Clinical features	Investigations
Drug-induced myalgias or myositis	➢Absence of prominent morning stiffness ➢Lack of proximally and symmetrically distributed symptoms	➢Elevations of the serum creatine kinase levels
Inflammatory myopathy	➢Symmetric proximal muscle weakness ➢Shoulder and hip pain is not as prominent	➢Elevated muscle enzymes ➢Abnormal electromyography ➢Abnormal MRI ➢Evidence of myositis on muscle biopsy

Disease	Clinical features	Investigations
Fibromyalgia	➢Widespread musculoskeletal pain, stiffness, aching, and fatigue ➢Symptoms of longstanding duration	➢A normal ESR, CRP, and hematocrit
Parkinson disease	➢Stiffness in an older adult ➢Abnormalities on neurologic examination, including tremor and rigidity	
Spondyloarthropathy	➢Proximal symptoms similar to PMR ➢Presence of enthesitis, dactylitis, anterior uveitis,	➢Sacroiliitis on imaging ➢ High prevalence of human leukocyte antigen (HLA)-B27

Disease	Clinical features	Investigations
Hypothyroidism	➢Aching, stiffness and arthralgias ➢Slow relaxation of deep tendon reflexes	➢A low serum thyroxine T4 ➢An elevated TSH
Multiple myeloma	Bone pain	➢Elevated ESR ➢Presence of a monoclonal protein in the serum or urine
Hyperparathyroidism	Proximal stiffness and aching	➢Hypercalcemia ➢Elevated parathyroid hormone
Osteomalacia	Widespread musculoskeletal pain.	Multiple microfractures
Depression	➢Weight loss ➢Somatic symptoms, including aching and stiffness	

Management

- Corticosteroids (ie, prednisone) are considered the treatment of choice.
- Recommend starting corticosteroid therapy with 12.5-25 mg/day of prednisone or the equivalent.
- A slow tapering of the prednisone, less than 1 mg/month.
- Once prednisone is tapered to 10 mg/day, a slow taper by 1 mg every 2 months until treatment discontinuation was associated with optimal control of disease activity.

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- **Steroid sparing agent**

✓ Tocilizumab : an interleukin-6 receptor antagonist.

✓ Methotrexate : no benefit alone or with low dose corticosteroid

✓ TNF-α inhibitors : not recommended

✓ Azathioprine : high frequency of adverse drug effects

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Consultations

Ophthalmologists, pathologists, and surgeons may be consulted on an as-needed basis should concern arise about the development of GCA.

Diet

Calcium and vitamin D supplementation should be initiated.

Activity

Generally, activity restriction is unnecessary. Physical therapy is recommended for those with difficulty achieving good mobility despite adequate medical therapy

Conclusion

- PM occurs in patients who on average are over 50 years of age.
- Cardinal symptoms are shoulder and hip girdle pain with pronounced stiffness lasting at least one hour.
- Clinicians must be alert to mimics, including infection, malignancy, metabolic bone disease, and elderly onset rheumatoid arthritis.
- Erythrocyte sedimentation rate or C reactive protein, or both is usually raised at disease onset.
- Giant cell arteritis is present in about 5 to 30% of patients.
- PM is treated with glucocorticosteroids, starting at 15 mg prednisone a day.


Thank You!