Polymyalgia rheumatica

Dr Katya Dolnikov
D_katya@rambam.health.gov.il
2017

Introduction

- Polymyalgia rheumatica (PMR) is an inflammatory rheumatic condition
- Characterized clinically by :
  - aching and morning stiffness in the shoulders, hip girdle, and neck.
- It can be associated with giant cell (temporal) arteritis (GCA), and the two disorders may represent different manifestations of a shared disease process

Epidemiology

- Disease of adults over the age of 50, with a prevalence that increases progressively with advancing age
- The peak incidence of PMR occurs between ages 70 and 80
- PMR is relatively common. The lifetime risk of PMR is second only to rheumatoid arthritis (RA) as a systemic rheumatic disease in adults
- Women are affected two to three times more often than men
- Cases of familial aggregation are recognized
- The incidence is highest in Scandinavian countries and in people of northern European descent
- PMR appears to be uncommon in Asian, African-American, and Latino populations, though all racial and ethnic groups may be affected.

Association with GCA

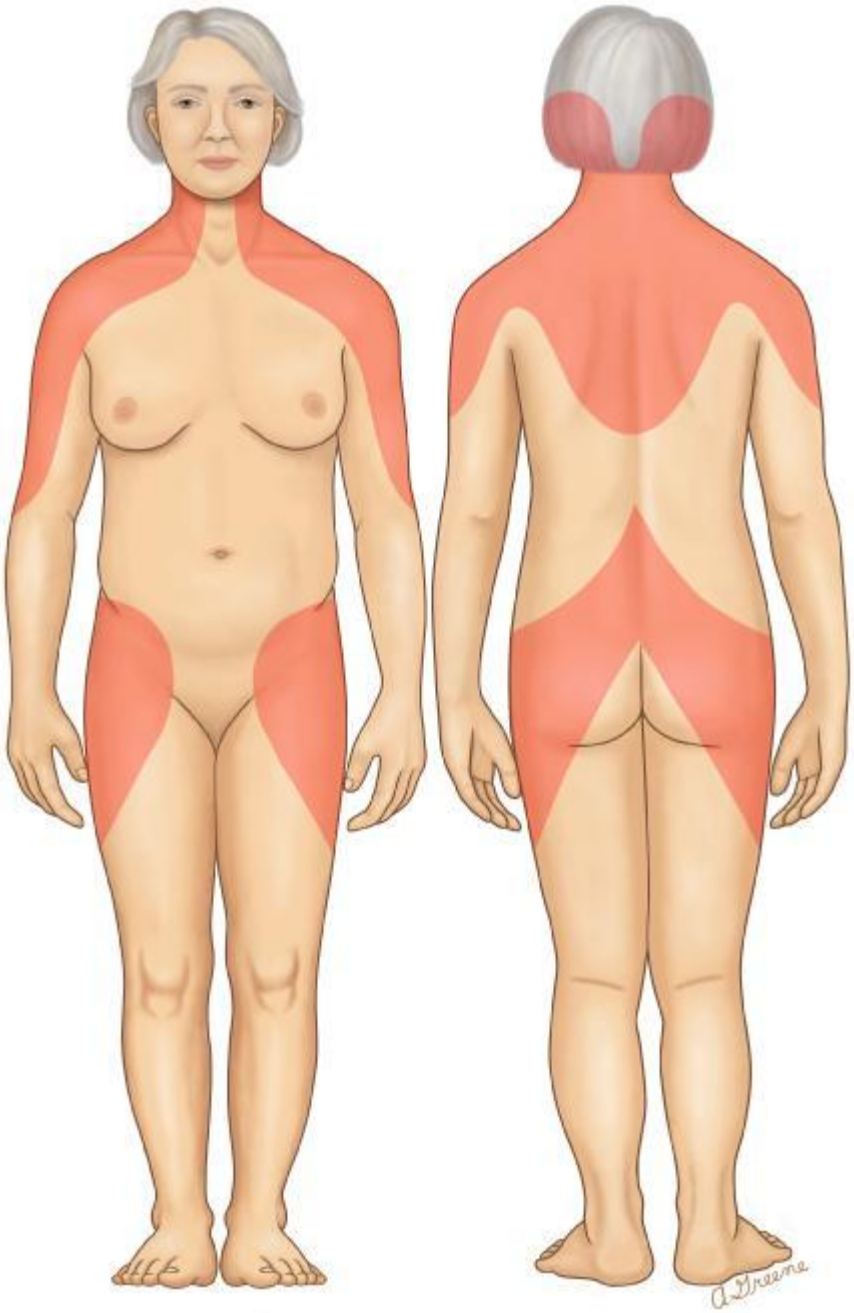
- PMR occurs in about 50% of patients with GCA
- The percentage of patients with PMR who experience GCA at some point is ~10%
- The two disorders may not be active synchronously

Pathogenesis

- The cause of polymyalgia rheumatica (PMR) is unknown
- Both environmental and genetic factors appear to play a role
- Both PMR and GCA are associated with specific alleles of human leukocyte antigen (HLA)-DR4
- Some studies have suggested a cyclical pattern in incidence and seasonal variation

Signs and Symptoms

- Aching and morning stiffness
- Shoulders, hip girdle, neck, and torso are involved
- Patients over the age of 50
- Symptoms are usually symmetric
- Recent, discrete change in musculoskeletal symptoms



Signs and Symptoms

- Morning stiffness > 30 minutes
- Stiffness at the shoulders and hips may cause trouble with dressing
- Stiffness may be so severe that there is difficulty turning over in bed at night or arising from bed in the morning.
- The ‘gel’ phenomenon, stiffness after inactivity, is often notably severe in PMR
- An inability to actively abduct shoulders past 90 degrees because of stiffness is a typical finding

Signs and Symptoms

- Shoulder pain is more common at presentation than hip pain
- Pain is worse with movement and may interfere with sleep
- Synovitis and bursitis - in peripheral joints, such as the knees, wrists, and MCPs
- Synovitis and bursitis are thought to be the causes of the discomfort and stiffness
- Swelling and tenosynovitis – Some patients develop swelling and pitting edema of the hands, wrists, ankles, and top of the feet
- Tenosynovitis can also cause carpal tunnel syndrome
- Decreased range of motion – There may be decreased active and passive range of motion of the shoulders, neck, and hips.
- Muscle tenderness – not a prominent feature, and what tenderness there may be about the shoulders is more likely due to synovial or bursal inflammation than muscle involvement
- Normal muscle strength
- Systemic signs and symptoms – malaise, fatigue, depression, anorexia, weight loss, and low-grade fever.

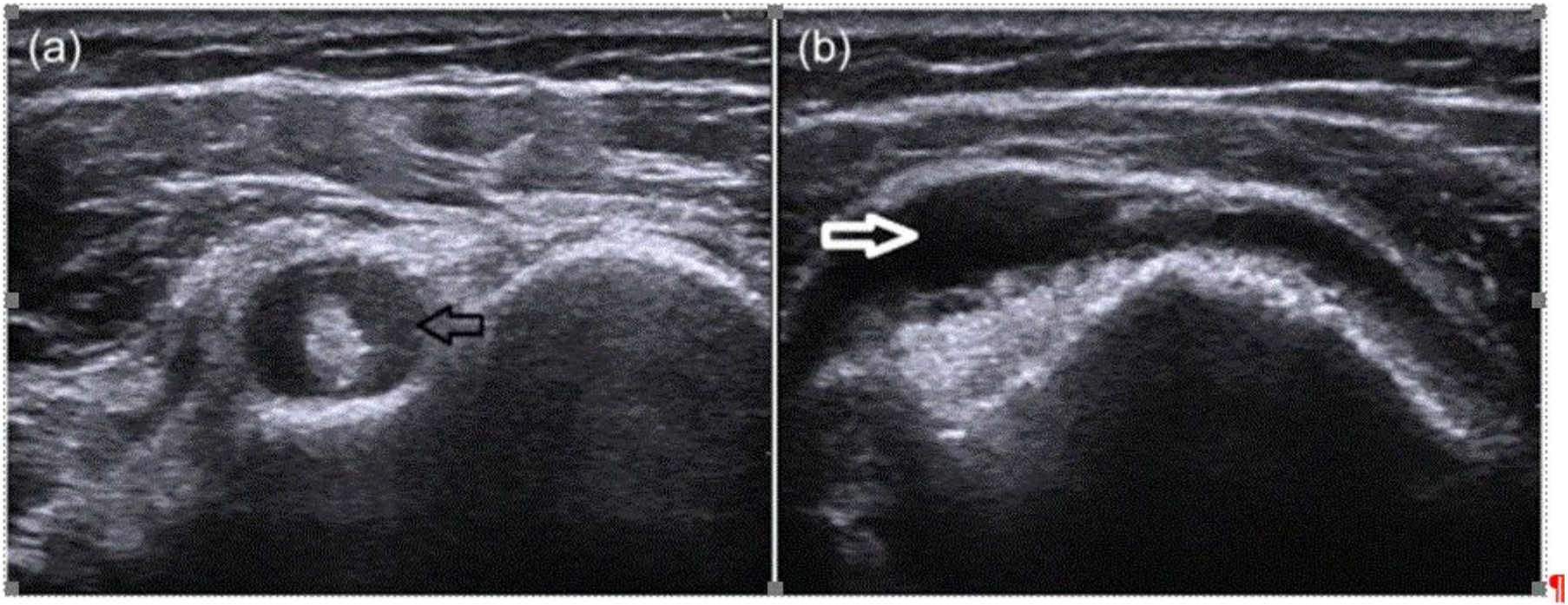
Laboratory findings

- Elevated ESR, CRP (although can be normal)
- Normocytic anemia
- Thrombocytosis
- Serologic tests, such as ANA, RF, ACPA are typically negative
- Increase in liver enzymes, especially alkaline phosphatase, although these abnormalities are more common in patients with GCA than PMR alone

Imaging

- Routine radiographs of inflamed joints do not show abnormalities in patients with PMR
- MRI and US can demonstrate synovial inflammation, with a predilection for extra-articular synovial structures (bursitis, tenosynovitis)
- Synovitis is never erosive

Extra-articular involvement



Biceps tenosynovitis

subdeltoid bursitis

Evaluation of the patient

- Medical history
- Anemnesis
- Physical examination
- Assessment of the response to low-dose glucocorticoids.
  - Symptoms are generally 50 to 70 % better within 3 days of prednisone (10 to 20 mg/day) and
  - Almost all patients respond completely within two weeks of initiation of therapy
- MRI or US may be helpful to assess whether there is underlying bursitis or other evidence for inflammation

Diagnosis

- No pathognomonic test or established diagnostic criteria for polymyalgia rheumatica (PMR)
- Presence of all of the following, after exclusion of alternative disease:
  - Age 50 years or older at disease onset
  - Proximally and bilaterally distributed aching and morning stiffness for at least two weeks
  - 2/3 areas: neck or torso, shoulders or proximal regions of the arms, and hips or proximal aspects of the thighs
  - ESR ≥40 mm/h
  - Rapid resolution of symptoms with low-dose glucocorticoids.

Evaluation for GCA

- Patients with clinically "pure" PMR lack the classic findings of GCA:
  - temporal artery tenderness,
  - headache,
  - jaw pain, visual symptoms
  - arm claudication.
- GCA may appear at any point during the clinical course of PMR
- At every follow-up visit the patient should be monitored for GCA signs and symptoms
- Evaluation, including biopsy, should be performed if symptoms of GCA develop, even if patients are on glucocorticoids

Diffrential Diagnosis

- **Rheumatoid arthritis** - symmetric polyarthritis of the small joints of the hands and feet, which is persistent and only partially responsive to low doses of prednisone. In PMR fewer joints are swollen, and swelling subsides completely in response to low-dose prednisone
- **RS3PE syndrome** - **R**emitting **S**eronegative **S**ymmetrical **S**ynovitis with **P**itting **E**dema, also described as the puffy edematous hand syndrome or distal extremity swelling with pitting edema
- **Inflammatory myopathy** - Dermatomyositis or polymyositis present with symmetric proximal muscle weakness
- **Fibromyalgia**
- **Infective endocarditis**
- **Lyme disease** may present with nonspecific constitutional symptoms that include myalgias and arthralgias
- **Malignancy**
- **Vasculitis**

Treatment

- Practically all patients with PMR alone will respond to 12.5 to 25 mg/day of prednisone
- Persistent aching and stiffness/ elevated CRP or ESR under prednisone mandate pursuit of an alternative diagnosis
- Slow tapering down