POLYMYALGIA RHEUMATICA – A BRIEF REVIEW

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Polymyalgia rheumatica (PMR) is an inflammatory disorder which is characterized by vague pain and aches along with morning stiffness affecting mainly the shoulder girdle and the pelvic girdle. It exclusively affects patients aged 50 years or over although younger age group can be involved. Giant cell arteritis (GCA) is a frequent accompaniment of this disorder. A widespread inflammatory reaction and a characteristic response to corticosteroid therapy are typical features of both the diseases.

Classification criteria for PMR-

Many sets of criteria are available to classify PMR but none of them has been fully validated .American College of Rheumatology (ACR) and European League Against Rheumatism (EULAR) has defined new classification criteria for PMR which has a widespread usage now.(1)

ACR/EULAR criteria

- Patients 50 years or older, Bilateral shoulder aches, and Abnormal C-reactive protein and/or erythrocyte sedimentation rate, plus at least 4 points (without ultrasound, US) or 5 points (with US) from the following:
- Morning stiffness > 45 minutes (2 points)
- Hip pain or limited range of motion (1 point)
- Absence of rheumatoid factor or anti-cyclic citrullinated peptide antibodies (2 points)
- Absence of other joint pain (1 point)
- If US is available, at least one shoulder with subdeltoid bursitis and/or biceps tenosynovitis and/or glenohumeral synovitis (either posterior or axillary) and at least one hip with synovitis and/or trochanteric bursitis (1 point).
- If US is available, both shoulders with subdeltoid bursitis, biceps tenosynovitis, or glenohumeral synovitis (1 point)

Epidemiology

Polymyalgia rheumaticais considered a disease of adults over the age of 50 and its prevalence rises with advancing age. The peak incidence of PMR is known to occur between ages 70 and 80 [2]. Women are affected twice more often than men.

Pathogenesis

The aetiology of PMR is unknown. However, evidence suggests that both genetic susceptibility and environmental risks factors may play a role in the development of the disease

Clinical manifestations

The hallmark features of PMR are vague aches and early morning stiffness in the neck, shoulder and pelvic girdle, typically, early morning stiffness may last 30 min or longer. The patients also describe stiffness after periods of rest known as the 'gel' phenomenon which is quite severe in PMR.

Shoulder pain is reported by 75–99% of patients, whereas the hip girdle and neck are less frequently involved (50–90% of cases). It can be initially unilateral at presentation but becomes bilateral in later stages of the disease.

It is inflammatory in nature and gets aggravated by active movements and can interfere with activities of daily life like dressing or rising from a chair. Muscle tenderness is also a common accompanying feature .There may be decreased active and passive range of motion of the shoulders and other axial joints .Constitutional features like fatigue, low grade fever, depression and weight loss occur in up to 40% of patients. Many patients exhibit distal joint synovitis, mainly wrists, knees and metacarpophalangeal joints. PMR-associated arthritis is non- erosive and usually remits after corticosteroid therapy. (3)

Laboratory findings:

The characteristic laboratory finding in PMR is an elevated ESR. Elevated serum C-reactive protein level and normocytic normochromic anaemia are also present. Rheumatoid factor, antinuclear antibodies and cyclic citrullinated peptide antibodies are typically negative in PMR. MRI and ultrasound may demonstrate underlying synovial inflammation .PET scanning has no proven clinical value in the care of patients with PMR.

Differential diagnosis- Although the symptoms of PMR are very characteristic, several other diseases like autoimmune disorders, infections and malignant disorders can present with similar complaints. Inflammatory arthritis like Late onset Rheumatoid arthritis, RS3PE (**R**emitting **S**eronegative

Symmetrical **S**ynovitis with **P**itting **E**dema), Metabolic bone disorders, Malignancies like Multiple myeloma, Myopathy, Viral myalgias Thyroid disorders need to be ruled out on priority for establishing the diagnosis.(4)

Treatment

Corticosteroids remain to date the cornerstone of treatment for PMR. In PMR, to start with an initial dose of 10–20 mg of prednisone is sufficient in most of the cases to control the inflammatory symptoms and stiffness although optimal dosing regimen is not well defined and need to be tailored on case to case basis.(5) Patients report dramatic relief after the initiation of therapy even after a single dose. The rapid response in 24–72 h can help to confirm the diagnosis. Lack of clinical improvement after treatment with prednisone 20 mg daily for a week should prompt the physician to question the diagnosis of PMR and consider other diagnoses. After 2–3 weeks of corticosteroid therapy, the doses should be slowly tapered to lower levels with monitoring of clinical symptoms and inflammatory markers specially ESR. In general, a treatment course of 1–2 years often suffices but some patients might need corticosteroids for years and even indefinitely, often maintained on 2.5–5 mg of prednisone daily or the lowest possible dose which keeps the disease in remission.

Results are not very encouraging for clinical studies of methotrexate (MTX) and tumor necrosis factor (TNF) inhibitors. Methotrexate can be added as steroid sparing agent as is suggested by some studies but not all.(6) Role of TNF inhibitors is still under investigation and some studies suggest that they could be effective in some patients suffering from long-standing PMR, but not in newly diagnosed PMR.(7) Tocilizumab, a humanized IL-6 receptor monoclonal antibody, has also been used to treat PMR, and has shown favourable evidence but further studies are required for the same.(8)

Prognosis

PMR affects the quality of life because of the pain and functional limitation but increased mortality due to the disease is not reported. Patient needs to be monitored for Side effects related to the glucocorticoid therapy i.e osteoporosis, glucose intolerance, and hypertension.

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