Polymyalgia rheumatica

General information

Polymyalgia rheumatica (PMR) and giant cell arteritis (GCA) may be different points on a continuum of the same disease. Both have an increased frequency of HLA-DR4 and systemic monocyte activation. 15% of patients with PMR eventually develop GCA.

Epidemiology

Both GCA & PMR occur in people ≥ 50 years old. The incidence increases with age and peaks between 70 and 80 years and is higher at higher latitudes. PMR is more common than GCA. Prevalence: 500/100,000).Incidence: 52.5 per 100,000 people ≥ age 50, higher in females (61.7) than males (39.9).

Features

● an inflammatory condition of unknown etiology

● clinical characteristics

a) aching and morning stiffness in the cervical region and shoulder & pelvic girdles lasting > 1 month. The pain usually increases with movement

○ shoulder pain: present in 70–95% of patients. Radiates toward elbow

○ hip & neck pain: 50–70%. Hip pain radiates towards knees

b) age ≥ 50 years

c) ESR ≥ 40 mm/hr (7–20% have normal ESR)

d) usually responds rapidly to low dose corticosteroids (≤ 20mg prednisone/day)

e) systemic symptoms (present in ≈ 33%): fever, malaise or fatigue, anorexia and weight loss

● favorable prognosis: usually remits in 1–3 years

Differential diagnosis

The crowned dens syndrome (CDS), also known as periodontoid calcium pyrophosphate dehydrate crystal deposition disease, is typified clinically by severe cervical pain, neck stiffness and atlantoaxial synovial calcification which could be misdiagnosed as meningitis, epidural abscess, polymyalgia rheumatica, giant cell arthritis, rheumatoid arthritis, cervical spondylitis or metastatic spinal tumor.

Crystalline deposition on cervical vertebrae is less well known disease entity and only a limited number of cases have been reported to date [1)]

Treatment

PMR responds either to low doses of steroids (10–20 mg prednisone/day) or sometimes to NSAIDs (response to steroids is much more rapid). The initial dose of steroids is maintained for 2–4 weeks, and then by ≤ 10% of the daily dose every 1–2 weeks while observing for signs of giant cell arteritis (GCA).

Case reports

Blecher et al. described a 48-year-old woman suffering from bilateral upper-extremity numbness and axial radiating pain. Magnetic resonance imaging revealed soft-tissue edema and enhancement surrounding the dorsal tip of the C7 spinous process. Excisional biopsy of the lesion revealed a mildly inflamed bursa, with no evidence of an active infection. Removal of the inflamed bursa resulted in complete resolution of the upper-extremity numbness and improvement in her neck pain. Although similar cases have been reported to be associated with rheumatologic conditions, most notably polymyalgia rheumatica (PMR), the current report underlines the presentation of radicular-like complaints associated with interspinous bursitis in the absence of other conditions affecting the musculoskeleton [2)].

A 52-year-old male on chronic prednisone for polymyalgia rheumatica presented with a subacute history of headaches, nausea, phonophobia, intermittent diplopia and gait instability. He was hospitalized 2 weeks prior to presentation with extensive evaluations only notable for leptomeningeal inflammation on MRI. His symptoms progressively worsened and he developed aphasia. He was transferred to our facility where extensive spinal fluid examinations were repeated and were again nondiagnostic. Ultimately, a diagnostic skull-based biopsy was performed which demonstrated Blastomyces dermatitidis fungal meningitis. Despite extensive sampling and cultures, only 1 of the intraoperative samples yielded diagnostic results. This underscores the low sensitivity of current methods to diagnose CNS blastomycosis. This case suggests that a neurosurgical biopsy may be necessary and should be considered early in the diagnostic process, especially if a definitive diagnosis is elusive. If a biopsy is performed, sampling should be ample and from multiple areas. Following the diagnosis, our patient was treated with liposomal amphotericin B and then voriconazole with a good clinical response [3)].

A 72-year-old man was admitted to our hospital due to severe headache. Two months prior to admission, the patients had exhibited recent-onset stiffness and myalgia of shoulder and pelvic girdle that was compatible with PMR. Magnetic resonance imaging revealed a mass lesion in the pituitary fossa with focal hemorrhage. Endocrinologic studies demonstrated hypopituitarism. The headache and myalgia were improving with corticosteroid treatment; however, a trans-sphenoidal surgery was performed due to visual field loss. A white-colored mass was resected, and histologic examination showed diffuse infiltration of lymphocytes and plasma cells consistent with lymphocytic hypophysitis.

Post-operatively, the headache and visual field loss resolved completely. This is the first documented case of apoplectic lymphocytic hypophysitis complicating PMR, and a possible mechanism for this rare association was discussed [4].

Polymyalgia rheumatica presenting after successful treatment of Cushing's disease [5].

Polymyalgia rheumatica: a systemic vasculitis. Review of the literature and presentation of 2 clinical cases [6].

References

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