A Polymyalgia Rheumatic-Mimicking Case with Young Adult Onset
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Abstract
The diagnosis of polymyalgia rheumatica (PMR) is made primarily on clinical features and laboratory evidence of inflammation. Among the criteria for diagnosing PMR, disease onset at an age of 50 years or over is one of the major premises. The present case with 35-year-old onset showed clinical features of symmetric proximal myalgias (shoulder and pelvic girdle) with pronounced stiffness for more than 3 weeks, morning stiffness lasting more than 45 minutes combined with laboratory abnormalities (elevated CRP and ESR), and a dramatic improvement with steroid treatment at a low dosage (20mg/day), which is quite compatible with PMR except for the age of onset of the disease. This case shows that PMR-mimicking conditions can occur in patients even under 50 years of age.

Introduction
Polymyalgia rheumatica (PMR) clinically consists of severe pain and stiffness in the shoulders, often in the neck and pelvic girdle, and morning stiffness lasting for more than 45 minutes, together with laboratory abnormalities including elevated C-reactive protein (CRP) and erythrocyte sedimentation rate (ESR) and a dramatic response to steroid [1-4]. Almost all cases of PMR occur in adults of 50 years or over, and the incidence of the disease increases with each decade of life after 50 years, peaking around 75 years [5]. Among the criteria for diagnosing PMR, disease onset at an age of 50 years or over is one of the major premises [2,3]. This is a case report on a 35-year-old man who fulfills all the criteria of PMR except age at disease onset.

Clinical Report
The course of clinical symptoms and laboratory investigations in this case is depicted in Figure 1. A 35-year-old man had a fever (38 °C), sore throat, headache and pains in the whole body for 3 consecutive days without the symptoms of upper respiratory tract infection. Two days before the appearance of the symptoms, his 3-year-old son was diagnosed with streptococcal infection. The patient saw a physician at our clinic. Physical examination revealed slight fever (37.2 °C) and redness in the pharyngeal tonsils without swelling. Streptococcus Group A was detected from the swab of pharyngeal tonsils. He was prescribed amoxicillin for 10 days, resulting in complete improvement of pharyngeal tonsillitis, headache, and body ache. Approximately 2 weeks later, he developed new-onset severe pain and stiffness in both shoulders, the neck, and the pelvic girdle. His symptoms were worst in the morning, and he had great difficulty rising from bed or a chair, or even turning over in bed. Morning stiffness of the body and extremities lasted for more than one hour. He also noticed exacerbation of pain in the right hip joint when he stood up. He consulted with an orthopedist in another hospital and underwent X-ray examinations of the shoulder and hip joints, revealing no abnormality including chondrocalcinosis. MRI examination in the pelvic girdle and lumbar spine revealed no abnormality including inflammatory signs such as bursitis or joint synovitis. He suffered from these symptoms for 3 weeks before coming back to our hospital to consult the author. Physical examination was unremarkable: there was no fever, pitting edema in the bilateral dorsalis pedis and dorsum of the hand, skin eruption, or specific tender points. Neurological examination showed no abnormality either, including cranial nerves, extremity muscle strength, deep tendon reflexes, plantar responses, abnormal movements, or position sensation and vibratory sensation in the feet. Laboratory examination showed increased CRP (4.01). Serum chemistry was normal, including complete blood cell count, CK, urea and electrolytes, calcium, creatinine and blood glucose, urinalysis, and thyroid hormone. Antineutrophil cytoplasmic antibodies (ANCA) that target the proteins myeloperoxidase (MPO) and proteinase 3 (PR3), rheumatoid factor (RF), and anti-cyclic citrullinated peptide antibody (anti-CCP antibody) were all negative. Infectious diseases, malignancy, rheumatic arthritis, polymyositis, ANCA-associated vasculitis and fibromyalgia were ruled out from the clinical manifestations and laboratory investigations. The patient was diagnosed with polymyalgia rheumatica by clinical features and elevated inflammatory markers including CRP. Giant cell arteritis was not considered to be present concurrently. Prednisolone at a low dosage (20mg/day) was started, which resulted in dramatic improvement of clinical features such as morning stiffness and pain in the shoulders and hip girdle about 4 days after medication. One week after treatment, his inflammatory markers improved: CRP and ESR were 0.41 and 13mm/h, respectively. Two weeks after the beginning of prednisolone treatment, his inflammatory markers were completely normal (CRP: ≤ 0.30, ESR: 10 mm/h) (Figure 1). The dose of prednisolone was slowly tapered and discontinued over 10 months. More than 8 months after steroid discontinuation, PMR had not relapsed.

Keywords: Polymyalgia rheumatica; Polymyalgia rheumatica-mimicking; Criteria for polymyalgia rheumatica; Young adult onset

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Discussion

PMR is a relatively common inflammatory disease in elderly people and a common indication for long-term corticosteroid therapy [6]. Given the lack of a gold standard diagnostic test for PMR, the diagnosis is made primarily on clinical features and laboratory evidence of inflammation [7,8]. Polymyalgia rheumatica has non-specific features and therefore a wide differential diagnosis is crucial in order to exclude conditions mimicking PMR such as active infection, cancer and other inflammatory diseases.

The present case showed acutely developing clinical features of symmetric proximal myalgias (shoulder and pelvic girdle) with pronounced stiffness for more than 3 weeks, morning stiffness lasting more than 45 minutes combined with laboratory abnormalities (elevated CRP and ESR), and a dramatic improvement with steroid treatment, all of which are consistent with the hallmarks of clinical manifestations of PMR [1-4]. Based on the provisional European League Against Rheumatism/American College of Rheumatology (EULAR/ACR) classification criteria for PMR (2012) [3], and on the British Society for Rheumatology (BSR) and the British Health Professionals in Rheumatology (BHPR) guidelines (2009) [2], this case is quite compatible with PMR except for the age of onset of the disease. Rapid symptom resolution and reduction of the CRP and ESR to normal ranges after low-dose corticosteroid treatment in this case is also characteristic of PMR [1,4,9]. Moreover, certain conditions mimicking PMR could be ruled out from clinical manifestations, as well as from laboratory and imaging examinations: inflammatory disorders such as rheumatoid arthritis, synovitis, spondyloarthropathy, systemic lupus erythematosus, systemic vasculitis and inflammatory myopathy, and non-inflammatory disorders such as osteoarthritis, adhesive capsulitis (frozen shoulder), infection, fibromyalgia, and endocrine and metabolic diseases.

Although the cause of PMR is unknown, both genetic and environmental factors contribute to disease susceptibility and severity. Some studies show a cyclical pattern in incidence, which indicates that PMR may be triggered by an environmental infection, such as parvovirus B19, Mycoplasma pneumoniae, and Chlamydia pneumoniae [1]. Thus, the author could not exclude the possibility that streptococcal infection of pharyngeal tonsils might have triggered PMR in this patient.

Regarding the patient’s age at the onset of the disease, almost all previous reports noted that PMR affects the elderly and that its prevalence increases with age, peaking at 70-80 years [5]; indeed, an age of 50 years or older is considered a criterion for diagnosis [2,3]. PMR usually appears in patients over 65 and is almost never seen in people under age 50 [1]. This case shows that PMR-mimicking conditions can occur in patients even under 50 years of age.

Conclusion

This case suggests that the occurrence of a PMR-mimicking case in a patient under 50 should be noted in relation to the criteria for diagnosing PMR. If and when similar case reports accumulate, the age criterion of 50 years for the onset of the disease should be reviewed and modified.

References


