A 24-Year-Old Man with Symptoms and Signs of Polymyalgia Rheumatica

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Most physicians regard polymyalgia rheumatica (PMR) as a disease that affects only the elderly. This case report of a 24-year-old man with proximal limb girdle muscle pain, stiffness, tenderness, weakness, and an elevated erythrocyte sedimentation rate, who had a dramatic response to steroids, supports the premise that PMR may affect a wider range of our population. Increased physician awareness that this illness does occur in patients younger than 50 may prevent delays in diagnosis, and decrease the needless suffering and incapacitation of younger patients with this disorder.

KEY WORDS. Polymyalgia rheumatica; giant cell arteritis; prednisone. *(J Fam Pract 1998; 47:68-71)*

Polymyalgia rheumatica (PMR) is a disorder characterized by morning stiffness, bilateral shoulder or hip muscle pain and tenderness, and an erythrocyte sedimentation rate (ESR) >40 mm/h. It is often associated with fatigue, weight loss, anorexia, and depression, and can be incapacitating, even to the point of preventing affected individuals from standing up once they have assumed a seated position. The diagnosis of PMR should not be entertained unless the morning stiffness lasts for more than 1 hour each day, and the symptoms and signs described above have persisted for more than 2 to 3 weeks. There is no pathognomonic confirmatory test for PMR. It is a diagnosis of exclusion. Although it is predominantly a disease of the elderly (in whom prevalence estimates run as high as 1%[1]), six case reports in the literature, as well as the following account of a 24-year-old man, lend support to the premise that PMR may affect a younger population than previously thought.

Case Report

A 24-year-old man awoke one morning with sharp pain and stiffness in his thighs and groin that failed to improve with massage, stretching, or position changes. He denied any other motor or sensory symptoms, problems with bladder or bowel function, skin changes, visual problems, alcohol, drug, or medication use, or any significant personal or family medical history. Physical examination disclosed bilateral thigh (adductor and quadriceps) tenderness and weakness (strength 4/5). Despite the lack of trauma, adductor muscle strain seemed to be the most likely diagnosis. The patient was provided with crutches because of his obvious difficulty with ambulation, given a prescription of ibuprofen, and referred to a physical therapist for assistance with rehabilitation.

Three weeks later, the patient had not improved, and he sought a second opinion from the medical center emergency department (ED) physicians for his thigh pains, stiffness, and weakness. Radiographs of the hips, pelvis, and femurs at this time were normal. The ED physician diagnosed muscle spasms, and added diazepam to the patient's regimen of ibuprofen. The patient returned the next day, unimproved. A repeat physical examination at this time documented normal color to the extremities, normal reflexes, no sciatica elicited with straight-leg raises, but mildly decreased strength in both hip flexor and extensor muscles. Laboratory data obtained at this visit included a normal creatine phosphokinase (CPK) and an elevated ESR of 101 mm/h.

Over the next few months, the patient was referred to multiple specialty clinics for evaluation, in the following order: rehabilitative medicine, orthopedics, internal medicine, neurology, and rheumatology. Accumulated laboratory data, radiography results, and other studies are summarized in Table 1. Persistent elevation of the ESR and a mild normochromic, normocytic anemia were the only abnormalities. The rheumatologist's differential diagnosis included viral reactive arthritis and spondyloarthropathy, and he treated the patient with indomethacin and range of motion exercises.

The patient's condition failed to improve over the next month, and he sought care once again from his primary care provider. Now, 6 months after initial presentation, the diagnosis of polymyalgia rheumatica was entertained, despite his young age, and the despondent, nonfunctional patient was treated with prednisone 50 mg/day. Within 2 days he was symptom-free, and he began running 2 miles per day.

Discontinuation of steroids after 7 days led to a complete relapse, despite concurrent use of indomethacin. He was restarted on prednisone 20 mg/day, and the dosage tapered to 10 mg/day for the next 6 months to

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maintain a normal ESR level. The provisional diagnosis was polymyalgia rheumatica.

DISCUSSION

The exclusion of patients younger than 50 in most studies of PMR, a condition with a peak incidence of disease near age 68, is arbitrary.[2] Polymyalgia rheumatica is suspected when patients complain of morning stiffness, difficulty standing from a seated position, and fatigue. A provisional diagnosis of PMR should not be made unless patients meet at least three of the positive criteria in Table 2.[1] If a patient has the first three criteria on this list (bilateral pelvic or shoulder pain/stiffness, symptoms for more than 2 weeks, and an ESR >40 mm/h), along with two additional negative criteria (normal muscle enzymes and no swelling of the hand joints), these findings become 93% sensitive and 98% specific for PMR.[3] The etiology of PMR is unconfirmed at present, but is thought to be triggered by a precedent viral infection, such as adenovirus or respiratory syncytial virus.[8] An infectious cause is supported by the report of another case in which a married couple simultaneously developed PMR.[9]

Polymyalgia rheumatica is a possible variant of seronegative rheumatoid arthritis.[10] It would stand to reason, then, that seronegative juvenile rheumatoid arthritis might also cause a synovitis that presents as PMR in younger patients.

The differential diagnosis of PMR is outlined in Table 3. Quinidine and more recently topical minoxidil (for hair growth) have been incriminated as inducing a PMR-like illness with proximal muscle weakness or pain.[5,6]

Laboratory and imaging tests that may be useful for supporting the diagnosis of PMR, or for revealing other causes for the symptoms, are: complete blood count with ESR (a normal sedimentation rate virtually excludes the diagnosis of PMR; sensitivity 90%, specificity 97%), rheumatoid factor (for PMR, the sensitivity is only 7.5%, specificity 98%, but a positive test may suggest lupus erythematosus, mixed connective-tissue disease, or rheumatoid arthritis), antinuclear antibody, thyroid-stimulating hormone, creatine phosphokinase, C-reactive protein (CRP), Borrelia titers, lupus preparation, chest film, and radiography of any possibly affected joints. Magnetic resonance imaging of the affected muscles can exclude tumor, rhabdomyolysis, and inflammatory illness, and may be useful to direct the site of muscle biopsy, if one is planned.[7] Among the diagnostic possibilities, rheumatoid arthritis, Lyme disease, and giant cell arteritis (GCA) are the conditions most commonly confused with PMR.

Factors pointing toward PMR and away from reactive arthritis in our 24-year-old patient include proximal, instead of distal, joint involvement; persistence of symptoms for over a month instead of progression; a negative test for rheumatoid factor; ESR >90 mm/h; sudden onset compared with insidious onset; lack of joint swelling; and rapid complete response to steroids.[11]

TABLE 1

Accumulated Laboratory Data from Multiple Specialty Clinics Evaluating a 24-Year-Old Patient

Laboratory findings with abnormal test results

ESR = 101 mm/h

Hematocrit = 36%*

Hemoglobin = 12 mg/dL*

Laboratory and radiology data obtained with normal test results

Electrolytes
Blood urea nitrogen (BUN)
Creatinine, glucose

Rheumatoid factor
Calcium, phosphorus
Liver function tests
Thyroid-stimulating hormone

Borrelia titers

Radiographs of chest, hips, pelvis, femurs
Bone scan
Nerve conduction studies
Electromyography

Urinalysis
Angiotensin-converting enzyme (ACE)
Creatine phosphokinase (CPK)

Aldolase
HLA-B27, anti-nuclear antibody (ANA)
Sickledex
Serum protein electrophoresis (SPEP)

*Hematocrit and hemoglobin were 42% and 13 mg/dL, respectively, prior to illness.

TABLE 2

Polymyalgia Rheumatica Diagnostic Criteria

Positive criteria

Bilateral pelvic or shoulder pain and stiffness
Symptoms lasting more than 2 weeks
ESR >40 mm/h
Stiffness lasting more than 1 hour
Age >65 years
Depression or weight loss
Bilateral upper leg or arm weakness

Negative criteria

Normal muscle enzymes (CPK, aldolase)
Absence of hand joint swelling

CPK denotes creatinine phosphokinase.

TABLE 3

Possible Differential Diagnoses of Polymyalgia

Inflammatory disorders	Endocrinopathy
Rheumatoid arthritis	Hypothyroidism
Polymyositis	
Giant cell arteritis	Neurologic disorders
Fibromyalgia	Parkinson's disease
Lupus erythematosus	
	Toxins
Infectious diseases	Quinidine
Bacterial endocarditis	Minoxidil
Lyme disease	
	Myopathies
Malignancy	Becker's muscle dystrophy
Multiple myeloma	Eaton-Lambert syndrome
	Carnitine deficiency

Lyme disease presents as migratory painful arthralgias and myalgias and is associated with a rash (erythema migrans) and occasional blurred vision. Borrelia titers will be high on serum analysis. Of note is that both Lyme disease and PMR can show elevations of the ESR >80 mm/h. Treatment for Lyme disease involves either doxycycline or amoxicillin. Falling Borrelia titers following antibiotic administration supports the diagnosis.[12] The ability of Lyme disease to mimic classic idiopathic rheumatologic entities is well known.[4]

Giant cell arteritis is frequently present along with PMR. Severe headache, jaw claudication, or hearing changes may be manifestations of GCA. Temporal artery biopsy showing granuloma infiltration of the vessel lumen will confirm the diagnosis. Immediate treatment with high-dose steroids is indicated to reduce the risk of blindness.

Once other diseases have been reasonably excluded, and a diagnosis of PMR is satisfactorily made, low-dose prednisone is the treatment of choice. The typical starting dose is 10 to 20 mg/day by mouth. Response is dramatic, and symptoms should resolve within 48 hours.[11] If symptoms do not abate, then another diagnosis should be sought. Once symptoms are controlled, the successful dose of prednisone should be maintained for 1 month before tapering. Tapering should be prolonged, with periodic checks for a rising ESR, and decreases of only 2.5 mg prednisone every 2 weeks until 10 mg is reached, then 1 mg every 6 weeks, leveling off at 5 mg/day. In one study of 13 patients with PMR and elevated CRP, the CRP fell to normal at a rate that precisely reflected the patients' clinical improvement. The ESR also fell, although at a slower rate.[13] Treatment may continue for 2 to 7 years. Nonsteroidal anti-inflammatory drugs have been tried in very mild cases with a much less dramatic response. Azothioprine may be of use in patients unable to tolerate prednisone.

The prognosis for patients with PMR is generally good, with rapid improvement being the rule and a 50% cure rate after 2 years of therapy. Those patients in whom CRP normalizes within 1 week of starting prednisone appear to have the best outcomes.[14] The prognosis is worse in patients whose initial ESR is >90 mm/h,[15] and 40% of patients require steroids for more than 4 years.[16] Prognosis is also worse in men, who seem to have an increased mortality rate from vascular disease within the first 2 years of diagnosis.[17] Recurrences are more frequent in those patients treated for less than 20 months.[15] Patients with polymyalgia rheumatica are at increased risk for developing hypothyroidism, and 6% have (or if untreated may develop) temporal arteritis requiring higher doses of steroids.[16] A positive test result for the anticardiolipin antibody suggests that a PMR patient is more likely to develop subsequent arteritis.[18]

The complications of therapy can be worse than the original disease. Fifty percent of patients with PMR experience problems on a long-term regimen of steroids, including osteoporosis-induced fractures, glucose intolerance, hypertension, weight gain, cataracts, gastrointestinal bleeding, infections, and psychosis.

CONCLUSIONS

The incidence of PMR is much lower in adults younger than 50 years of age, but it does occur. Diseases and disorders do not always meet textbook age restrictions or definitions. Physicians should, however, consider PMR after ruling out myopathy, arthropathy, and infection in any adult presenting with persistent proximal extremity muscle pain and tenderness. Associated complaints of headache or tender temples should mandate a temporal artery biopsy or a consideration of higher doses of steroids. Although the dramatic response to steroids in this patient does not completely confirm the diagnosis, it is possible that he represents the youngest patient reported to have polymyalgia rheumatica.

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