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Polymyalgia Rheumatica

Common Disease, Elusive Diagnosis

Diana R. Mager, DNP, RN-BC

Polymyalgia rheumatica (PMR) is a common inflammatory rheumatic disease with little known about its etiology or incidence. Frequently found in older adult women, this disease can be debilitating, painful, and dangerous. Diagnosing PMR can be elusive due to lack of specific laboratory tests, and treatment with use of long-term glucocorticoids can be difficult due to side effects. The following article describes the pathophysiology, diagnosis, signs and symptoms, and treatment of PMR, as well as implications for home healthcare.

Overview

Although polymyalgia rheumatica (PMR) is frequently referred to as the most common inflammatory rheumatic disease (Ameer & McNeil, 2014; Mackie et al., 2014; Matteson et al., 2012), little is known about its etiology, and reported incidence rates vary greatly. PMR is rarely seen in those under age 50, and incidence increases progressively after age 50, peaking in those aged 70 to 79; thus, the main known risk factor is age (Kennedy, 2012; Mackie & Pease, 2013; van Hecke, 2011). Approximately twice as common in women, the estimated lifetime risk of developing PMR is 2.4% for women and 1.7% for men (Mackie et al.; van Hecke). Found most commonly in Caucasian older women (Mackie & Pease) and in people of Scandinavian descent (van Hecke), PMR may occur in either gender and throughout adulthood, making awareness of the disease presentation important to home care nurses, so that atypical cases will not be missed. Often exhibiting an unpredictable course, PMR, if left untreated, may result in devastating health outcomes (Mackie et al.). However, with prompt diagnosis and appropriate treatment PMR responds well and prognosis is excellent (Ameer & McNeil).

Pathophysiology and Diagnosis

Environmental, genetic, and infectious factors may contribute to the development of PMR, though the exact etiology is uncertain (Kennedy, 2012; van Hecke, 2011). The disease is characterized by intense systemic inflammation and an imbalance between endothelial injury and repair (Pirro et al., 2012). There is no specific diagnostic

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testing for PMR, so diagnosis is derived from a combination of clinical and laboratory findings, and by excluding other rheumatologic conditions such as rheumatoid arthritis (Mackie & Pease, 2013). Unfortunately, the lack of specific testing and the painful yet vague symptoms closely resembling other diseases are factors that contribute to misdiagnosis (Ameer & McNeil, 2014; Kennedy). Although several sets of diagnostic criteria have been proposed in the past, a more recent set published by the American College of Rheumatology/European League Against Rheumatism has not yet been validated for clinical practice (Ameer & McNeil; Mackie & Pease). The lack of both a standard set of classification criteria and reliable outcome measures has created challenges to developing regimens to treat this complicated disease (Matteson et al., 2012).

There are, however, several nonspecific diagnostic laboratory tests useful for PMR diagnosis. Several acute-phase reactants such as the erythrocyte sedimentation rate (ESR), C-reactive protein (CRP), and interleukin-6 may be significantly elevated in the presence of PMR. Of these, CRP tends to be a more sensitive disease indicator than ESR (Al Rashidi et al., 2013; Mackie & Pease, 2013). An ESR > 40 coupled with the presence of related symptoms may indicate PMR (Nothnagl & Leeb, 2006). Complicating matters is that elevations of these markers also occur during infection and/or with the presence of cancer and other inflammatory diseases, necessitating the ruling out of these possible differential diagnoses. Some patients also present with normochromic, normocytic anemia, and/or elevated alkaline phosphatase (van Hecke, 2011). Additionally, patients with

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PMR may present with normal acute-phase reactants, forcing some clinicians to base diagnosis solely on symptoms and on the patient's rapid response to glucocorticoid treatment (Kennedy, 2012; Mackie et al., 2014; Schmidt & Warrington, 2011). Although treatment with low-dose prednisone is common, some researchers warn that response to steroids alone should not be used as a definitive PMR diagnosis (Ameer & McNeil, 2014; Matteson et al., 2012).

Contributing to the elusive PMR diagnosis are the unremarkable clinical findings upon physical examination. Findings often include normal muscle strength and lack of joint swelling, although there may be some restriction in range of motion in shoulders, hips, and/or neck, related to pain (van Hecke, 2011). Ultrasounds and magnetic resonance imaging are sometimes indicated to locate inflamed tissue, and to rule out other injuries (Ameer & McNeil, 2014). Given the lack of specific diagnostics, the commonly reported symptoms, and the insignificant physical findings with PMR, compiling a thorough patient history is key to confirming the diagnosis.

Signs and Symptoms

Symptoms tend to appear abruptly with PMR, and pain often presents symmetrically (van Hecke, 2011). The most common symptoms occur as persistent, severe aching pain and stiffness in the neck, shoulders, upper arms, hips, and pelvis (Mackie & Pease, 2013; Pirro et al., 2012; Schmidt & Warrington, 2011). Other symptoms include proximal muscle pain and weakness in the arms and hips (Kennedy, 2012), and in the axial joints (shoulders and hips) (van Hecke). Patients often report morning stiffness with difficulty getting out of bed, or moving from sitting to standing positions due to synovitis of proximal joints, and/or inflammation of extra-articular synovial structures (Mackie & Pease; van Hecke). Stiffness may be severe enough that patients cannot raise their arms above their shoulders to comb their hair (Ameer & McNeil, 2014). Daytime fatigue may occur due to sleep disorders related to pain (Schmidt & Warrington). Along with these painful symptoms, patients may also experience anorexia, weight loss, low-grade fever, anemia, and depression (Kennedy).

Approximately 10% to 15% of patients with PMR will also develop giant cell arteritis (GCA), a vasculitis that affects medium- to large-sized blood vessels (Ameer & McNeil, 2014; Mackie & Pease, 2013). Although it is not uncommon for a patient to have both diseases, not all patients with PMR will go on to develop GCA. Further, GCA can occur at any point during the course of PMR, either as a presenting symptom, or during the treatment phases. Similar to PMR, risk factors such as female gender, Caucasian race, and age over 50 years also contribute to the risk of developing GCA (Mackie & Pease), though the relationship between GCA and PMR is not clearly understood (Ameer & McNeil). Symptoms of GCA include a constant and severe headache with scalp tenderness. Head pain is usually temporal but may be in the frontal, parietal, or occipital areas, as well as in the eyes, ears, face, jaw, or neck with jaw claudication. Serious effects can result from untreated GCA, such as transient loss of vision, blindness, limb claudication, stroke, and other artery occlusions, making early diagnosis and treatment critical (Mackie & Pease).

Treatment

The treatment of choice for PMR is long-term use of medium-dose (15–20 mg/day) glucocorticoids (Kennedy, 2012; Mackie & Pease, 2013), due to their anti-inflammatory properties (Pirro

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et al., 2012). Success is best accomplished by balancing the need to comfortably treat symptoms while concurrently avoiding the potential adverse effects of glucocorticoids. Fortunately, PMR symptoms tend to respond rapidly to the steroids, alleviating symptoms within 12 to 48 hours (Schmidt & Warrington, 2011), and leveling acute-phase reactants within a week (Kennedy). Several weeks after initiation of steroids, patients are reassessed for clinical response and for inflammatory markers (CRP and ESR). After approximately 1 month of glucocorticoid use, doses are tapered gradually by 2.5 mg every 2 to 4 weeks (Mackie & Pease). When 10 mg/day is reached, the dose is then decreased by 1 mg/month, a regimen that takes approximately 1 year to complete (Kennedy), though the length of treatment can vary to 3 or even 5 years (van Heck, 2011).

During the glucocorticoid-tapering regimen, patients may have a relapse necessitating an increase in the dose until symptoms subside. Tapering prednisone slowly (<1 mg/month) has been associated with fewer relapses and earlier termination of steroid treatments. Relapse occurs in approximately 23% to 33% of patients (Kennedy, 2012). Overall PMR is a self-limited disease, and will eventually dissipate, at which time the patient becomes asymptomatic and steroids are discontinued (Kennedy; van Hecke, 2011). Because patients may react differently to the treatment regimen, providers must be willing to be flexible to ensure that doses are adjusted according to patient response (van Hecke).

According to Mackie et al. (2014), there is insufficient evidence regarding the efficacy of using regimens other than glucocorticoids to treat PMR. Methotrexate (MTX), a disease-modifying anti-rheumatic agent (DMARD), has been used with PMR (Kennedy, 2012; van Hecke, 2011), but study results are contradictory (Mackie & Pease, 2013; Schmidt & Warrington, 2011). However, although MTX is most often used as a chemotherapeutic agent and has only been minimally studied for use with PMR, it has been considered the most promising of the DMARDs in decreasing cumulative steroid dose and relapses (Kennedy). Despite its potential usefulness in treating PMR, MTX use must be closely monitored for adverse effects ranging from minor to life threatening. Side effects may include fatigue, dizziness, anorexia, nausea, severe bone marrow depression, renal toxicity, and increased risk for lymphoma (Joseph et al., 2014; Karch, 2014). Finally, using nonsteroidal anti-inflammatory drugs with glucocorticoids has shown no advantage over using steroids alone regarding duration of therapy (van Hecke). (See case study.)

CASE STUDY

H. Smith, a 78-year-old male with a history of chronic obstructive pulmonary disease and hypertension, reported a sudden onset of severe bilateral upper arm pain and stiffness with achy pain in the hips and shoulders. A full diagnostic work-up was completed to rule out cardiac disease, Lyme disease, and arthritis. Blood work revealed an elevated erythrocyte sedimentation rate (ESR) (76) and C-reactive protein (CRP) (1.5). Oral prednisone (15 mg) was initiated for 4 weeks, with almost immediate relief of pain and stiffness, after which a slow taper of the prednisone was initiated. Despite being on the steroid, Mr. Smith began having breakthrough arm pain and a flare-up of the polymyalgia rheumatica (PMR) symptoms when he reached a level of 5 mg/prednisone per day. Steroids were then increased to 20 mg/day for 4 weeks, and symptoms resolved until a slow taper was again initiated. This regimen of increasing and then tapering down prednisone with relapsing symptoms continued over 3 years, at which time the physician suggested a small weekly dose (7.5 mg) of oral Methotrexate (MTX). Despite mixed clinical findings about MTX efficacy in treating PMR, Mr. Smith had total symptom control within 4 weeks and, over several months, was able to tolerate a tapered prednisone dose to 2 mg/day without PMR flare-ups, and with normal to borderline high acute-phase reactants. The long-term goal is to discontinue both MTX and prednisone, and reevaluate symptoms and markers.

This case is somewhat atypical in terms of the client's gender, unusually lengthy disease course, and repeated symptom flare-ups during the prednisone taper. In this case, MTX appears to have been successful in managing symptoms and in allowing for the decrease in the glucocorticoid dose. Potential complications related to MTX such as immunosuppression, bone marrow suppression, and drug interactions were carefully monitored by clinicians, as were laboratory values such as complete blood cell counts as well as liver and kidney function tests.

Implications for Home Care

The majority of home care consumers in the United States are women over the age of 65 (National Association of Home Care and Hospice, 2010), making this population at risk for developing PMR as well as GCA. Home care nurses should be able to recognize the signs and symptoms related to these serious but treatable diseases. Given the vague nature of the PMR symptoms (stiffness and aches), patients

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and nurses alike may tend to incorrectly dismiss complaints as normal for older adults. Prompt recognition and referral to one's provider will be essential for early diagnosis and effective treatment initiation.

Patients at home with PMR may need additional help with their activities of daily living given the degree of disability caused by the pain and stiffness that accompanies PMR, especially in the morning when rising from bed. Careful patient monitoring is also essential regarding the adverse effects of corticosteroid use, including elevated blood sugar and blood pressure, as well as sleep disturbances and bone density changes over time (Kennedy, 2012). These patients will require frequent blood work to monitor ESR, CRPs, and blood sugars, and will benefit from medication education to avoid unnecessary medication errors as the doses taper over time.

Conclusions

Although PMR is challenging to diagnose, it tends to respond well to corticosteroids, quickly freeing patients from the pain and discomfort that accompany the disease. Although there is no gold standard on classifying criteria for diagnosing PMR, the disease is diagnosed by a combination of reported symptoms, exclusion of other diseases, and elevated markers such as ESR and CRP. Stiffness, pain, and other complications such as GCA may occur, rendering nursing assessment and intervention critical components for improving patient outcomes at home. ⌂

Diana R. Mager, DNP, RN-BC, is an Assistant Professor, Fairfield University School of Nursing, Fairfield, Connecticut, and a Home Care Nurse and Consultant, Visiting Nurse and Hospice of Fairfield County, Norwalk, Connecticut.

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Address for correspondence: Diana Mager, DNP, RN-BC, 1073 North Benson Rd., Fairfield, CT 06824 (dmager@fairfiel.edu).

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