Polymyositis: Essential Information for Primary Care Providers

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ABSTRACT

Polymyositis is one of a rare group of skeletal muscle diseases known as idiopathic inflammatory myopathies. The etiology is not fully understood, and its clinical presentation is often vague yet similar to more common neuromuscular diseases, making diagnosis difficult. A number of different tests are available to assist providers in making an accurate diagnosis. Once a diagnosis is made, there are a number of various treatment modalities available. Nurse practitioners must be familiar with treatment protocols and follow-up. The focus of this article is on polymyositis; its presentation, signs, and symptoms; the process of accurate diagnosis; and common treatment strategies.

Keywords: Bohan and Peter criteria, electromyography, elevated creatinine kinase, idiopathic myopathies, muscle biopsy, polymyositis, proximal muscle weakness

Idiopathic inflammatory myopathies (IIMs) include a rare group of acute, chronic, and subacute acquired diseases of skeletal muscle characterized by moderate to severe muscle weakness and inflammation.[1] There are 3 major classifications of inflammatory myopathies: dermatomyositis (DM), polymyositis (PM), and inclusion body myositis (IBM). All of these myopathies have a similar and often vague set of signs and symptoms at initial onset, making diagnosis rather difficult. Patients suffering from symptoms associated with myositis initially present to their primary care provider's office, and in today's world many people use a nurse practitioner (NP). After diagnosis, the NP may be responsible for monitoring the care of these patients; therefore, it is imperative for NPs to recognize the symptoms of this disease and be familiar with the diagnostic and treatment processes. The purpose of this article is to define polymyositis; its presentation, signs, and symptoms; the process of making an accurate diagnosis; and common treatment strategies.

OVERVIEW

IIMs are a group of systemic rheumatic autoimmune disorders characterized by inflammation of the skeletal muscle, muscle weakness, muscle fatigue, and often pain.[2] Along with muscles, organs can also be affected, including the skin, cardiac, gastrointestinal, and pulmonary systems. The IIM group of disorders is not anything new to the field of medicine. They were first described in the literature more than a century ago as generalized muscle disorders primarily affecting the muscles of the trunk and the proximal limbs.[2] In 1903, Steiner[3] made a clear delineation between idiopathic polymyositis and other forms of myositis that are caused by bacteria and/or parasites. In the 1940s, it was recognized that polymyositis may occur without cutaneous lesions, muscle pain, or any other constitutional symptoms.

The 3 most common disorders within the group of IIMs include PM, DM, and IBM. PM was initially defined as symmetric proximal myositis with the absence of histopathologic signs of other myopathies. DM is characterized by muscle weakness and the presence of certain types of skin rash. The muscle involvement of PM is clinically indistinguishable from that of DM, but PM and DM differ histologically. IBM is typically characterized by its later age of onset and refractory nature to any treatment.

PM and DM are rare disorders that have been found worldwide and are thought to be increasing in frequency. The current incidence of PM and DM is

approximately 2-10 cases per 1,000,000 population per year.[4] Both PM and DM have a peak prevalence in childhood (7-15 years old) and also in midlife (30-50 years old). IBM differs in that the peak prevalence is later in life (after the age of 50). There is approximately a 3:1 female predominance in both PM and DM. Individuals of African and Latino descent tend to be at a higher risk for developing an IIM and tend to have poorer outcomes when compared with those of European descent.

ETIOLOGY/PATHOGENESIS

The cause of PM and other IIMs remain unknown, but evidence suggests it is autoimmune in origin. There are a number of factors that are believed to be involved, but genetic and environmental factors are considered the most likely culprits. "Anecdotal clusterings in time and space suggest environmental influences on polymyositis and dermatomyositis, but the specific agents remain elusive."[4(p1716)] Common environmental agents connected to polymyositis include both infectious agents and noninfectious agents. Select viral and bacterial infections have been identified, and certain drugs, cytokines, dietary supplements, vaccines, medical implants, and occupational exposures have been implicated in case reports and some epidemiologic studies.[4]

Nagaraju and Lundberg[2] suggest there is "an association with immune response genes and occasional reports of familial clustering that supports the role of genetic factors in these diseases."[(p1404)] Like other autoimmune diseases, PM is a complex disorder involving human leukocyte antigen immune response genes, non–human leukocyte antigen immune response genes (eg, cytokines, tumor necrosis factor receptor, interleukin 1, and tumor necrosis factor receptors), complement components (eg, C4 and C2), immunoglobulin allotype, and T-cell receptors.[2] The paucity of information regarding the genetic contribution to this disease is partially because of its rarity, the small number of subjects in any study, and the heterogeneity in the disease phenotype.

CLINICAL PRESENTATION

Although most patients with PM typically present with muscle fatigue and weakness, it is a connective tissue disease and can involve other body systems; therefore, a full history and physical examination is imperative. The most predominant symptoms of PM are muscle weakness and impaired muscle endurance. The classic initial clinical finding of PM is the slow, progressive development of a symmetric proximal muscle (eg, neck, pelvic, thigh, and shoulder muscles) weakness that develops over weeks to months.[2,4,5] Typically, patients report increasing difficulty with everyday tasks, such as rising from a seated position, climbing stairs, walking uphill, lifting objects, and working with arms above their head.[4,5] Patients generally experience more problems with performing repetitive movements than with single strength exercises. The muscle weakness progresses slowly if untreated, and, in the most severe cases, patients may become wheelchair dependent.[4] In addition to weakness, some patients have also experienced chronic aching pain in their proximal muscle groups.

Patients with PM often experience joint pain and arthritis. The most common form of arthritis is symmetric arthritis in the small joints of the hands and feet.[5] It is typically nonerosive; however, in rare cases, it has been found to be erosive and destructive.[4,5] Some patients may also experience general symptoms, such as fatigue, weight loss, fever, and Raynaud phenomenon.

Skin involvement is typically seen only in DM, although there is one particular skin pathology found with PM known as mechanic's hands. This rash is a hyperkeratotic, scaling, and fissuring of the fingers, particularly along the radial side of the index fingers.[2,5]

As the disease progresses, additional muscle groups are affected. Dysphagia and poor nutrition may occur because of impaired contractility of the tongue and pharyngeal musculature.[2] Damage to the pharyngeal musculature can lead to aspiration and potential pneumonia.[4,5] Other striated muscles that may be involved include the lower esophageal musculature, causing gastric reflux, or the sphincter ani, causing fecal incontinence.[2] Constipation, diarrhea, and stomach pain are other common gastrointestinal symptoms that may result from disturbed motility of the gut and/or inflammation of the gastrointestinal tract.[2] There are rare cases in which patients develop difficulty with breathing related to weakness of the diaphragm and thoracic muscles. If the disease