Polymyositis, Dermatomyositis, and Autoimmune Necrotizing Myopathy: Clinical Features

Sabiha Khan, MD[a], Lisa Christopher-Stine, MD, MPH[b,*]

KEYWORDS

• Myositis • Polymyositis • Dermatomyositis
• Inflammatory myopathy

Idiopathic inflammatory myopathies (IIMs) are a heterogeneous group of autoimmune disorders predominantly affecting skeletal muscles, resulting in muscle inflammation and weakness. Along with the musculoskeletal manifestations, involvement of other organ systems is seen, including the skin, cardiac, gastrointestinal, and pulmonary systems. The 3 most common inflammatory myopathies are polymyositis (PM), dermatomyositis (DM), and inclusion body myositis (IBM). Several much rarer syndromes are also described under the broad spectrum of inflammatory myopathies.

This review details the clinical findings noted in PM, DM, and the emerging entity of autoimmune necrotizing myopathy.

The Bohan and Peter[1,2] criteria have been traditionally used to define and diagnose PM and DM. IBM was not part of the original classification but has been included over time. The Bohan and Peter criteria combine clinical, laboratory, and pathologic features to define PM and DM. More recently, newer criteria have been proposed for the classification of myositis but are not yet widely accepted.

EPIDEMIOLOGY

There is a variation in the age at which PM and DM occur. Although DM has a bimodal incidence pattern, peaking during childhood and then again between 50 and 70 years

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[a] Division of Rheumatology, Department of Medicine, Johns Hopkins University School of Medicine, 5200 Eastern Avenue, Mason F. Lord Center Tower, Suite 4100, Baltimore, MD 21224, USA
[b] Division of Rheumatology, Department of Medicine, Johns Hopkins University School of Medicine, 5200 Eastern Avenue, Mason F. Lord Center Tower, Suite 4500, Baltimore, MD 21224, USA
* Corresponding author.
E-mail address: lchrist4@jhmi.edu

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of age,[3] PM is rare in childhood and occurs mainly after the second decade of life.[4] Both conditions are more common in women. The reported incidence of both PM and DM is 4 to 10 cases per million population per year.[5]

DEFINITION

The most commonly used criteria for the diagnosis and classification of PM and DM are those defined by Bohan and Peter: (1) symmetric proximal muscle weakness; (2) elevation of serum skeletal muscle enzyme levels, including creatine kinase and aldolase; (3) electromyographic (EMG) evidence of the classic pattern of muscular impairment, with polyphasic, short, small motor unit potentials, fibrillation, positive sharp waves, increased insertional irritability, and repetitive high-frequency discharges; (4) muscle biopsy specimens with typical histopathologic findings of degeneration, regeneration, necrosis, and interstitial mononuclear infiltrates; and (5) characteristic cutaneous manifestation of DM, including heliotrope rash or Gottron sign (**Box 1**).[1,2] As mentioned earlier, over time, IBM has been included in these criteria as its own entity.

MUSCULAR MANIFESTATIONS

The classic clinical finding of both PM and DM is the progressive development of symmetric proximal muscle and truncal weakness that develops relatively slowly, over the course of weeks to months. Patients usually report progressive difficulty with everyday tasks requiring the use of proximal muscles, such as rising from a chair, climbing steps, lifting objects, or combing their hair. Fine motor movements that require the use of distal muscles, such as buttoning shirts and writing, are affected only late in the course of the disease, and when found early in the illness, these affected movements should prompt a search for another neuromuscular disorder.[6] Facial muscles remain unaffected; however, pharyngeal and respiratory muscles can become affected, resulting in complications that are discussed in detail later in this review.[6] Often erroneously thought to be painless diseases, both PM and DM may have associated myalgias and muscle tenderness early in the disease course, more often seen in DM.

The exception to this pattern of muscle involvement is amyopathic DM (ADM), in which patients have the classic dermatologic findings of DM without the accompanying muscular findings.[7]

Box 1
The Bohan and Peter classification criteria

1. Symmetric proximal muscle weakness
2. Elevation of skeletal muscle enzyme levels
3. Abnormal EMG results[a]
4. Muscle biopsy abnormalities[b]
5. Typical skin rash of DM[c]

[a] Polyphasic, short, small motor unit potentials; fibrillation; positive sharp waves; insertional irritability; and bizarre, high-frequency, repetitive discharges.
[b] Degeneration/regeneration, perifascicular atrophy, necrosis, phagocytosis, fiber size variation, and mononuclear inflammatory infiltrate.
[c] Gottron sign and heliotrope rash.