



Updates in Diagnosis and Managements of Polymyositis: Simple Review

**Laila Abdullah S. Alanazi^{1*}, Abdullah Mohammed A. AL Madshush¹,
Omniyyah Mohammed S. Alatawi¹, Asmaa Hamed M. Albuhairey¹,
Jomana Khalid M. Aljohani¹, Asmaa Muslim Alfahimani¹,
Jalawi Talal A. Alotaibi¹, Abrar Atallah O. Alatawi¹,
Mohammed Ibrahim F. Bin Ibrahim¹, Saud Mohammed S. Alrofydi²
and Marwan Fahad H. altemani³**

¹University of Tabuk, Saudi Arabia.

²King Khalid University, Saudi Arabia.

³King Salman Armed Force Hospital, Saudi Arabia.

Authors' contributions

This work was carried out in collaboration among all authors. All authors read and approved the final manuscript.

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ABSTRACT

Idiopathic inflammatory myopathies (IIMs) includes an unusual group of acute, chronic, and subacute developed diseases of skeletal muscle characterized by moderate to severe muscle weakness and inflammation. Polymyositis is generally considered to be a prototypic T cell-mediated autoimmune myopathy, while DM was traditionally associated with a humoral-driven microangiopathy, though the putative autoantibodies and their targets have yet to be identified, and there is increasing evidence implicating the type I interferon pathway in the pathogenesis of the disease. Women between the ages of 50 and 70 are the most typically affected. Proximal muscular weakness is the most common clinical symptom. Inflammatory arthritis, Raynaud's phenomenon, myocarditis, and interstitial lung disease are all examples of extramuscular involvement. In this review, we overview updates in diagnosis and managements of polymyositis.

*Corresponding author: E-mail: Laila16ae@gmail.com;

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1. INTRODUCTION

Idiopathic inflammatory myopathies (IIMs) includes an unusual group of acute, chronic, and subacute developed diseases of skeletal muscle characterized by moderate to severe muscle weakness and inflammation. [1] Inflammatory myopathies are mainly dermatomyositis (DM) and polymyositis (PM). Polymyositis causes muscle weakness, usually in the muscles closest to the trunk of your body. Dermatomyositis causes muscle weakness, plus a skin rash. Doctors may use a physical exam, lab tests, imaging tests and a muscle biopsy to diagnose myositis [2, 3] All of these myopathies have a similar and often vague set of signs and symptoms at initial onset, making diagnosis rather difficult.

Although the clinical presentation of DM and PM is similar, it usually manifests as sub-acute painless proximal limb weakness plus distinctive skin manifestations of DM, immunopathological observations, and more recent gene expression and proteomic analysis proved convincingly different from each other in terms of the underlying pathogenesis mechanisms. Recently, an alternate classification system was presented, based on the main histological characteristics of each condition [4].

Polymyositis is generally considered to be a prototypic T cell-mediated autoimmune myopathy, while DM was traditionally associated with a humoral-driven microangiopathy, though the putative autoantibodies and their targets have yet to be identified, and there is increasing evidence implicating the type I interferon pathway in the pathogenesis of the disease.

1.1 The Epidemiology of PM and DM

As documented by numerous study organisations, the incidence of IIMs is fairly low. In infancy polymyositis is rare and generally occurs after the first 10 years of life. The most common presentation period is 45-60 years old. DM affects both children and adults with an overall female/male ratio of about 2:1 [5]. In the last twenty years, epidemiological studies have shown a higher incidence and prevalence than historically reported. This may be because of

more detailed criteria for diagnosis and better patient access and services.

It should be noted that, depending on changes in study methods, the reported prevalence and incidence of PM and DM vary widely, but female sex and urban life appear to be coherent risk factors.

2. CAUSES AND RISK FACTORS

The causes of PM and DM remain uncertain, while the origin has involved both an autoimmune process and hereditary as well as environmental variables. There are several variables that can pose risks of polymyositis; hereditary factors is well accepted that autoimmunity results from susceptible genes, environmental factors and a dysregulated/dysfunctional immune system [6]. Many previous case reports, family studies, studies of animal models, candidate gene case-control studies, and whole genome investigations supports a great role for genetic factors in the etiology of autoimmune disease and specially PM [7].

In the 1950s, the earliest documented examples of family IIM were described and there is a scarcity of affected sibling pairs and twins. [8, 9] Candidates for genetic investigations have found common genetic sensitivities with other autoimmune illnesses [10, 11]. To date the Major Histocompatibility Complex (MHC) has been revealed to be the strongest genetic associations with IIMs.

Also environmental factors In recent years, evidence has shown that environmental factors play also play a role in the development of autoimmunity. Environmental factors include infection, gut microbiota, drugs, chemicals, pollutants and physical agents [12,13]. Animal models of myositis have been developed that are induced by viruses, drugs, or parasites, providing additional evidence for the likely role of environmental agents in the pathogenesis of IIMs [14].

Environmental factors also were discussed in recent years and there have been suggestions that environmental variables also play a part in autoimmune development. Infections, gut

microbiota, medicines, chemical substances, pollutants and physical agents are examples for environmental factors [12, 13]. Animal models of myositis that are induced by viruses, drugs, or parasites, providing additional evidence for the likely role of environmental agents in the pathogenesis of IIMs [14].

2.1 Clinical Considerations

The presence of PM as an isolated object has been through vigorous debate, especially since the separation of IBM from PM [15, 16]. The controversy discussed was whether PM can occur as a primary musclespecific disease or just as part of a more widespread connective tissue disease or autoimmune disorder, considering their recurrent coexistence.

Although isolated PM was reported to be more common than was previously reported [16], several studies have found that it is not as common as DM and IBM [17,18] except in Japan where PM outnumbers the other varieties of IIM [18,19].

PM usually expresses as a painless proximal myopathy with subacute onset and is usually responsive to corticosteroids and immune suppression. It occurs predominantly in adults but can also occur in children infrequently, although it is significantly less frequent than juvenile DM [20]. Besides this basic phenotype, a distal variant, predominantly in the upper limbs and strong therapeutic response, has been reported [21]. Other restricted versions of the paraspinal muscles include those with dropped head or camptocormy [22,23].

Both PM and DM patients present with a varying degree of muscle weakness, typically emerging slowly over several weeks to months, but can be acutely in rare cases [24]. The weakness is fairly symmetric, mainly proximal and unassociated with sensory loss or ptosis with sparing of extraocular muscles which are characteristics of myasthenia [24]. Distal muscle weakness develops in the late stages of PM and DM with affection of motor movement.

In contrast, this feature is an early and prominent finding in sIBM [25]. In PM and DM, the neck extensor muscles may also be involved, causing difficulty in holding up the head and rarely causing a dropped head syndrome (DHS) [26].

Primary weakening of the diaphragm and muscles of the accessories, especially in advanced cases of pharyngeal muscles, can contribute to air failure, or dysphagia, nasal discourse and heart attacks and nasal regurgitation and pneumonia of aspiration [27,28]. The tendon reflexes are normally retained but are not present in seriously atrophied or weaker muscles. In patients with polymyositis, myalgia is occurring in fewer than 30% [29].

PM has often been misinterpreted as a single clinical characteristic is lacking and exclusion is nevertheless identified [30, 31]. PM is best defined as a proximal myopathy subacute that develops muscle weakness for weeks or months, is associated with adults but seldom has an impact on children and excludes people with rash, neuromuscular family history, exposure of myotoxic medicines (e.g. statins, penicillamine, and zidovudine). PM mimics many other myopathies and may also be diagnosed incorrectly in cases of DM, sIBM, NAM, overlap syndrome associated with a connective tissue disease, muscular dystrophies, myalgia syndromes, or toxic and endocrine myopathies [31, 32, 33].

3. DIAGNOSIS AND CLASSIFICATION

According to the Bohan and Peter diagnostic criteria, muscle strength and/or weakness should be the first step in determining a diagnosis. Muscle strength can be measured in several ways but the patient is asked to sit or squat without support in a fast and straightforward test of weakness in proximal leg muscles. [34, 35, 36] Early in the course of the disease, it can be achieved by patients with a minor difficulty, but as the disease develops, patients will not be able to get up without pulling their arms or lying forward in order to gain momentum [34].

Patient with or without skin rash should be suspected of having PM or DM if they are complained of muscle weakness, fatigue and myalgia. PM/DM criteria excluding neuromuscular, endokrinous, or neurogenic illnesses, myotoxic exposure to medications, musculoskeletal dystrophies, and metabolic myopathy, SIBM, NOM or infection [2, 37, 38]. These criteria are not ideal and sometimes do not rule out IBM because of studies with small patient cohorts and perhaps wrong illness classifications. The pathologic characteristics of

existing criteria were added in 2003 by Dalakas and Hohlfeld [30].

First, the important role of the muscle biopsy test was repeated and the following was proposed: Primary CD8/MHC-I complex inflammation and no definitive PM vacuoles, omnipresent MHC-I expression, but no CD8⁺ cell infiltrates or vacuoles for likely PM, peripascular, perimysial or perivascular infiltrates, perifascular atrophy and rash for specific DM; no likely DM rash present. Additionally, ADM is diagnosed if there is an acute rash, but biopsy findings are unspecific or labelled for DM and there is no weakening.

In 2003, the MSG and ENMC international workshop, many neurologists, rheumatologists, and statisticians worked together to suggest classification criteria for IIMs [3]. This workshop provided information on how to use these criteria to each category of myositis, including definite and probable PM/DM, ADM, possible DM sine dermatitis, non-specific myositis and NAM. At the same time, this workshop also pointed out the unmet needs in treatment due to difficulties in the study design and the low incidence and prevalence of patients. The workshop also promoted the development of valid, sensitive, and reliable outcome measures for (randomized controlled trial) RCTs in myositis.

Lately, muscle immunopathology, myositis specific autoantibodies testing, and new methods of muscle imaging such as contrast-enhanced ultrasound or Magnetic Resonance Imaging (MRI) have been used in the diagnosis of patients with IIM, contributing to enhance diagnostic skill [39].

The Myositis Activity Profile (MAP) and an outcome measure core set established by the IMACS group, which assesses disease activity, damage, and quality of life, are two newer evaluation methods. [40, 41] The IMACS outcome measures have not yet been validated in longterm trials, but they may still be useful for tracking PM improvement. [34] Alexanderson et al [8] conducted a study to assess the MAP's measuring properties in adult PM patients. They came to the conclusion that the MAP has potential measurement capabilities for assessing constraints in daily activities and will be utilised more frequently in the future [41].

In order to properly evaluate for PM, laboratory data is required in addition to muscle strength testing. Measuring serum levels of skeletal

muscle enzymes is one of the most important laboratory tests. A high amount of muscle-derived enzymes indicates that muscle injury is ongoing. Alanine aminotransferase, aspartate aminotransferase, lactic acid dehydrogenase, creatinine kinase (CK), and aldolase are examples of these enzymes. In up to 75% of individuals with PM, any combination of these enzymes may be increased; consequently, each of these enzymes should be examined to increase the odds of detecting an abnormality [42].

The primary stage in the assessment of these patients has usually been the measurement of CK. It appears to be the most sensitive and specific predictor of muscle fibre injury when compared to other blood muscle enzymes. [34] It's worth noting that significant muscular dysfunction can occur with little to no enzyme increase, necessitating the use of additional PM criteria and methodologies. [43] Myoglobin in the blood is a sensitive indicator of muscle fibre membrane integrity, making it a helpful tool for assessing disease activity.

The serum myoglobin assay has the disadvantage that patients with myositis frequently exhibit a large range of myoglobin values due to circadian fluctuation. [34] Serum myoglobin is likewise more difficult to come by than serum CK.

3.1 Differential Diagnosis

When contemplating the diagnosis of PM, a comprehensive list of differential illnesses must be ruled out. There are a number of myopathies that are similar to PM, but their responses to therapeutic treatment might vary greatly. The differential diagnosis of polymyositis and dermatomyositis (PM/DM) includes: Other idiopathic inflammatory myopathies (inclusion body myositis and immune-mediated necrotizing myopathy), Drug-induced myopathy, Motor neuron disease, Myasthenia gravis, Lambert-Eaton syndrome, Hypothyroidism, Muscular dystrophy, Myotonic dystrophy, Amyotrophic lateral sclerosis, Amyloid myopathy, Sarcoid myopathy and Diabetic amyotrophy [34,42].

3.2 Treatment

The goal of PM treatment is to reduce inflammation in the afflicted muscles, relieve symptoms, and restore muscular strength and endurance. The use of high-dose corticosteroids

to reduce inflammation in the muscles is the first line of treatment [44].

It has been found that corticosteroids alone do not work for up to 50% of patients, and stopping corticosteroids without further immunosuppressive medication is linked to illness return within a year [34].

Nonglucocorticoid immunosuppressants such as methotrexate (MTX) and azathioprine (AZA) may be added to glucocorticoid therapy in severe disease or may be used in place of steroids to reduce morbidity associated with steroid use. [34, 35] Lazarou and Guerne⁶ report observing the induction of full remission with no or minimal glucocorticoid use when adequate immunosuppressant medications were introduced early. There have been no randomized placebo-controlled prospective studies completed on the effectiveness of MTX. Dosing of MTX typically starts at 15 mg/wk and can be cautiously increased to 50 mg/wk. [34, 45] There are concerns with the long-term use of MTX, such as hepatic toxicity and the potential for pulmonary toxicity. These are not absolute contraindications; however, patients taking MTX must be carefully monitored [45].

AZA is another immunosuppressant commonly used to treat PM. Nagaraju and Lundberg [34] found in their double-blind, placebo-controlled trial that the combination of AZA and glucocorticoids compared with prednisone alone was associated with better functional ability and a lower requirement for prednisone after 1 and 3 years. [34, 35] The recommended dosing of AZA is 2 mg/kg/d. In patients with refractory PM, the combination of AZA and MTX has been shown to be more successful in some cases than AZA alone. [34] An additional treatment option for PM is intravenous immunoglobulin (IVIG). When combined with corticosteroid therapy, IVIG has been shown to improve muscle strength and reduce serum CK levels as found in a Cochrane systematic review by Gordon [46]; however, there is insufficient evidence supporting the use of IVIG as a first-line treatment. IVIG is often used in combination with oral corticosteroids or other immunosuppressive drugs. Availability and cost may also deter its use. Emerging treatment protocols have shown promise in the treatment of PM. Immunosuppressants, such as tacrolimus, have recently been shown to be effective in the treatment of PM when used in combination with corticosteroids. [47] The coadministration of tacrolimus with prednisone appears to be the

best tolerated first-line therapy when compared with prednisone alone regarding therapeutic effects and safety [46,47].

Additionally, the initiation of tacrolimus may also lead to a rapid reduction of prednisone, which is particularly important for patients with various complications associated with corticosteroid use in the long-term. [47] The patient's rheumatologist will monitor the majority of these medications, but it is important for NPs, as the primary care provider, to recognize medications these patients may be prescribed along with potential side effects and drug-drug interactions.

monitoring medication side effects and the improvement of symptoms is very important in the follow up period. Routine monitoring of muscle enzymes, typically CK, is one of the easiest methods for measuring improvement; however, it is possible for CK levels to normalize without any clinical improvement. [34] As a result, it is essential to make sure the patient's muscle strength and endurance correlate with the normalizing CK levels. There are a number of muscle strength tests, as noted earlier in this article, that can be used to measure a patient's improvement.

3.3 Prognosis

In 2012, a study of 160 p.m./DM patients in the United States found a 62 percent 10-year survival rate [48]. Cardiovascular (22%) and pulmonary (22%) problems, infections (15%), and cancer (11%) are the leading causes of death [48]. Gender, age at diagnosis, presence of Raynaud phenomenon, ILD, dysphagia, respiratory muscle involvement, and heart involvement affect prognosis at any point in the clinical course [49], but the predictive impact of autoantibodies requires more long-term research. Long-term data on JDM is still lacking.

Adults with JDM experienced lower quality of life and fitness, as judged by maximal oxygen uptake as a marker of muscle function, as compared to age-matched controls [50,51].

Only a few studies have looked at short- and long-term effects in PM/DM [52,53]. Previous studies have found a wide range of PM/DM remission rates, ranging from 25% to 70% of patients [54, 55, 52, 53]. In a study of 30 patients with PM/DM, Uthmanet al. [53] found that up to

77 percent of patients obtained PM/DM remission.

Most other series, on the other hand, have emphasised a less fortunate end. In fact, only 40% of 77 PM/DM patients obtained PM/DM remission, with 43 percent improving and 17 percent worsening their clinical state [56]. Another recent study of 131 PM/DM patients found that 20% of patients stayed in remission and were off medicines after a median follow-up of 5 years, whereas the other 80% had a polycyclic or chronic, continuous course [57].

4. CONCLUSION

PM is a striated muscular inflammatory condition that is idiopathic. Women between the ages of 50 and 70 are the most typically affected. Proximal muscular weakness is the most common clinical symptom. Inflammatory arthritis, Raynaud's phenomenon, myocarditis, and interstitial lung disease are all examples of extramuscular involvement. During periods of active disease, serum muscle enzymes are frequently increased.

In the serum of PM patients, a range of autoantibodies are frequently identified. On EMG and muscle MRI, certain anomalies are frequently identified. Muscle biopsy is used to get a definitive diagnosis. Although corticosteroids are the basis of treatment, a variety of other immunomodulatory medications are also utilised to treat this condition. Although the majority of individuals respond to treatment, long-term muscular injury is not uncommon.

5. THE ASSOCIATION WITH MALIGNANCY

Symptoms of PM/DM can be a signal of developing cancer. Known risk factors for cancer in patients with PM/DM are older age, male gender, dysphagia, skin necrosis, cutaneous vasculitis, rapid onset of the disease, elevated creatinine kinase (CK) and C reactive protein (CRP), and an increase in the erythrocyte sedimentation rate (ESR).

DISCLAIMER

The products used for this research are commonly and predominantly use products in our area of research and country. There is absolutely no conflict of interest between the authors and producers of the products because we do not intend to use these products as an avenue for

any litigation but for the advancement of knowledge. Also, the research was not funded by the producing company rather it was funded by personal efforts of the authors.

CONSENT

It is not applicable.

ETHICAL APPROVAL

It is not applicable.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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