Dermatomyositis - Presentation, Diagnosis, and Management


1Dermatology Consultant, King Fahad General Hospital, Jeddah, Saudi Arabia
2King Fahad General Hospital, Jeddah, Saudi Arabia
3University of Hail, Hail, Saudi Arabia
4Medical University of Warsaw, Poland
5Medical University of Lodz, Poland
6Umm Al-Qura University, Mecca, Saudi Arabia
7Taibah University, Al Madinah, Saudi Arabia
8Almaarefa University, Riyadh, Saudi Arabia
9Alfarabi Colleges, Riyadh, Saudi Arabia
10Prince Sultan Military Medical City, Riyadh, Saudi Arabia
11King Saud Bin Abdulaziz University for Health Sciences, Riyadh, Saudi Arabia

*Corresponding Author: Ahmed Abdulfattah Darwish, Dermatology Consultant, King Fahad General Hospital, Jeddah, Saudi Arabia.

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Abstract

Introduction: Dermatomyositis (DM) is a disorder of skin and muscles, which is a chronic inflammatory in nature. It is commonly considered as an autoimmune disease. However, the etiopathogenesis of the disease still remains unidentified. An increased risk of associated malignancy is seen in patients suffering from DM by about 3 to 8-fold as opposed to that of the general population. It is important that these patients are diligently evaluated at the time of diagnosis to correctly identify the stage of the disease.

The Aim of Work: The study is aimed to assess the knowledge and practices regarding the presentation, diagnosis, and management of the disease.

Methodology: The review is comprehensive research of PUBMED from the year 1975 to 2012.

Conclusion: In this review, the clinical presentation, diagnosis, management of Dermatomyositis has been summarized. Adequate knowledge regarding the stage and severity of the disease will help to lessen the incidence of severe complications. Important clinical manifestations can shed light on the systemic involvement or potential to turn into a malignancy. However, the optimal treatment for the condition remains questionable. Research continues in the field of susceptibility, heterogeneity, and progression of the disease.

Keywords: Cutaneous Manifestations; Skin Disease; Topical Corticosteroids; Autoimmunity

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Introduction

Dermatomyositis (DM) is one of the inflammatory idiopathic inflammatory myopathies [1-3]. Their etiology still remains uncertain and belongs to a heterogeneous group of rare systemic disorders.

Their usual characteristics include [3]:

- Subacute onset of symmetrical proximal muscle weakness,
- Common involvement of other organ systems such as lung and skin,
- Strong association with autoantibodies, and
- Responsiveness to immunosuppression.

Skeletal muscle diseases like polymyositis (PM), and inclusion body myositis (IBM) share many common features with dermatomyositis (DM) but are different in terms of presentation and pathophysiology [4]. As opposed to DM and PM, patients with IBM are not very responsive to immunosuppressive therapies and rarely have other extra muscular involvement or autoantibodies [4].

The term 'dermatomyositis' was coined in 1891 by Unverricht describing patients with inflammatory myopathic patients with dermatological findings [5].

The involvement of extra muscular organs like skin, heart, lungs and gastrointestinal tract may be seen with dermatomyositis [3].

Another type of DM, named amyopathic DM (ADM, or dermatomyositis sine myositis), is a similar disorder seen in patients who have distinctive skin findings as that of DM without weakness or abnormal muscle enzymes.

The most commonly used diagnostic criteria for this disease was given by Bohan and Peter in the year 1975. They have suggested a set of criteria that can help to diagnose and classify Dermatomyositis and Polymyositis [6].
**Epidemiology**

Dermatomyositis and Polymyositis has a worldwide occurrence, affects all races, and usually has a bimodal pattern of incidence. The first one is seen in childhood (5 - 14 years of age) and between 50 - 70 years of age. Women are usually affected 2 - 3 times more than men [3,7].

**Clinical presentation**

Clinical manifestations of dermatomyositis can be divided into [1]:

1) Cutaneous Manifestations
2) Muscle diseases
3) Systemic features
4) Myositis and malignancies

**Cutaneous manifestations**

Dermatomyositis has to distinguish pathognomonic cutaneous features, which are the heliotrope rash and Gottron's papules.

The heliotrope rash is violaceous to dusky erythematous, with the presence or absence of edema in proportion, which involves the periorbital skin. Rarely the heliotrope manifests as dilated veins of the eyelids. Sometimes these eruptions may be overlooked in darker skin tones [1,7].

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Gottron’s papules or signs are erythematous macules and patches over the metacarpophalangeal joints, the proximal interphalangeal joints, and the distal interphalangeal joints. They are commonly seen covering the elbows, knees, and feet. They can vary from a slight to thick psoriasiform scale [1,7].

Some other features like poikiloderma (the combination of atrophy, dyspigmentation, and telangiectasia), alopecia, malar erythema, violaceous erythema on the extensor surfaces such as the back of the neck are often present. Likewise, facial erythema, cuticular and periungual changes can be seen.

Scalp involvement with psoriasiform dermatitis is also seen. Oral findings such as gingival telangiectasia and angiokeratomas are seen in patients affected with Dermatomyositis during childhood [1].

Figure 3: Heliotrope eruption with periorbital changes [1] Gottron’s Papules [1].

Figure 4: Telangiectatic macule and papule [7].

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Patients with Dermatomyositis-sine myositis (ADM) have no muscle weakness, and serum enzyme levels are normal. They have abnormal muscle images in ultrasound, magnetic resonance imaging (MRI), or muscle biopsy [1].

Sometimes it is difficult to distinguish between patients of dermatomyositis and those of Subacute cutaneous Lupus Erythematosus. Lesions occur more over the bony prominences and are associated with severe pruritis in the former, whereas lesions are usually between the knuckles and are asymptomatic in the later. In dermatomyositis the distribution of the lesion on the face is around commonly around the nasolabial folds and eyelids as opposed to the later [1].

Exfoliative erythroderma, vasculitis, erosive lesions, and vesiculobullous lesions, an eruption that simulates pityriasis rubra pilaris are other rare cutaneous manifestations.

In children calcinosis of the skin/muscle/fascia, insulin resistance and lipodystrophy are also seen [7].

**Figure 5:** A Mechanic Hands B. Cuticles with hemorrhages c. Multifocal alopecia [7].

**Muscle diseases**

Variable degrees of muscle weakness are seen in Dermatomyositis, which are insidious in onset and worsens progressively over a period of time. However, cases of rapid progression of disease can also be seen [8]. The muscle involved presents itself in asymmetric and typical manner, and in the later course of the disease, distal muscle weakness is seen. Patients have trouble performing activities such as lifting objects, climbing stairs, getting up from chair, washing the hair. Difficulty with manipulating and holding objects can be seen due to distal muscle involvement. Patients can be seen with a head drop if there is involvement of the extensor muscles of the neck. If the striated muscles of the upper esophagus or pharynx are involved, difficulty in swallowing and symptoms of aspiration can be seen. Dysphonia, dysphagia, and weakening of respiratory muscles are seen with rapid progression of the disease and could be associated with poor prognosis [1]. About one-third of the patients present with myalgias and muscle tenderness [8].

**Systemic features**

Dermatomyositis is a multiorgan disorder in about 20% of the patients with inflammatory myopathy, arthralgia or arthritis is seen. The joints of the wrists, hands, and ankles are affected associated with morning stiffness [7].

Dysphagia (proximal or distal) is seen in 15 - 50% of the patients. The difficulty of swallowing is directly related to the severity of muscle involvement. Pulmonary diseases are common in patients with esophageal dysfunction and seen in 15% - 65% of patients. Hypoventilation or aspiration can be because of dysphagia or opportunistic infections or drug-induced hypersensitivity pneumonitis [7].

Cardiac involvement is not commonly seen in these patients, but if it is present, the prognosis is poor. Less frequently seen are congestive heart failure, pericarditis, and valvular disease. A radiographic examination can reveal calcification of muscles. This manifestation is however rare [7].

**Myositis and malignancy**

In 2001 the relationship of dermatomyositis to malignancy had been confirmed. It can vary in occurrence from 6 to 65% [11]. Malignancies can occur before during or after the onset of the disease. Apart from lung, pancreatic, stomach, colorectal cancer, gynecologic, or ovarian malignancies can be seen commonly. These are usually seen in the older age group [1].

**Diagnosis**

The scope and nature of the disease are determined by the initial diagnostic evaluation. This will help to confirm the diagnosis and exclude other disorders with similar manifestations. After a careful history and physical examination, a suitable laboratory testing is chosen.

Symmetric proximal muscle weakness and elevated muscle enzymes are the characteristic clinical and laboratory finding. The presence of Myositis-specific or myositis-associated antibodies is seen. The cutaneous findings seen in dermatomyositis can be differentiated from other diseases by skin biopsy. Characteristic skin lesions along with muscle weakness can help in more accurate diagnosis. Enzymatic elevations of creatine kinase, aldolase, lactic dehydrogenase, or alanine aminotransferase are seen. Following the response to therapy the two useful tests are Creatine kinase and lactic dehydrogenase. In patients tested are nonconclusive additional electromyography, muscle biopsy, ultrasound, or magnetic resonance imaging can be done [12,13].

Serological tests such as Antinuclear antibody testing are usually positive in such patients. Certain subsets of the disease can be recognized by myositis-specific antibodies. Anti-Jo-1 antibody (and the 6 other antisynthetase autoantibodies) is an indication of pulmonary involvement but is less common in Dermatomyositis as compared to Polymyositis. About 25 - 30% of the patients Anti-Mi-2 is seen. A new antibody to a 155-kD antigen or Se antigen (90 - 95 kd) seen as a marker of ADM. This antibody can be seen in juvenile DM and could indicate the possibility of a chronic course [14].

An elevated Willebrand factor can be associated with active disease in children, whereas in adults, it is associated with muscle weakness, fatigue, and fever. However, the markers for DM are not very consistent in their appearance. A thorough evaluation must be done after the confirmation of the disease to assess the severity, predict prognosis, and identify any other associated disorders. Patients should be thoroughly checked for pulmonary, cardiac and esophageal involvement with the help of chest X-ray/pulmonary function studies, electrocardiogram, or esophageal motility studies, respectively. Evaluation for malignancy in patients can be done with the appearance of any new symptoms or once a year for the first three years after the initial diagnosis [1].

**Management**

**Aim of the therapy**

The goals of management include [15]:

1) Confirm the true diagnosis (e.g. distinguish drug-induced dermatomyositis-skin lesions from authentic dermatomyositis skin disease);

2) Educate the patient about short and long-term disease considerations;

3) Treatment of skin lesions and symptoms;

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4) Observe for disease/treatment complications (e.g. the appearance of interstitial lung disease, methotrexate hepatotoxicity);
5) Disease associations (e.g. occult internal malignancy).

Course and therapy

In patients with progressive weakness, bed rest can be helpful accompanied by a range-of-motion exercise program to avoid contractures. Those who complain of dysphagia should avoid having meals immediately before going to bed and elevate the head of their bed.

Pharmacological methods

Systemic corticosteroids have been the most commonly used mode of therapy. Prednisone is given as initial therapy with a dosage of 0.5 - 1 mg/kg and continued for six weeks after the inactivation of the enzyme.

It is possible for 25% of the patients to be unresponsive to systemic steroids and about 25 - 50% to develop severe steroid-related side effects.

Therefore, early intervention with steroid-sparing immunosuppressive agents, such as methotrexate, azathioprine, cyclophosphamide, cyclosporin, induce or maintain remission if intervened early [16].

Pulse methylprednisolone therapy, combination immunosuppressive therapy, etanercept, infliximab, or rituximab are other alternatives that can be used if the patient fails to respond to immunosuppressants. In pregnant patients with DM, intravenous immune globulin (IVIG) can be used safely. However, Tachyphylaxis is seen as a side effect with repeated IVIG infusions [1].

Severe mortality and morbidity are seen if lung disease is seen as a complication of DM. Aggressive modalities like tacrolimus, combination therapy with corticosteroids, cyclosporin and cyclophosphamide, azathioprine, and autologous peripheral blood stem cell transplantation are being used to manage such patients.

Management of cutaneous lesions

Even when myositis responds to corticosteroids or immunosuppressive drugs, the persistence of cutaneous lesions is usually there. Most of these patients are photosensitive, and the use of broad-spectrum sunscreen with a high sun-protective factor that can block UVB and UVA rays is suggested [16]. Topical methods of using corticosteroid or nonsteroidal immunomodulators (tacrolimus ointment, pimecrolimus cream) are recommended. Patients who do not respond to Hydroxychloroquine HCl (dose of 200 to 400 mg/day which is effective in about 80% of the patients) can be switched to chloroquine phosphate (Dosage of 250 to 500 mg/day) [15].

Antimalarial therapy can have ophthalmic side effects, which should be monitored regularly.

Topical or systemic antibiotics can help to treat subepidermal vesicles or blisters. Telangiectasias can be treated with pulse dye or diode laser. Aluminum hydroxide suspension 15 - 20 ml q.i.d, probenecid 250 mg/day, potassium para-aminobenzoic acid 15 - 25 g/day, warfarin 1 mg/day and diltiazem 30 - 90 mg/day can be used to treat cutaneous calcinosis. The inflammation that occurs with the calcinosis can be suppressed successfully by EDTA [17].

In patients with recalcitrant cutaneous disease, use of Mycophenolate mofetil has been proved effective since 2006. It starts with a dosage of 1g b.id. then can be increased up to 1.5g if tolerated well. The onset of action is slow, and hence the therapy should be reduced after observing the response for 4 - 8 weeks [18].

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The monthly administration of intravenous immune globulin helps to cure patients with cutaneous lesions.[1] A common complication seen in children and adolescents is calcinosis. Early aggressive treatment can prevent its progression. The use of intravenous methylprednisolone can reduce the frequency and severity of the disease. Literature suggests the delay incorrect diagnosis of the disease is what leads to the occurrence of calcinosis. Though spontaneous regression is uncommon, immunosuppressants can reduce the chance of calcinosis. Diltiazem has also proved to be effective in the treatment of calcinosis. The usage of extracorporeal shock-wave therapy in other forms of soft tissue calcification (such as calcific tendonitis of the shoulder has made progress in the recent past, since pharmacological treatments are usually insignificant in curing the lesions [15].

Prognosis of the disease

Prognosis of recovery from the disease depends on [15]:

1) Age of the patient
2) Severity of disease
3) Presence or absence of dysphagia
4) Cardiopulmonary involvement
5) Presence of any associated malignancy
6) Responsiveness to treatment

Conclusion

Patients with active dermatomyositis experience skin symptoms such as pruritus, skin pain and skin burning, muscle stiffness, and other systemic manifestations. Cutaneous ulceration/scarring, poikilodema, and cutaneous calcinosis can be seen in patients with prolonged skin inflammation. Patients suffering from the disorder can have cardiopulmonary involvement or associated malignancies. So, it is of extreme importance to diagnose the condition early and aggressively treat the condition in the earlier stages to avoid complications. Several confirmatory diagnostic measures have evolved over the course of time, which helps in the differential diagnosis of similar conditions as well. Once the diagnosis is confirmed the management of the myositis must be taken with immediate effect depending on the age group, severity, and prognosis of the condition.

Since DM can cause psychosocial and occupational morbidity/disability, both topical and systemic antipruritic therapy can offer symptomatic relief to the patients. Pharmacological therapy with traditional broad-spectrum immunosuppressive therapy such as systemic corticosteroids, methotrexate, azathioprine, cyclosporin, or mycophenolate or biological therapy with high-dose IV can help in refractory dermatomyositis skin inflammation.

Future topical pharmacological drugs like antimalarial drugs, Thalidomide, Tacrolimus and Pimecrolimus. Systemic therapy with recombinant biological therapy are gaining popularity in the treatment of the condition.

Bibliography


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