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Clinical Presentation and Evaluation of Dermatomyositis

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ABSTRACT

Dermatomyositis is a rare and complex disease that has been often confused with other connective-tissue diseases and has been poorly treated. It is crucial for doctors to distinguish it from other muscle diseases as it often responds to treatment. However, death can occur in dermatomyositis due to severe infection or swallowing muscle damage, leading to pneumonia. The condition is uncommon and affects a specific group of people, with peak onset occurring just before the age of forty and after. Scleroderma and myositis are more common in women, while the juvenile form of myositis affects more boys than girls. The disease's onset is often delayed due to the difficulty in recognizing the patient's symptoms. Symptoms are widespread and come on gradually, with many different arrangements making diagnosis difficult. However, characteristic skin rashes may prompt further tests for the disease. Testing for the disease presents more difficulty than diagnosing it, as early stages may not show abnormalities in blood or raised muscle chemicals. However, a muscle biopsy can confirm the diagnosis.

INTRODUCTION

Dermatomyositis is a type of inflammatory myopathy, a long-term condition that causes muscle inflammation, weakness, and a characteristic skin rash. Inflammatory myopathies are a group of diseases that involve chronic muscle inflammation, which can cause muscle weakness. Although dermatomyositis is a rare condition, it is considered the most common inflammatory myopathy in children. Dermatomyositis got its name based on the following: "dermato-" comes from the Greek word for skin, which is "derma," and "myositis" means inflammation of the muscles. The term "classic dermatomyositis" describes a disease with a particular rash and inflamed muscles. The first detailed description of this condition was made by the British physician Thomas Bankes in 1891. However, it was not until 100 years later when the myositis-specific autoantibodies (MSAs), antibody groups that can be detected in the blood and are very helpful in diagnosing myositis, were described. There are no specific estimates for the prevalence of dermatomyositis. However, it is known that dermatomyositis is rare, particularly in adults - although slightly less so in the elderly - and somewhat more common in children. It can affect people of any age, from children to older adults, but most patients are diagnosed between the ages of 40 and 60. Also, women are twice as likely as men to be affected. Patients from different ethnic backgrounds and races can develop dermatomyositis, and where the disease occurs in people from specific ethnic backgrounds, such as in African Americans and Asians, somewhat different patterns of disease are seen. This condition is not contagious, so patients do not need to be isolated. The causes are not well understood, although it is thought that when an individual's muscles are damaged, the body's immune system responds in some way to try to clear a perceived infection or initiate repair. But in dermatomyositis, for reasons that are not yet known, this immune response seems to be overactive and cause inflammation in the muscle and skin. Also, genetic and environmental factors seem to have some roles in causing the condition.(1)

DEFINITION OF DERMATOMYOSITIS

There is no cure for dermatomyositis at the moment, but the condition can be managed effectively with the right treatment. The severity of the condition varies from person to person and ranges from mild cases with little disease activity or impact on daily life, to severe cases which can have a large impact on mobility and function and can even be life-threatening. Some people might experience flares of the disease activity, where symptoms get worse for a time, followed by periods of remission, where they feel better or symptoms are

entirely absent. Symptoms of dermatomyositis are caused by inflammation and might vary from person to person. Typically, it presents with a skin rash and muscle weakness, but it can also cause joint pain, lung problems or inflammation of the blood vessels. The timing of when different symptoms develop can vary: sometimes the rash or skin changes can precede muscle weakness by some time, but in other cases muscle problems might come first or there might be no skin manifestations at all.

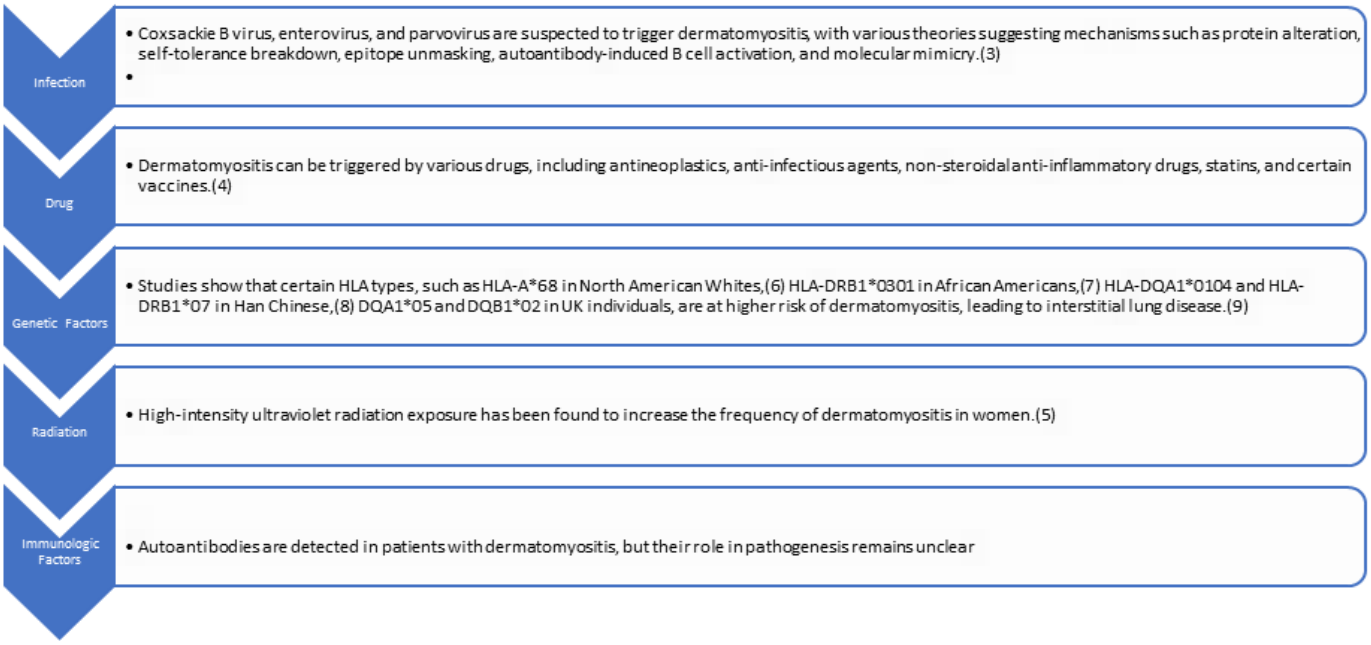
Dermatomyositis can affect anybody, but it most commonly presents in two different age ranges: it affects children between the ages of 5 and 15 and adults between the ages of 40 and 60. In adults, women are slightly more likely to develop the condition than men.

Dermatomyositis is a type of inflammatory myopathy or muscle disease. Specifically, dermatomyositis primarily affects the skin and the muscles. It is a type of autoimmune disorder, which means that the body's immune system - which normally helps fight infections - begins to attack healthy tissue. This leads to inflammation of the skin and underlying muscle tissue. It is not entirely clear what causes the immune system to start acting in this way, but it is thought that a viral infection may trigger the immune response in genetically susceptible individuals. It is not contagious and you cannot catch it from or pass it on to another person. (2)

ETIOLOGY

Dermatomyositis is a condition characterized by various genetic, immunologic, and environmental factors, although its exact cause remains unknown.

ENVIRONMENTAL FACTORS



EPIDEMIOLOGY

Dermatomyositis is a rare condition with an incidence rate of 9.63 per 1,000,000 people in Olmsted County, Minnesota. It primarily affects individuals aged 40-50, with a mean age of 44.0 ± 18.3 years at diagnosis. Women are more likely to be affected, with incidence rates of 3.98 and 4.68 per 1,000,000, respectively.(10)(11)

Southern Europe in Europe has a higher prevalence of dermatomyositis compared to Northern Europe. (12) A Quebec study revealed a higher prevalence of dermatomyositis in urban areas, while a Pennsylvania cohort study found clusters of clinically amyopathic dermatomyositis in regions with high airborne pollution, suggesting environmental factors may trigger the condition.(13)(14)

PATHOPHYSIOLOGY

Dermatomyositis is a condition resulting from a humoral attack on muscle capillaries and arterioles. The attack is initiated by completer factor-3, forming C3b and C4b, followed by the formation of neoantigen C3bNEO and the C5b-C9 membrane attack complex (MAC). This causes inflammation on vascular walls, leading to hypoxic injury to muscle fibers, particularly those at the periphery. Over time, capillary density decreases, leading to necrosis and degeneration of muscle fibers.

HISTOPATHOLOGY

Skin Biopsy

Dermatomyositis, a skin biopsy condition, shares similarities with systemic lupus erythematosus, characterized by vacuolar changes, increased lymphocytic infiltrate, and increased mucin deposition in the dermis. (15)

Muscle Biopsy

Muscle biopsy can diagnose dermatomyositis by examining the perivascular and interfascicular regions, which are characterized by a concentrated inflammatory infiltrate containing B cells, CD4+ T helper cells, macrophages, and plasmacytoid dendritic cells. Perifascicular atrophy, a hallmark histopathological feature, is observed in the perifascicular region, where degenerating and regenerating muscle fibers may be observed. Microangiopathy, a form of intramuscular blood vessel injury, can be observed through immunoglobulin and complement deposits on endomysial capillaries. (16)(17)

HISTORY AND PHYSICAL

In suspected cases of dermatomyositis, a comprehensive history and physical exam should be conducted to identify typical muscular and cutaneous symptoms, exclude other causes like inherited, infectious, or endocrine myopathy, and review organ systems to determine if other organ systems are involved. The main presenting symptoms are muscle weakness and skin findings, with the disease onset being insidious or acute with a waxing and waning course.

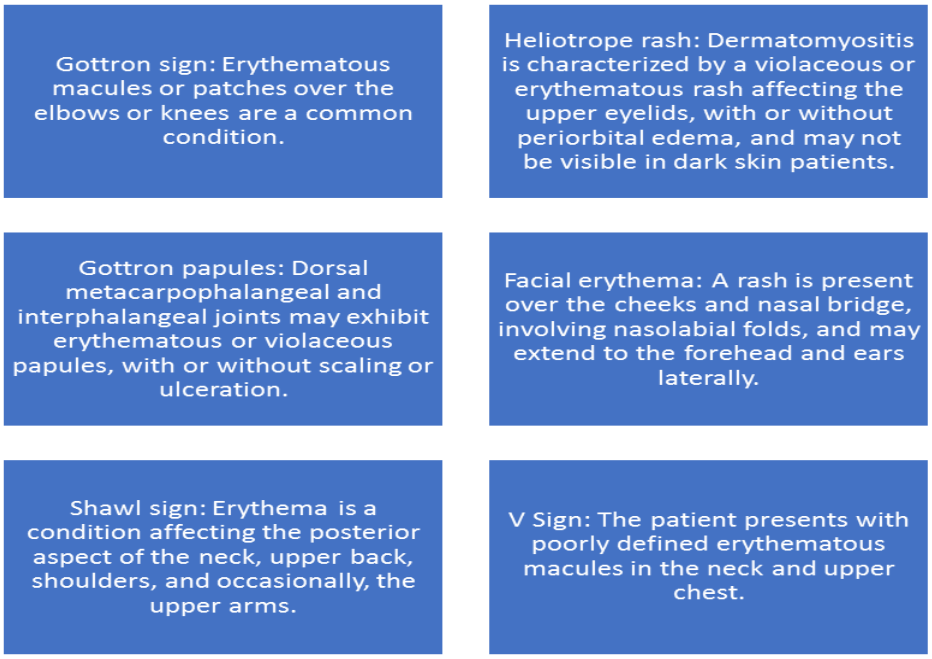
Muscular

Dermatomyositis is a common symptom characterized by muscle weakness, often onset sub acutely and gradually progressing. Patients may experience difficulty in activities like climbing stairs, lifting objects, and combing hair. Distal muscle weakness, pain, and stiffness are rare, and in severe cases, dysphagia or dysphonia may occur. Examination may reveal reduced muscle strength in proximal muscles like deltoids, hip flexors, and neck flexors. Muscle tenderness is usually mild, while distal muscle strength remains intact. Depressed deep tendon reflexes and muscle atrophy are not typically observed in severe cases.

Cutaneous

Skin changes may precede or coincide with muscular symptoms, presenting with rashes, photosensitivity, pigmentation changes, pruritis, nail changes, and alopecia, and can also cause nail changes.

Dermatomyositis is characterized by pathognomonic findings such as:



Joints

Dermatomyositis is a condition that can lead to non-erosive polyarthritis or arthralgia of small hand joints, causing joint pain or swelling.

Respiratory

Patients with underlying interstitial lung disease may experience exertional dyspnea, exercise intolerance, and non-productive cough, with bilateral dry crackles and reduced chest movement due to respiratory muscle weakness.

Esophageal

Patients with oropharyngeal and upper esophageal weakness may experience difficulty swallowing solids and liquids, and may also experience symptoms of gastroesophageal reflux.

Other findings

Dermatomyositis may present with Raynaud's phenomenon, gastrointestinal ulcers, and cardiac symptoms. Systemic symptoms like fever, malaise, and weight loss may indicate occult malignancy. Factors predicting malignancy include male gender, older age, dysphagia, and absence of interstitial lung disease. Patients should inquire about drug intake and undergo age-appropriate cancer screening exams. (18)

EVALUATION

LAB INVESTIGATIONS

Muscle Enzymes

Initial testing for suspected dermatomyositis should include muscle enzymes like CK, aldolase, LDH, AST, and ALT. These tests guide diagnostic studies and assess therapy response, and may occur before muscle weakness appears. Elevated enzymes may occur before muscle weakness.

AUTOANTIBODIES

Antinuclear antibodies (ANA) are common in dermatomyositis patients but do not diagnose the condition. Instead, testing should focus on detecting myositis-specific autoantibodies (MSA), present in 30% of cases. These antibodies provide valuable information for prognosis and organ involvement patterns. Aminoacyl-transfer (t) ribonucleic acid synthetase (antisynthetase) and anti-Jo are the most common autoantibodies associated with dermatomyositis. These autoantibodies are associated with specific complications and findings.

Anti-Jo	Anti-Mi2	Anti- MDA5	Anti- SRP	Anti- TIF-1 gamma	Anti-SAE	Anti-NXP2
Antisynthetase syndrome is a condition characterized by interstitial lung disease, mechanic's hands, Raynaud phenomenon, sclerodactyly, and arthritis.	V-neck sign, shawl rash, and Acute onset disease	Severe cutaneous involvement, amyopathic dermatomyositis, and rapidly progressive ILD are common conditions	severe myositis, resistant to treatment	malignancy	Dysphagia is a skin disease that precedes myositis.	calcinosis cutis(19)

Electromyography aids in identifying affected muscle groups, guiding biopsy selection, and distinguishing dermatomyositis from neuropathic conditions, but its findings are not specific and may be absent in 11% of patients. (20) Findings suggestive of dermatomyositis include the following:

- Complex repetitive discharges
- Early recruitment
- Low amplitude, short polyphasic motor unit potentials
- Increased insertional activity
- Spontaneous fibrillations
- Positive sharp waves

RADIOLOGY

Chest radiography: Patients with dermatomyositis should undergo chest radiography to screen for interstitial lung disease. If respiratory symptoms or abnormal findings are present, high-resolution computer tomography (HRCT) and pulmonary function tests should be performed. HRCT findings suggest interstitial lung disease.

Magnetic resonance imaging (MRI): Magnetic resonance imaging is a non-invasive, sensitive test for evaluating myositis, revealing muscle edema, hyperintense inflammation areas, and fat suppression. (21)

Barium swallow: If esophageal dysfunction is present, certain actions may be taken.

HISTOPATHOLOGY

Muscle biopsy is a crucial test for confirming the diagnosis of dermatomyositis, excluding other causes of muscle weakness or skin rash. It should be obtained on weak muscles identified by physical exam or contralateral to abnormal muscles identified by electromyography. Patients with suspected dermatomyositis but lacking characteristic skin findings should undergo a muscle biopsy, while those with characteristic skin manifestations but lacking muscle weakness should undergo a skin biopsy procedure. (15)

OTHER INVESTIGATIONS

Baseline lab investigations for dermatomyositis include a complete blood count, creatinine, liver function tests, and inflammatory markers like ESR and CRP. ESR in dermatomyositis is usually normal or mildly elevated. Serum Thyroid-stimulating hormone may be ordered to exclude hypothyroidism. Electrocardiography may be ordered to look for subclinical conduction abnormalities. Pulmonary function tests may be conducted to assess the severity of pulmonary involvement. Patients with interstitial lung disease show a restrictive defect on pulmonary function tests.

INVESTIGATIONS FOR MALIGNANCY

Dermatomyositis patients are at a higher risk of developing underlying malignancy within the first five years of diagnosis. Therefore, they should undergo age and sex-appropriate cancer screening, including colonoscopy, urine analysis, mammography, and pap smears. Women at high risk of ovarian cancer should also be screened. There is no consensus on the frequency and extent of cancer screening, but earlier studies suggest a focus on history, physical exams, and basic lab tests. (22) Recent research suggests that blind computer tomography testing of the chest, abdomen, and pelvis in asymptomatic patients may be effective in detecting occult malignancies.(23)

TREATMENT

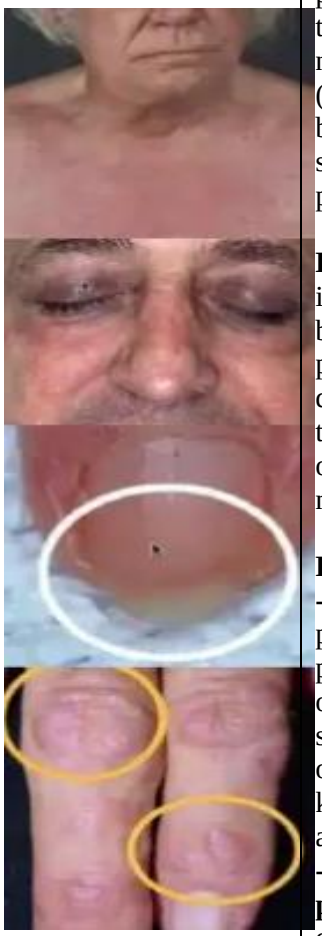
Dermatomyositis is a skin disease that affects the skin and can be treated with systemic glucocorticoids, immunosuppressants, and other treatments. The first-line treatment for muscle disease in dermatomyositis involves high doses of prednisolone for several months until muscle enzyme levels decline and strength improves. Patients should be evaluated regularly for an adequate response, which may take six weeks or three months. Once an adequate response occurs, the administration of systemic steroids is gradually tapered off over time.

If patients do not respond satisfactorily to therapy with steroids and azathioprine or methotrexate, they are considered resistant. Treatment options include rituximab, mycophenolate mofetil, calcineurin inhibitors, intravenous immunoglobulin (IVIG), and cyclophosphamide. Rituximab is the recommended first-line agent in resistant cases, while mycophenolate mofetil and tacrolimus are useful in refractory cases, especially if there is a

concomitant interstitial lung disease. Cyclophosphamide is preferred in cases of rapidly progressive interstitial lung disease.

Immunosuppressants can alleviate the adverse effects of long-term systemic steroids, such as osteoporosis, increased susceptibility to infections, cushingoid features, and secondary diabetes. Monitoring patients for any adverse effects of immunosuppressants is essential, including stomatitis, hepatotoxicity, and leucopenia. Cotreatment with folic acid or leucovorin can help minimize these effects. Azathioprine can cause flu-like reactions, myelosuppression, and pancreatitis, while cyclophosphamide increases the risk of malignancy and should be avoided unless multiple drug therapies have failed.

Skin disease in dermatomyositis is managed with general measures, physiotherapy, and medical therapy. Sun-protective measures, such as sunlight avoidance, use of sun-protective clothing, and sunscreen with a sun protective factor of 30 or higher, are recommended. Medical therapy for skin disease includes topical agents and systemic medications, with hydroxychloroquine and methotrexate being the most commonly used systemic agents. Calcinosis, which occurs more frequently in juvenile dermatomyositis, can be managed with calcium channel blockers like diltiazem or surgical removal of calcinotic nodules. Physical therapy and rehabilitation play an essential role in management, with patients with mild disease encouraged to participate in active exercise programs to prevent contractures. Anti-resorptive therapy may be indicated in patients on long-term systemic corticosteroids to prevent osteoporosis. Prophylaxis against Pneumocystis Jirovecii with trimethoprim and sulfamethoxazole is also recommended.

Dermatoses	Morphology	Etiology Pathogenesis	Treatment/Management
DERMATOMYOSITIS 	The preference sites for certain patterns include the nape of the neck, upper chest (V) pattern, upper back, neck, and shoulder (shawl) pattern. **Heliotrope rash** is characterized by edema and pink violet discoloration in the striated orbicularis oculi muscle. **Pathognomonic** **-Gottron sign:** A pink to reddish-purple eruption, often atrophic or scaling, is often observed over the knuckles, knees, and elbows. **-Gottron papules:** DM is characterized by flat-topped, polygonal violaceous papules over the knuckles, which are less common but highly common.	**-F>M;** common in blacks; relatively rare -Evolve through multiple sequential phases: The process of autoimmune disease involves three phases: genetic susceptibility, an induction phase triggered by environmental stimuli, an autoimmune expansion phase, and an injury phase involving multiple immunologic effector mechanisms. Characterized by **Inflammatory myositis** and **skin disease** -Variant: a) **Amyopathic DM**: Absent or subclinical myopathy b)**Polymyositis PM:** Muscle involvement without skin changes -Musele involvement: **weakness of proximal muscle groups in characteristic** **Diagnostic criteria** • **Dermatomyositis:** Patients with the CUTANEOUS lesion - 4 remaining • **Polymyositis:** Patients with NO CUTANEUS lesion or 4 remaining ➢ Cutaneous lesions ➢ **Heliotrope rash:** pinking violet edematous erythema on the upper palpebral ➢ **Gottron's papules or sign:** Pinkish violet flat-topped papules, atrophy, or erythema on extensor surfaces and finger joints. ➢ **Proximal muscle weakness** ➢ Myogenic changes on EMG	**-Prednisone:** The primary treatment for acute musele disease involves doses starting at 1mg/kg/day until the severity decreases and musele enzymes reach near normal levels. **-Methotrexate and mycophenolate mofetil** are used as steroid-sparing agents -Daily used sunscreen with high SPF - Antimalarials like hydroxychloroquine are effective in reducing the severity of DM. **Prognosis** - Major causes of death include cancer, ischemic heart disease, and lung disease. -Independent risk factors • Older age • Shorter disease history • WBC above 10,000/mm^3 • Failure to induce clinical remission • Temperature greater than 38 degrees Celsius at diagnosis

DIFFERENTIAL DIAGNOSIS

Muscle weakness can be caused by various conditions that should be excluded through history, physical exams, and investigations before a definitive diagnosis of dermatomyositis can be made.

Inclusion body myositis is a type of muscle weakness that is usually asymmetric and involves distal muscles like the wrist and finger flexors. It is rare and refractory to treatment with corticosteroids. Drug-induced myopathy, which can cause mild myalgia or severe rhabdomyolysis, is a condition that requires a drug history to rule out. Hypothyroidism, which can present with proximal weakness and elevated muscle enzymes, can also be differentiated from dermatomyositis. Myasthenia gravis, which predominantly causes muscle weakness of the ocular and bulbar muscles, is associated with anti-acetylcholine receptor antibodies and does not cause elevation of muscle enzymes. Polymyalgia rheumatica, which presents with pain and stiffness of muscles around the shoulder and pelvic girdle, can be differentiated from dermatomyositis by the presence of inflammatory markers, absence of elevated muscle enzymes, and normal muscle strength. Other differential diagnoses include muscular dystrophies, motor neuron disease, neuropathy, inherited metabolic myopathies, and myasthenia gravis. The presence of characteristic skin findings, symmetric muscle weakness, electromyography, and muscle biopsy can help distinguish dermatomyositis from these conditions.

PREDICTION

Dermatomyositis has a mortality rate of 10%, with the highest rate occurring in the first year of the disease. (24)

- Pulmonary involvement in the form of respiratory muscle weakness or interstitial lung disease
- Cardiac involvement
- Underlying malignancy
- Advanced age
- Initiation of treatment more than six months following the onset of symptoms
- Severe muscle weakness on presentation
- Presence of dysphagia (25)(26)(27)(28)

85% of survivors have normal strength, 34% have mild disability, and 16% have no disability. 20% achieve remission with treatment, while 80% have chronic or polycyclic conditions. (29)

COMPLICATIONS

ESOPHAGEAL DISEASE

Dysphagia, malnutrition, aspiration risk, calcinosis, muscle atrophy, and contractures are common complications of patients with oropharyngeal and esophageal muscle weakness.

HEART DISEASE

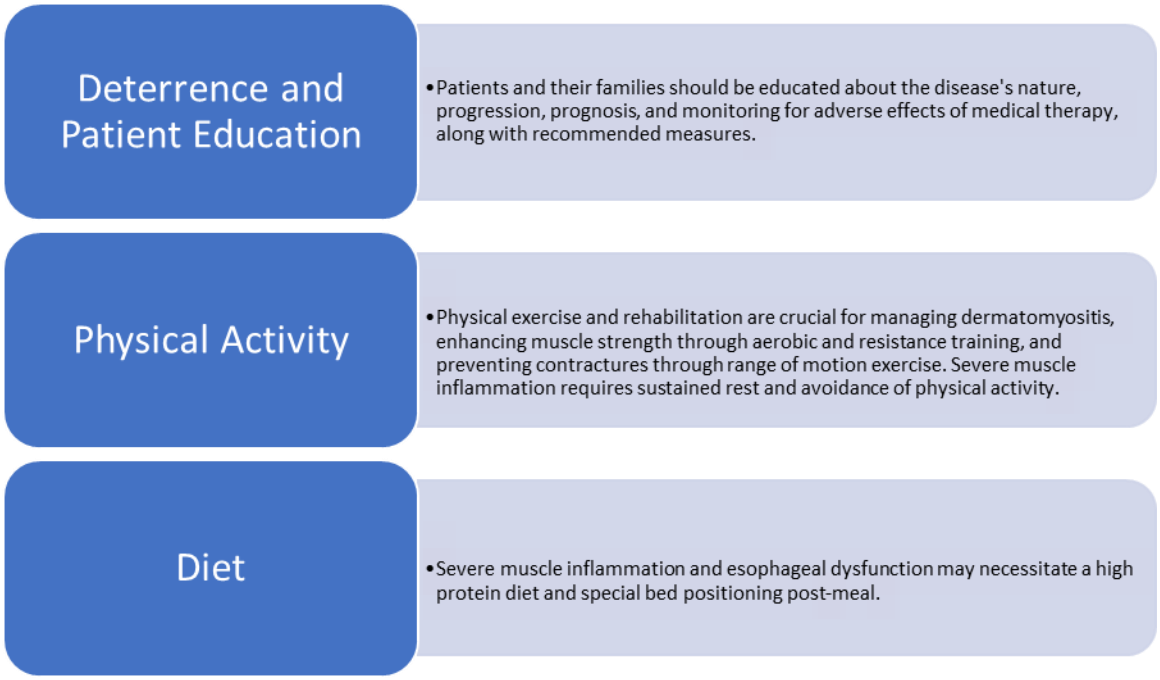
Dermatomyositis typically presents subclinical cardiac involvement, with ECG revealing conduction abnormalities and arrhythmias, potentially leading to myocarditis, congestive heart failure, or coronary artery disease. (30)

RESPIRATORY DISEASE

Pulmonary involvement in dermatomyositis can lead to hypoventilation, aspiration pneumonia, or interstitial lung disease. Interstitial lung disease (ILD) is prevalent in about a third of patients and is linked to anti-histidyl transfer ribonucleic acid synthetase antibodies. Aspiration pneumonia, caused by respiratory muscle weakness, can cause significant morbidity and mortality.

MALIGNANCY

Dermatomyositis patients are at a higher risk of malignancies, occurring in 24% of cases. (31) A study of patients in Sweden, Denmark, and Finland found a 3.0 incidence ratio for malignancy. Risk factors included old age, absence of interstitial lung disease, severe cutaneous involvement, anti-155/140 antibodies, myositis-specific antibodies, treatment resistance, and previous malignancy with relapse. Common malignancies included ovary, lung, pancreas, stomach, and colon adenocarcinomas. (32) A study in Sweden, Denmark, and Finland revealed a 3.0 malignancy incidence ratio among patients, with risk factors including old age, absence of interstitial lung disease, severe cutaneous involvement, anti-155/140 antibodies, treatment resistance, and previous malignancy with relapse.(33)



SUN PROTECTIVE MEASURES

Patients should avoid sunlight, use sunscreen, and wear sun-protective measures like wide-brimmed hats and full-body-covered clothing to prevent worsening photosensitive skin rash.

ENHANCING HEALTHCARE TEAM OUTCOMES

Dermatomyositis is a rare condition that requires a multidisciplinary team to manage and may require referral to a tertiary care center. It is typically diagnosed by a rheumatologist, internist, family physician, or pediatrician, depending on age. The EULAR/ACR criteria for idiopathic inflammatory myopathies is a sensitive and specific classification system used to identify cases. Initial workups for suspected dermatomyositis include testing for muscle enzymes and myositis-specific autoantibodies. If the diagnosis is uncertain, electromyography, muscle, and skin biopsy may be performed. Consultations with other specialties may be necessary depending on the type of associated complications. Early recognition and prompt treatment can reduce morbidity and mortality. The first-line treatment for dermatomyositis is prednisolone. (34) Patients resistant to glucocorticoids alone or with extra muscular complications or severe weakness should receive concurrent treatment with glucocorticoids and immunosuppressants. Sun avoidance and sun-protective measures are recommended for all skin disease patients. Early resistance exercise rehabilitation can improve daily functioning. (35)

CONCLUSION

Dermatomyositis is a rare inflammatory myopathy that causes muscle inflammation, weakness, and a characteristic skin rash. It is the most common inflammatory myopathy in children and is more common in adults. The condition is rare, particularly in adults, and more common in children. Symptoms vary from person to person and can include skin rash, muscle weakness, joint pain, lung problems, or inflammation of blood vessels. The exact cause of dermatomyositis is not entirely understood, but it is thought that a viral infection may trigger the immune response in genetically susceptible individuals. Dermatomyositis is characterized by various genetic, immunologic, and environmental factors, with autoantibodies detected in patients but their role in pathogenesis remains unclear. Studies show that certain HLA types, such as HLA-A*68 in North American Whites, HLA-DRB1*0301 in African Americans, HLA-DQA1*0104 and HLA-DRB1*07 in Han Chinese, and DQA1*05 and DQB1*02 in UK individuals, are at higher risk of dermatomyositis, leading to interstitial lung disease.

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