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Review Article

Dermatomyositis: a narrative review

Rahul Shil[1]*, Shivaleela P. Upashe[2], Shreyasee Das[3]

[1]Department of MSN (Neuroscience), Sapthagiri NPS University, Bengaluru, Karnataka, India
[2]Department of Pediatric Nursing, Pravara Institute of Medical Sciences (Deemed to be university), Maharashtra, India
[3]Department of Psychiatric Nursing, Dr. B. R. Ambedkar Institute of Nursing, Bengaluru, Karnataka, India

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***Correspondence:**
Dr. Rahul Shil,
E-mail: Shil.rahul06@gmail.com

ABSTRACT

Dermatomyositis is a rare, idiopathic, immune-mediated inflammatory muscle disorder characterized by chronic muscle inflammation, muscle weakness, and pain. It is often accompanied by skin rashes, which appear patchily with red discoloration and are commonly found on the knuckles, cuticles, nail beds, palms, fingers, elbows, knees, upper eyelids, edematous areas, upper chest in a V shape, neck, and back of shoulders. Muscle weakness significantly impairs daily activities such as sitting upright, climbing stairs, washing hair, combing, and others. As an autoimmune disorder, dermatomyositis lacks a cure, but its progression can be mitigated by enhancing daily activity functioning and reducing extra-muscular symptoms like rashes, dysphagia, dyspnea, and fever. Immune suppressant medications can prove beneficial in managing the condition.

Keywords: Idiopathic inflammatory myopathy, Skin predominant dermatomyositis, Dermatomyositis

INTRODUCTION

Dermatomyositis is a severe illness characterized by muscle weakness and minimal skin rashes. It is a well-recognized form of myopathy that can impair swallowing and breathing. It is a specific type of polymyositis primarily affecting muscles and skin.[1] A skin rash that precedes or accompanies progressive muscle weakness is a hallmark of dermatomyositis. Although dermatomyositis cannot be cured, long-term medication and physical therapy can alleviate its symptoms. These interventions not only restore muscle strength and function but also aid in clearing the skin rash. Although dermatomyositis can affect individuals of any age or gender, women are more susceptible to developing it than men. It can occur at any age, particularly between the ages of 50 and 70. Compared to men, women are more likely to be affected by this medical condition. Individuals with dermatomyositis are also more prone to developing connective tissue disorders such as rheumatoid arthritis or lupus. Juvenile dermatomyositis is the term used to describe dermatomyositis in children aged 5 to 14. Approximately one in every million children is affected by this uncommon illness. Children with juvenile dermatomyositis often experience weakness, exhaustion, and shortness of breath, even without expressing pain.[2] The cause of dermatomyositis remains unknown, as it is idiopathic. Histopathologically, striated muscle swelling is a shared characteristic of this type of idiopathic inflammatory myopathy. There are three distinct types of idiopathic inflammatory myopathies that do not affect neuromuscular transmission: dermatomyositis, polymyositis, and inclusion body myositis.[3] In 1863, dermatomyositis was first documented in literature when a physician named Wagner published a patient's description detailing the patient's multiple rashes and symptoms consistent with dermatomyositis.[4] The incidence and prevalence rates of dermatomyositis are approximately 1 to 10 and 10 to 60

cases per million population. In juvenile dermatomyositis, the incidence rate is 1 to 3.2 cases per million children.[5]

TYPES

Dermatomyositis is classified into different types based on the extent of skin or muscle involvement. Some of the types of dermatomyositis are Classic dermatomyositis (CDM), amyopathic dermatomyositis (ADM), hypo myopathic dermatomyositis (HDM), clinically amyopathic dermatomyositis (CADM), adermatopathic dermatomyositis (Dermatomyositis sine dermatitis) (ADM) and juvenile dermatomyositis (JDM). The cutaneous expression of proximal muscle weakness within the first six months after the beginning of skin disease is known as CDM. ADM is also linked to cutaneous involvement, and it can happen six months or more after DM is diagnosed without any laboratory or clinical signs of a skin or muscle ailment.[6-9] Subjective muscular weakness does not exist in HDM, particularly after the first six months. However, more subjective information includes abnormal muscle enzymes and myopathy indicators on muscle biopsy electromyography. The subclinical signs of ADM and HDM are commonly referred to as clinical DM. Thus, there is evidence of muscle and skin illness in clinical DM. Furthermore, ADM has a has a cutaneous feature without muscle weakness for more than 6 months with subclinical muscular involvement which are often seen in more than 5-20% of patients with dermatomyositis.[9,10] JDM is the most common inflammatory myopathy in childhood. Typical rash and proximal muscle weakness is found in this disease condition of this type. this can be seen in the patients of less than 18 years old. Muscle weakness, calcinosis cutis, cutaneous vasculitis, ulcerations, and gastrointestinal tract vasculopathy are its primary characteristics. subtypes mainly include monocyclic, polycyclic chronic and ulcerative. Dermatomyositis in child shows greatest weakness in hip flexors extensors and abductors as well as neck flexors as compared to the adults.[11] In HDM, there is no subjective muscle weakness especially after the first 6 months. But a more subjective data includes abnormal muscle enzyme and signs of myopathy on electromyography of muscle biopsy.[2,3]

PATHOPHYSIOLOGY

Although the exact pathogenesis remains unknown but still autoimmune disease, some exogenous disorders (e. g., the viral infection) are some of the etiological factors of dermatomyositis. It is primarily believed to be the outcome of an attack by humor that targets the arteriole endothelium and muscle capillaries. The activation of the first component, C3, which creates C3b and C4b, is the beginning event. The C5b membrane attack complex and the neoantigen C3bNEO appear shortly after it. Edema results from this membrane's attack on complex deposits on the vascular walls. As time passes, the capillary density decreases, and the muscle fibers begin to degenerate and necrotize.[12]

CAUSES

The exact cause of dermatomyositis is still unknown but still there some causes related to the characteristics of the symptoms such as genetic abnormality, due to cancer specially in older people, autoimmune disease, an infection in the body, medication can be a cause, and another exposure in the environment which triggers the disease which include viral infections, sun exposure, certain medications and smoking.[2]

CLINICAL FEATURES

Knowing about the early sign of dermatomyositis is essential for the timely treatment. The symptoms may arise suddenly without any signs involving both skin and muscle issues. Patients should be mainly aware of the:

Muscle weakness

In the beginning the patient will have muscle abnormalities like muscle aches, weakness of the muscle in the trunk, upper arms, hips, thighs. The weakness affects both the left and right sides of the body, and tends to gradually worsen. Later the muscle may be stiff, tender, sore and show the sign of muscle degeneration. Muscle weakness might make it hard for the individual to do common motions including: raising their arms, climbing stairs, sitting upright, getting up from a seated position (like standing up from a chair or couch), getting up after lying down, it effects day to day activities like Washing your hair, combing, washing the hand etc.

Skin rashes

A dusky red rash develops, The most common locations for the rash are in the following areas i.e. across the shoulders and upper back, over the knuckles with a raised violaceous scaly eruption (Gatton's sign), often with changes to the cuticles and nail beds, on the palms and fingers, over the elbows and knees, around the upper eye lid and edema (heliotrope rash),on the upper chest in a V shape, neck and the back of your shoulders (a shawl sign rash). The rash, which can be itchy and painful, is often the first sign of dermatomyositis.

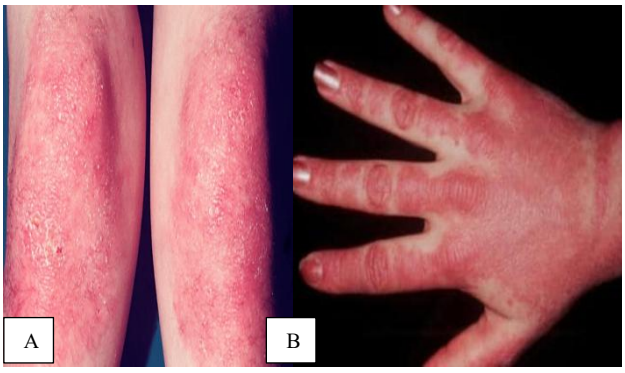

Figure 1 (A and B): Skin rashes.

Fatigue

Feeling tired without reason even without physical activities is common throughout.

Difficulty swallowing

Dermatomyositis can affect the muscle responsible for eating or drinking.

Joint pain

Some patients may experience pain or swelling in the joints.

Extra muscular manifestations

Systemic symptoms like fever, malaise, weakness, joint contractures, arthralgias, synovitis or deforming arthropathy with subluxation in the interphalangeal joints, cardiac disturbances, pulmonary dysfunction.[2-3,6,13]

CLINICAL DIAGNOSTIC TESTS

Wide ranges of diagnostic tests are generally done to confirm the diagnosis. Some of the diagnostic tests are below:

Blood analysis

A high amount of muscle enzyme, which mostly implies muscle injury, is checked by a blood test. Additionally, this blood test can identify the autoantibodies linked to various dermatomyositis symptoms, which aids in choosing the most appropriate medication and course of treatment.

Chest X-ray

This simple test is done to check the type of lung damage that sometimes occurs with the dermatomyositis.

Electromyography

The electrical activity in response to nerve and muscle stimulation will be measured by electromyography. Here, a tiny needle electrode will be put into the muscle through the skin. Muscle activity can be confirmed by changes in the pattern of electrical activity of the muscle, which is recorded while the patient relaxes or tightens the muscle. It can also help identify the muscles that are impacted.

MRI

This is done to create cross sectional image of the muscle by powerful magnetic field and radio waves. Unlike a muscle biopsy, an MRI can assess inflammation over a large area of muscle.

Skin or muscle biopsy

A biopsy involves removing a tiny sample of muscle or skin for laboratory examination, which can support the diagnosis of dermatomyositis. Muscle inflammation or other issues like injury or infection may be discovered by a muscle biopsy. However, a muscle biopsy might not be required if the skin biopsy confirms the diagnosis.[14]

TREATMENT APPROACHES

Though dermatomyositis is idiopathic and incurable still there are certain medication which helps in improving the muscle strength and function.

Corticosteroids

Drugs such as the prednisone (Rayos). The skin disease might get treated by the avoidance by sun exposure and by using sunscreen and photoprotective clothing as well as some type of topical corticosteroids, antimalarial agents, and immunomodulatory medications such as methotrexate, mycophenolate mofetil, or intravenous immunoglobin.

Corticosteroids sparing agents

These drugs are taken with the combination of corticosteroids to lower the dose and side effect of corticosteroids. The two most common medication for dermatomyositis is azathioprine and methotrexate.

Rituximab (Rituxan)

It generally helps in treating rheumatoid arthritis, it is an optional drug if the initial therapies does not work.

Antimalarial medications

It helps in persistent rash removal such as hydroxychloroquine (Plaquenil).

Sunscreens

This helps in protecting skin from skin exposure by applying sunscreen and wearing protective clothing and hats is important for managing the rash of dermatomyositis.[5,7,9]

THERAPEUTIC MEASURES

The main goal of managing dermatomyositis generally focused on treating weakness, skin disease and addressing any other underlying complications. Depending on the severity of disease the doctor might prescribe several therapeutic measures such as Physical therapy which helps in maintaining and improving the strength and flexibility. Speech therapy which improves the ability to talk and improves language ability. Additionally, intravenous immunoglobulin (IVIg), a refined blood

product containing healthy antibodies from thousands of blood donors, is occasionally recommended. In patients with dermatomyositis, these antibodies can eliminate skin lesions and prevent the harmful antibodies that target the skin and muscles. According to one study, the IVIG-treated group's muscular and cutaneous involvement dramatically improved at six months, and after a four-year follow-up period, their modified cutaneous dermatomyositis area and severity index (CDASI) scores significantly improved above their pretreatment values. In cases of dermatomyositis, subcutaneous IgG has also been beneficial. To get rid of uncomfortable calcium deposits and stop recurring skin infections, surgery can be a possibility.[13,14] Below are some other treatment measures.

Dietetic assessment

Chewing and swallowing may become more challenging as dermatomyositis progresses, but one can learn how to make foods that are easy to consume from a qualified dietitian.

General measures

For those with the most severe muscular inflammation, bed rest is the most important approach. Physical therapy is used to educate people with muscle weakness, particularly youngsters, about available treatment options. To preserve muscle strength, both adult and juvenile patients must engage in rehabilitation exercises.[4-6]

Surgery

To get rid of uncomfortable calcium deposits and stop recurring skin infections, surgery can be a possibility.

Self-care

Patients with dermatomyositis should wear protective clothing or high-protection sunscreen when going outside, as the rashes are more sensitive to the sun.[15,16]

ASSOCIATED CONDITIONS

Dermatomyositis might cause other conditions or put you at higher risk of developing them, including:

Raynaud's phenomenon

When exposed to cold temperatures, Raynaud's disease can cause fingers, toes, cheeks, noses, and ears to become pale.

Other connective tissue diseases

Dermatomyositis can coexist with other illnesses such lupus, rheumatoid arthritis, scleroderma, and Sjogren's syndrome.

Cardiovascular disease

Inflammation of the heart muscle can result from dermatomyositis. A tiny percentage of patients with dermatomyositis experience congestive heart failure and irregular heartbeats.

Lung disease

Dermatomyositis can cause interstitial lung disease. A collection of conditions known as interstitial lung disease results in lung tissue scarring, which makes the lungs rigid and inelastic. Breathlessness and a dry cough are some of the common symptoms.

Cancer

Adult dermatomyositis has been associated with a higher risk of malignancy, especially ovarian cancer in women. About three years after receiving a diagnosis of dermatomyositis, the risk of developing cancer seems to level off.[17-19]

COMPLICATIONS

Possible complications of dermatomyositis include:

Difficulty swallowing

Affected esophageal muscles can make swallowing difficult, which can lead to malnourishment and weight loss.

Aspiration pneumonia

This can be a cause of death in some patients due to weakened swallowing muscles causing patients to accidentally inhale food and liquids into the lungs.

Breathing problems

Breathing problems could be a serious issue as it affects the chest muscles which can create breathing problems, such as shortness of breath.

Calcium deposits

Calcium deposits ae more common in children with dermatomyositis and develop earlier in the course of the disease generally in muscles, skin and connective tissues as the disease progresses.[19,20]

CONCLUSION

In this review, we have comprehensively delineated the salient points pertinent to dermatomyositis. It is of paramount importance to recognize dermatomyositis as a distinct and uncommon disease entity characterized by idiopathic involvement of both skin and muscles. Given

the current lack of definitive causal elucidations, further research endeavors are warranted to unravel the precise etiology of the disease. This comprehensive understanding will facilitate the development of tailored treatment modalities. In the hope of future advancements, we anticipate the elucidation of the etiology of dermatomyositis, thereby illuminating its susceptibility, heterogeneity, and progression patterns.

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