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## Signs of Dermatomyositis

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**D**ermatomyositis, an idiopathic inflammatory myopathy with characteristic skin manifestations, is rarely seen and can be challenging to diagnose; however, certain salient features of this disease, if present, can make diagnosing the disease straightforward. The relative incidence rates of the various forms of inflammatory myopathy are not well established<sup>1</sup>; however, dermatomyositis is the most common and polymyositis the least common of the idiopathic inflammatory myopathies.<sup>2</sup> Clinically, the presence of characteristic skin rashes differentiates dermatomyositis from polymyositis. Histopathologic findings also are distinctive in dermatomyositis. Although the etiology of dermatomyositis is unknown, autoimmunity is suspected to be the underlying cause of inflammation, with unknown inciting events. This article reviews the clinical signs associated with dermatomyositis and briefly discusses the diagnostic evaluation and treatment approach.

### CLINICAL PRESENTATION

Dermatomyositis typically presents with proximal muscle weakness and skin rash, which develop slowly over weeks to months along with constitutional symptoms such as fever, malaise, arthralgias, and weight loss. Proximal muscle weakness is frequently seen, causing difficulty with tasks such as combing hair, arising from a chair, climbing stairs, and getting out of a bathtub. Weakness of the neck flexor muscles is often present. Weakness of ocular and facial muscles is possible but rare.<sup>1</sup>

Typical dermatomyositis rashes include heliotrope rash and Gottron's sign. *Heliotrope rash* is a violaceous discoloration of the eyelids associated with periorbital edema (**Figure 1**). *Gottron's sign* is a scaly and erythematous dermatitis seen on the dorsum of the hand, over the metacarpophalangeal and proximal interphalangeal joints (**Figure 2**). It may also be seen on the face, neck,

### RASHES OF DERMATOMYOSITIS

Heliotrope rash—eyelids  
Gottron's sign—hands and phalangeal joints  
Mechanics' hands—palms, radial aspect of fingers  
Shawl sign—posterior neck and shoulders  
V-sign—anterior neck and chest  
Poikiloderma vasculare atrophicans—torso

upper torso, elbow, knee, and medial malleolus. Other skin findings include mechanics' hands, the shawl sign, the V-sign, and poikiloderma vasculare atrophicans. *Mechanics' hands* refers to roughening or fissuring of the skin on the palm and radial surface of the fingers. The *shawl sign* refers to a macular erythema on the posterior neck and shoulders; the *V-sign* is a similar rash occurring on the anterior neck and chest. *Poikiloderma vasculare atrophicans* is a circumscribed violaceous erythema with associated telangiectasia, hypopigmentation or hyperpigmentation, and superficial atrophy; it is commonly found over the anterior neck, chest, posterior shoulders, back, and buttocks (**Figure 3**).

In approximately 10% of cases, dermatomyositis manifests only the classic cutaneous findings without muscle involvement, a condition referred to as *amyopathic dermatomyositis* or *dermatomyositis sine myositis*. Among these cases, one third to one half will not have muscle involvement later; the rest will develop muscle

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**Figure 1.** Heliotrope. Reprinted with permission from Sauer GC, Hall JC. *Manual of skin diseases*. 7th ed. Philadelphia: Lippincott-Raven; 1996:265.



**Figure 2.** Gottron's papules.

involvement. In approximately 30% of cases of dermatomyositis, the skin rash precedes muscle involvement by several weeks to months. Less frequently, the skin lesions precede the onset of muscle weakness by more than a year. In the remaining 60% of cases of dermatomyositis, skin and muscle changes appear concurrently.<sup>3</sup>

Besides the typical muscle and skin involvement, dermatomyositis also affects other organ systems. Dysphagia can develop from involvement of the skeletal muscle of the upper esophagus or from cricopharyngeal obstruction.<sup>2</sup> Pulmonary involvement is seen in 10% to 20% of patients with idiopathic myopathy, usually in the form of interstitial lung diseases such as interstitial pneumonitis with fine crackles on physical examination.<sup>4</sup> Another pulmonary manifestation is



**Figure 3.** Poikiloderma vasculare atrophicans.

dyspnea from thoracic muscle weakness. Cardiac involvement is rare, but may be present, causing myocarditis or cardiac arrhythmias. Rarely, Raynaud's phenomenon and arthralgias can be seen with dermatomyositis.

Distinctive clinical features in dermatomyositis can help to differentiate it from polymyositis. Subcutaneous calcinosis is seen in the elbows, back, and buttocks.<sup>5</sup> Also, in contrast to other idiopathic inflammatory myopathies, dermatomyositis may be associated with scleroderma and mixed connective tissue disease but not with other autoimmune or connective tissue diseases.<sup>2</sup>

Dermatomyositis is associated with a higher risk of malignancy. Although the prevalence of associated malignancy varies, the link between dermatomyositis and malignancy is well established.<sup>6</sup> The diagnosis of malignancy can be made before, concurrently, or after the diagnosis of dermatomyositis. There is a 6-fold increase in cancer risk in dermatomyositis patients. The risk of malignancy is highest early in the diagnosis, but some risk persists throughout the illness<sup>7,8</sup>; risk of malignancy rises with increasing age, capillary damage on biopsy, cutaneous necrosis, cutaneous leukocytoclastic vasculitis, and cutaneous mucinosis.<sup>4</sup> A broad range of malignancies have been reported with dermatomyositis, but certain malignancies are seen in increased frequency; these are ovarian, lung, pancreatic, stomach, and colorectal cancers, and non-Hodgkin's lymphoma.<sup>7,8</sup> Early search for a malignancy is important; work-up should be age and gender appropriate.

#### **DIAGNOSTIC EVALUATION**

Elevated serum muscle enzymes (eg, creatine kinase, aldolase, aspartate aminotransferase, alanine aminotransferase, lactate dehydrogenase) are helpful in the diagnosis of dermatomyositis. Creatine kinase is usually elevated (up to 50 times the normal limit); an

**Table 1.** Classification Criteria for Polymyositis and Dermatomyositis\*

1. Symmetric weakness of limb girdle muscles and anterior neck flexors, progressing over weeks to months, with or without dysphagia or respiratory muscle involvement
2. Positive muscle biopsy
3. Elevation of muscle enzymes
4. Electromyographic evidence with a triad of
  - i) Short, small polyphasic motor units
  - ii) Fibrillations positive sharp waves, insertional irritability
  - iii) Bizarre, high frequency repetitive discharges
5. Dermatologic features, including heliotrope rash and Gottron's sign

Adapted from Bohan A, Peter JB. Polymyositis and dermatomyositis. *N Engl J Med* 1975;292:344–7; and Bohan A, Peter JB. Polymyositis and dermatomyositis. *N Engl J Med* 1975;292:403–7.

\*For a diagnosis of dermatomyositis, 3 of the first 4 criteria plus criterion 5 must be present.

elevated muscle enzyme level is the only laboratory criterion included in the American College of Rheumatology criteria for the diagnosis of polymyositis and dermatomyositis.<sup>9,10</sup> However, creatine kinase levels can also be mildly elevated or normal. Aspartate aminotransferase and alanine aminotransferase are increased as well. Additionally, autoantibodies are usually seen in dermatomyositis—commonly aldolase, anti-histidyl tRNA (anti-Jo-1), and antibody against helicase activity (anti-Mi2). Anti-Jo-1 antibodies, rarely seen in patients with dermatomyositis, have been associated with interstitial lung disease. Anti-Mi2 antibodies, found in about 25% of patients with dermatomyositis, have been associated with good treatment response.<sup>5</sup>

Other useful tests in diagnosing dermatomyositis are electromyography (EMG) and magnetic resonance imaging. Typical EMG findings include increased spontaneous muscle activity with fibrillations, complex repetitive discharges, and positive sharp waves. These findings are not specific, but can be used to confirm a diagnosis of dermatomyositis (as well as other inflammatory myositis). EMG is also useful in selecting affected muscle for biopsy, as muscle involvement can be patchy. A biopsy is usually performed on the involved muscle of the opposite extremity. Biopsy performed on the same muscle on which EMG is performed can result in iatrogenic muscle abnormalities introduced by EMG. Previously, a false-negative rate of 10% to 25% in muscle biopsy was reported, primarily attributed to the uneven distribution of tissue pathology; however, using magnetic resonance imaging to detect

**Table 2.** Differential Diagnosis of Dermatomyositis

- Muscular dystrophy
- Drug toxicity (statins, azathioprine, cimetidine)
- Neurologic disease (myasthenia gravis, motor neuron disease, Parkinson's disease)
- Endocrine/immunologic disease (myxedema, stiff-man syndrome)
- Metabolic diseases (phosphofructokinase deficiency, phosphorylase B kinase deficiency)
- Mitochondrial diseases
- Miscellaneous: amyloidosis, cystic fibrosis, meningioma, Munchausen syndrome

Adapted from Christopher-Stine L, Plotz PH. Adult inflammatory myopathies. *Best Pract Res Clin Rheumatol* 2004;18:331–44.

inflammation and select biopsy site can reduce this false-negative rate.<sup>4</sup>

Muscle biopsy is most specific in diagnosing dermatomyositis. The typical histopathologic changes in dermatomyositis are perivascular infiltration of inflammatory cells (CD4+ T cells and B cells) and perifascicular atrophy. Less commonly, biopsy reveals inflammatory infiltrate of the endomysial capillaries. Skin biopsy can reveal similar findings in the dermis.<sup>4</sup> These histopathologic features are distinctive from the findings of polymyositis, in which the inflammatory cells are CD8+ T cells infiltrating the endomysium.

The diagnosis of dermatomyositis can be made using the American College of Rheumatology criteria for the diagnosis of polymyositis and dermatomyositis, established by Bohan and Peter in 1975 (**Table 1**).<sup>9,10</sup> For a definite diagnosis of dermatomyositis, 3 of the first 4 criteria plus the rash must be present.<sup>11</sup> (Similarly, a definite diagnosis of myositis can be made with 3 of the 4 criteria *without* the rash.)

## DIFFERENTIAL DIAGNOSES

Other diseases of muscle can present with clinical features similar to those of dermatomyositis. Features that suggest a diagnosis other than dermatomyositis or inflammatory myositis include: family history of a similar illness, which may suggest inherited disorders; weakness related to exercise, eating or fasting, which may suggest metabolic causes; neurologic signs, cranial nerve involvement, and fasciculations (suggesting neurologic causes); severe muscle cramping (rhabdomyolysis, drug induced myositis); early atrophy or hypertrophy, suggesting the muscular dystrophies; and a creatine kinase level more than 100 times above the normal limit, suggesting a metabolic myopathy. **Table 2** lists some of the differential diagnoses.

## TREATMENT

Corticosteroids are the mainstay of treatment of dermatomyositis. Although controlled studies have not been published, initial treatment with high doses of corticosteroids is recommended: either pulse methylprednisolone therapy for 1 to 3 days or 1 to 2 mg/kg body weight of prednisone in divided doses. A high dosage of prednisone is maintained for 4 to 6 weeks, followed by a slow taper (5–10 mg/wk) over the next 10 to 12 weeks. Approximately 90% of patients improve partially with corticosteroid therapy, and 50% to 75% of patients achieve complete remission. If a patient experiences a relapse after remission or while on a low dose of corticosteroid, high doses of prednisone (1–2 mg/kg) must be restarted.<sup>1,2,4,5</sup>

During the corticosteroid tapering, other immunosuppressive agents, such as azathioprine, methotrexate, cyclophosphamide, and cyclosporine, can be used to control disease activities or facilitate steroid tapering. If therapy with these agents is not successful, plasmapheresis and intravenous immunoglobulin can be tried with various degrees of success reported. Recently, anti-tumor necrosis factor therapy has been tried in patients with dermatomyositis.

Therapy for dermatomyositis rash includes topical corticosteroids, oral hydroxychloroquine (200–400 mg/d), emollients such as petroleum jelly and Eucerin cream, and antipruritic lotions. Sunscreen is also recommended for protection against ultraviolet light, which can exacerbate the rash.

Physical therapy is an important modality of any treatment plan. Physical therapy can help patients recover from the weakness seen in active disease, as well as inactivity and deconditioning.

During therapy, disease activity must be monitored. Muscle disease can be assessed by testing muscle strength and measuring serum creatine kinase. Skin disease can be assessed clinically. Treatment duration will depend on response to therapy, and many cases require lifelong treatment. The prognosis for patients with dermatomyositis is excellent; the 5-year

survival rate is approximately 85% to 90%. Poor prognostic factors are older age and association with malignancy.<sup>2</sup>

## CONCLUSION

Dermatomyositis typically presents with proximal muscle weakness and skin rash, primarily heliotrope rash and Gottron's sign. Because of the associated risk of malignancy seen with dermatomyositis, a malignancy work-up is indicated at the time of diagnosis. Corticosteroids are the mainstay of treatment. The prognosis is good for most patients with dermatomyositis who have no associated malignancy. **HP**

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