

SKIN FOCUS

DERMATOMYOSITIS

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Dermatomyositis is a rare systemic auto-immune disease that manifests with proximal inflammatory myopathy and a characteristic cutaneous eruption. The cutaneous eruption is characterised by heliotrope erythema of the peri-orbital area. There is also erythema of the face, neck, and upper trunk. Violaceous papules called Gottron's papules occur over the knuckles. Dermatomyositis may be associated with polymyositis, interstitial pneumonitis and vasculitis.

Epidemiology and aetiology

Dermatomyositis occurs in a bimodal age distribution. It occurs in adults 45-50 years of age and affects women twice as often as men. The juvenile form is found in the age group 10-15 years.

The aetiology is unknown. Cases in persons over 55 years of age may be associated with malignancy of the breast, lung, ovary and gastro-intestinal tract (GIT).

Clinical presentation

Photosensitivity and muscle weakness may occur. Patients may have difficulty in rising from a supine position, raising arms over their head and dysphagia.

The signs of dermatomyositis include Gottron's papules on the dorsum of the knuckles, peri-orbital heliotrope colouration, photosensitive violaceous eruption and poikiloderma (which includes hyperpigmentation, hypopigmentation, telangiectasia and epidermal atrophy), as well as peri-ungual telangiectasia. Subcutaneous calcification occurs above the elbows in the juvenile form.

Muscle weakness occurs in the proximal limb/girdle distribution. Oesophageal muscles may occasionally be involved.

Differential diagnosis

1. Systemic lupus erythematosus
2. Mixed connective tissue disease
3. Airborne or allergic contact dermatitis
4. Photodrug eruption
5. Cutaneous T-cell lymphoma
6. Atopic dermatitis
7. Trichinosis

Evaluation

Patients with cutaneous eruption should have a skin biopsy performed to confirm the clinical impression of dermatomyositis.

Chemical pathology. During the acute active phase, there is increase in creatine phosphokinase and aldolase. However aspartate, alanine transaminase and lactate dehydrogenase are often elevated as well.

Electromyography (EMG). Over 90% of patients have an abnormal EMG result. Muscle biopsy is an important test for confirming the diagnosis and excluding other inflammatory myopathies.

MRI and ultrasound of muscle are sensitive tests that can add greatly to patient evaluation.

Chest X-ray. Interstitial fibrosis may be present in some patients.

Histology

Skin. Dermatomyositis shows epidermal atrophy with an interface dermatitis with increased dermal mucin.

Muscle. One should biopsy the shoulder or the pelvic girdle (the one that is tender). Findings include necrosis within muscle fibres, and inflammatory cells (histiocytes, macrophages, lymphocytes, plasma cells). Vasculitis may occur in juvenile dermatomyositis.

Diagnosis

Proximal muscle weakness with two of three laboratory criteria, i.e., elevated serum 'muscle enzyme' levels, characteristic electromyographic changes, diagnostic muscle biopsy.

Course and prognosis

Adults with dermatomyositis are more likely to have an internal malignancy than age-matched controls. The prevalence has been estimated at 6-50% with the greater prevalence among older patients. Most malignancies occur within 2 years of the diagnosis of dermatomyositis. Prognosis is good except in patients with malignancy or pulmonary involvement. Successful treatment of the neoplasm is often followed by improvement of dermatomyositis. Two-thirds respond to steroid therapy. Skin or muscle involvement can occur alone initially, followed at some time by the other.

Management

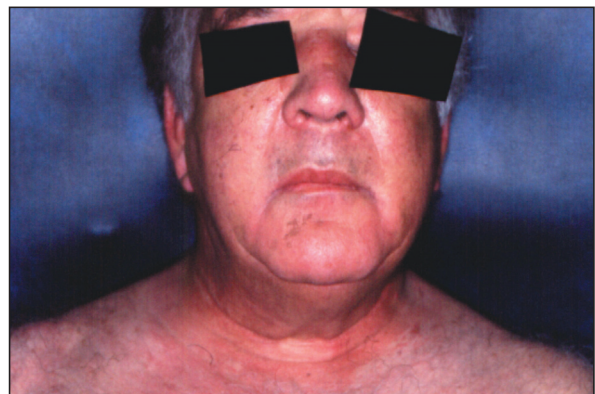
Systemic therapy

Prednisone. 1 mg/kg of body weight tapered to 50% over 6 months and stopped after 2-3 years. Best if combined with **azathioprine** 2-3 mg/kg per day.

Other treatments used. Low-dose weekly methotrexate, pulse cyclophosphamide and cyclosporin.

Cutaneous lesions

1. Sunscreens with high sun protection factor
2. Topical corticosteroids
3. Retinoids



Dermatomyositis presenting as photodermatitis.