2. Dermatomyositis

Dermatomyositis (DM) is a multisystem condition that falls into the larger category of inflammatory myopathies. The etiology of DM is thought to stem from an autoimmune reaction to internal (malignancy) or external (infections, drugs) factors in genetically predisposed individuals. The hallmarks of this condition are its characteristic cutaneous manifestations, proximal muscle weakness, and muscle inflammation.

There are adult and juvenile forms of DM. The adult form is associated with an increased risk of malignancy (10% to 50%), especially ovarian, breast, and lung cancer. However, the risk of malignancy might return to that of the general population 2 years from the onset of this condition. The juvenile form does not have an increased risk of malignancy but has an increased incidence of calcinosis cutis and small-vessel vasculitis.

Dermatomyositis affects women twice as often as men. Skin manifestations are the first presentation of disease in 40% of patients. The main characteristics of skin findings include heliotrope rash, Gottron papules, and Gottron sign. Heliotrope rash is a violaceous periorbital erythema, with or without edema of the eyelids. Gottron papules are a pathognomonic sign of DM. These dusky erythematous, flat-topped papules are most often found on the dorsal aspect of the knuckles, hands, and elbows, and might evolve to have depigmented white centres. Gottron sign is a violaceous discoulouration of the knuckles, elbows, or knees. Cuticular dystrophy and nail-fold telangiectasias are also commonly observed. Other cutaneous findings that have been described in DM, but which are nonspecific for the disease, include panniculitis, urticaria, and hyperkeratosis of the lateral fingers and palms, known as “mechanic’s hands.”

Systemic manifestations of DM include proximal muscle weakness, arthritis, arthralgia, dyspnea, dysphagia, dysphonia, and arrhythmia. Mortality is secondary to muscle weakness, cardiopulmonary involvement, or associated malignancy.

Diagnosis

The diagnosis of DM is based on the integration of clinical findings, elevated muscle enzymes (creatine kinase, aspartate aminotransferase, or lactate dehydrogenase), abnormal electromyography, or muscle biopsy. Magnetic resonance imaging might be useful in confirming inflammatory myopathy in patients without weakness. Serologic abnormalities are common and might support the diagnosis. Median prevalence of positive antinuclear antibodies in DM patients is about 40%. Anti-Jo-1 and anti-Mi-2 antibodies are highly specific for DM with respective median prevalence of 20% and 15% in DM patients.

Dermatomyositis might be difficult to differentiate from other skin conditions such as atopic or contact dermatitis, drug reactions, psoriasis, rosacea, seborrheic dermatitis, cutaneous T-cell lymphoma, systemic lupus erythematosus, and scleroderma. The diagnosis is often only confirmed with skin biopsy and muscle evaluation.

Management

Management of a DM patient should start with a systemic evaluation, including a chest x-ray examination, an esophageal motility study, a pulmonary-function test, and an electrocardiogram. Further investigations for malignancy are tailored based on age- and sex-specific risk factors. For example, mammography and endovascular ultrasound are recommended for female patients with DM. Investigations for occult malignancy are initially performed at diagnosis, then are repeated yearly for at least 3 years.

Patients with DM are usually followed by dermatologists, rheumatologists, or neurologists. A recent Cochrane review of immunosuppressant and immunomodulatory treatments for DM concluded that there have not been a sufficient number of high-quality studies to make firm recommendations regarding treatment. Currently, skin disease is managed with sun-protective measures, topical steroids, and systemic agents, including hydroxychloroquine (warn patients of increased incidence of drug reaction; ophthalmology follow-up required), low-dose weekly methotrexate, and intravenous immunoglobulin therapy.

Muscle and systemic disease can be managed with prednisone 1 to 2 mg/kg daily tapered to 50% over 6 months and to 0 over 2 to 3 years, steroid-sparing agents (azathioprine, methotrexate, cyclophosphamide, cyclosporine), or intravenous immunoglobulin 1 g/kg each day for 2 days monthly for at least 6 months.

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Competing interests

None declared

References