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The clinical features, diagnosis and classification of dermatomyositis

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ABSTRACT

Dermatomyositis (DM) is an idiopathic inflammatory myopathy (IIM) characterized by an inflammatory infiltrate primarily affecting the skeletal muscle and skin. Most common and peculiar cutaneous lesions include Gottron's papules, Gottron's sign and heliotrope rash. Different DM subsets have been identified until now encompassing classic DM, amyopathic DM, hypomyopathic DM, post-myopathic DM, and DM sine dermatitis.

Patients with DM have a higher incidence rate of malignancy than the normal population. In these patients cancer occurs in about 30% of cases with higher occurrence in men and in elderly people.

Bohan and Peter's diagnostic criteria, proposed in 1975, have been widely accepted and used until now. In the last ten years muscle immunopathology, myositis specific autoantibodies testing, and the use of new techniques of muscle imaging such as contrast-enhanced ultrasound or Magnetic Resonance Imaging have been introduced in the diagnostic work-up of patients with DM leading to the development of new diagnostic criteria.

1. Introduction

DM is an idiopathic inflammatory myopathy (IIM) characterized by an inflammatory infiltrate primarily affecting the skeletal muscle and skin with typical cutaneous lesions. Different DM subsets have been identified until now [1–3]. These includes classic DM, when muscular and skin involvement coexist, amyopathic DM (ADM), when the disease affects only the skin, hypomyopathic DM, when cutaneous manifestations of DM are associated with subclinical evidence of myositis, post-myopathic DM, when patients with previous classic DM present a recovery of myositis but skin rashes remain active, and DM sine dermatitis, when no rash is detected but histology feature of the muscular biopsy sample is indicative of DM.

DM affects both children and adults with an overall female/male ratio of about 2:1. Patients with IIMs have a higher risk of malignancy than the normal population which in DM occurs in approximately 30% of cases with a higher occurrence in men and in old age [1–4]. DM is a rare disease, although it seems to be the most common IIM in all age groups. The exact incidence and prevalence of the disease is unknown. The reported incidence of DM ranges from 1.2 to 17 new cases per 1,000,000 inhabitants with a prevalence between 5 and 11 cases per 100,000 individuals [2,3,5,6]. An increasing incidence from the 1940's until now has been observed which is probably due not only to a real increase in disease incidence but also to the development of classification criteria (Bohan and Peter's criteria [7,8] were published in 1975), as well as to new diagnostic tools which contribute to improving diagnostic capability.

2. Clinical features

The onset in DM may be acute (days) or insidious (several months). The cardinal muscular symptom is muscle weakness, mainly affecting the proximal muscles; myalgias can be observed less frequently. The most common clinical sign is the decrease of strength in the proximal muscles associated with contractures. Muscular atrophy (40% of cases) tends to appear late in the course of the disease. In severe cases, respiratory and oropharyngeal muscle involvement can cause dysphagia, respiratory difficulties and *ab ingestis* pneumonia.

Skin manifestations sometimes concur, but more often precede by several months or years muscle involvement [1–3,7]. Euwer and Sontheimer [9] proposed a classification in which DM skin manifestations are subdivided into pathognomonic, highly characteristic and compatible skin lesions (Table 1). The most common and peculiar manifestations, including Gottron's papules, Gottron's sign and heliotrope rash, are shown in Fig. 1.

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Table 1
Cutaneous manifestations of dermatomyositis [9].

Pathognomonic skin lesions of DM
1. *Gottron's papules*
   Papules having a violaceous hue overlying the dorsal-lateral aspect of interphalangeal and/or metacarpophalangeal joints. When fully formed, these papules become slightly depressed at the center which can assume a white, atrophic appearance. Associated telangiectasia can be present.
2. *Gottron's sign*
   Symmetrical macular violaceous erythema with or without edema overlying the dorsal aspect of the interphalangeal/metacarpophalangeal joints, olecranon processes, patellae, and medial malleoli.

Highly-characteristic skin lesions of DM
1. Periorbital violaceous (heliotrope) erythema with or without associated edema of the eyelids and periorbital tissue.
2. Grossly-visible periungual telangiectasia with or without dystrophic cuticles.
3. Symmetrical macular violaceous erythema overlying the dorsal aspect of the hands and fingers (where it can track the extensor tendon sheaths), extensor aspects of the arms and forearms, deltoids, posterior shoulders and neck (the shawl sign), V-area of anterior neck and upper chest, central aspect of the face and forehead.

Compatible skin lesions of DM
1. Poikiloderma vasculare atrophicans (poikilodermatomyositis)
Circumscibed violaceous erythema with associated telangiectasia, hypopigmentation, hyperpigmentation, and superficial atrophy most commonly found over the posterior shoulders, back, buttocks, and V-area of the anterior neck and chest.
2. Calcinosis cutis

2.1. Other clinical features

General symptoms include fever, malaise, weight loss and arthralgias. Raynaud's phenomenon is more common in patients with idiopathic DM and in DM associated with other connective tissue diseases. Cardiac involvement includes heart failure, left ventricular diastolic dysfunction, and hyperkinetic left ventricular contraction [10]. Interstitial lung disease (ILD) is commonly associated with anti-tRNA synthetase antibodies [11,12].

3. Diagnosis

Skin manifestations are easy to recognize by physical examination. Gottron and heliotrope rashes are DM specific manifestations and usually do not require histological confirmation. When muscle involvement is suspected, muscle biopsy is indicated before beginning treatment. Biopsy is usually performed in an area with active muscle involvement in the proximal muscles of legs or arms.

3.1. Electromyography

Needle electromyography provides a functional view of muscle damage. Although nonspecific, abnormalities may be observed in 70–90% of patients [2]. Increased spontaneous and insertional activity with fibrillation potential, complex repetitive discharges, positive sharp waves, small polyphasic motor units potentials, and early recruitment reflect ongoing muscle abnormalities. Late in the course of the disease, insertional activity may be decreased as a consequence of muscle fibrosis.

3.2. Muscle imaging

Muscle Magnetic Resonance Imaging (MRI) is the gold standard of the imaging study of muscle diseases, providing a detailed anatomic view of the extent of muscle involvement. In DM, T2-weighted images and short tau inversion recovery (STIR) show symmetric muscular edema, particularly in the musculature close

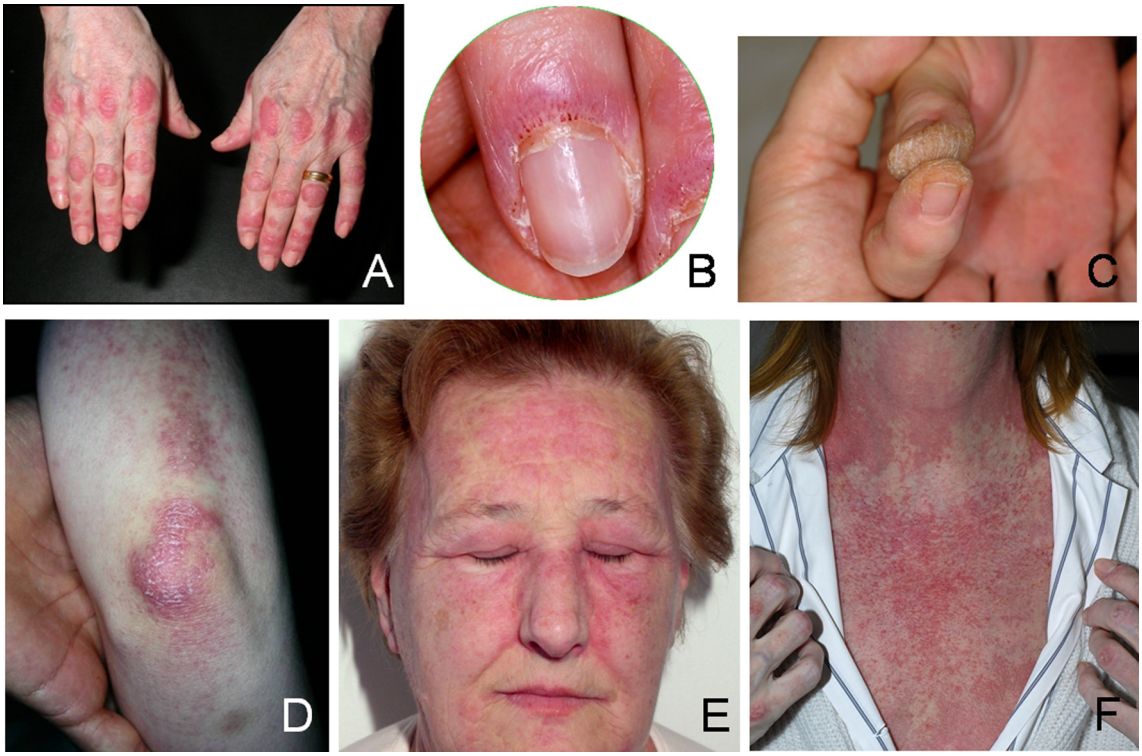

Fig. 1. Skin manifestation of DM A. Gottron's papules; B. Periungual telangiectasia with cuticular hemorrhage and distrophy C. Mechanic's hand; D. Gottron's sign; E. Heliotrope rash; F. Poikilodermatomyositis. For definitions see Table 1.