

# Dermatomyositis

Advances in Recognition, Understanding and Management

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The first detailed descriptions of patients with rare muscle disease (acute form of myositis) with cutaneous lesions were reported by E. Wagner [1] in 1863 and P. Potain (1875) [2]. These authors introduced to the medical community a new group of IIMs, characterized by the damage of many skeletal muscles and by skin manifestations. These disorders are rare, but increasingly recognized. They have a variety of clinical manifestations, immunological abnormalities and courses, and form a diverse group of diseases with unclear causes. The most common forms of these disorders are dermatomyositis (DM) and polymyositis (PM) [3].

In 1887, Professor Hans Unverricht published a description of a peculiar muscle disease with fatigue and malaise, muscle pain and weakness, swelling of face, and bluish lesions over eyelids [4]. He reported a 27-year-old stonemason, who developed acute onset of weakness, stiffness, and pain in the proximal arm, leg, and back muscles. A week later, diffuse swelling of face and extremities, accompanied by a low-grade evening fever and a bluish rash over his eyelids, had developed. Over the next few days the patient noticed shortness of breath, thickening of the voice, and dysphagia. After several weeks, the patient died with cyanosis and clinical manifestation of a pulmonary arrest. Autopsy showed fluid-filled lungs and swollen proximal muscles. Histopathological examination showed various stages of degeneration of muscle fibers, and focal round-cell interstitial infiltration of the affected muscles. In 1891, Unverricht reported a second case and gave a name to this disorder — **dermatomyositis** [5].

Up to the end of the nineteenth century, several authors published independently clinical descriptions of these disorders. Twenty-eight cases had been reported, and the name of the disease was coined variously as myositis universalis acuta, polymyositis, pseudotrichinosis, or DM [6–10].

In the first decade of the twentieth century, Jacoby observed a patient with skin atrophy, edema of the face and eyelids, arthralgias, and weakness of muscles, and reported the case as poikiloderma vascularis atrophicans [11].

In 1916 the first two cases of DM with association of malignancy (i.e., paraneoplastic DM) [12, 13] were published. Stertz [12] reported a patient of DM associated with gastric cancer, and Krenkeleit reported DM associated with breast cancer [13].

Petges and Clejat (1926) reported a case with idiopathic atrophic sclerosis of the skin, subcutaneous calcinosis, and myositis [14]. Gottron [15] in 1930 provided an extensive

description of cutaneous manifestations of DM, and introduced erythematous papules and macular lesions covering bony prominences as a specific hallmark of the disease. Mills in 1993 [16] noted that no extensive clinical experience was reported in English literature until that of O'Leary and Waismann (1940) from the Mayo Clinic, who analyzed 40 cases.

The first cases of DM in children were reported by Hecht in 1940 [17]. The vascular pathology of juvenile DM was initially recognized by Wedgewood et al. [18] in 1953. Later on, Everett and Curtis (1957) [19] and Banker and Victor (1966) [20], emphasized the differences between juvenile and adult DM.

Keil (1942) [21] was among the first who differentiated DM from systemic lupus erythematosus (SLE) and accepted the idea that cutaneous manifestations of DM may precedes the muscle disease.

The first cases of DM in Bulgaria were described in 1941 and 1943 by V. Ganev [22] and L. Popov [23].

Two periods are recognized in the development of the concept of DM in the twentieth century: the first is up to the mid-1970s, when the main parameters of the diseases were identified and the criteria for diagnosis of myopathy were formulated [24]. In 1977 Bohan and Peter defined five diagnostic criteria for the idiopathic nonsuppurative inflammatory disorders of striated muscles, which became useful for accurate diagnosis in clinical practice [24]. Although empirically derived, these criteria are useful to include cases within a well-defined range and to exclude patients in which the diagnosis may be in doubt in prospective and retrospective studies [25].

DM sine myositis was first described in six patients by Krain in 1975 [26], but the term amyopathic DM was introduced by Pearson in 1979 [27] for the patients who had typical cutaneous findings of DM but did not have any clinical or laboratory signs of muscle disease for at least 2 years after the onset of the skin pathology.

The second period in the development of the concept of DM in the twentieth century started with the determination of cutaneous signs and formulation of new clinical forms [28–32]. At the same time, some MSAs were identified [32–35], and new therapeutic modalities were introduced.

Serologic studies during the past 25 years have demonstrated the presence of autoantibodies in DM/PM patients, and have helped to classify some of these patients into specific subsets (i.e., the antisynthetase syndrome) [36]. Between 1976 and 1985 Reichlin, in collaboration with Nishikai, Arnett, and Targoff, detailed the multiplicity of autoantibodies in DM and PM, including recognition of anti-Mi-2 and anti-Jo-1 antibodies, in a series of articles [33–38].

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