AUTOIMMUNE CONNECTIVE TISSUE DISEASES

THE POLYMORPHOUS CLINICAL SPECTRUM OF DERMATOMYOSITIS: AN UPDATE

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Background: Dermatomyositis (DM) belongs to a group of rare autoimmune diseases characterized by a variable degree of skin symptoms and myopathy. The diagnostic hallmark of DM is the characteristic heliotrope rash, Gottron's papules and weakness of the proximal muscles. Along with pathognomonic, characteristic and compatible cutaneous features, several uncommon and rare skin manifestations have been described.
Wong-Type DM (WTDM) is a rare clinical subset of DM, characterized by the coexistence of DM and pityriasis rubra pilaris such as hyperkeratotic, erythematous, follicular confluent papules on the backs of the hands, arranged in a linear way over the bony prominences. Flagellate dermatitis has been also described as a rare manifestation in DM patients. Such manifestations can be also seen in adult-onset Still's disease, bleomycin-induced eruptions, and dermatitis from ingestion of shiitake mushrooms.
Recently described skin lesions are inverse Gottron's papules, and digital pulp ulcerations. Inverse Gottron's papules, a very rare manifestation of DM, are located on the palmar surface (as opposed to classical Gottron's papules) of finger interphalangeal joints and present as localized white triangular hyperkeratosis. Digital pulp ulcerations have been also rarely reported in DM; they are usually associated with vasculopathy or vasculitis, and with anti-MDA5 autoantibodies.

Observation: We reviewed the German-language and the English-language scientific literature using the key words "dermatomyositis", "auto-antibodies", and "clinical features" alone or in combination, focusing on particular cutaneous symptoms and their association to defined auto-antibodies profiles. Furthermore, we focused on peculiar clinical features of DM, including WTDM.

Key message: DM is known for its polymorphous skin manifestations. Several subsets of serum auto-antibodies have been described, which correlate with different clinical cutaneous and systemic features. Furthermore, several new clinical features have been recently described, including inverse Gottron's papules and digital pulp ulcerations.