

NEW ASPECTS IN DERMATOMYOSITIS ETIOPATHOGENY AND SIMPTOMATHOLOGY

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Abstract:

Dermatomyositis is part of major collagenosis, characterized as a nonspecific inflammatory process that interested striated muscles and skin (inflammatory myopathy). The disease is most common in female sex, sex ratio is 2:1, can occur at any age but most commonly between 40-60 years or children aged between 5 and 15 years the predisposing causes of the are: genetic predisposition, immunological abnormalities, infectious agents. Dermatomyositis is a disease that primarily affects the muscles and skin but may affect other systems and organs (digestiv system, respiratory system).

Key words: dermatomyositis, skin, rash, muscles.

Dermatomyositis is part of major connectivitis, characterized as a nonspecific inflammatory process that interested striated muscles and skin (inflammatory myopathy). According to the classification of Bohan and Peter, there are several types of inflamatory myopaties:
-Type I - primary idiopathic polymyositis (myositis without being accompanied by a rash)

-Type II - primary idiopathic dermatomyositis characterized by myositis and specific skin rash

-Type III - polymyositis and dermatomyositis associated with malignant neoplasia (paraneoplastic form).

-Type IV - juvenile dermatomyositis or polymyositis (myositis associated with rash in children, younger than 18 years)

-Type V - dermatomyositis or polymyositis associated with other collagen diseases (with SLE, sclerodermia, vasculitis) making Sharp's syndrome.

Subsequently, Witaker add the classification of Bohanwith type VI that includes various other types of myositis: eosinophilic myositis, localised nodular myositis. Recently studies have demonstrated the existence of a subgroup of patients who highlighted the presence of a typical rash but without the involvement of muscles. This condition was called amyopatic dermatomyositis(ADM).

The disease is most common in female sex, sex ratio is 2:1, it can occur at any age but it is most commonly between 40-60 years or in children, aged between 5 and 15 years. Predisposing causes of the disease are:
1) genetic predisposition, as evidenced by this atg. HLA DR3, DR 52, DR 6
2) immunological abnormalities

3) infectious agents: bacteria - mycobacterial, Rickettsia, parasites - Toxoplasma, Triquinella, Cysticercus, viruses - Echovirus, Coxsackie, HIV, hepatitis B, influenza virus, varicella-zoster virus.

4) drugs, Cyclosporine, Cimetidine, Zidovudine, Rifampicin, Hydroxyurea, Vincristine, Penicillamine, Colchicine, Corticosteroids, Danazol, etc. From the pathophysiological point of view, dermatomyositis is considered an autoimmune disease, the trigger mechanism being a lymphocytotoxic hyperergic reaction, addicted to antibodies and directed against skeletal muscle and skin. Dermatomyositis is characterized by various dystrophic and degenerated lesions and by lymphocytic infiltration of the muscle fibers and the dermal-epidermal junction. The perimysium is also infiltrated by the same cells (lymphocytes and macrophages) and it shows fibrinoid and mucoid degeneration. There were identified deposits of immune complex and C3 complement fraction at the level of dermal-epidermal junction.

Dermatomyositis is a disease that primarily affects the muscles and skin but may affect other systems and organs (digestive system, respiratory system). Pathognomonic for dermatomyositis are heliotrope skin rash and Gottron papules. The skin heliotrope eruptions are purple colored, located on the eyelids, forehead, nose and symmetric periorbital (giving of „the mask with glasses “).

The Gottron papules are light purple, located on the bony protuberances, at the level of metacarpal-falanx joints and proximal or distal interphalangeal joints. They are also metacarpal at the level of elbows, knees or feet. Another sign is a peri-nail rash which leads to a hypertrophy of the cuticula, with the appearance of small hemorrhagic infarction. Peri-nail teleangiectasia may be clinically apparent or may be discovered only by capillaroscopy (capillary microscopy). The skin also presents areas of skin atrophy, depigmented areas, pigmented areas and teleangiectasia giving of an aspect of poikyloderma. Skin and muscle calcinosis (calcium deposits in the skin and muscles) is usually encountered in juvenile dermatomyositis (40% of patients) and characterized by the presence of yellow nodule on the skin or bone protuberances. Muscle damage is the second sign of the disease and is reflected in a decrease of muscle force, predominantly in the proximal muscles.

In conclusion, dermatomyositis prognosis depends of myopathy severity, and the association with a malignant disease and / or cardiopulmonary affection. Even in patients whose disease is favorable to healing, a muscle residual weakness still persists.

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