Dermatomyositis

Dermatomyositis a polymyositis are idiopathic chronic inflammatory diseases of the striated muscles. They are manifested by symmetrical muscle weakness, mainly in the proximal part of the upper and lower limbs. Typical skin symptoms may also accompany dermatomyositis. It occurs mainly in childhood or in the fifth decade and affects women more often.

Etiology

The cause is unidentified. One possibility could be a autoimmune reaction against skeletal muscle, most likely triggered by an infectious agent. In 40-80%, we find autoantibodies against the nuclear and cytoplasmic components of cells (Anti Mi-2, anti-Jo-1). The disease's occurrence is mostly observable during viral infections (coxsackies, echoviruses, paramyxoviruses, herpes viruses) - viruses trigger a defensive reaction, which later degenerates into an autoimmune process. There is also an increased incidence of neoplasia.

Clinical picture

The disease begins with flu-like symptoms, arthralgias and Raynaud's phenomenon. In the acute phase, red-violet eyelid swelling - heliotropic erythema - may occur. Gottron's papules are purple erythematous spots up to teleangiectasia most often found on the dorsum of the hand. Area livid erythema also on the forehead, cheeks, neck and upper torso; photosensitivity is present; the dominant feature is progressive muscle weakness and soreness - especially the proximal muscles of the limbs, torso and neck (typical difficulties when combing, placing objects in height); in severely ill patients, dysphagia is added - swallowing of the swallowing muscles; dyspnoea in respiratory muscle damage.

Rare form: *amyopathic dermatomyositis* – typical skin symptomatology without muscle damage

Complication

Pulmonary involvement (aspiration pneumonia, interstitial pulmonary fibrosis), joint involvement, rarely the heart.

Diagnosis

1. Symmetrical proximal muscle weakness
2. High creatine kinase (MM), aldolase or myoglobin
3. EMG signs of myopathy
4. Muscle biopsy with signs of inflammatory myopathy
5. Typical skin manifestations

Differential diagnostics

Muscular dystrophies, myopathies - drug, toxic, endocrinological; toxoplasmosis, trichinosis

Therapy

Total corticoids, possibly with immunosuppressants (MTX)

Progress

In half of the cases, complete remission in children and chronic treatment is usually necessary for adults.

Malignant disease is present in 15-20%!

Links

References

- Jiří Štork et al. Dermatovenerologie. Galén, Karolinum. 2008;
- presentation on the website http://kozni.lf1.cuni.cz;
- lectures by teachers at the Department of Dermatovenerology, 1st Faculty of Medicine, Charles University and General Hospital;
- Kuba Holešovský, otázky z dermatovenerologie