Myasthenic Syndrome

Myasthenia literally means **muscle weakness. Myasthenic syndrome** is the name for a set of symptoms characteristic of muscle activity disorders caused by damage to the neuromuscular disc. This includes autoimmune diseases - Myasthenia gravis, Lambert-Eaton syndrome (including paraneoplastic syndromes, e.g. in small cell carcinoma), and also botulinum toxin, which affects transmission on the neuromuscular disc.

Clinical picture

A typical manifestation is **muscle fatigue** and **weakness**, which is particularly pronounced after exertion. It can also affect respiratory muscles and phonation . Sensory impairment and pain are not manifested. During the examination, the tone and reflexes are normal, the muscles are not atrophic.

Damage to the respiratory muscles can lead to a life-threatening myasthenic crisis accompanied by respiratory failure.

Symptoms are at their peak in the evening. Transient paresis may occur after repeated movements, which disappear after a certain period of rest.

Eyelid ptosis in a patient with Myasthenia gravis

Comparison of myasthenia and myopathy

Parameter	Myasthenia	Myopathy
Name	asthenia=weakness	pathy=disorder
Level of disorder	neuromuscular disc	muscle
Location	generalized	proximal parts of limbs
Sense impairment	no	no
Pain	no	yes
Atrophy	no	yes
Fasciculation	no	no
Proprioceptive sense	normal	normal or reduced
Tone	normal	reduced

Diagnostics

- EMG;
- antibodies to ACh;
- antiMuSk antibodies;
- muscle neck test, where the patient does not keep the chin at the sternum;
- a test of muscle strength, when the patient opens the HK and we count the time it takes for the limbs to drop.

Treatment

Symptomatic

- Cholinesterase inhibitors: pyridostigmine, neostigmine
- regimen measures: do not exert yourself, ventilation support
- do not use ATBs that reduce MG, or benzodiazepines

Immunomodulatory

- corticosteroids: prednisone, prednisolone, methylprednisolone (necessary to think about NU - steroid diabetes, ulcer disease, osteoporosis, depression, hypertension, Na retention, Cushing's syndrome)
- immunosuppressants: azatiprine (most used), cyclosporine A, mycophenolate mofetil, tacrolimus, cyclophosphamide, methotrexate, rituximab,

Intravenous immunoglobulins

Drug of choice in myasthenic crisis. Improvement within a week.

Therapeutic plasmapheresis

The effect lasts 4-10 weeks. The disadvantage is venous access and non-selective removal of blood proteins. A variant of Plasmapheresis can be immunoadsorption, when Ig is removed, or only some class of Ig.

Thymectomy

Increases the likelihood of remission or improvement. It also reduces overall mortality.

Links

Related articles

- Myopathic syndrome
- Myasthenia gravis
- Neuromuscular diseases
- Neuromuscular diseases (pediatrics)

Reference

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2. ↑ Skočit nahoru k:a b JECH, Robert. *Základy obecné neurologie* [přednáška k předmětu Neurologie, obor Všeobecné lékařství, 1. LF UK v Praze]. Praha. 14.05.2012. Dostupné také z <https://www.neuro.lf1.cuni.cz/vyuka/soubory/5r/Obecna_neurol_CZ.pdf>.

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