Myasthenia gravis

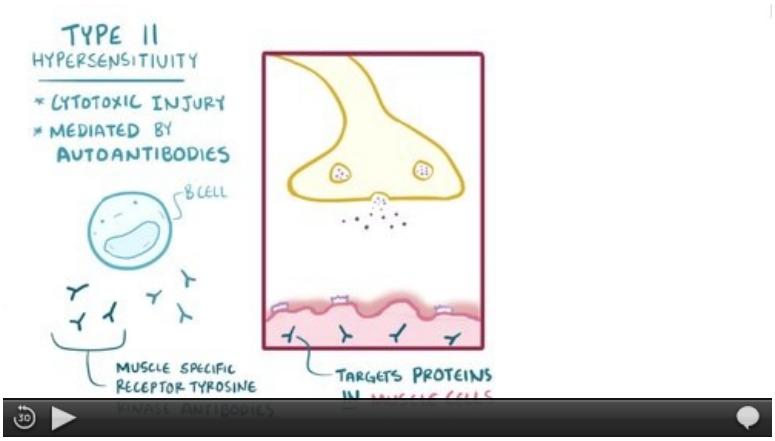
Myasthenia gravis is an autoimmune disease , the cause of which is a disturbance in the transmission of excitement between the nerve and the muscle in the area of the neuromuscular plate.

Pathogenesis

In *acquired myasthenia*, the cause is a humoral autoantibody that binds to the postsynaptic receptor for acetylcholine and destroys it. The formation of autoantibodies is caused by acetylcholine-positive ectopic muscle cells in the thymus.

In the case of the **congenital form**, the immunological character of the disease was not proven. It is a presynaptic or postsynaptic neuromuscular transmission defect.

The neonatal form is a transitory form based on the transplacental transfer of antibodies from maternal blood to the fetus. It occurs in approximately 12% of children born to mothers with myasthenia gravis.



Video in English, definition, pathogenesis, symptoms, complications, treatment.

Clinical division

Juvenile form

Ptosis of eyelid

It manifests itself until puberty, but rarely before the age of 5. It is characterized by **abnormal fatigue of muscle function.**

First, the motor cranial nerves are affected: diplopia, unilateral ptosis of the eyelid, drooping of the corners of the mouth, reduced facial expression, dysphagia, weak voice, dysarthria. Weakness of other skeletal muscles gradually sets in, which becomes more pronounced after exertion and in the evening. In some patients, the disease manifests itself in the insufficiency of the respiratory muscles and the diaphragm.

Myasthenic crises with failure of the bulbar and respiratory muscles, which are often provoked by an infectious process, represent the greatest danger.

The incidence of autoimmune diseases, e.g. thyrotoxicosis, is higher. Thymoma occurs in 10–15%.

Neonatal myasthenia

It usually manifests itself already in the first 48 hours after birth with a disorder of the sucking reflex, a weak cry and weakness of the limbs. It can result in respiratory insufficiency.

Congenital myasthenia

It manifests itself from birth to 2 years. Eye muscle paresis dominates.

Familial infantile form

It is characterized by episodes of severe respiratory insufficiency at birth or in infancy. Responds well to treatment with acetylcholinesterase blockers.

Diagnostics

- **medical history** and **clinical neurological examination** ;
- **EMG** – rapid reduction of the amplitude of action potentials after repeated stimulation;
- **pharmacological tests** – tensilone and prostigmine test;
- **determination of antibodies against acetylcholine receptor or anti-MuSK antibodies ;**
- CT of the mediastinum to rule out thymoma.

Differential diagnosis of elevation of "muscle" enzymes

- **skeletal muscle involvement** → increased CK-MM, increased LDH 2, CK/AST > 10, increased S-myoglobin;
- **myocardial involvement** → increased CK-MB, increased CK-MB/CK ratio, increased LDH 1, increased troponin T;
- **acute liver damage** → increased ALT, AST and LDH 5.

Therapy

We try to stabilize the patient with anticholinesterase drugs (neostigmine, pyridostigmine), while paying attention to overdose and the occurrence of cholinergic crises (= nausea, vomiting, sweating, salivation, flushing, bradycardia, miosis, diarrhea). In case of hypotension, slow administration of atropine is also recommended.

In the case of a thymoma and an unstable form of myasthenia, we prepare the patient for thymectomy, which has a positive effect on the course of the disease in juvenile myasthenia. It is controversial in the infantile and congenital form. If the desired effect does not occur after thymectomy, we will consider appropriate immunosuppression (corticoids, azathioprine, methotrexate, ciclosporin A). In case of failure of this treatment or in the case of refractory myasthenic crisis, we proceed to the administration of intravenous immunoglobulins or plasmapheresis, which is a life-saving procedure in the most severe conditions.

Myasthenic crisis

This is a life-threatening condition. Diplopia, eyelid ptosis, dysarthria, dysphagia, inability to hold limbs and the worst is respiratory muscle weakness when the patient cannot breathe on their own. In the case of a myasthenic crisis, the patient must be intubated and provided with UPV. IVIG or plasmapheresis are given in crisis.

Links

Related articles

- Neuromuscular diseases

References

- AMBLER, Zdeněk. *Základy neurologie : [učebnice pro lékařské fakulty].* 7. edition. Galén, c2011. ISBN 9788072627073.

External links

- Myasthenia gravis (czech wikipedia)
- Myasthenia gravis (english wikipedia)

Source

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