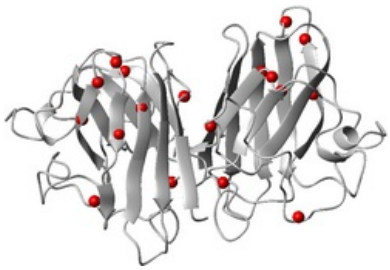
Amyotrophic Lateral Sclerosis

Amyotrophic lateral sclerosis (ALS) is a systemic neurodegenerative disease, which affects all **motor neurons** of a body. Motor neurons in a spinal cord are called *"lower"* and these in a brain are called *"upper"* . ALS destroys both of them and patients gradually lose their ability to do voluntary movements. ALS is also called *Charcot´s disease* or *Lou Gehrig's disease*.

ALS is one of the most common motor neuron disease, because its incidence is 5 per 100,000 people. Typical age of the onset is between 40 and 60 years and men are affected twice as likely. ALS is a fatal disease and people usually die within 3 or 4 years.

Etiology

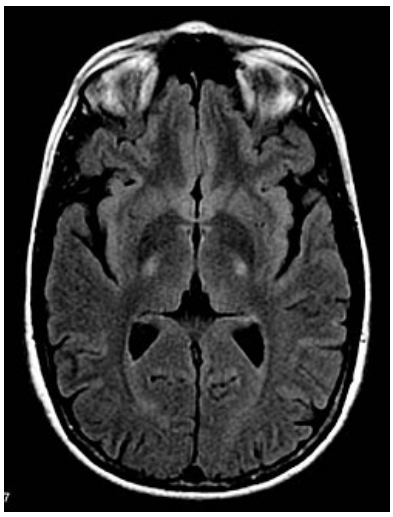
There are few opinions why ALS occurs, but he fact is that the reason of motor disabilities is **motor neurons death**. Why are they going under the apoptosis? We are still not sure. In affected motor neurons we can find the increased level of **ubiquitin** and edema of a neuron body. In patients with ALS were found ubiquitin antibodies, which can probably attack the cells. Autoimmunity is higly important in ALS syndromes, which are connected with some neoplasms.



SOD1 investigated mutations

Another theory suppose an influence of excitatory aminoacids - especially **glutamate** - and their higher extracellular concentration, which also kill motor neurons.

From 5 to 10 percent of ALS are **genetic-based** (familiar form). In 1993 the mutation of gene, which coded superoxiddismutase 1 (SOD1), was discovered in these people. This enzyme is one of the most powerful antioxidant in our body and protect our cells from oxygen radicals.



MRI - increased T2 signal in capsula interna (typical for ALS)

Symptoms

ALS starts very inconspicuously on small muscles of the hand (*musculi interossei*). The first affected muscle is usually *m. adductor pollicis*. At the beginning we can see **fasciculations**, **atrophies**, **hyperreflexia** (mainly masseter reflex) and **irritative phenomena**, which are the typical signs of **periferal motoric lesion** (although ALS is a combined disability of central and periferal motor neurons). Typical is **muscle weakness**.

One of the most important symptom is located on a tongue and can help us to diagnose ALS. As a reaction on damage of bulbar region the fasciculations and atrophy of the tongue occur at the beginning. In later stage patients are **not able to stick the tongue out**! This fact has to warn us that they suffer from ALS. Thanks to bulbar region atrophy the patients have also problems with **swallowing**, **speech** (*dysarthria*). Swallowing problems leads to **kachexia**, so we have to use nasogastric tube or PEG (percutaneous endoscopic gastrostomy) to feed them.

As the disease progresses people lose their ability to move their arms, legs or body and can communicate just by their eye movements. Unfortunatelly their **mental health** remains normal and they appreciate everything. Depressive feeling are normal for them, so their family support can be usefull.

Diagphragma is also just a muscle, which is controlled by motor neurons. When the progression of ALS affects it, people die because of **breath insufficiency**.

Remember that the patients have to have both of the motor neurons symptoms - **central** (spasticity, irritative phenomena, hyperreflexia) and also **periferal** (atrophy, fasciculations, muscle weakness). ALS is a combined disability!

Diagnosis

There is **no specific test**, which can assure us about ALS, so we have to based our diagnosis on specific symptoms. **Electromyography** (EMG) can be used to differential diagnosis - ALS is connected with high-voltage potentials of motor units. **MRI** in patients with ALS is normal, but we can exclude different origin of the symptoms, which can be for example some tumors or cervical myelopathy.

Also some infectious diseases (HIV or Lyme disease) can look like ALS, so we have to make **blood and urine tests** to find some inflammatory elements.

However, the **conventional neurological examination** can help us the most!

Therapy

There is **no specific therapy of ALS**, so people die on within 3-4 years after its onset. However, we can help them with the symptoms. PEG and tube for nutrition or rehabilitation can relieve their situation. There is also one medication, which reduces the production of oxygen radicals, called *riluzol*.

Links

Related articles

- Neurodegeneration
- Motor Neurons
- Link Title
- Motoric Lesions
- Irritative Phenomena
- PEG
- Neurological Tumors
- Cervical Myelopathy
- Electromyography
- MRI
- Ubiquitin
- Oxygen Radicals

External links

- National Institute of Neurological Disorders and Stroke (http://www.ninds.nih.gov/disorders/amyotrophiclateralsclerosis/detail_ALS.htm)
- Neurology.org (http://www.neurology.org/content/68/13/E17.full)

Bibliography

- JEDLIČKA, KELLER,, et al. *Speciální neurologie.* 1st edition. 2005. ISBN 80-7262-3142-5.