Primary Lateral Sclerosis

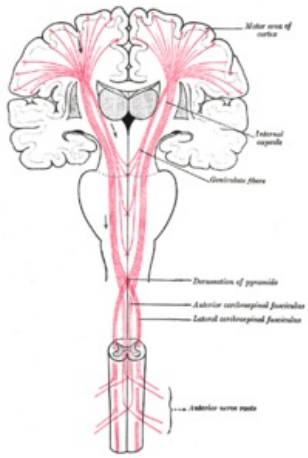
Primary lateral sclerosis(PLS) is a rare neuromuscular disease, which is sorted into motorneuron diseases. It affects the upper motorneuron – corticospinal tract – which is the main control centre of motoric movement of the muscles. So the movements of the patient are very slow and hard for them.

A typical patient is usually between 40 and 60 years old and men are affected twice likely than women. The onset of PLS should be very variable, but its progression is very slow and can take also decades. Primary lateral sclerosis is not a fatal disease, althought the patient usually need some medical help – rehabilitation, wheelchair or some assistive help.

Etiology and Patogenesis

There is one special form of PLS, which occurs in childhood, called **juvenile primary lateral sclerosis**. Nowadays, we know that there is a gene transmited from parents to their children, called *ALS2*. These gene is responsible for protein called "alsin". If there is some mutation present, the protein is not stable, but we still do not know how does this influece the motoric neurons. The onset of the juvenile form is between 15 and 20 years. The pattern is autosomal recessive.

Patogenesis is based on a neurodegeration of upper neurons in spinal cord and also in the brain. **The lower motorneuron is not affected**, so there are no fasciculations. It is important in differential diagnosis of amyotrophic lateral sclerosis, which affect both the lower and upper.



Tractus corticospinalis

Symptoms

- muscle weakness (leg, arms and tongue)
- spasticity (*hypertonia*)
- balance problems
- slow movements
- depression
- slurred speech
- stiffness
- hyperreflexia (Babinski´s sign present)

Diagnosis

There is **no specific test** for primary lateral sclerosis. Diagnosis of PLS is a long-term matter and we usually have to monitor the patient for 3 or more years, if we want to know if we are right. At the beginning primary lateral sclerosis should be mistaken for any other motorneuron disease (e.g.:Amyotrophic lateral sclerosis). In fact we have to mainly exclude other diseases, so it is called diagnosis "*per exclusionem*". For this reason we can also use some specific methods (MRI, CT or EMG) for the differential diagnosis (tumour, muscle diseases, infections).

Therapy

As with other motorneuron diseases there is no specific treatment of PLS. The symptomatic therapy includes some **medication**, as central myorelaxants (baclofen), analgetics (against pain) or anti-depressant. **The physical therapy** is also important, because we want to prevent joint contractures and muscle atrophy.

Links

Related articles

- Motor Neurons
- Motorneuron Diseases
- Hypertonia
- Amyotrophic Lateral Sclerosis

- MRI

External links

- NINDS (http://www.ninds.nih.gov/disorders/primary_lateral_sclerosis/primary_lateral_sclerosis.htm)
- Mayo Clinic (http://www.mayoclinic.com/health/primary-lateral-sclerosis/DS01115)
- Genetics Home Reference (http://ghr.nlm.nih.gov/condition/juvenile-primary-lateral-sclerosis)