

the muscles of the arm and shoulder will be involved. The spread of dysfunction is towards the spine. Only in the progressive stages there will be paralysis of the lower limb muscles as well as the upper limbs – here it is a lesion of both upper and lower motor neurons (the period around 5–6 years from the beginning of illness).

In cases of **amyotrophic lateral sclerosis** the main affected neuron was the upper motor neuron. The so called lateral sclerosis means the degeneration of the pyramidal tracts and the cortical motor cells are involved as well. The lesion will be manifested with motor disturbances of the limbs and from the early stages, disturbances of muscles of the face and neck. The corticospinal tract will gradually degenerate and gliosis will appear. Only in rare cases there is a pure injury of the upper motor neuron. When the disease progresses there will be a consequent lesion of the lower motor neuron.

6.21.1 Progressive bulbar paralysis and pseudobulbar paralysis

Lower motor neuron lesion with fasciculation and atrophy and upper motor neuron lesion without muscular atrophy. In these diseases and as shown from the headline the condition is worsened by the progressing degeneration of the motor neurons. A critical condition occurs when the bulbar nuclei are involved, and as a result of this the organism is unable to remove the secretions from the respiratory tract, and there will be an aspiration bronchopneumonia and sometimes there will be an acute asphyxia.

6.22 Peripheral nerves

Peripheral nerves can be classified into different groups according to some criteria – e.g. into **myelinated and non myelinated fibers**, according to nerve thickness, into **sensory and motor** etc. In most diseases all those groups are affected, but we know some diseases that are selective for certain types of nerve fibers.

Neuropathies – this term is preferred more than neuritis, because most of those diseases are not inflammatory in type. We might divide them into parenchymal and interstitial.

In **parenchymal neuropathies** the axon and the myelin sheath are the primary affected parts (the myelin sheath is composed of Schwann cells). Most of those diseases are consequences of toxic and nerve fibers – **polyneuropathies**. According to the etiology we can divide those diseases into:

1. **toxic** (diphtheria, arsen, some drugs)
2. **current** (e.g. beri-beri due to vit B deficiency)
3. **metabolic** (diabetes mellitus, porphyria, metabolic leukodystrophy in some malignancies, uremia).

Degeneration can be of two types: first of all affects the axon with the consequent demyelination (similar to the Wallerian degeneration upon neurotmesis) or there will be a primary degeneration of the Schwann cells with the consequent axonal degeneration. The diseases might of course affect only one nerve, or one group of nerve fibers. In interstitial neuropathies the blood vessels and connective tissue are the primary part affected (epineurium, perineurium and endoneurium). In this type of diseases only certain types of nerves or nerve groups can be affected according to the type of the disease. The most common is neuropathy. Nerve fibers can be injured during their compression. The most common cause of injury is ischemia. The carpal tunnel syndrome is an example, where the median n. is compressed between the carpal bones and ligaments. The cranial nerves are vulnerable to injury in cases of increased intracranial pressure, and this might end up with their degeneration. The degenerative ischemic changes of the nerve fibers can occur due to arteritis. No neuronal changes occur in the area of vasculitis, yet those changes present in the area supplied by this vessel (below the vasculitis). In the area of neuronal degeneration, the inflammatory changes are not always present.