transversal lesion of the spine all the long tracts degenerate upwards (sensory tracts) and dawnwards (motor tracts). The most common example of **a descending degeneration** is the condition following brain infarction in the area of internal capsule. The degeneration spreads from the lesion along the corticospinal axons till their terminals in the anterior horns of the spine. After a period of time we might register this degeneration in the form of demyelination along the whole corticospinal tract. Those changes actually involve the whole pyramidal system. After a long time there will be gliosis and scarring of the tissue.

### 6.20.2 Injury of the motorpathways

The division of motor neurons and pathways into **the upper motor neuron** (UMN) and **the lower motor neuron** (LMN) based on anatomical and functional characteristics has got a great clinical and diagnostic value.

**The upper motor neuron** is formed of the cortical motor neurones (the precentral gyrus), the motor pathways in the brain that (pass through the internal capsule), motor pathways in the brain stem (where decussation of tracts on to the contralateral side take place) to meet the cranial nerves of the contralateral side. This crossing takes place in the pyramidal decussation, where the anterior corticospinal tract (non crossed) is separated from the pyramidal tract (being the crossed part of the motor pathway).

**The lower motor neuron** is composed of the carnial nerve nuclei (the motor nuclei) and their axons in the carnial nerve fibers, as well as the anterior horn cells with their axons in the spinal nerves. It is important to realize that during the long course of the motor neuron from the cerebral cortex till the anterior horn of the spine represent the upper motor neuron and this could be injured by multiple disease processes. The lower motor neuron can be injured in the spine, as well as in the peripheral nerves.

The most common cause of injury to the UMN is hemorhage in the area of internal capsule, different levels of this pathway could be involved in cases of multiple sclerosis or in case of other diseases that cause demyelination of the motor from the cortex till the spinal cord and its segments.

A typical example for LMN injury is poliomyelitis that affects the motor neurons in the anterior horn of the spine, or a peripheral neuropathy that destroys axons and their coverings and hence results in disturbances of transmission.

In both cases i.e. upper motor neuron and lower motor neuron lesion (UMNL and LMNL) the clinical picture will reveal paralysis, yet there are some important changes that differ in their quality and spectrum of the present reflexes in both the mentioned cases. When the lesion is in the upper motor neuron (central paralysis) the lower motor neurons will escape the control of the higher centers so the muscle tonus will increase, the tendon and other spinal reflexes are increased, and the extensory (Barbinsky) reflex will appear. This is why we call this type of paralysis the spastic type. Following a lower motor neuron lesion there is actually an error in the connection to the effector, so the reflexes are absent, and there will be muscular atrophy. This type is known as **the hypotonic paralysis**.

## 6.21 Diseases of the motor neuron

Are usually of unknown etiology. They appear more frequently in adult patients and mainly in men. The basic lesion is a progressive degeneration of the cortical and the spinal motor neuron, that will be manifested by their dysfunction. The variability of symptoms depends on the ratio of affected upper motor neuron to the affection of the lower motor neuron, and on the site of the clinicial lesion. The evaluation of this ratio is possible in the initial stages of the disease when the changes are bound to a certain localization, but later on there will be a diffused affection of tissue and here the differentation becomes much harder.

**A progressive muscular atrophy** is the main sign of neuronal atrophy (mostly due to lower motor neuron injury) due to the degeneration of neurons in the anterior horn. This degeneration worsens within many years, and the cells will become necrotic. Signs and symptoms of the initial stages appear within (1–4) years and are manifested as the affection of fine movement of the fingers, their fasciculation, later on the muscles of the hand will be affected, and at last

the muscles of the arm and sholder 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 illnes).

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 prefered more than neurities, because most of those diseases are not inflammatory in type. We might devide them into parenchymal and interstitial.

In **parenchymal neuropathies** the axon and the myelin sheeth are the primary affected parts (the myelin sheeth is composed of Schwann cells). Most of those diseases are consequences of toxic and nerve fibers – **polyneuropathies**. According to the etiology we can devide 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, metachromic 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 vesseles 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 syndrom is an example, where the median n. is compresed 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.