CLINICAL FOCUS

General CNS Disorders

INFECTIONS

Encephalitis (en-sef-ă-lī′tis) is an inflammation of the brain; it is most often caused by a virus and less often by bacteria or other agents. A large variety of symptoms may result, including fever, paralysis, coma, or even death.

Myelitis (mī-ĕ-lī′tis) is an inflammation of the spinal cord caused by trauma, multiple sclerosis, or a number of infectious agents, including viruses, bacteria, or other agents. A large variety of symptoms may result depending on the extent and level of injury or infection.

Meningitis (men-in-jī′tis) is an inflammation of the meninges. It may be virally induced but is more often bacterial. Symptoms usually include stiffness in the neck, headache, and fever. Pus may accumulate in the subarachnoid space, block CSF flow, and result in hydrocephalus. In severe cases, meningitis may also cause paralysis, coma, or death.

OTHER CNS DISORDERS

An **aneurysm** (an′ū-rizm) is a dilation, or ballooning, of an artery. The arteries around the brain are common sites for aneurysms, and hypertension can cause one of these "balloons" to burst or leak, causing a hemorrhage around the brain. With hemorrhaging, blood may enter the epidural space (epidural hematoma), subdural space (subdural hematoma), subarachnoid space, or the brain tissue. Blood in the subdural or subarachnoid space can apply pressure to the brain, causing damage to brain tissue. Blood is toxic to brain tissue, so blood entering the brain can directly damage brain tissue.

A **concussion** is a blow to the head producing momentary loss of consciousness without immediate detectable damage to the brain. Often, no more problems occur after the person regains consciousness; however, in some cases, **postconcussion syndrome** may occur a short time after the injury. The syndrome includes increased muscle tension or migraine headaches, reduced alchol tolerance, difficulty in learning new things, reduction in creativity; as well as motivation, fatigue, and personality changes. The symptoms may be gone in a month or may persist for as much as a year. In some cases, postconcussion syndrome is the result of a slowly occurring subdural hematoma that may be missed by an early examination. The blood may accumulate from small leaks in the dural sinuses.

Cerebral compression may occur as a result of hematomas, hydrocephalus, tumors, or edema of the brain, which can occur as a result of a severe blow to the head. The intracranial pressure increases, which may directly damage brain tissue. The cerebellum may compress the fourth ventricle, blocking the foramina and causing internal hydrocephalus, which further increases intracranial pressure. The greatest problem comes from compression of the brainstem. Compression of the midbrain can kink the oculomotor nerves, resulting in dilation of the pupils with no light response. Compression of the medulla oblongata may disrupt cardiovascular and respiratory centers, which can cause death. Compression of any part of the CNS that results in ischemia for as little as 3–5 minutes can result in local neuronal cell death. This is a major problem in spinal cord injuries.

CLINICAL FOCUS

Peripheral Nervous System Disorders—Cranial Nerves

General issues of PNS disorders are described in chapter 12. This chapter addresses only those specific to the cranial nerves.

Trigeminal neuralgia, also called tic douloureux, involves one or more of the trigeminal nerve branches and consists of sharp bursts of pain in the face. This disorder often has a trigger point in or around the mouth, which, when touched, elicits the pain response in some other part of the face. The cause of trigeminal neuralgia is unknown.

Facial palsy (called Bell palsy) is a unilateral paralysis of the facial muscles. The affected side of the face droops because of the absence of muscle tone. Facial palsy involves the facial nerve and may result from facial nerve neuritis. The facial nerve passes from deep to superficial through the parotid gland. Although the cause of facial palsy is often unknown, it can result from a stroke or tumor in the cerebral cortex or brainstem (see chapter 14). Temporary facial palsy can result from inflammation of the parotid gland or from anesthesia accidently introduced into the gland during dental anesthesia. Temporary facial palsy can even result from extreme cold in the face, where the superficial branches of the facial nerve are located.

INFECTIONS

Herpes simplex I is usually characterized by one or more lesions (sores) on the lips or nose. The virus apparently remains dormant in the trigeminal ganglion. Eruptions are usually recurrent and often occur in times of stress or of reduced resistance, such as during a case of the common cold. For this reason, they are called cold sores or fever blisters.

CLINICAL GENETICS

Neurofibromatosis

Neurofibromatosis type 1 (noor′ō-fī-brō-mă-tō′sis; von Recklinghausen disease) is an autosomal-dominant trait localized to chromosome 17. Neurofibromatosis type 1 is characterized by hyperpigmented skin present at birth and by multiple benign tumors (neurofibromas), which grow on nerves just under the skin and along nerves throughout the body. The skin of affected individuals is covered with characteristic large, flat, dark brown spots called café au lait spots, which are a type of birthmark. The neurofibromas increase in size and abundance with age and can cause severe disfigurement. Neurofibromatosis type 1 results from a mutation in the *neurofibromin 1* (NF1) gene, which encodes the protein neurofibromin. Neurofibromin is produced by neurons and glial cells, such as Schwann cells and oligodendrocytes. Neurofibromin is a tumor suppressor protein, which means it plays a role in keeping cells from growing uncontrollably. A mutated NF1 gene produces a defective neurofibromin protein, which then allows the glial cells to grow unchecked and form neurofibromas. Neurofibromatosis type 1 has an incidence of 1:2500–3300 individuals. Approximately 50% of the mutated genes are inherited from an affected parent, but the remaining 50% are caused by new mutations. Two mutated copies of the gene are necessary to cause the disorder, but most individuals born with one NF1 mutation eventually develop a second mutation and begin to develop neurofibromas and café au lait spots.

A second form of neurofibromatosis, **neurofibromatosis type 2** (NF2), has the same inheritance characteristics of NF1, but is much rarer (incidence of 1:50,000–120,000 individuals). NF2 is localized to chromosome 22 and encodes the protein merlin (schwannomin), which is also a tumor suppressor protein. Individuals with neurofibromatosis type 2 do not develop café au lait spots. Instead, the disease is characterized by the development of bilateral tumors surrounding the vestibular division of CN VIII (vestibular schwannomas). Tumors on CN VIII cause ringing in the ears (tinnitus), hearing loss, and vertigo from the pressure of the tumor on the nerve as it travels through the internal acoustic meatus. Treatment of neurofibromatosis consists of surgery to remove tumors that are causing the patient severe pain, loss of function, or disfigurement or to remove tumors that are thought to have become malignant.