Syringomyelia

Syringomyelia is a designation for the presence of a cavity in the spinal cord filled with fluid , such a cavity is called a syrinx. The disease is manifested to a greater or lesser extent by a neurological deficit, which tends to be progressive. Syringomyelia is often described in association with Chiari malformation[1].

Epidemiology

Syringomyelia is a relatively rare disease with a prevalence in the (American) population of 8.4/100,000 population. Appears after the age of 30, more often in men. A familial occurrence has also been described. [2]

Pathophysiology

The pathophysiology of the disease is not fully understood. There are several theories about the origin of syringomyelia. [2]

Gardner's hydrodynamic theory says that an obstruction in the outflow of cerebrospinal fluid from the 4th ventricle through the foramen of Magendi is responsible for the formation of cavities. [2] During each systole of arterial pressure, the pressure is transmitted to the cerebrospinal fluid, which is subsequently propagated to the canalis centralis (central canal), the so-called "water-hammer effect". Increased pressure in the canalis centralis causes a cavity to form.[1] Simple distension of the canalis centralis is referred to as **hydromyelia**. [3]

Williams' theory points out that under some circumstances a gradient between intracranial pressure and intramedullary pressure may arise due to impaired circulation of cerebrospinal fluid through the foramen magnum. These circumstances are, for example, coughing or the Valsalva maneuver, which worsen venous return, increase intracranial pressure, and in the case of anatomical predisposition (Chiari malformation), obstruction occurs in the foramen magnum by a mechanism similar to that of valves. There is an increase in pressure in the cisterna magna simultaneously with a relatively lower pressure in the spinal subarachnoid space. Pressure propagates caudally from the fourth ventricle, forming a syrinx. [2]

Oldfield's theory explains the formation of intramedullary cavities. During systole, pressure is transmitted to the cerebellum, which propagates caudally with its tonsils (this can be demonstrated by dynamic MRI). Thus, the cerebellum oscillates rhythmically depending on systole and transmits pressure to the spinal subarachnoid space. CSF propagates perivascularly and through the interstitial space into the spinal cord due to the higher pressure. Cavities are thus created in the spinal cord, which cause neurological symptoms by their oppression of surrounding structures (pathways, neurons, microcirculation). [2]

The intramedullary pulsatile pressure theory claims, on the contrary, that intramedullary the pulsation pressure is relatively higher than the pressure in the spinal subarachnoid space. As a result of these forces, a cavity filled with extracellular fluid is created in the spinal cord. [2]

Types of syringomyelia

Syringomyelia communicating with the 4th ventricle

It occurs in approximately 10% of cases, and is sometimes associated with impaired circulation of the cerebrospinal fluid. In such a case, the situation is solved with a shunt. [2]

Syringomyelia due to blockage of CSF circulation (without communication with the 4th ventricle)

The most common form of syringomyelia, occurs in 50% of cases. The reasons are as follows

- Chiari malformation is the most common cause of this type of syringomyelia;
- **Basal arachnoiditis** (post-infectious, inflammatory, post-radiation, blood in the cerebrospinal fluid);
- **Basilar impression or intussusception**
- **Meningeal carcinomatosis**
- **Pathological structures** (arachnoid cyst , pannus in rheumatoid arthritis , occipital encephalocele, tumors). [2]

Syringomyelia from spinal cord involvement

Less than 10% of syringomyelia cases are of this type. Syringomyelia can arise on the basis of various insults:

- Spinal trauma
- Radiation necrosis
- Bleeding from a ruptured aneurysm, arteriovenous malformation or tumorous mass
- Infections (spinal abscess , HIV , transverse myelitis)

- Cavitation superimposed on ischemic insult or degenerative disease. [2]

Syringomyelia due to spina bifida

Spina bifida can be caused by a variety of mechanisms, including those already mentioned above. Resolution of spina bifida prevents the development of syringomyelia. [2]

Syringomyelia due to intramedullary tumors

The tumors most often associated with syringomyelia are ependymoma and hemangioblastoma . The fluid in the cavities most often comes from secretory active tumor cells, or the cavity is filled with blood (related to the mentioned tumors). Extramedullary tumors (intradural or extradural) have a somewhat different pathogenesis, as the cavities are formed due to obstruction of the circulation of the cerebrospinal fluid. [2]

Idiopathic syringomyelia

The cause is not known. Surgical intervention can correct the neurological deficit. [2]

Clinical Picture

The development of syringomyelia is usually slow, chronic. Acutely, syringomyelia can occur with involvement of the brainstem . Syringomyelia mostly affects the cervical spinal cord. Neurological symptoms depend primarily on the location in the spinal cord.

The syringomyelitic cavity primarily interrupts the crossing of the spinothalamic tract, causing **sensory dissociation**. There is a loss of perception of heat and pain, while touch, vibration and proprioception are preserved. In this way, one or both upper limbs can be affected or it can be manifested in a so-called "shawl-like" distribution (shoulders and upper front and back chest, as if you throw a large scarf over yourself - "shawl"). During progression, the enlarging syrinx can also affect the posterior cord and thus cause a loss of vibration sensation and proprioception. The patient may complain of dysesthesia . [4]

When the cavities spread to the anterior corners of the spinal cord, there may be **impairment of motor skills**, diffuse **muscle** atrophy starting from the hands and then proximally through the arms to the shoulders. If the lumbar spine is affected, there are similar manifestations on the lower limbs. Sometimes there is respiratory insufficiency depending on the change in body position. On physical examination, there are reduced reflexes in the upper limbs. There is spasticity, hyperreflexia, and irritating extensor symptoms in the lower limbs . [4]

In the late stages, there is also a disorder of the autonomic system manifested, for example, in the activity of the intestines, bladder or as sexual dysfunction. Damage to sympathetic neurons can manifest as Horner's syndrome. [4]

Sometimes the syndrome manifests as so-called **syringobulbia** , in which there is dysphagia, dysarthria, asymmetric weakness, atrophy of the tongue, nystagmus, and typical dissociation of sensation (see above). [4]

In 65% of cases, syringomyelia occurs simultaneously with Chiari **malformation type I**. The symptoms are thus modified by this malformation. While in adults **headache** dominates as the main symptom, in children the main dominant symptom is apnea. **Furthermore, segmental muscle weakness** and Lhermitt's sign may occur. The occurrence of tribal symptoms is also common. Propagation of the cavity into the trunk is manifested by cranial nerve disorders and cerebellar symptoms. [4]

Diagnostics

Cavities formed in the spinal cord are best seen on MRI . Because the cavities are usually filled with cerebrospinal fluid, they appear as **hypodense** foci in the T1-weighted image, and as **hyperdense foci in the T2 -weighted image**. An **exception** occurs if the syrinx is the result of a tumour process. [1] The fluid in the cavity has a higher protein content and therefore the MRI image is different. In addition, the wall of the cavity is formed by tumor cells or densely packed glial tissue. The diagnosis of such a cavity is then mediated by a contrast MRI examination. [5]

If we cannot use MRI for any reason, then we have alternatives such as myelography and CT-myelography and **CT-myelography.** [4]

Therapy

The therapy of syringomyelia depends on the clinical picture and type of syringomyelia. [1]

Conservative treatment includes avoiding heavy physical exertion associated with significant isometric contraction and Valsalva expiration (against a closed glottis), sleeping with an elevated head and keeping the neck in a neutral position. [6]

An essential part of the therapy is **neurorehabilitation**, which is important in preserving the remaining neurological functions and preventing complications in quadriparetic patients such as infection or pressure ulcers . [7]

Surgical intervention in the form of **decompression** or **shunt** operations is recommended in case of worsening neurological symptoms and unmanagable central pain. [6]

The following actions are considered:

- *Suboccipital and cervical decompression* − suboccipital craniectomy with laminectomy C1, C2, or C3 and dura plasticity. [7]
- *Laminectomy and syringotomy* − dorsolateral myelotomy to the so-called DREZ zone (dorsal root entry zone), usually at the C2-C3 level. [7]
- *Short-circuit operations (Shunts)* − there are several options:
  - *Ventriculoperitoneal shunt*
  - *Lumboperitoneal shunt*
  - *Syringosubarachnoid DREZ shunt* [7]
  - *Syringoperitoneal shunt* [7]
- *Fourth ventriculostomy* .
- *Terminal ventriculostomy* – the extended section of the central canal at the tip of the conus medullaris is referred to as the terminal chamber. It is used only in patients without Chiari malformation and only in patients with syringomyelia that extends into the lumbar region. [7]
- *Neuroendoscopic methods* .
- *Surgical methods ensuring better mobility of the spinal cord in the spinal canal* in case of post-traumatic adhesions. [7]

Pain and paraparesis have the best response to surgery. Conversely, loss of sensitivity, symptoms of lower motor neuron involvement and cerebellar symptoms have a smaller response. [6]

Analgesics (ibuprofen, acetylsalicylic acid, naproxen, etc.) and muscle relaxants (methocarbamol) are used for **symptomatic treatment**.

Links

Related Articles

- Chiari malformation

External Links

- ASAP (http://asap.org/)
- Chiari & Syringomyelia Foundation (http://www.csfinfo.org/)

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