Tumors of the central nervous system (pediatrics)

Content

Epidemiology

CNS tumors are the most common solid tumors in childhood. The highest incidence of tumors is under the age of five. The incidence is approximately 2.5:100,000 (in the Czech Republic, about 50-60 new cases per year). Boys are affected 1.3 times more often than girls. CNS tumors are the second most common cause of death in childhood (after injuries).

Etiology

The etiology of CNS tumors is unknown, connections with some endogenous and exogenous factors are assumed. An association with neurocranial irradiation in the treatment of ALL is reported .

Division

- According to localization, supratentorial 45-50% and infratentorial 50-55%.
- According to the place of origin into primary and metastatic.
- According to the biological nature and pathology - half of the primary tumors arise from glial cells, then from meningeal cells, from cells of the choroid plexus, nerve sheaths, blood vessels, from germinal and embryonic cells.

Astrocytoma

See the Astrocytoma page for more detailed information .

- It occurs both infratentorially and supratentorially.
- Low grade most common 35%.
- High grade only about 8% - glioblastoma multiform , anaplastic astrocytoma.
- There is a piloid astrocytoma in the cerebellum, it grows from the vermis, it is benign, it fills IV. ventricle, has few blood vessels.

Medulloblastoma

- Occurs infratentorially.
- At the age of 1–10 years, they account for 20% of CNS tumors.
- One of the most malignant childhood tumors, it most often occurs around 3-8 years of age. year of life.
- It grows from the lower part of the vermis, infiltrates the diapers and often metastasizes through the cerebrospinal fluid, mainly to the spine.
- It is radiosensitive.

Ependymoma

See the Ependymoma page for more detailed information .

- It occurs both infratentorially and supratentorially.
- They make up about 8%.

Pineal tumors and germinal tumors of the CNS

- 4% of CNS tumors.
- They are mostly malignant tumors, they have a varied histogenetics.
- The main clinical manifestation is intracranial hypertension with hydrocephalus (Parinaud's sign - palsy looking up).
- In germinal tumors, premature puberty also occurs .

Hemangioblastoma

- It occurs either as a spontaneously arising tumor (80%) or as part of the genetically determined von Hippel-

Lindau syndrome (20%), in which hemangiomas of the cerebellum, spinal cord and retina, cysts of parenchymatous organs, more often occur in the kidneys . pheochromocytoma and pancreatic ca.
- It is benign and more often affects younger individuals.
- The most common location is the cerebellum.
- It can be cystic, this is the most dangerous form because it can grow quickly and threaten the patient's life with acute occipital herniation .
- Furthermore, the form is solid, usually richly vascularized.
- Clinical manifestations - cerebellar syndrome and intracranial hypertension , polyglobulia occurs in 20% of cases , due to ectopic secretion of erythropoietin by the tumor.
- Cystic forms must be operated on immediately.

Tumors in IV. cerebral ventricles

- We classify according to the place of origin: central – fills the entire ventricle, lateral – spreads to the pons of the cerebellum, in the ceiling.
- It can be benign, often localized in the ceiling of the chamber, malignant forms are lateral or infiltrating.

Craniopharyngioma

For more detailed information, see the Craniopharyngioma page .

- 7% of CNS tumors.

Oligodendroglioma

See the Oligodendroglioma page for more detailed information .

- 2% of CNS tumors.

Tumors of the meninges

- Very uncommon in children.

Choroid plexus tumors

- 2% of CNS tumors.
- Papilloma of the choroid plexus occurs most often in children under 2 years of age, it fills the chamber and thus creates obstructive, hypersecretory hydrocephalus , it is a benign tumor.

Clinical manifestations

Clinical manifestations depend on the location and histological type of the tumor.

- The general symptoms in children are dominated by intracranial hypertension syndrome :

1. headache (pressure, generalized or retroorbital, variable intensity, worse in the morning, if the tumor is infratentorial, then the pain in the head is typical);
2. morning projectile nausea and vomiting;
3. visual disturbances - advanced symptom, papilledema ;
4. disorders of consciousness up to coma, seizures.

Obstructive triventricular hydrocephalus is caused by cancer originating infratentorially.

- Focal symptoms : are the result of irritation or destruction of the given brain tissue (stem symptoms, cerebellar symptoms, disorders of cognitive and sensory functions, cranial nerve palsies, vision and hearing disorders, side-specific hemiparesis can also be a disorder);
- the peculiarity of the CNS in children is that it can adapt to the tumor for a long time and recede, so clinical symptoms may be poor at the beginning of the disease;
- in infants and small children whose cranial sutures have not yet fused, the main symptom is macrocephaly, followed by changes in behavior - apathy or increased irritability, crying, failure to thrive.

Diagnostics

- Neurological and ophthalmological examinations, especially of the fundus, show congestion in up to 90% of cases.
- Today, MRI is a superior imaging method for tumors in the CNS, it is often administered with a contrast agent, and it detects even tumors only a few millimeters in size.
- CT with iv contrast, this method is not suitable for some tumors in the brainstem and in the posterior cranial fossa, 5% of tumors are iso-dense with the surrounding brain tissue and do not show up on CT even after administration of a contrast agent.
- In infants who do not yet have a closed fontanel, an ultrasound examination is performed first.
- On an X-ray of the skull, we can see the symptoms of intracranial hypertension, such as the spacing of the seams, wear of the dorsum of the saddle.
- Positron emission tomography (PET) is a suitable method in cases where the findings with standard imaging methods are unclear or in cases where we cannot histologically verify the tumor, it will help us determine its aggressiveness - grade.

- cerebrospinal fluid examination , we send the sample for biochemistry, cytology, microbiology and flow cytometrywith DNA analysis.

Treatment

- The basis of the treatment of CNS tumors is surgery, the extent of the intervention is determined by the prognosis (cytoreduction alone can prolong survival or increase the effectiveness of adjuvant treatment). It is necessary to perform an imaging examination immediately after the operation (no later than 72 hours after the operation) in order to detect any residual tumor.
- Chemotherapy is only used to a limited extent in the treatment of CNS tumors, and the success of this treatment can only be assumed for small tumor masses. There are only a few substances (used in chemotherapy) that are able to pass through the intact blood-brain barrier (nitrosourea, hydroxyurea).
- Radiotherapy has a proven effect on most CNS tumors and is often indicated after diagnosis or after surgery. It is not indicated before the 3rd year of life, it would be especially destructive for immature brain tissue. Myelination of the CNS is not complete until around 2-3 years of age.

Differences from adults

- Different clinical manifestations - symptoms resulting from intracranial hypertension syndrome dominate, non-specific symptoms are common in young children.
- Different histogenetic origin – in children, gliomas of low degree of malignancy and tumors of embryonic origin are the most common.
- A frequent tendency of the tumor to spread through the cerebrospinal fluid.
- Other predominant localization – more often infratentorial in the midline, less often supratentorial.
- Malignant tumors more often, especially in the earliest period of life.

Links

pediatrics. neurology