Neuroblastoma

Neuroblastoma is an **embryonic malignant tumor of early childhood,** originating from cells of nervous tissue. It is a solid tumor with extracranial localization. It is caused by a malignant reversal of immature sympathetic cells that come from the neural crest. It is characterized by its variable behavior - spontaneous regression (more than 10% of tumors), maturation in benign ganglioneuroma (less common) but also the occurrence of highly malignant forms with the spread of metastases. The primary tumor can be located in the abdominal cavity (total 65% - adrenal gland 40% and retroperitoneum 25%), in the mediastinum (15%), in the cervical region (5%) and in the pelvic sympathetic ganglia (5%). In approximately 1% of cases, the location of the primary tumor cannot be determined. Neuroblastoma metastasizes via **the lymphatic and blood vessels**, establishing metastases in the lymph nodes, bone marrow, cortical bones, dura mater, orbit, liver and skin. Less often, they can also metastasize to the lungs or intracranially.

Epidemiology

In the Czech Republic, neuroblastoma is diagnosed in approximately **20–30 children per year**. It is the most common malignancy in infants. 90% of all neuroblastomas are diagnosed by the age of 5. Occurrence in adolescents is rare, in young adults very rare.

Histological picture

Microscopically, it is a tumor of small blue round cells. The nucleus is hyperchromatic, the border of the cytoplasm is narrow, tumor cells tend to form rosettes. Based on histology, prognostically favorable and unfavorable tumor types can be distinguished.

Clinical picture

Non-specific general symptoms are common:

- fatigue and weakness,
- anorexia,
- weight loss,
- failure to thrive,
- behavioral changes,
- elevated temperatures,
- anaemia,
- edema,
- hypertension.

Other symptoms are based on **umor location (or metastases):**

- abdominal cavity - tactile resistance or even deformation of the abdominal wall, abdominal pain, anorexia, constipation,
- head and neck − exoftalmus, Horner syndrome, strabismu, papillary edema, optic atrophy, unilateral nasal obstruction,
- intraspinal localization− back pain, sensory disorders, motor defects and paraplegia,
- mediastinum − dyspnoe, dysphagia, recurrent respiratory infections,
- pelvic localization − urination disorders and / or defecation,
- skin metastases - resistance with purple appearance,
- bone metastases - bone pain, refusal to walk and lameness,
- metastases in orbit - hematomas of eyelids, protrusion and deviation of bulbs.

Uncommon but **typical manifestations:**

- acute cerebellar encephalopathy,
- lower limb paresis - intraspinal tumor spread (primarily paraspinal tumor),
- severe diarrhea - production of vasoactive intestinal pepite,
- Horner's syndrome - affliction of the sympathetic ganglia of the neck and upper chest,
- hypertension with sweating and redness − catecholamine production.

Diagnostics

Approximately 90% of neuroblastomas in children are diagnosed in the first 5 years of life. It is based on physical examination, imaging methods, laboratory examination and surgical biopsy. Histopathological diagnostics with tumor classification is absolutely essential for determining the risk group, prognosis and treatment scheme.

Classification

It is classified into clinical stages according to internationally valid recommendations (International Neuroblastoma Staging System − INSS) up to 4 levels: I., II.A, II.B, III., IV., IV.S. Furthermore, a new international classification is used (International Neuroblastoma Risk Groups − INRG) which is based on the patient's age (≤18 and> 18 months), disease stage (localized vs. metastatic), genetic changes (MYCN proto-oncogene status) and the presence of risk factors of surgical and imaging methods. According to risk factors, the disease is divided into low, medium and high risk.

Differential diagnostics

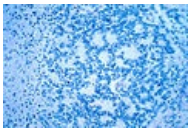
We must take into account Wilms' tumor, other germ cell tumors, then also Non-Hodgkin's lymphoma, Ewing's sarcoma, rhabdomyosarkoma and other cancers of childhood age. We can't forget about non-cancerous causes - such as cysts.

Therapy

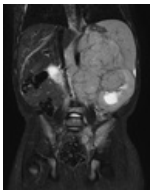
Most localized forms I. and II. stages are treated only surgically, some are spontaneously regressing and surgery is not necessary. Therapy of III. stage tumors combines surgery and chemotherapy. In the most risky forms, megachemotherapy with subsequent autologous **bone marrow transplantation** is included in the therapy. .

Prognosis

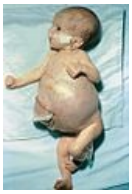
The prognosis **depends on the degree** of risk derived from the degree of **malignancy of the tumor**. In the low degree of malignancy, the three-year survival is 95–100%, in the middle 85–90%, in the high degree of risk it falls below 35%. Age less than 1 year is considered a positive prognostic factor because these patients are more likely to develop a tumor. Approximately 40% of patients have a generalized form of the disease with metastases at the time of diagnosis.



Neuroblastoma - small cells arranged in rosettes.



MRI image - neuroblastoma in the retroperitoneum in a 2-year-old patient.



A child with hepatomegaly and abdominal wall deformity caused by neuroblastoma metastases.

Odkazy

Related articles

- Mediastinal tumors
- Neuroectoderm tumors
- Spinal tumors
- CNS tumors (pediatrics)
- CNS tumors

Used literature

- ŠTĚRBA, Jaroslav – MAZÁNEK, Pavel – BAJČIOVÁ, Viera. *Pokroky v diagnostice a léčbě neuroblastomu u dětí* [online]. [cit. 2012-01-07]. <https://zdravi.euro.cz/clanek/postgradualni-medicina/pokroky-v-diagnostice-a-lecbe-neuroblastomu-u-deti-162702>.
- ŠNAJDAUF, Jiří – ŠKÁBA, Richard. *Dětská chirurgie.* 1. edition. Praha : Galén, 2005. ISBN 807262329X.
- LEBL, Jan – JANDA, Jan – STARÝ, Jan. *Klinická pediatrie.* 1. edition. Praha : Galén, 2012. 698 pp. ISBN 9788072627721.