Wilms' tumor

File:Nefroblastom, 6ti leta pacientka.png

Wilms tumor or **nephroblastoma** (WT, OMIM: 194070 (https://omim.org/entry/194070)) is a relatively common solid tumor of childhood. Specifically, it is a kidney cancer (unilateral or bilateral) characterized by clusters of embryonic nephrogenic blastoma cells. They make up about 5-6% of childhood tumors and are **the sixth most common tumor in children**. It typically affects preschool children, with a maximum incidence between the 2nd and 3rd year of life. It affects both kidneys in about **7 %** of cases.[1][2]

It is very often **associated with congenital defects**. These include urogenital tract defects, aniridia, pseudohermaphroditism, macroglossia.

Etiopathogenesis

Only about 1% of cases have a hereditary cause, the vast majority of cases arise sporadically. **Hereditary form** is mainly associated with mutated ***WT1*** gene on 11th. chromosome (11p13). Rarely, mutations in other genes may be involved (*WT2*–*WT5*). *WT1* tumor-supressor gene that codes a transcription factor (type *zinc finger*) is involved in the differentiation of the urogenital tract. Hereditary syndrome sometimes occurs in association with sporadic aniridia (lack of iris), sometimes also in association with neurofibromatosis type 1, mutations of *BRCA1* gene či Bloom syndrome. Congenital syndromes that have a high risk of developing WT include Beckwith-Wiedemann - hemihypertrophy of the limbs and Drash's. Children with these syndromes must be monitored by an oncologist for at least the first 6 years of life.[1][2]

Clinical picture

- The most common manifestation is painless resistance in the abdomen; parents may sometimes notice an increase in tummy tuck ("tight pants");
- non-specific problems - anorexia, constipation, vomiting, fever;
- approximately in **20 %** of children the first manifestation is hematuria[2] or abdominal pain.

Metastases

- **hematogenous spread** – mainly the lungs, then the liver, the brain. Rarely to the bone;
- **Lymphogenous spread** – to regional lymph nodes (hilar, paraaortic).

Diagnostics

- Ultrasound examination of the abdomen;
- CT examination of the abdomen;
- X-ray and CT of lungs to rule out metastases;
- the definitive diagnosis is histological (surgical solution usually follows chemotherapy)[2];
- the hereditary form can be confirmed by targeted molecular genetic testing (available in the Czech Republic).

Therapy

- Usually **neoadjuvant chemotherapy** – 4 weeks;
- **surgical** removal of the entire kidney with tumor and regional nodes;
- **chemotherapy** – actinomycin D, vincristine, cyclophosphamide..;
- event. **radiotherapy**[2];
- high-dose chemotherapy followed by bone marrow transplantation – treatment of recurrences after reaching 2nd or 3rd remission.

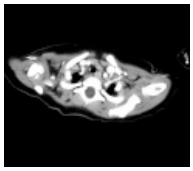
Special approach required in **treatment of bilateral tumors**:

- nephrectomy of a more affected kidney and partial nephrectomy on the other hand are indicated;
- bilateral nephrectomy with kidney transplantation.

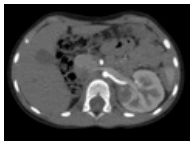
Prognosis

- It can be cured in **90 % of children**, if the tumor is inside the kidney;
- if the tumor is bilateral or spread beyond the kidney, **60 %** of children survives.[2]

Wilms' tumor (nefroblastoma)



CT scan – Wilms' tumor



CT scan - nephroblastoma in a 4-year-old patient with a solitary double kidney

Summary video

Video in English, definition, pathogenesis, symptoms, complications, treatment.

Links

Related articles

- Hereditary cancer syndromes
- Neurofibromatosis

Source

- ŠÍPEK, Antonín. *Geneticky podmíněná nádorová onemocnění* [online]. The last revision 8. 6. 2007, [cit. 4. 2. 2010]. <http://www.genetika-biologie.cz/hereditarni-nadorove-syndromy>.

Reference

1. KLEIBL, Zdeněk – NOVOTNÝ, Jan. *Hereditární nádorové syndromy.* 1. edition. Praha : Triton, 2003. 31 pp. ISBN 80-7254-357-1.

2. Klinika dětské onkologie FN Brno. *Kidney tumors - nefroblastoma (Wilms tumor)* [online]. [cit. 2011-01-02]. <https://www.fnbrno.cz/detska-nemocnice/klinika-detske-onkologie/informace-pro-pacienty/t2698>.