A CASE OF ADULT NEPHROBLASTOMA

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Clinical data

- In 52-year-old man, a tumor of the upper pole of the right kidney was detected on routine abdominal ultrasound check-up (1998).
- Ultrasound examination showed mostly retroperitoneal mass.
- Computerized tommography revealed irregular expansive formation on the ventral side of the upper pole of the kidney toward the liver.
The rest of the clinical examination was unremarkable as well as laboratory findings.

After right sided nephrectomy patient received radiotherapy (TD 45 Gy) and four cycles of polychemotherapy treatment with Dactinomycin and Vincristine.

After 16 years of regular surveillance, the patient is alive, with no signs of malignant disease.
The resected kidney with perirenal tissue and adrenal gland contained well defined tumor measuring 6 x 6 x 5 cm that only scantily affected subcapsular kidney tissue. Most of the tumor was located in the perirenal adipose tissue. Adrenal gland was unaffected.

The cut surface of the tumor was reddish pink in color, soft in consistency with areas of necrosis.
Histologically tumor was nodular, showed mixed, “triphasic” histological pattern consisting of:
small cells with dark, round nuclei resembling blastema, which tend to form large sheets,
ribbon like structures and rosettes
These areas showed diffuse, intensive cytoplasmic immunohistochemical positivity for WT–1, CD56 (100X)
Some areas had strongly positive cytoplasm for CD99, and S 100 protein.

Some areas were negative for CD99.
There was focal, scant positivity for synaptophysin. The proliferation rate (Ki67) was high.

Synaptophysin, 40x  
(Ki67, 100x)
There were nests of epithelial cells immunohistochemically positive for pan CK and WT1.
Stroma with eosinophilic, syncytial cells, in some areas resembling astrocytes, showed strong positivity for GFAP.

Proliferation rate was very low in these areas (Ki67, 40x)
Stromal SMA positive reaction was detected in bundles between nodules and sheets of small round cells.

Fibrillary stromal elements showed IH positivity for WT-1.
There were large areas of necrosis and abundant lymphoid tissue, mostly peripherally.
Our diagnosis

- Adult nephroblastoma, Tumor Wilm's (1998)

Wilm’s tumor is an embryonal renal tumor with a classical triphasic histology with varying proportions of blastemal, epithelial and stromal cells recapitulating the fetal kidney

Foci of metanephric blastemal cells that persist after birth are considered as potential Wilms tumor precursors

Adult tumor Wilm's– teratoid variant with extensive neuroepithelial differentiation (2014).

- It represents only 0.5% of all renal neoplasm in adults. Up to date, about 300 well-documented cases of adult WT have been reported in the literature.

- Teratoid Wilms' tumor (WT) is a rare variant of nephroblastoma which had been reported in pediatric patients. Fewer than 30 cases of teratoid WT have been reported until date.

Adult tumor Wilm’s– teratoid variant

- The term teratoid variant was first used by Variend et al. in 1984. Fernandes et al. proposed the criteria of more than 50% heterologous component for the diagnosis.

- There were 2 reported case of adult teratoid WT with extensive neuroepithelial differentiation.

References:
Differential diagnosis

- neuroblastoma
- peripheral primitive neuroectodermal tumor
- malignant ectomesenchymoma
- primary renal teratoma

Profesor C. Fletcher‘s second opinion:
high grade primitive neuro–epithelial neoplasm showing bidirectional glial and neuronal differentiation (1998).
Thank you for your attention