Metanephric adenoma

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Summary. In a recent survey of more than one hundred childhood renal tumors in our Laboratory files, we identified a unique case characterized by an unusual degree of differentiation and cell maturity.

Histologically this case was notable for an orderly array of small and uniformly-packed tubules with a rosette-like configuration. The nuclei were oval, smooth and of a bland appearance. Mitoses were absent. Many glomerular figures were intermingled. This renal tumor picture is somewhat different from that known as tubular Wilms' tumor because of the well-differentiated adenomatous pattern and the absence of any blastema.

The term metanephric adenoma is suggested for this tumor, which may represent the benign counterpart of Wilms' tumor.

Key words: Wilms' tumor, Metanephric adenoma

Materials and methods

Clinical data

A seven-year-old boy was seen for polycythemia. Red blood cell count was 9,000,000. High values of erythropoietin were detected. A mass of the upper pole was found in the right kidney. Ecography scans disclosed an oval outline and an uneven texture (Fig. 1). The patient was treated according to the SIOP 6 protocol which includes therapy for four weeks with Vincristine 1.5 mg/m² body surface every week, and in addition two cycles of Actinomycin-D 15 µg/Kg for three days in weeks one and three. The response to the treatment was negligible and the mass volume persisted unmodified. The tumor was excised by nephrectomy. The boy is in good health five years after surgery.

Microscopy

The entire tumor was sampled and several blocks were also obtained from the nontumorous renal tissue. The material was formalin-fixed and paraffin-embedded. All sections were stained with haematoxylin-eosin and additional staining included periodic acid-Schiff before and after diastase digestion and Masson trichrome.

Results

The kidney measured 4x9 cm and the tumor nodule was 3.5x5 cm in diameter. The aspect was of a pink solid tissue of uniform consistency. The nodule was encapsulated and had sharp margins. The histological examination revealed a process of uniform general architecture given by a small rosette-like configuration often acquiring a distinct tubular pattern (Fig. 2). Rosettes and tubules were closely packed with scanty stromal component. In some places they were more detached acquiring a more definite shape. They consisted of small uniform cells with oval nuclei and scanty cytoplasm (Fig. 3). The rosettes had a fibrillar...
Fig. 1. Ecogram showing an upper polar mass of the kidney with clear-cut margins and an uneven structure.

Fig. 2. Low-power view showing a strictly uniform adenomatous structure of small cellular nests with a rosette-like configuration. Haematoxylin-eosin. x 100
centre without a definite lumen. The nuclei had uniform, thin chromatin. Atypia and mitoses were absent. The cells had a bland appearance. Undifferentiated blastema was never seen. Glomerular bunches abutted in the larger and more dilated tubules. The tumor pattern did not change in the various specimens from different sites and maintained the benign appearance. The entire tumor was viable and no single focus of necrosis was detected.

Coarse vacuoles gave the fibrillary centre of the rosettes a tubular configuration.

When the glomerular bunch was at the centre of the lesion, a ribbon-like structure resulted. Small calcospherites were scattered. The glomerular buds had a flat epithelial lining and a cellular mesangium.

Nephrogenic rests were not observed in the unaffected renal tissue.

Discussion

In this case the diagnosis of Wilms’ tumor might be suggested by clinical and histological criteria (Bennington and Beckwith, 1975; Beckwith, 1983; Dehner, 1987). Yet the process was dominated by cell maturity; mitoses and atypia being virtually absent. The strikingly uniform and mature appearance of small rosette-like tubules resulted in a monophasic adenomatous pattern, suggesting to us the term of metanephric adenoma (MA). Distinction from the tubular variant of nephroblastoma is easily accomplished because the latter shows disorderly-arranged tubules layered by atypical embryonal cells (Beckwith, 1983) often associated with foci of blastema.

In 1977 Stambolis reported a benign epithelial nephroblastoma coexisting with a Wilms’ tumor (Stambolis, 1977). In that case the benign nephroblastoma was considered as originating from primitive blastema. Our case seems to be different from that of Stambolis because of the absence of blastema.

We feel that the present tumor is benign and this report is aimed to prompt further contributions and to warn those involved in the diagnosis and treatment of nephroblastoma. MA might represent another expression of low-grade renal masses in addition to mesoblastic nephroma, cystic nephroblastoma, and ossifying tumor of infantile kidney (Chatten, 1976; Chatten et al., 1980).

MA and metanephric hamartoma have been reported as diffuse processes involving the cortex and related to nephroblastomatosis (Liban and Kozenitzky, 1970; Bove et al., 1979). These entities show immature tubules with glomeruloid and papillary forms. One such case was
reported as secondary to in-utero aspirin toxicity (Bove et al., 1979). Our case is different because it was represented by a polar mass with a clear-cut contour and was histologically characterized by a high degree of structural and cellular maturity.

Dehner stated his experience with renal tumors in children older than the average age for Wilms' tumor and which were composed of immature tubules arranged in alternating compact and loose areas, and contained psammoma bodies (Dehner, 1987). He suggested the diagnosis of immature tubular adenoma for his cases that have done well with only a nephrectomy.

Adenomatous and metanephric nodules have been recognized as a component in the wide spectrum of nephrogenic rests and nephroblastomatosis and are distinguished from Wilms' tumor because of their cell maturity (Heideman et al., 1985; Holm-Nelsen and Olsen, 1988; Vogler et al., 1988; Beckwith et al., 1990). The belief that these adenomatous nephrogenic rests are benign is further supported, according to Beckwith et al. (1990), by the fact that they are unaffected by chemotherapeutic treatment (Beckwith et al., 1990).

The term metanephric hamartoma has been used by Vogler et al. (1988) but, as far as we are aware, in all cases it was the result of progressive maturation of nephrogenic rests or nephroblastomatosis (Vogler et al., 1988). Conversely, the present case was notable for its nodular appearance in the upper pole of the kidney, mimicking a Wilms' tumor, and for the lack of nephrogenic rests and nephroblastomatosis in the unaffected kidney.

The origin of Wilms' tumor from nephroblastomatosis is now understood, as well as the evolution of nephrogenic rests (nodular renal blastema) to adenomatous metanephric hamartoma. Therefore, it seems logical to conclude that the present case might represent a total conversion from renal blastema to mature tubular epithelium. This conclusion is likewise supported by the clinical and anatomical findings, so that we are inclined to regard this MA case in the spectrum of Wilms' tumor, probably representing the extreme degree of monophasic epithelial differentiation by far overwhelming the tubular pattern which may be encountered in any Wilms' tumor.

Of course, MA is entirely different, from adenoma secondary to arteriolosclerosis, renal scarring and to long-term dialysis (Budin and McDourell, 1984).

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References


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