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Case Report

Benign Epithelial Nephroblastoma

A Contribution to Its Histogenesis*

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Summary. The occurence of a malignant Wilms'tumor in the right kidney and of a benign epithelial nephroblastoma in the left kidney of a 5-year-old girl is reported. Both kidneys contained foci of persistent well differentiated blastema. In the left kidney a direct transformation of the primitive metanephric epithelium into a benign nephroblastoma is shown. This finding suggests an origin of epithelial nephroblastoma from persistent nephrogenic tissue and indicates that the former represents the benign counterpart of the malignant Wilms'tumor.

Key words: Wilms'tumor – Benign epithelial nephroblastoma – Persistent renal blastema/nephroblastomatosis complex.

Introduction

Congenital and infantile renal tumors have been generally considered as "congenital Wilms'tumors". They represent however, a heterogenous group of benign renal lesions which consists of three distinct clinico-pathological entities: 1. The congenital mesoblastic nephroma, also called fetal mesenchymal hamartoma, 2. the epithelial nephroblastoma with its two subgroups of (a) the benign multilocular cystic nephroma and (b) the tubulopapillary nephroma. These neoplasms are not only rare but also poorly documented (Bolande, 1974). The third entity comprises the nodular renal blastema-nephroblastomatosis complex.

All these tumors, especially the nodular blastema-nephroblastomatosis complex have been observed in association with typical malignant Wilms'tumors. The present report deals with the histologic findings in a case of Wilms'tumor in which a benign epithelial nephroblastoma in the opposite kidney and many foci of persistent blastema in both kidneys were found.

* Dedicated to Prof. Dr. Max Marcus on his 85th birthday

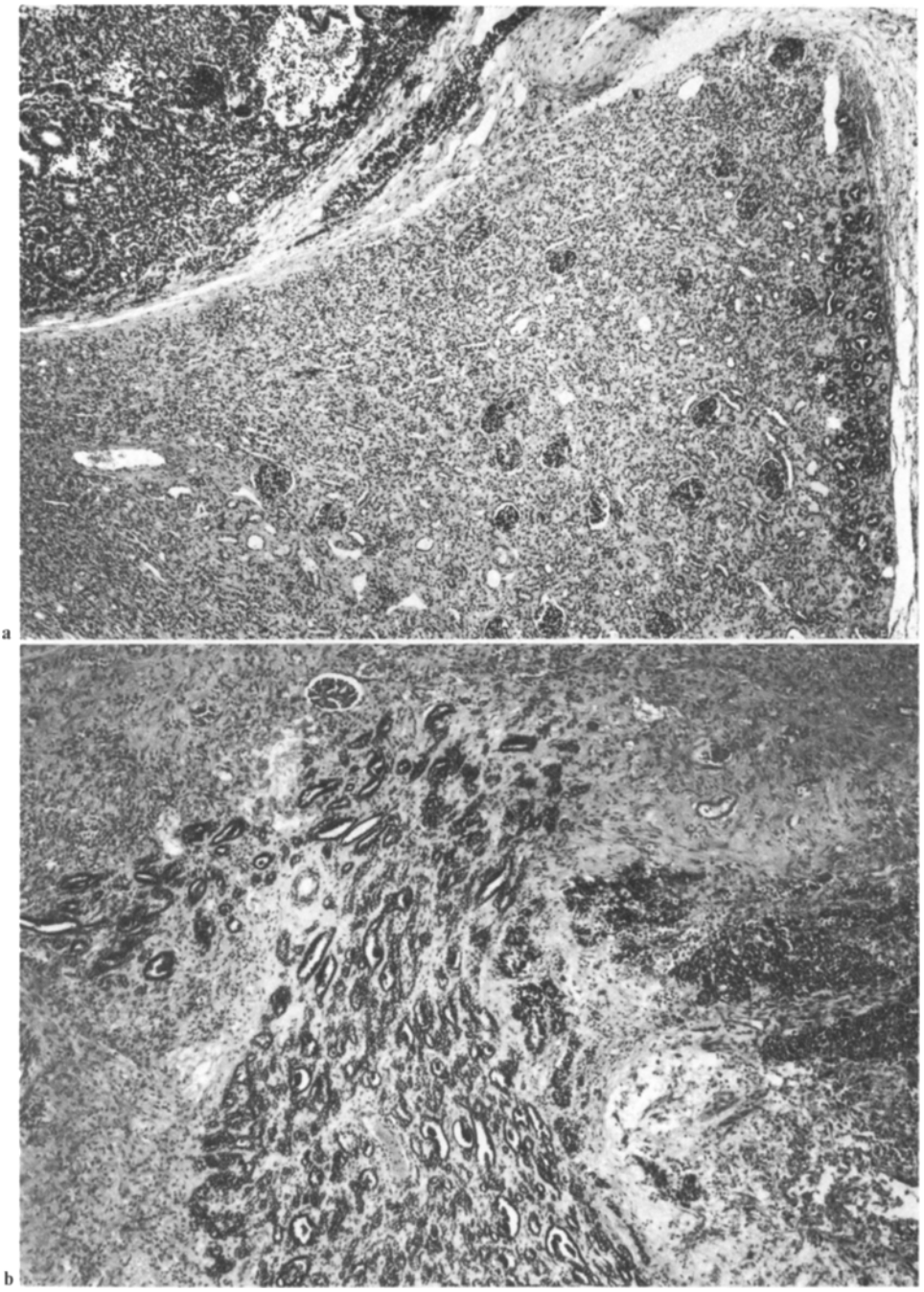

Fig. 1 a and b. Foci of well differentiated persistent blastema in the right kidney. **a** In a subcapsular area. **b** In a column of Bertin. a and b: HE, $\times 50$

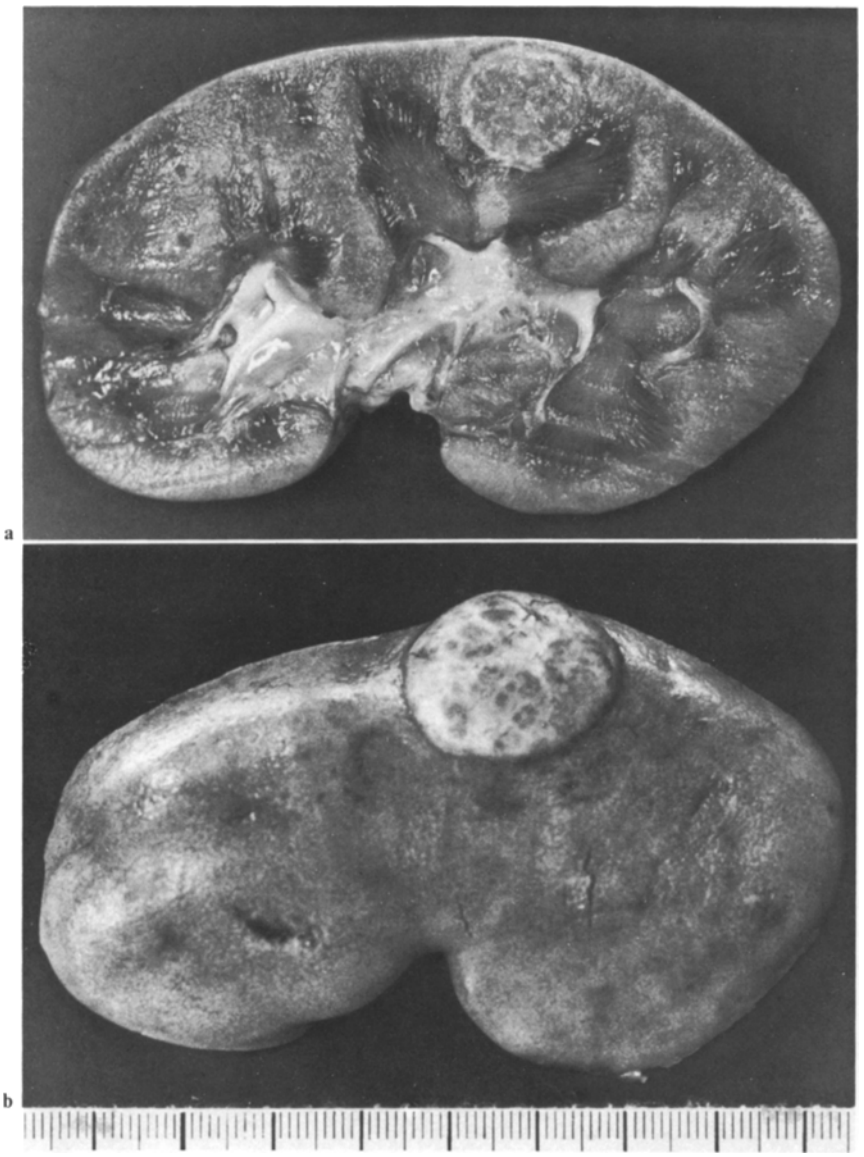

Fig. 2a and b. Left kidney. Gross specimen shows solitary benign nephroblastoma. **a** Cut section, **b** posterior surface

Case Report

Ch. K., 5 years old, was hospitalized for abdominal pain. A right renal tumor was diagnosed and a nephrectomy was carried out (E.Nr. 14620/76). The gross specimen showed a multi-nodular tumor with haemorrhagic and necrotic areas. Histologically the typical epithelial, blastomatous and mesenchymal sarcomatous elements of Wilms'tumor were found. No other structures were present. In many subcapsular regions and in the columns of Bertin, foci of well differentiated

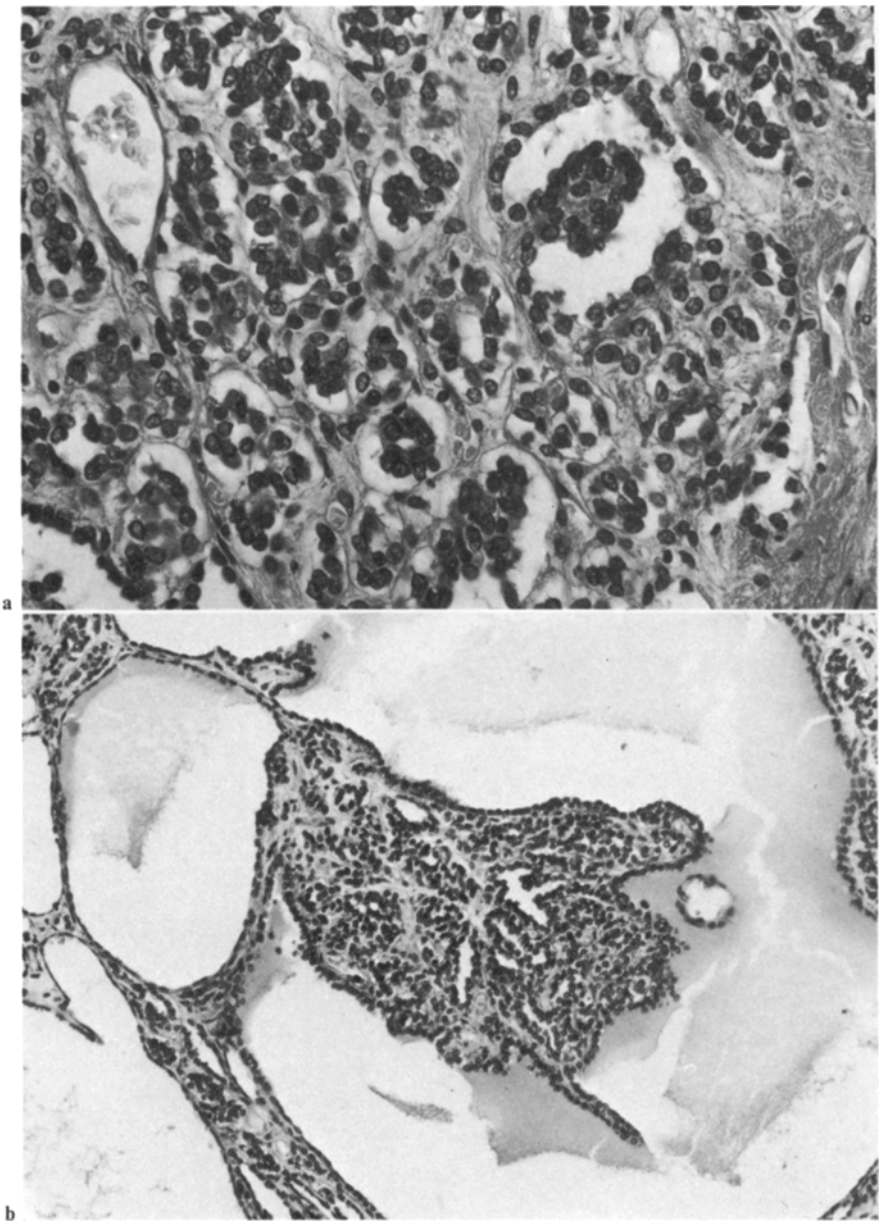

Fig. 3a and b. Benign nephroblastoma of the left kidney. **a** Tubular and glomeruloid structures. HE, ×300. **b** Cystic formations and papillary infoldings. HE, ×125

persistent blastema were seen. These foci showed no histological relationship to the Wilms'tumor (Fig. 1a and b).

On the 15th postoperative day the child died. A complete necropsy was performed (S.Nr. 383/76). No metastases were found. Malformations were absent.

The left kidney harboured a subcapsular solid tumor mass 2 cm in diameter (Fig. 2a and b), sharply demarcated from the normal renal parenchyma by a capsule. The entire tumor was

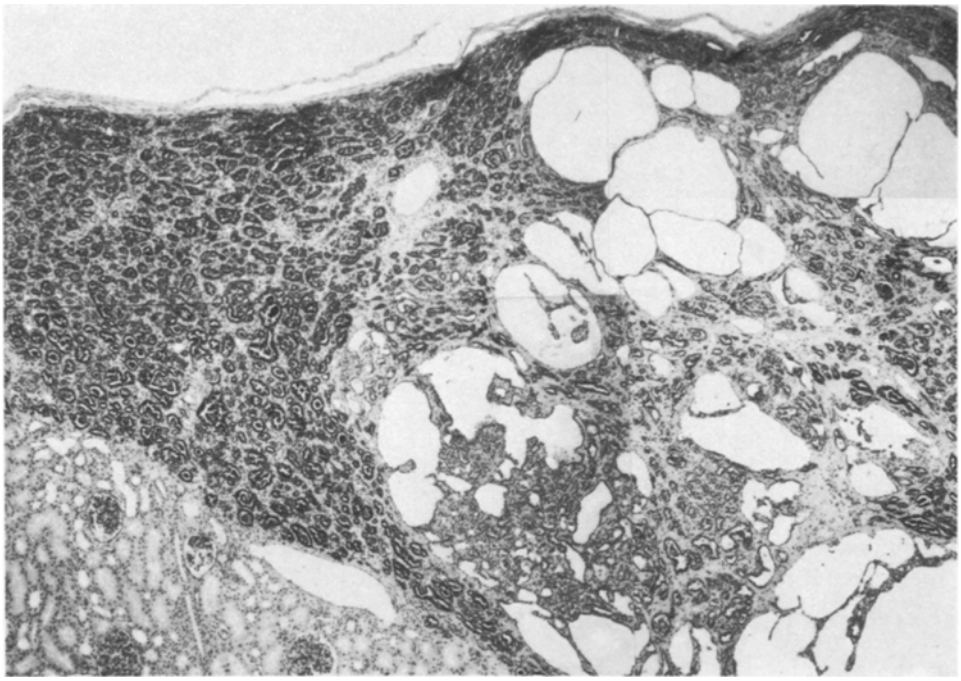

Fig. 4. Primitive metanephric epithelium in direct anatomical continuity to the nephroblastoma of the left kidney. HE, ×50

epithelial, consisting of tubulary and glomeruloid structures and of abundant papillary infoldings and festoons (Fig. 3a and b). Mitoses were not found. At the periphery the tumor possesed no capsule, but a subcapsular aggregate of primitive metanephric epithelium was present which showed a direct anatomical continuity to this tumor nodule (Fig. 4). The subcapsular zone of metanephric tissue entirely consisted of well differentiated but immature tubules and glomeruloid structures showing no atypicality or mitoses.

Discussion

The following constellation of findings was seen in the 5 year old girl: In the right kidney a malignant Wilms'tumor was present. Additionally, many foci of persistent metanephric blastema were found in the subcapsular areas and the columns of Bertin. These foci had no anatomical relationship to the Wilms'tumor. The left kidney harboured a benign epithelial nephroblastoma. A subcapsular zone of well differentiated primitive metanephric epithelium in this kidney bore a direct histo-topographic relation to the nephroblastoma.

Foci of persistent blastema are considered by most authors to be the possible origin of Wilms'tumor and for this reason they have been called "nephroblastoma in situ" (Shanklin and Sotelo-Avila, 1969). Statistical findings are in favour of this hypothesis, since the relationship of persistent blastema to Wilms' tumor has been shown to be a very significant one (Bove et al., 1969).

Bennington and Beckwith (1975) found, in a systematic search, 12 cases with persistent metanephric blastema in 2452 consecutive pediatric necropsies. The frequency was greater among children in this series who were younger than three months, where from 1035 cases positive findings were noted nine times. The estimated occurence of foci of persistent blastema in young infants is thus about 1%. This rate however, is much higher than the expected incidence of Wilms'tumor, the latter given as 1 per 10000 live births (Cochran and Froggatt, 1967). This shows that most of the foci of persistent blastema regress spontaneously and do not serve as source of Wilms'tumor. However, children with persistent blastema are doubtless running a much greater risk of developing Wilms'tumor than blastema-free individuals (Bove and McAdams, 1976).

Recently, histological evidence showing direct transformation of the nodular renal blastema – nephroblastomatosis complex into Wilms'tumor has been observed (de Chadarévian et al., 1977). In our case, no such relationship could be found in the right kidney. In the left kidney however, a direct transformation of the subcapsular primitive epithelium into a benign epithelial nephroblastoma is shown. Our findings demonstrate the origin of epithelial nephroblastoma from persistent nephrogenic tissue and suggest that the former represents the benign counterpart of the malignant Wilms'tumor.

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