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Inguinal nephroblastoma

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Summary. A case of an extrarenal nephroblastoma is reported which was located in the inguinal region. In the surrounding fat tissue normal ectopic glomerular and tubular structures and two more nephroblastomas were found. Therefore and because of negative clinical investigations in regarding of a renal malignant tumour the inguinal mass was thought to be the primary tumour and not a metastasis. This conclusion is important for the diagnosis and therapy.

Key words: Extrarenal nephroblastoma – Inguinal localisation – Metanephric blastema.

Introduction

Nephroblastoma or Wilms tumour is the most frequent renal tumour in childhood (Truckenbrodt 1972) constituting 7–20% of malignant tumours other than leukemias and lymphomas in patients under the age of 15 (Bennington 1975; Bodian 1964). Nephroblastoma is commonly a unilateral but in 5.8% of cases a bilateral tumour is present according to the Wilms Tumour Study (Bennington and Beckwith 1975). Extrarenal cases are rare and are not mentioned by Gutjahr in his synopsis (1981). It is obvious that extrarenal nephroblastoma needs renal blastema from which to develop. Harms and Löhr (1978) reported on a case of inguinal nephroblastoma and listed six other cases of extrarenal nephroblastoma arising in the mediastinum (1 case), the retroperitoneum (2 cases), in a sacrococygeal teratoma (1 case) and in the inguinal region (2 cases). Two further cases are presented by Madanat et al. (1978). Harms and Löhr (1978) pointed out the importance of recognizing the tumours as primary extrarenal nephroblastoma and not as metastasis of a renal nephroblastoma, although according to Bennington and Beckwith (1975) 25% of cases have already metastasized at the time of diagnosis.

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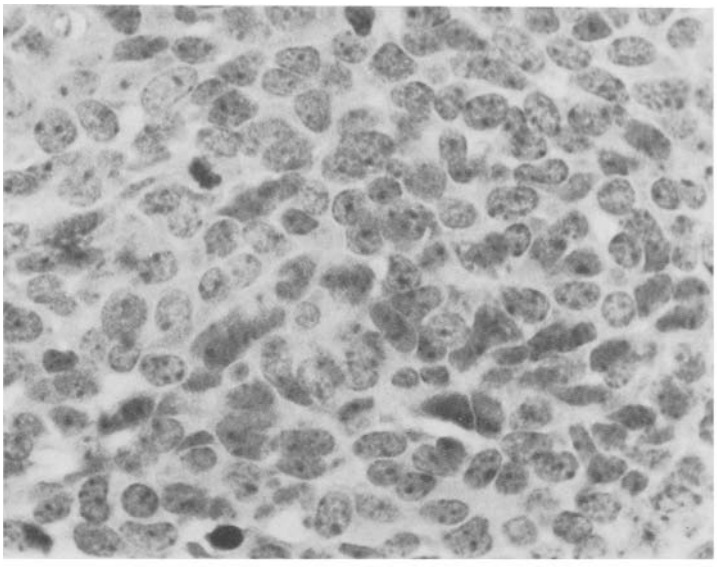
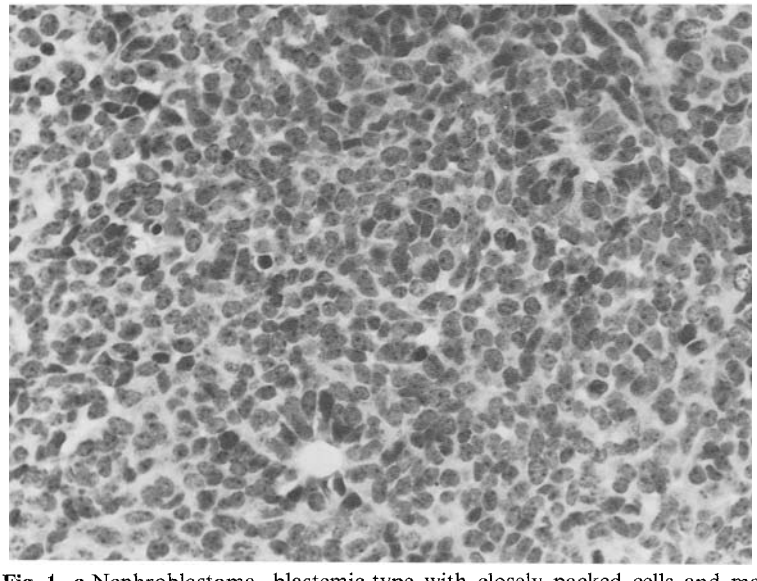


a



b

Fig. 1. a Nephroblastoma, blastemic-type with closely packed cells and many mitosis (PAS × 420). **b** Nephroblastoma with some tubular structures (Haem-Eosin × 260)

For this reason and because of the formal pathogenesis, we report this case.

Clinical features

A fourteen month old female child had suffered from B-Streptococcensepsis and meningitis in the newborn period with complete recovery after treatment. At the age of three months

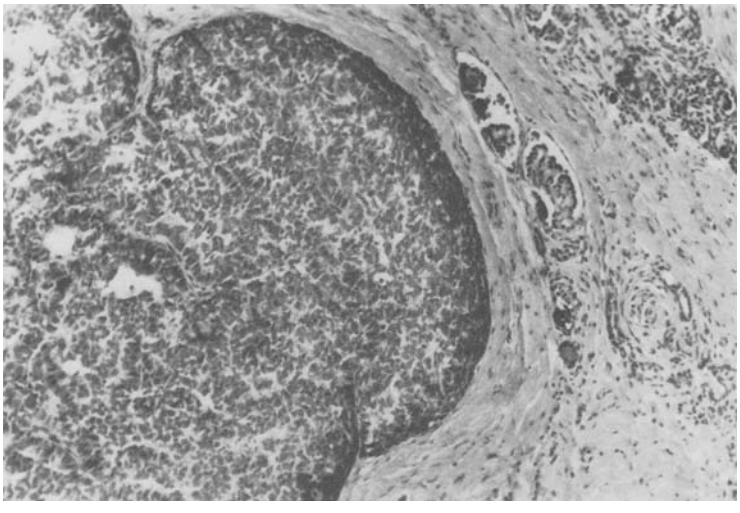


Fig. 2. Nephroblastoma and dysplastic renal tissue (Haem-Eosin × 600)

a swelling as big as a pea was noticed in the left inguinal region and taken to be a lymph node reaction after BCG-vaccination. This swelling increased in size and was 2 cm in diameter in admission to the hospital. The tumour was covered by normal skin, painless and not attached to the skin or structures beneath it. The mass was thought to be an incarcerated ovary in an inguinal hernia. At operation a solid tumour with necrotic areas, believed to be a lymph node, was removed.

Pathological findings

A round, non-encapsulated tumour of 2 cm in diameter with areas of necrosis and pale gray solid components on cut surface was studied by light microscopy using normal and special stains (PAS, Goldner, Gomori). The tumour was composed of closely packed slightly ovoid cells with very scanty cytoplasm (Fig. 1). A delicate network of collagen fibres existed and occasionally tubular structures were found (Fig. 2). On the margins of resection some glomerulae, appeared with broad mesangial matrix but otherwise with normal structure. The tumour showed many mitosis and moderate anaplasia. The diagnosis of Nephroblastoma, blastemic-type (Beckwith and Palmer 1978) was made. No lymphatic tissue was found but the most reasonable explanation was that of metastasis of a renal nephroblastoma of the kidney.

Clinical investigation in search of a renal nephroblastoma proved to be negative. Although it seemed unlikely, we assumed that this was a primary nephroblastoma of the inguinal region. Since histological examinations showed that the tumour had not been removed completely, a second operation was performed. This exstirpated soft tissue showed dysplastic renal tissue (Fig. 2) and two completely separated microscopically small nephroblastomas (Fig. 3a and b). In the surrounding fat tissue normal glomerular and tubular structures (Fig. 4) were found. Considering the results of clinical and pathological investigations we diagnosed a primary nephroblastoma of the left inguinal region.

A third operation was made to remove all fat tissue from the fossa inguinalis, which showed no more renal or tumour tissue. Combined chemotherapy according to the Study on Wilms Tumours of the Society for Pediatric Oncology, was given. No radiotherapy was given in consideration of the tumour location. The child is now 4 years old and in the best of health.

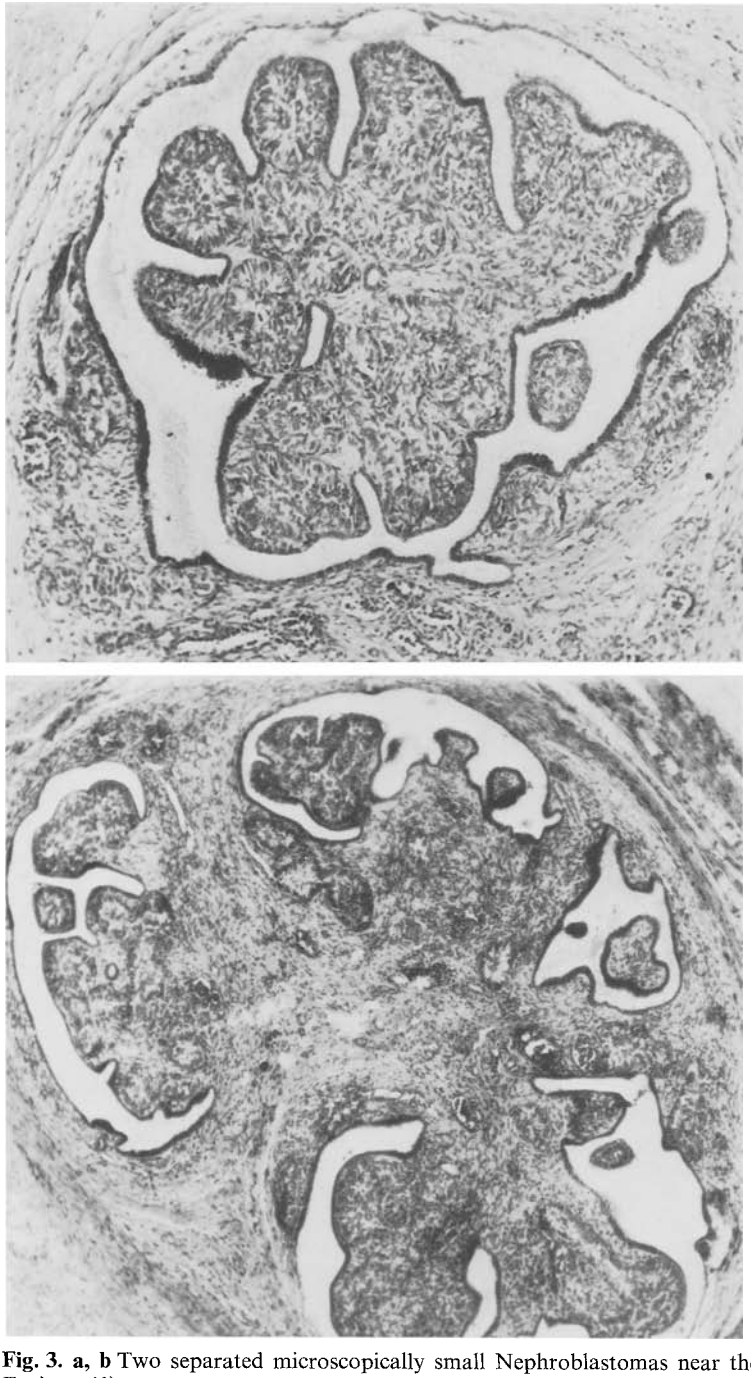


Fig. 3. a, b Two separated microscopically small Nephroblastomas near the tumour (Haem-Eosin ×40)

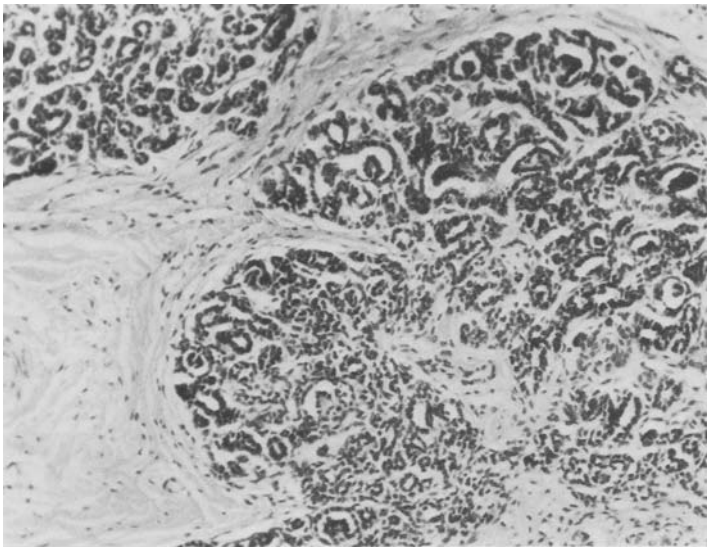


Fig. 4. Normal glomerular and tubular structures in the surrounding tissue of the tumour (Haem-Eosin ×65)

Discussion

Five cases of inguinal nephroblastomas had been published by 1978. We observed the sixth case. Harms and Löhr (1978) pointed out that renal tissue must be present for development of a nephroblastoma. Most authors now presume that the origin of this tumour is from metanephric blastoma (Bennington and Beckwith 1975) which explains also the existence of nephroblastoma in a teratoma, as these can contain renal tissue (Willis 1951). Renal tissue is not normally found in the inguinal region, but R. Meyer (1931) showed that this tissue can occur in this area by studies performed on an embryo. His results make it possible to explain the occurrence of a primary nephroblastoma in the inguinal region. Our case shows that ectopic and dysplastic renal tissue was present in the surroundings of the nephroblastoma and therefore that the tumour developed from this renal tissue. It is not possible to decide whether the nodule first noticed at the age of three months pea size was already a nephroblastoma or just the ectopic dysplastic renal tissue that in the next 11 months gave rise to the tumour.

The name extrarenal nephroblastoma seems not to be very appropriate, considering that these tumours actually arise from ectopic renal tissue. The term "renal" applies to the kidney itself and not to renal tissue. "Extrarenal nephroblastoma" is therefore not correct for tumours originating outside the kidney but from metanephric blastoma.

If an "extrarenal nephroblastoma" is diagnosed, one must take care, that the surrounding tissue be completely removed and carefully examined since it possibly contains renal tissue with toumorlets as shown in our case.

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