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A Case of Retroperitoneal Teratoma with Nephroblastoma

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We present a case of retroperitoneal teratoma in a 4-year-old girl in which a Wilms' tumor-like element was predominant, unlike the usual pattern of the immature or malignant teratoma. Mature elements were composed of adipose tissue, neural plexus and ganglia, cartilage, smooth and skeletal muscles, and glandular epithelium of the respiratory and gastrointestinal types. Three months after complete excision of the mass, a recurrent tumor developed. It consisted of only nephroblastomatous elements without teratomatous components. Theories for the histogenesis of this rare tumor are discussed.

Key words : *Teratoma, Nephroblastoma(Wilms' tumor), Retroperitoneum*

INTRODUCTION

Wilms' tumor (nephroblastoma) and neuroblastoma are among the two most frequent solid malignant tumors of childhood. Unlike neuroblastoma, Wilms' tumor almost exclusively arises from the kidney and is contained, in part, by the renal capsule (Ward & Dehner, 1974). Approximately 17 cases of extrarenal Wilms' tumor have been reported in world medical literature (Koretz et al., 1987).

We have observed recently a recurrent retroperitoneal teratoma in a child in which the dominant element was nephroblastomatous. This peculiar combination of histologic patterns raises several interesting points with regard to histogenesis. Clinically this unusual tumor posed a considerable therapeutic challenge to the oncologist.

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CASE REPORT

The patient was a 4-year-old girl, who was admitted to the hospital because of a palpable abdominal mass. At the age of 8 months, the mass was first noticed on the left upper guadrant and it began to enlarge thereafter. Perinatal and family histories were noncontributory. On physical examination, the abdomen was markedly distended with a 64㎝-long abdominal circumference. A large, firm, nodular, and nontender mass occupied the entire left side of the abdomen. The physical examination was otherwise unremarkable. All laboratory findings were unremarkable. Tumor markers including serum human chorionic gonadotropin, alpha-fetoprotein, carcinoembryonic antigen, and urine vanillylmandelic acid levels were within normal limits. Chest PA revealed elevation of both diaphragms, and an intravenous pyelography showed a fatty mass in the left abdomen without involvement of the left kidney. The calyceal patterns of both kidneys were normal. An ultrasonographic examination of the abdomen showed heterogeneous echogenicity in the left fatty mass. Abdominal computed axial tomography showed a left retroperitoneal huge mass with a fatty Hounsfield nu-

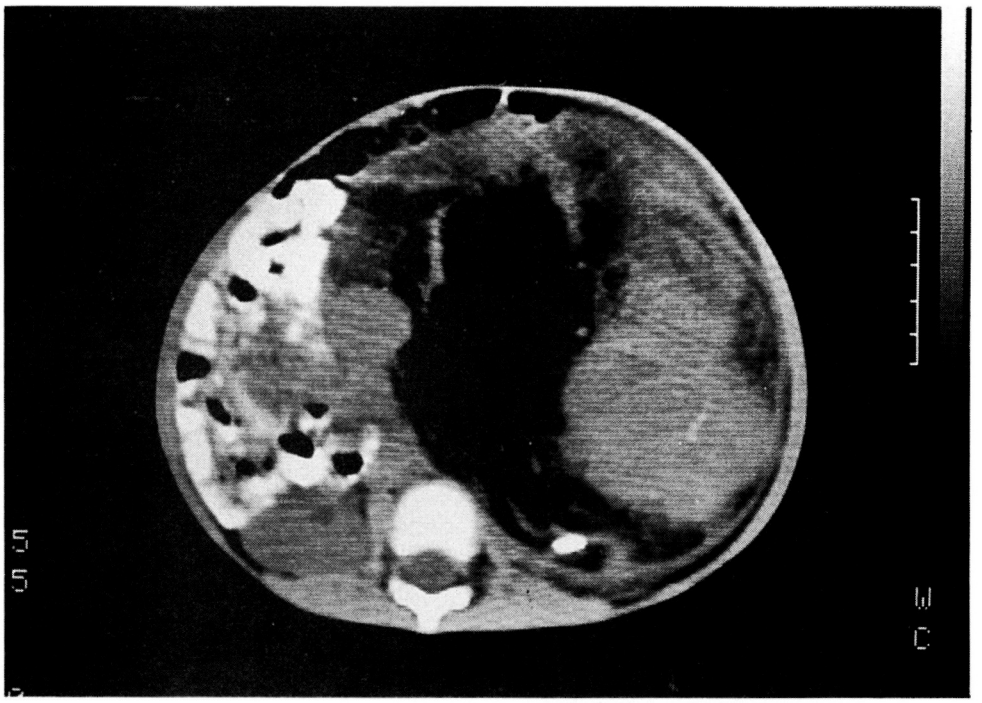

Fig. 1. Abdominal CT scan shows an irregular huge mass of fatty and solid densities with septation in the left retroperitoneum.

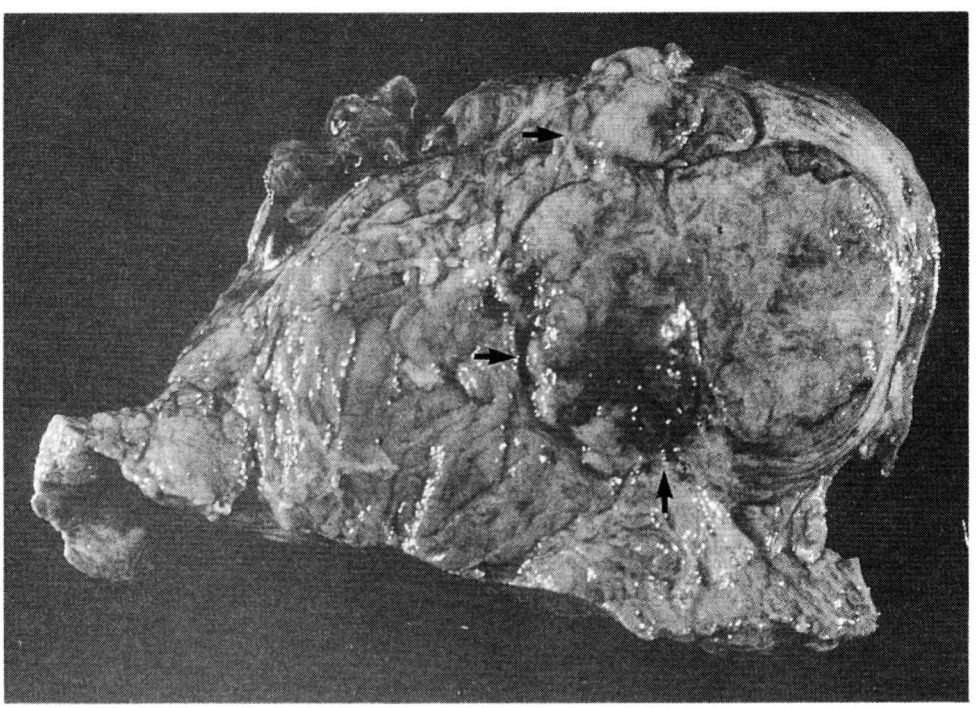

Fig. 2. The cut surface of excised specimen is composed of lobulated adipose tissue and irregular nodules (arrows) of myxoid fleshy tumor with multifocal hemorrhages and necroses.

mber, but the lower part of the tumor revealed a solid density with displacement of the small and large bowels (Fig.1).

Under the clinical impression of left retroperitoneal teratoma, exploratory laparotomy was performed. A well-encapsulated retroperitoneal mass was excised completely, the blood supply of which originated from the left renal pedicle and two lumbar arteries. The left kidney was displaced to the right side but was easily dissected free of the mass. Since the kidney was not visibly involved, nephrectomy was not performed.

Postoperatively it was recommended that the patient receive chemotherapy but it was refused. Three months after diagnosis, a large recurrent tumor mass was palpable in the left abdomen. Chest PA also revealed some elevation of both diaphragms, and intravenous pyelography showed a round solid mass shadow with nonvisualization of the left kidney. Abdomianl sonography revealed a large echogenic mass with septation in the retroperitoneal cavity. Tumor markers including alphafetoprotein, human chorionic gonadotropin, and carcinoembryonic antigen were within normal limits.

Exploratory laparotomy disclosed an irregular large recurrent tumor, which measured 15×8×5㎝ in size and revealed severe adhesion to the left kidney. Only a biopsy was performed.

The girl was discharged without chemotherapeutic regimens due to the parents' refusal.

PATHOLOGIC FINDINGS

The resected specimen was a well-encapsulated multinodular soft mass, measuring 20×15×12㎝ in size and 2㎏ in weight. The cut surface was composed of lobulated yellow mature adipose tissue divided with fibrous septae and interspersed cartilagenous islands. The remainder consisted of several nodules of yellow-gray, solid fleshy tumor, occasionally exhibiting a variegated appearance with hemorrhage and necrosis. It constituted approximately 30% of the excised tumor mass (Fig.2).

Microscopic examination of the lobulated adipose tissue portion revealed mature teratoma composed of adipose tissue, fibrovascular septa, neural plexus and ganglia, cartilage, skeletal and smooth muscles, and glandular and cystic structures lined with respiratory and gastrointestinal-type epithelial cells (Fig.3). The remaining 30% of the tumor was composed of compact cellular aggregates surrounded by thin fibrovascular septa (Fig.4). The small darkly stained cells in the compact nests had scanty cytoplasm, ill-defined borders, and abundant mitoses, resembling the undifferentiated renal blastema found in Wilms' tumor. Scattered zones of hemorrhage and necrosis were found. Occasional structures suggesting imperfectly formed tubules were noted within the solid cellular nests. However, differentiation into renal glomeruli was not obvious. The surrounding mesenchyme had a loose-textured appearance without specific differentiation. No other malignant elements such as neuroblastoma or embryonal carcinoma were identified.

The biopsy specimen of the recurrent tumor was only composed of previously recognized nephroblastomatous elements without components of the other teratoma. All three elements of typical nephroblastoma including undifferentiated blastema, mesenchymal tissue, and embryonic tubular structures were noted (Fig.5).

Immunohistochemical PAP (peroxidase-antiperoxidase) stain for vimentin (BioGenex Laboratories, USA) revealed focal positive cytoplasmic granules on fibroblast-like mesenchymal and blastemal elements (Fig.6). Embryonic tubular structures were negative for both cytokeratin (BioGenex Laboratories, USA) and vimentin.

Electron microscopic examination (Zeiss 109) was performed with paraffin-embedded blocks of nephroblastomatous component after reprocessing. Individual tumor cells were compactly aggregated without evidence of obvious cellular junctional structures. The nuclei were small. round-to-oval, and with a condensed chromatin pattern. The cytoplasm was little in amount, and cytoplasmic organelles were not precisely identified due to severe destructive changes of fixation.

DISCUSSION

Nephroblastomas are generally accepted as mesodermal in origin, being derived from the metanephrogenic blastema, which normally develops into glomeruli and the secretory part of the renal tubules (Hou & Azzopardi, 1967). The presence of other mesodermal components such as muscle, bone, and cartilage is explained, in part, through the nephrotome hypothesis by the incorporation of somite elements which may represent fanciful embryology (Ward & Dehner, 1974). Potter, on the other hand, states that metaplasia of metanephric tissue may be possible. The presence of keratinizing

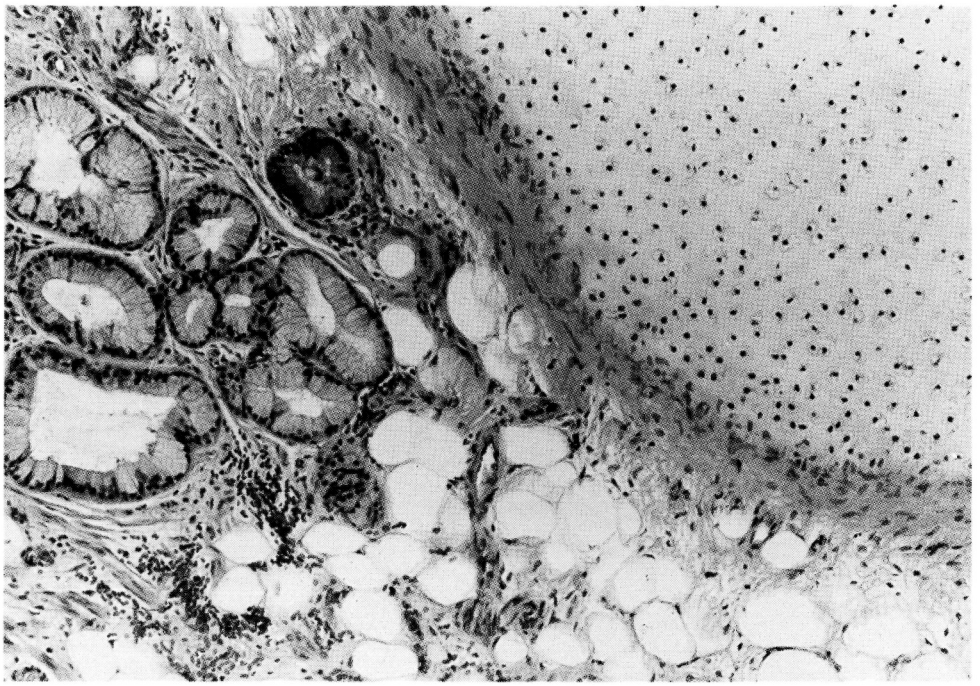

Fig. 3. One portion of mature teratoma is composed of an island of mature cartilage, grouped mucinous glands, and intervening adipose tissue (H&E, ×100).

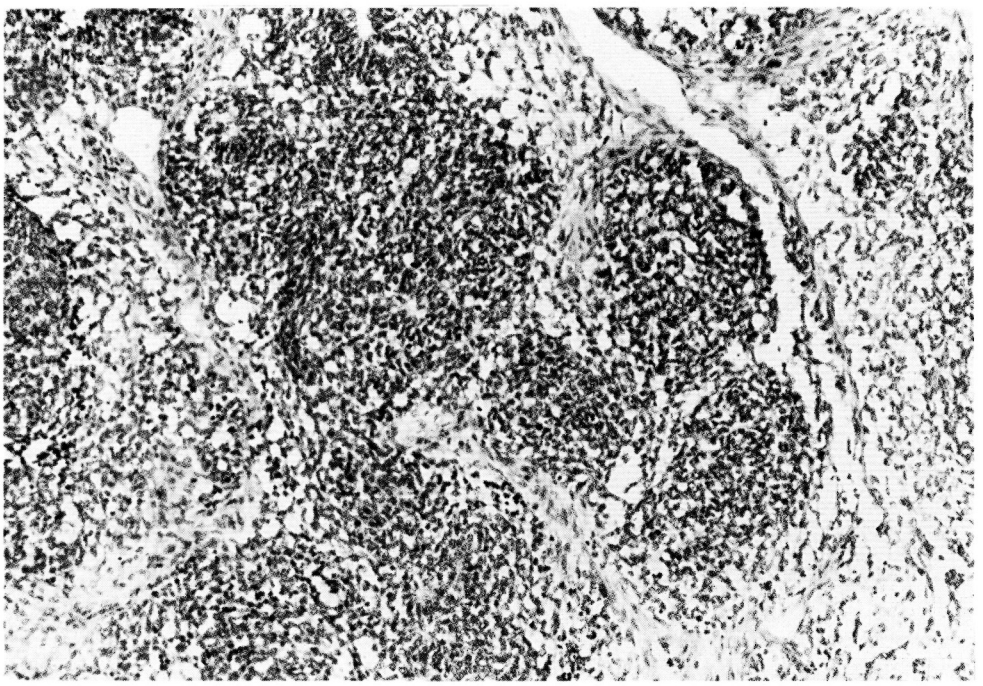

Fig. 4. The fleshy nodular portion of tumor reveals lobulated solid sheets of small dark undifferentiated cells and intervening mesenchymal tissue, resembling nephroblastoma (H&E, × 100).

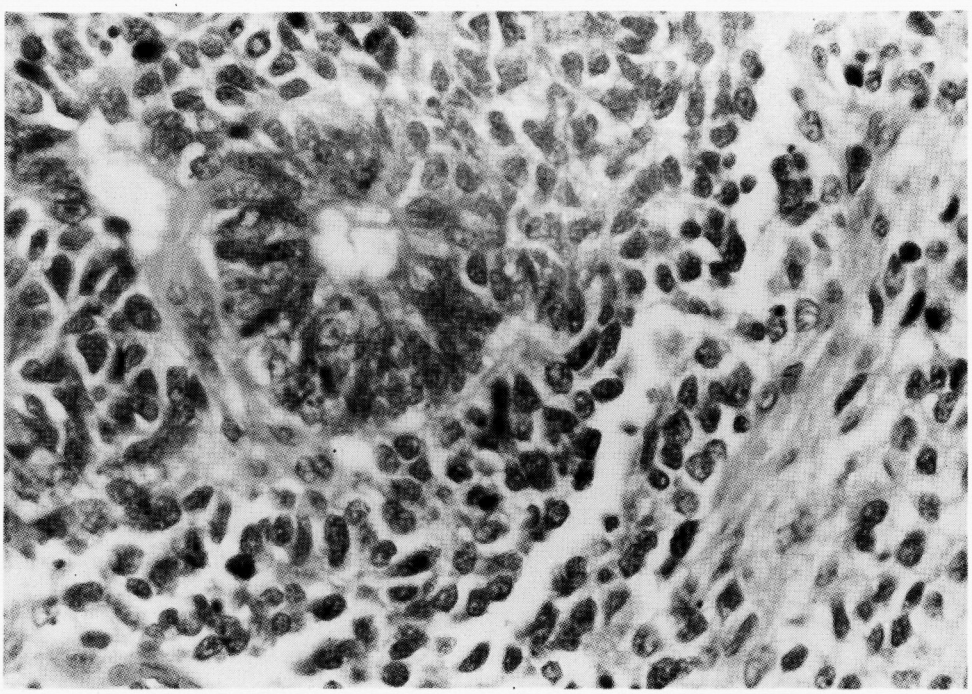

Fig. 5. An embryonic tubular structure is noted in the background of undifferentiated blastema and surrounding fibroblast-like mesenchymal elements (H&E, ×100).

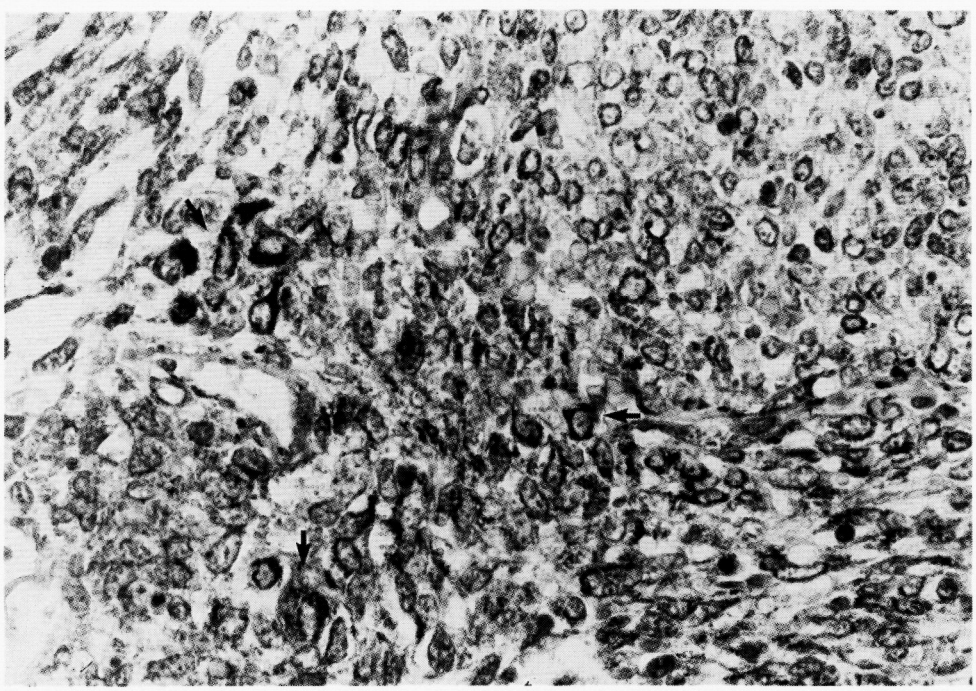

Fig. 6. Immunohistochemical stain for vimentin reveals focal positivity(arrows) on mesenchymal and blastemal elements (PAP, ×400).

squamous epithelium, mucin-secreting epithelium, and argentaffin cells in a nephroblastoma is considered as metaplastic derivatives of collecting tubules (Hou & Azzopardi, 1967).

Extrarenal Wilms' tumor is an extremely rare lesion and has been reported in different locations (Koretz et al., 1987). The extrarenal location favors Wilms' theory which postulates that the tumor arises from remnants of undifferentiated mesodermal tissue before the mesodermic derivatives of myotome, sclerotome, and nephrotome had developed (Edelstein et al., 1965 ; Tebbi et al., 1974). Some authors have eported nephroblastic tissue in the teratomas, usually in retroperitoneal or sacrococcygeal locations (Malik et al., 1967 ; Koretz et al., 1987). This has led to debate as to the embryologic origin of the nephroblastoma. When extrarenal Wilms' tumor occurs in locations where germ cells can be expected to exist, it may be argued that the failure to identify derivatives from all 3 germ layers is the result of inadequate sectioning. The finding of extrarenal Wilms' tumors outside the course of germ cell migration, such as those arising in the inguinal area, would suggest that they may not necessarily be teratoid and may have originated from misplaced primitive nephrogenic blastema (Hansmann & Budd, 1932 ; Tang et al., 1981). Though the nephrotome hypothesis may explain a few of the extrarenal nephroblastomas, a teratomatous histogenesis is strongly suggested by our own case and others (Tebbi et al., 1974 ; Ward & Dehner, 1974).

In a review of 91 gonadal and extragonadal teratomata in infancy and childhood (Berry et al., 1969), the most common sites of teratoma were seen to be in the sacrococcygeal area. Retroperitoneal teratoma was only one case in that review. The retroperitoneum is the primary site for 4% to 5% of germ cell neoplasms of all types in childhood. Unlike the sacrococcygeal teratoma, the retroperitoneal counterpart is seen with equal frequency in males and females and progression to a malignant teratoma such as an endodermal sinus tumor is very rare(Dehner, 1987). Beyond the age of 2−4 months, most sacrococcygeal teratomas are malignant and histologically, the most common and readily recognizable portion of malignancy is the endodermal sinus tumor or embryonal carcinoma (Chretien et al., 1970). Sacrococcygeal teratomas in infants and children were graded histologically as mature teratomas (grade 0), immature teratomas (grade 1, scanty amounts of atypical neuroblastic elements), immature teratomas (grade 2, moderate amount of embryonic tissue, predominantly neuroectoderm), and immature teratomas (grade 3, yolk sac carcinoma or abundant neuroectodermal tissue) by Gonzalez-Crussi et al. (1978) Some pathologists adopted a grading system of ovarian immature teratoma by Norris et al. (1976) We believe that it is important to emphasize the morphological type of the malignant tissue since it would be expected to represent a major determinant in the prognosis and the response to various therapeutic modalities.

Nephrogenic tissue is an uncommon component of teratomas in general, with only 7 available reports with details on 9 tumors (Ward & Dehner, 1974 ; Dische & Johnston, 1979). Among them, there were two sacrococcygeal tumors and one retroperitoneal neoplasm composed, in part, of immature renal tissue. In 2 cases of sacrococcygeal teratomata, embryonic renal tissue was noted in concert with other mature differentiated teratomatous components (Berry et al., 1969). Of a total of 82 teratomas reported by Willis (Malik et al., 1967), only 1 contained nephroblastic tissue, and this was not a retroperitoneal tumor. In Korea, a similar case of retroperitoneal teratoma with feature of Wilms' tumor in a 10−month−old girl was discussed at the KSP monthly slide conference (Kim et al., 1990).

In therapeutic modalities, a case of sacrococcygeal teratoma with nephroblastoma was successfully treated with chemotherapeutic regimens of vincristine and actinomycin D (Ward & Dehner, 1974). But the rarity of these cases does not allow us a simple conclusion for the best treatment. Our patient was advised to receive chemotherapeutic regimens of Wilms' tumor, but the family refused due to economic problems.

In differential diagnosis of mixed tumor of these 2 components, teratoid Wilms' tumor arising in the kidney should be included. There are several possible explanations for the pathogenesis of this complex renal tumor, i.e., a "collision" between a true teratoma and a nephroblastoma, as well as a nephroblastoma arising within a true renal teratoma. But a more plausible explanation is a direct origin of the diverse epithelia, together with mesenchymal elements, and possibly also ganglion cells, from totipotent primitive nephric blastema in the case of teratoid Wilms'tumor arising in the kidney (Va-

riend et al., 1984). A similar case of left renal teratoid Wilms' tumor was reported in a 3-year-old girl in Korean literatures (Suh et al., 1986). This tumor consisted predominantly of otherwise typical Wilms' tumor irregularly mixed with teratoid tissue elements such as intestinal tract, mucous glands with argentaffin cells, goblet cells and transitional epithelium. These heterologous elements were regarded as diverse epithelial differentiation of totipotent cells in a certain nephrogenetic period. Squamous epithelium, respiratory and intestinal-type epithelium, melanin-containing cells, skeletal muscle, and cartilage have all been previously described in hepatoblastoma (Dehner, 1978).

Immunohistochemically, the blastematous elements of nephroblastoma are known to show only positivity for vimentin ; the epithelial elements react for keratin, epithelial membrane antigen, and various lectins ; the mesenchymal elements show a reactivity pattern consonant with their morphologic appearance (i.e., myoglobulin, desmin, or neuron-specific enolase) (Rosai, 1989). In our case, the blastematous and mesenchymal components revealed focal positivity for vimentin. Negative reaction of the epithelial tubular components for keratin was interpreted as some spectral specificity with molecular weight of primary antibody.

Ultrastracturally, blastemal foci consist of sheets of undifferentiated cells having nuclei that are uniformly round to oval and cytoplasm containing few organelles. Epithelial differentiation is recognized by the development of basal lamina and an increase in cytoplasmic organelles, microfilaments, and cilia (Ward & Dehner, 1974 ; Dehner, 19 87). Regrettably, we could observe only compact nuclei of small undifferentiated cells without further evidence of specific differentiation due to severe artefact of poor fixation.

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