Journal of Korean Medical Science
Vol 6, No. 4, 367-371 December 1991

Malignant Rhabdoid Tumor of the Kidney

— A Case Report —

Tae Jin Kim, Tae Jung Kwon, and Je G. Chi

Department of Pathology, Seoul National University Children's Hospital
Seoul National University College of Medicine, Seoul, Korea

Malignant rhabdoid tumor is a distinct renal tumor in children. It had been regarded as a rhabdomyosarcomatoid variant of Wilms' tumor, but it is now thought as a separate entity. We report a case of malignant rhabdoid tumor of the kidney in a 26-month-old girl who presented with a left abdominal mass. Grossly, a large mass in the lower pole of the left kidney was well encapsulated and measured 4×4×3.5cm. On cross section, it was soft and yellowish white and showed multifocal necroses. The mass was mainly located in the medial medullary portion and compressed the renal pelvis laterally. Microscopically, the tumor masses were hypercellular and anaplastic without definite blastematous elements. In larger portion, the tumor cells had abundant eosinophilic cytoplasm and hyaline globules. In addition to the classic "rhabdoid" feature, alveolar, sclerosing, and lymphomatous patterns were seen. Ultrastructurally, tumor cells with abundant cytoplasm contained tangles of intermediate filament corresponding to vimentin in immunostaining.

Key Words: *Malignant rhabdoid tumor, Kidney, Vimentin, Childhood, Tumor*

INTRODUCTION

Rhabdoid tumor of the kidney was first described to specify a highly aggressive neoplasm having characteristic morphologic features (Haas et al., 1981). Prominent nucleoli, PAS-positive cytoplasmic inclusions, and light microscopic features mimicking rhabdomyoblastic differentiation are hallmarks of this tumor. It was initially thought to represent a rhabdomyosarcomatoid pattern of Wilms' tumor, but subsequent failure to demonstrate evidence of myoblastic differentiation (Fung CHK et al., 1981) and a lack of morphological or clinical linkage to Wilms' tumor supported a view that rhabdoid tumor was a separate entity. The "malignant rhabdoid tumor" was proposed and widely accepted as a descriptive term with regard to its clinical behavior (Fung CHK et al., 1981.)

Address for correspondence: *Je G. Chi, Department of Pathology, Seoul National University Children's Hospital, 28 Yongon-dong, Chongno-gu, Seoul 110-744, Korea (Tel: 02-760-3540)*

The features of malignant rhabdoid tumor include the presence of polygonal cells with roughly ovoid vesicular nuclei; prominent, usually single and fairly central nucleoli; variable amounts of eosinophilic cytoplasm and hyaline globular cytoplasmic inclusions in scattered cells. Ultrastructurally, cytoplasmic inclusions are of filamentous nature and have no diagnostic rhabdomyoblastic elements (Haas et al., 1981). They are composed of parallel 6 - 9 nm filaments, packed in concentrically whorled arrays and are not membrane-bound. Immunohistochemical study and Western blot analysis revealed that these cytoplasmic inclusions consisted of two distinct intermediate filament proteins; vimentin, a mesenchymal-specific protein, and a 54-kilodalton (kd) cytokeratin, present in nonsquamous epithelium but absent from squamous epithelium (Vogel AM et al., 1984).

We recently experienced a case of malignant rhabdoid tumor of the kidney and report here with review of current histogenetic opinions.

CASE REPORT

A 26-month-old girl presented with transient hematuria for three days, accompanied with fever and upper resipiratory infection. On physical examination, a firm, non-tender mass was detected in the left upper quadrant of the abdomen. Enlarged left cervical lymph nodes were also palpated, measuring 1.5×1.5cm. Abdominal computed tomography and intravenous pyelography revealed a left renal mass. Laboratory data showed no specific findings. There was no hypercalcemia or endocrine dysfunction. Suspicious metastatic lesions were noted in the right lung by chest computed tomography. On operation, a large mass in the lower and mid portions of the left kidney was detected and measured 7×8cm. The tumor was well encapsulated and had a smooth elastic firm consistency. It was severely adherent to the posterior psoas muscle at the lower pole. A careful examination revealed several enlarged paraaortic lymph nodes, but no other metastatic lesions were found.

PATHOLOGIC FINDINGS

Grossly, a large mass was noted in the lower pole of the left kidney. The mass was mainly located in the medial medullary portion and compressed the renal pelvis laterally. It was well-encapsulated and measured 4×4×3.5cm. On cross section, it was soft and yellowish white and showed multifocal necroses (Fig. 1). A few whitish separate nodules were seen outside the tumor capsule. There was also a large extrarenal mass over the renal capsule, measuring 5×1.5×1.5cm. It was well delineated from the adjacent normal tissue, but showed infiltrative growth in areas.

Microscopically, the tumor masses were hypercellular and anaplastic without definitive blastemal features. In large portions, the tumor exhibited solid proliferation of monotonous tumor cells with vesicular nuclei, prominent nucleoli, abundant eosinophilic cytoplasm, and occasional intracytoplasmic inclusions (Fig. 2). Nucleoli

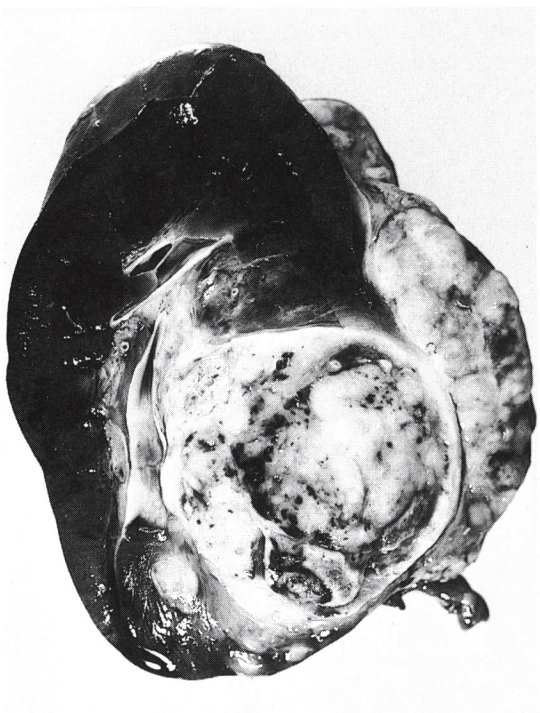

Fig. 1. Cut section of the left kidney reveals a well-encapsulated, soft, yellowish white mass in the medial medullary portion with a daughter nodule. An extracapsular mass is also noted. The tumor shows a locally invasive growth pattern with pushing margin.

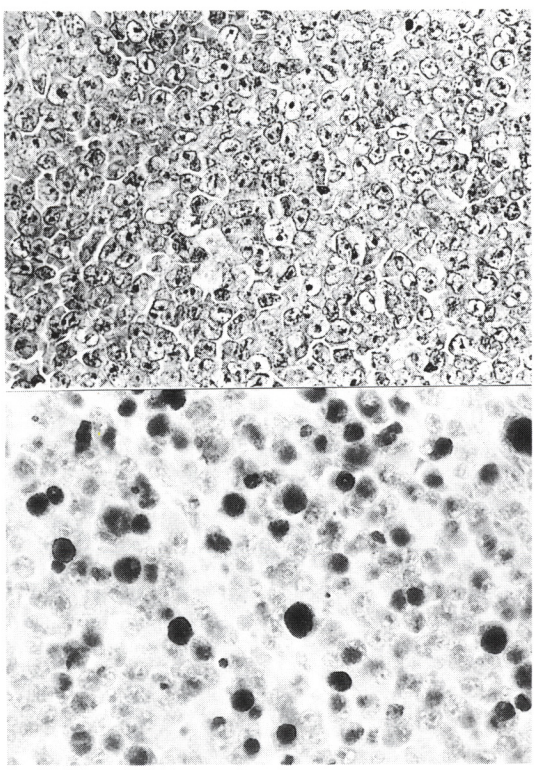

Fig. 2. Classic rhabdoid pattern (above; H&E, ×400). Note abundant cytoplasm, vesicular nuclei, and prominent nucleoli. Immunoperoxidase staining for vimentin shows vimentin-positive intracytoplasmic globules (below; avidin-biotin, ×400).

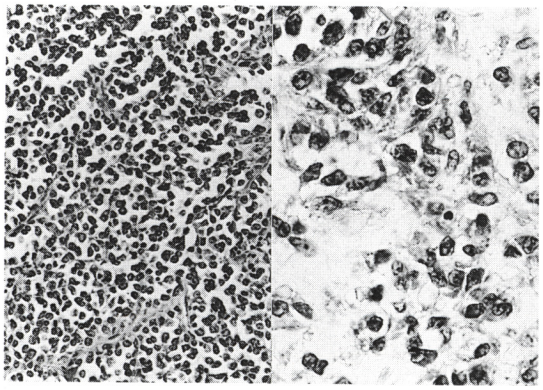

Fig. 3. Alveolar pattern reminiscent of alveolar rhabdomyosarcoma (left; H&E, ×200) and slerosing pattern showing scattered tumor cells in hyalinized stroma (right; H&E, ×400).

were usually single and centrally located. Mitoses were numerous. However, the other areas revealed various different patterns. In some areas, the tumor showed an alveolar pattern, reminiscent of alveolar rhabdomyosarcoma (Fig. 3), but no cross striations were found and immunostaining for desmin was negative. In this area, the tumor cells had also abundant eosinophilic cytoplasm and occasional hyaline intracytoplasmic globules. In the peripheral portion of the tumor, the tumor was characterized by dense hyaline stroma separating nests, cords, or single tumor cells (Fig. 3). Besides the above sclerosing area, tumor cells became spindle-shaped and showed somewhat myxoid appearance (Fig. 4). Spindle tumor cells blended into the surrounding connective tissue. Focal cyst formation was seen in this portion, where blood vessels were prominent. In other areas, the tumor consisted of monotonous tumor cells with vesicular nuclei, but was devoid of abundant cytoplasm. This area showed features similar to those of large cell lymphoma (Fig. 4). The tumor exhibited extreme local invasiveness. Renal pelvis involvement and multifocal renal tubular invasion with tumor cell cast were evident (Fig. 5).

Immunohistochemical study for anti-vimentin using avidin-biotin-complex method showed a well-demarcated vimentin-positive cytoplasmic globule in classical "rhabdoid" cells (Fig. 2). The cytoplasmic globule was also positive for PAS staining after distase treatment. The other cells were faintly positive in the cytoplasm. Staining for desmin was negative in all tumor cells and cytokeratin was not expressed except for entrapped renal tubules.

Ultrastructural examination revealed the filamentous nature of the cytoplasmic inclusions, but no definite rhabdomyoblastic elements. The inclusion was of large

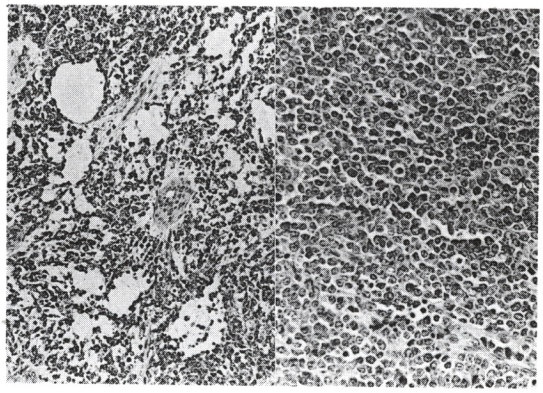

Fig. 4. In areas, the tumor consists of spindle cells in myxoid background (left; H&E, ×100). In some portions, the tumor cells resembling lymphoma cells have scanty cytoplasm and vesicular nuclei (right; H&E, ×200).

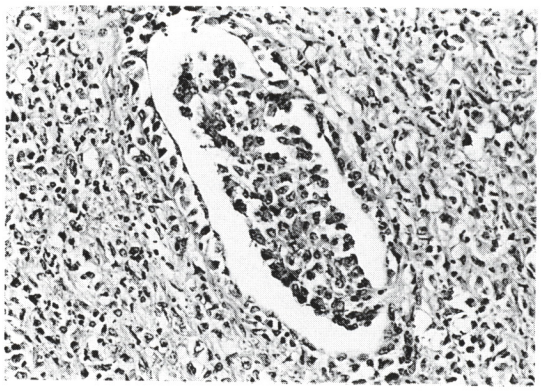

Fig. 5. The tumor cells invade the tubular lumens (H&E, ×200).

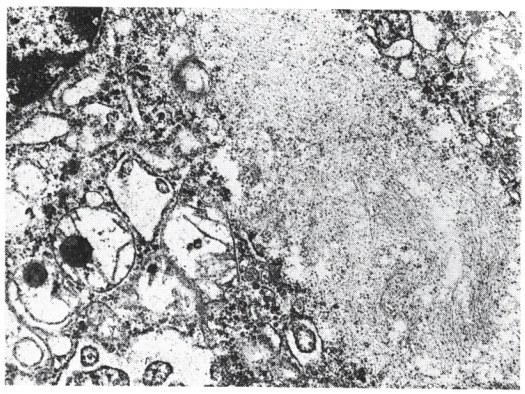

Fig. 6. Electron-micrograph of classic rhabdoid cell reveal whorled cytoplasmic filaments, mitochondria, and dilated endoplasmic reticulum (×30,000).

whorled masses of intermediate filaments, and was thought to be correlated with the vimentin-positive globules (Fig. 6). Dilated rough endoplasmic reticulum was observed. No basal lamina was seen. Nucleus was compressed by the cytoplasmic inclusion, but had a prominent nucleoli. The nucleoli were single, large, and centrally located.

DISCUSSION

We described a case of malignant rhabdoid tumor showing a classic "rhabdoid" pattern combined with variable patterns such as alveolar, sclerosing, and lymphomatous pattern. The classic "rhabdoid" cells had a vesicular nuclei with prominent nucleoli and contained concentrically whorled mass of intermediate filaments, which corresponded to well-defined vimentin-positive globules.

Following the first description of rhaboid tumor in 1981, many reports of similar tumors in the kindney and extrarenal sites have appeared. The rhabdoid tumor became very common in many organs and tissues including the liver, chest wall, paravertebral tissue, heart, pelvis, prostate, brain, and soft tissues (Parham DM et al., 1988; Tsokos M et al., 1989; Tsuneyoshi M et al., 1985; Schmidt D et al., 1982; Frierson HF et al., 195; Biggs PJ et al., 1987). These reports denied the consideration that rhabdoid tumor was a variant of Wilms' tumor. Myogenous origin was also discarded in view of no skeletal muscle differentiation in ultrastructural and immunohistochemical study. Haas et al. suggested a neural crest origin of the tumor on the basis of the similarity of the filamentous cytoplasmic inclusions to those of some APUD tumors (1981). Histicoytic, epithelial, and mesenchymal origins have also been suggested (Gonzalez-Crussi F et al., 1982; Parham DM et al., 1988; Frierson et al., 1985). A recent review of 111 cases of rhabdoid tumor on the National Wilms' Tumor Study argued that rhabdoid tumor was a distinct entity probably arising from primitive cells involved in the formation of the renal medulla (Weeks DA et al., 1989). According to the report, median age at diagnosis was 11 months, with a range from 0 to 106 months. Clinically, the tumor showed very poor prognosis. Mortality rate was about 79% and distant metastases frequently developed in the lung, abdomen, liver, brain, and bone. Nodal metastases were also common. Curiously, in 13.5% of the cases, brain tumors were associated and four patients showed hypercalcemia (Rousseau-Merck M-F et al., 1983; Bonnin JM et al., 1984). In comparison with rhabdoid tumor of the kidney in children, similar tumors of other sites were heterogeneous (Weeks DA et al., 1989). They were reported in older age and showed favorable prognosis.

Previously concentric filamentous structure consisting of vimentin and low-molecular-weight cytokeratin was considered to be very specific to the rhabdoid tumor (Vogel AM et al., 1984). However, as discussed above, such structures were described in several carcinomas such as renal cell carcinoma, and breast, lung, and liver carcinomas and sarcomas such as epitheliod sarcoma and synovial sarcoma (Datta BN, 1977; Dekker A et al., 1973). It might be considered as a peculiar expression of intermediate filament organization not specific for rhabdoid tumor. Although vimentin is mainly distributed in cells of mesenchymal origin, it can also be expressed by cells in culture and transiently by developing cells (Steinert PM and Liem RKH, 1990). In this regard the concentric filamentous structure is thought to be nonspecific. Some authors coined "pseudorhabdoid" tumor to represent such tumors showing rhabdoid appearance, i.e. PAS-positive inclusions in the cytoplasm, but not corresponding to classic rhabdoid tumor (Weeks DA et al., 1986). However, all rhabdoid tumors of extrarenal sites or in older age are not discarded. Hunt (1990) described rhabdoid tumor of the liver in children showing classic morphology and poor prognosis. Rhabdoid tumor may, therefor, not be confined to the kidney, but a histological entity that can occur anywhere in the body.

Our case showed classic morpholgy of the rhabdoid tumor of the kidney including "rhabdoid" cells, invasiveness, lymph node metastasis and probable lung metastasis. We believe that this case was the first case of malignant rhabdoid tumor reported in Korea. According to many authors, rhabdoid tumor must be separated from Wilms' tumor and be a distinct entity having a characterstic histolgy and uncertain origin.

REFERENCES

Biggs PJ, Garen PD, Powers JM, Garvin AJ: *Malignant rhabdoid tumor of the central nervous system. Hum Pathol 18:332-337, 1987*

Bonnin JM, Rubinstein LJ, Palmer NF, Bechwith JB: *The association of embryonal tumors originating in the kidney and in the brain. Cancer 54:2137-2146, 1984*

Datta BN: *Hyaline intracytoplasmic globules in renal carcinoma. Arch Pathol Lab Med 101:391, 1977*

Dekker A, Krause JR: *Hyaline globules in human neoplasms. A report of three autopsy cases. Arch Pathol Lab Med 95:178-181, 1973*

Frierson HF, Milla SE, Innes DJ: *Malignant rhabdoid tumor*

of the pelvis. Cancer 55:1963-1967, 1985

Fung CHK, Gonzalez-Crussi F, Yonan T, Martinez N: *"Rhabdoid" Wilms' tumor. Arch Pathol Lab Med 105:521-523, 1981*

Gonzalez-Crussi F, Goldschmidt RA, Hsueh W et al.: *Infantile sarcoma with intracytoplasmic filamentous inclusions: Distinctive tumor of possible histiocytic origin. Cancer 49:2365-2375, 1982*

Haas JE, Palmer NF, Weinberg AG, Beckwith JB: *Ultrastructure of malignant rhabdoid tumor of the kidney. A distinctive renal tumor of children. Hum Pathol 12:646-657, 1981*

Hunt SJ, Anderson WD: *Malignant rhabdoid tumor of the liver. Am J Clin Pathol 94:645-648, 1990*

Parham DM, Peiper SC, Robicheaux G, Ribeiro RC, Douglass EC: *Malignant rhabdoid tumor of the liver. Arch Pathol Lab Med 112:61-64, 1988*

Rousseau-Merck M-F, Nogues C, Nezelof C, Marin-Cudraz B, Paulin D: *Infantile renal tumors assocaited with hypercalcemia. Arch Pathol Lab Med 107:311-314, 1983*

Schmidt D, Harms D, Zieger G: *Malignant rhabdoid tumor of the kidney. Histopathology, ultrastructure and comments on diffential diagnosis. Virchows Arch (Pathol Anat) 398:101-108, 1982*

Steinert PM, Liem RK II: *Intermediate filament dynamics. Cell 60:521-523, 1990*

Tsokos M, Kouraklis G, Chandra RS, Bhagavan BS, Triche TJ: Malignant rhabdoid tumor of the kidney and soft tissues. Arch Pathol Lab Med 113:115-120, 1989

Tsuneyoshi M, Daimaru Y, Hashimoto H, Enjoji M: *Malignant soft tissue neoplasms with the histologic features of renal rhabdoid tumors. An ultrastructural and immunohistochemical study. Hum Pathol 16:1235-1242, 1985*

Vogel AM, Gown AM, Caughlan J, Haas JE, Beckwith JB: *Rhabdoid tumors of the kidney contain mesenchymal specific and epithelial specific intermediate filament proteins. Lab Invest 50:232-238, 1984*

Weeks DA, Beckwith JB, Mierau GW: *'Pseudo-rhabdoid' tumors of kidney, abstracted. Lab Invest 54:10p, 1986*

Weeks DA, Beckwith JB, Mierau GW: *Rhabdoid tumor. An entity or a phenotype? Arch Pathol Lab Med 113:113-114, 1989*

Weeks DA, Beckwith JB, Mierau GW, Luckey DW: *Rhabdoid tumor of kidney. A report of 111 cases from the National Wilms' Tumor Study Pathology Center. Am J Surg Pathol 13:439-458, 1989*