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Clear Cell Sarcoma of the Kidney
—A case report—

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Clear cell sarcoma of the kidney (CCSK) is a highly malignant childhood tumor, distinguished from classic Wilms' tumor by its propensity to metastasize to the skeletal system. Authors described a case of CCSK from a 3-year-old boy in the right kidney, showing various histologic features, such as classic, epithelioid, trabecular, neurilemmoma-like, cystic and entrapped collecting tubular pattern. Ultrastructurally epithelial differentiation was absent. Immunohistochemically, none of the intrinsic tumor cells showed positive staining with the antibodies against the keratin, S-100 protein, carcinoembryonic antigen, vimentin, desmin and myoglobin, suggesting primitive mesenchymal cell in origin.

Key Words: *Clear cell sarcoma of the kidney, Wilms' tumor*

INTRODUCTION

Clear cell sarcoma of the kidney (CCSK) is recently recognized and thought to be a highly malignant childhood tumor of uncertain origin. It is distinguished from classic Wilms' tumor by the aspects of morphological and clinical behaviours (Beckwith and Palmer, 1978). This tumor constitutes about 4% of all childhood renal tumor and is apparently the same one that was reported as "the bone metastasizing renal tumor of childhood (Marsden and Lawler, 1978; Lawler and Marsden, 1979) because of its predilection for bone metastases. Beckwith and Palmer (1978) and Morgan et al. (1978) classified the neoplasm as a variant of Wilms' tumor with different clinicopathologic behavior. However, many authors favor the term" clear cell sarcoma of the kidney" as a seperate entity. Exact histogenesis of this tumor still remains uncertain, although theory of primitive mesenchymal origin is highly appreciated.

We experienced this rare tumor in a 3-year-old boy involving the right kidney and herein reported.

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CASE REPORT

A 3 year-old-boy was admitted to the Department of Pediatrics, Chung-Ang University Hospital with an abdominal mass incidentally detected by his father. He had suffered from generalized aching pain with nausea and fever of mild degree for 10 days, which were partially improved by symptomatic management at the private clinic. A sharply demarcated hard mass was palpable at the right side of abdomen without tenderness on physical examination.

Simple abdominal X-ray and ultrasonography showed a huge solid mass occupying the right side of abdomen. Computerized tomography showed a large, well demarcated renal mass with homogeneous low density and a peripheral fluid density with laterally displacing renal parenchyme (Fig. 1). Intravenous pyelography revealed delayed excretion. However, bone scan, chest X-ray and other laboratory findings were all within normal limit.

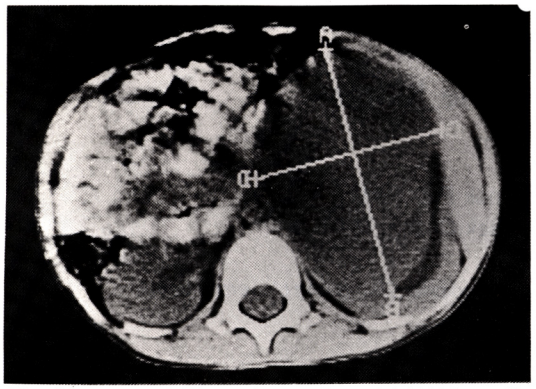

Fig. 1. Abdominal CT reveals an enlarged right kidney with homogeneous density and peripheral fluid density with laterally displaced renal parenchyme.

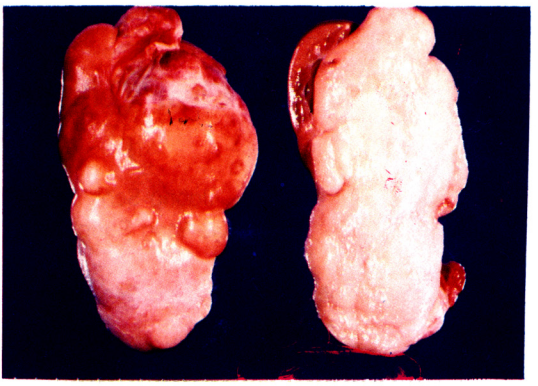

Fig. 2. Markedly enlarged kidney with bosselated surface and homogeneously pale tan to grayish white, bulging cut surface with partly microcystic areas.

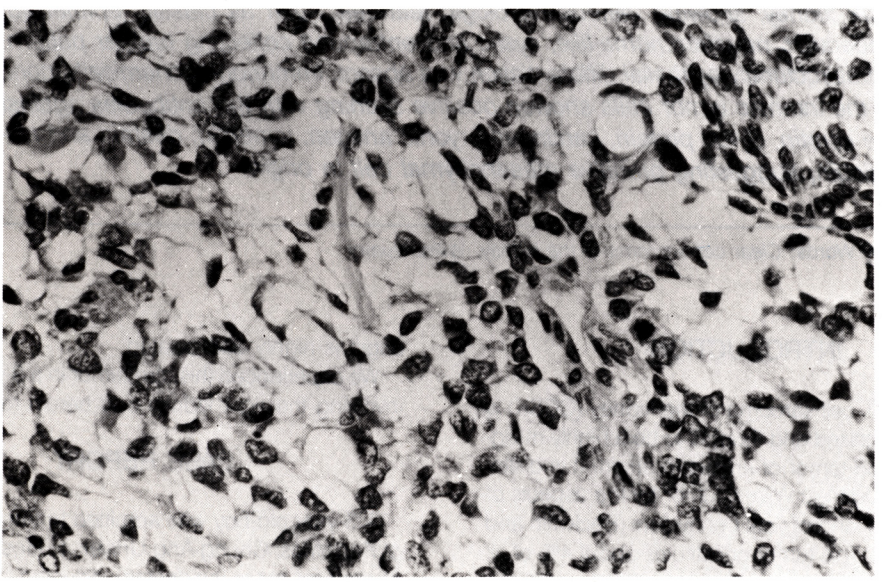

Fig. 3. Classic pattern of polygonal clear cells having oval to round nuclei and poorly stained, vacuolated cytoplasm associated with aborizing small vessels (H & E, ×200).

Two days later, right nephrectomy was performed under the impression of Wilms' tumor. The lower portion of tumor mass was firmly adhered to retroperitoneum and inferior vena cava, and several enlarged mesenteric and hilar lymph nodes were also noted. Metastasis to other organs was not evident. After operation, he was treated with radiation therapy (total 3,000 Rad) and combination chemotherapeutic regimen including actinomycin D, vincristine sulfate, adriamycin and cyclophosphasmide. Postoperative outcome was uneventful and he is now disease free for six months.

Removed kidney was markedly enlarged with bosselated surface, especially at the lower pole and hilar lesion, and was mostly replaced by the tumor. The kidney containing tumor weighed 605gm and measured 18.0×9.5×7.0cm. The cut surface of the tumor was buldging out and revealed homogeneously pale tan to pale yellow or grayish white appearance with partial lobulation and discretely minute cystic areas (Fig. 2). The renal parenchyme was spared only in the upper lateral aspect. The perinephric fatty tissue was free of tumor. Microscopically the tumor showed varying histology from area to area, composed of

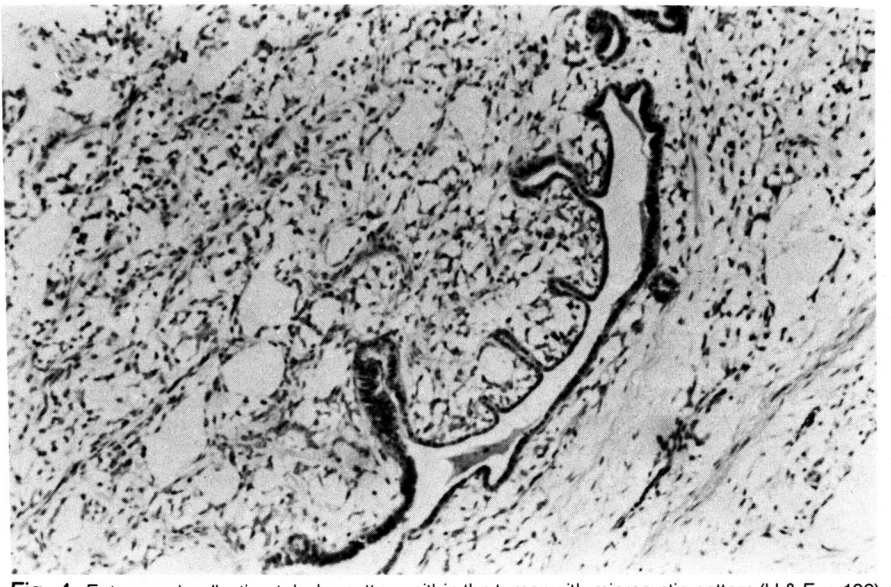

Fig. 4. Entrapped collecting tubular pattern within the tumor with microcystic pattern (H & E, ×100).

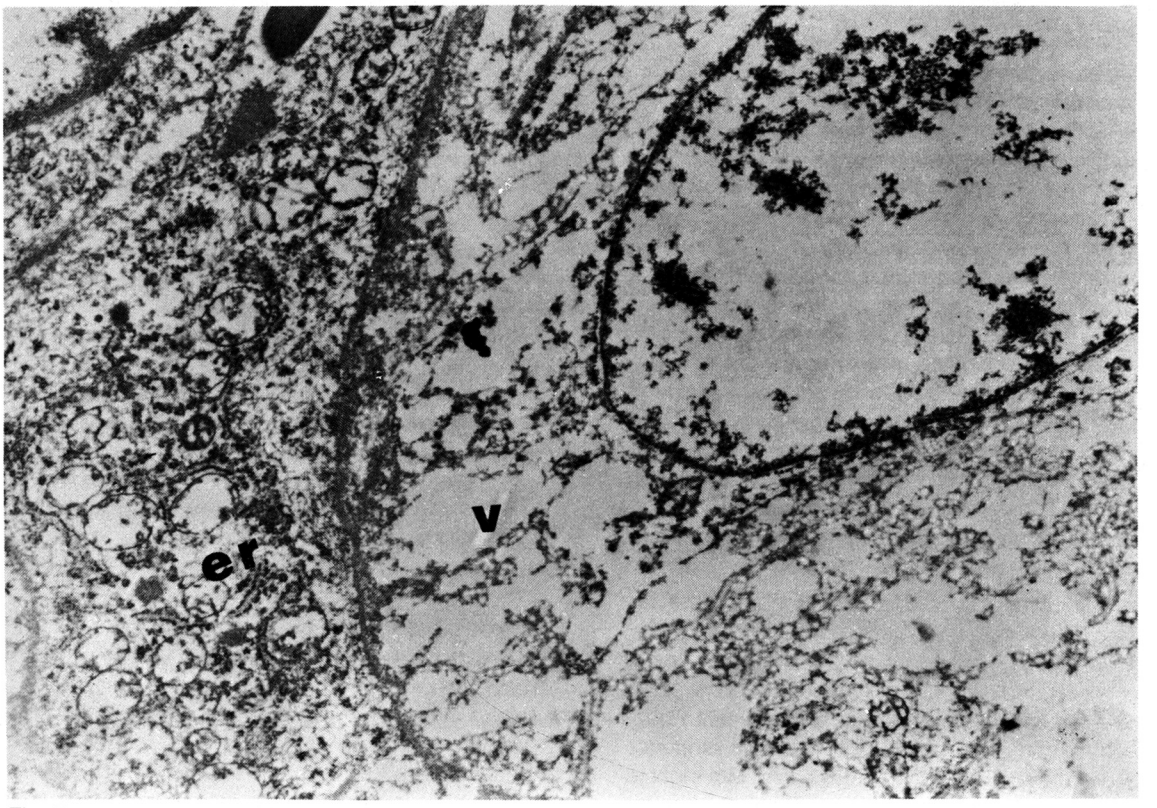

Fig. 5. Electronmicroqraph of clear tumor cells show vacuolar changes (v) in cytoplasm and moderately developed endoplasmic reticulum (er) with glycogen particles (E.M. ×13,600).

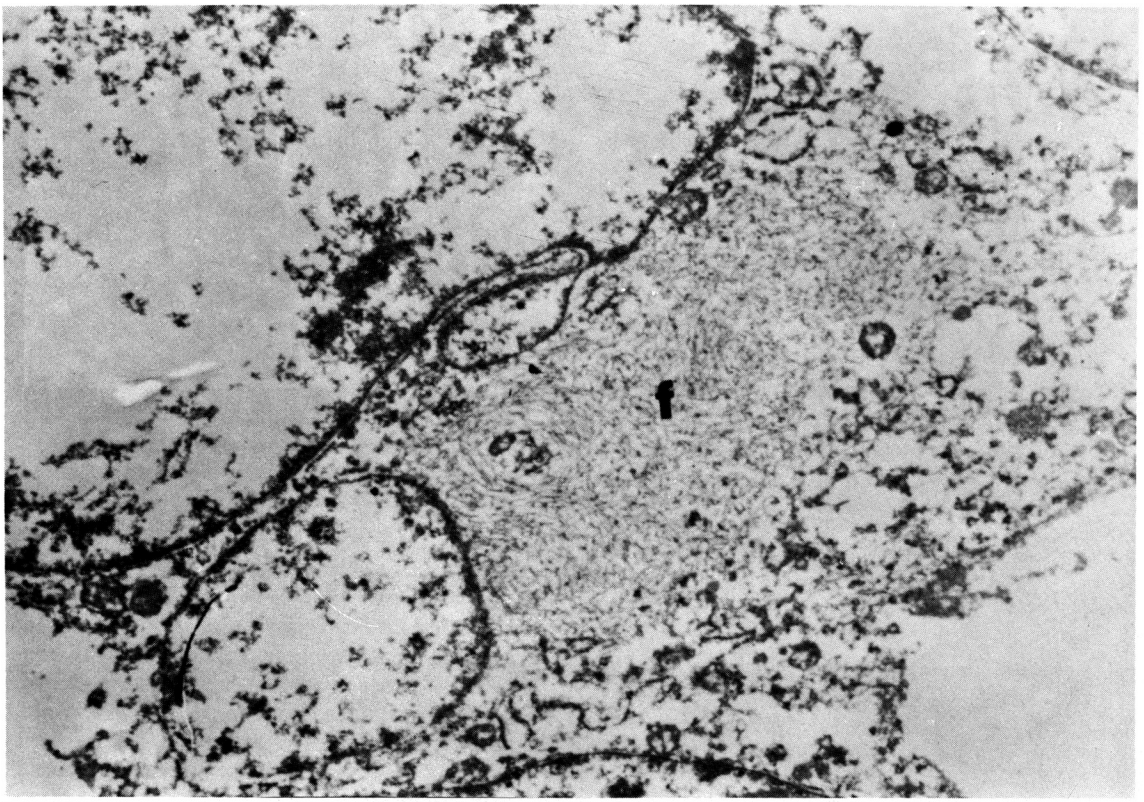

Fig. 6. Perinuclear low molecular keratin-like microfilaments (f) within a few tumor cells (E.M. ×13,600).

diffuse and solid proliferation of monomorphous polygonal clear or epithelioid cells having poorly stained, scanty cytoplasm and indistinct cell borders (Fig. 3). They have uniformly oval to round nuclei with fine, evenly granular chromatin. Mitotic fegures were occasionally noted. Another distinct microscopic feature of this tumor was nest or cord-like arrangement by evenly distributed, arborizing network of small vessels. In addition to the above classic patterns, some variant patterns were also present such as epithelioid, trabecular pattern, neurilemmoma-like nuclear palisading pattern, cystic pattern (Fig. 4) and entrapped collecting tubules (Fig. 4).

Electron microscopic features by previous paraffin embedded tissue revealed that clear tumor cells showed large and small cytoplasmic vacuoles suggestive of previous lipid content. Mitochondria and glycogen particles were also noted in moderate amount. Rough and smooth endoplasmic reticulum were moderately developed (Fig. 5). Nuclei were oval with slight indentation. Spindle cells show ed spindle shaped nuclei with scanty intracellular microorganelles and thick membrane without any desmosome-like structure. A few tumor cells containing perinuclear clumps of low molecular keratin-like microfilaments were also noted (Fig. 6). Basal lamina, desmosome, and melanosome were not oserved.

Immunohistochemical studies were performed by using the peroxidase antiperoxidase technique. The antibodies used in this study were keratin, S-100 protein, CEA, vimentin, desmin and myoglobin. None of the tumor cells showed positive staining with any above antibodies although false negative results could not be ruled out because of the materials were not fresh.

DISCUSSION

A clear cell sarcoma of the kidney (CCSK) is a highly malignant tumor which develops in young childhood and has a tendency to metastasize to the bone. Recent interest in histopathologic correlation of various histologic patterns of Wilms' tumor had led to the identification of at least one distinct tumor type which could not be included in the category of Wilms' tumor. The term CCSK used since 1978 is also far from satisfactory, but many authors propose to retain it until the

cell of origin of this enigmatic entity can be clarified (Beckwith and Palmer, 1978; Haas et al, 1984).

Incidence of CCSK is very rare, constituting only 4% of all childhood renal tumor (Beckwith, 1983; Finegold and Bennington, 1986) and only one case has been reported by Seo et al, 1984 in Korea. The age at presentation of CCSK is similar to that of classic Wilms' tumor and male to female ratio is 1.7 to 1 in general, suggesting male predominance (Haas et al. 1984; Okawa et al. 1987; Marsden et al. 1987).

Grossly, CCSK presents no distinguishing characteristic, and the cut surface is usually homogeneously tan or gray tan. Cystic spaces are often present. The margins of these tumors are sharply circumscribed and differ from those of most Wilms' tumors by that the latter infiltrate the renal parenchyme instead of a pseudocapsule by compression (Haas et al, 1984).

Microscopic appearance is usually uniform (classic pattern) but may vary in a same tumor (variant pattern) as seen in this presented case. The classic pattern is characterized by monomorpous proliferation of polygonal cells, which have oval to polygonal nuclei with fine, evenly granular chromatin and indistinct nucleoli, and poorly stained cytoplasm with indistinct cell boundaries. In addition delicate fibrovascular stroma with evenly arborizing capillary networks is seen coursing through the nest of cord-like arrayed tumor cells. The entrapped tubules often become lined by basophilic cuboidal to columnar cells with an embryonal appearance, which was the one reason that we could not have excluded a relationship of this tumor with Wilms' tumor. However, it has subsequently become apparent that such tubules do not occur in the metastatic foci or in the extrarenal extensions of CCSK. Variations are: epithelioid, trabecular pattern, neurilemmoma-like nuclear palisading pattern, fibrosis and stromal hyalinzation, cyst, and angioectatic pattern (Finegold and Bennington, 1986; Beckwith, 1986).

Ultrastructurally, the clear cells show vacuoles and a few mitochondria and glycogen particles while the spindle cells show fewer organelles. Epithelial subcellular structures such as desmosome, melanosome, basal lamina are not obaserved as well as mesenchymal structures, suggesting that the origin of this tumor is primitive mesenchymal cells.

Immunohistochemical studies on this case such as keratin, vimentin, desmin, S-100 protein and myoglobin were all negative, so that the histogenetic differentiation of CCSK can not be clarified (Okawa et al, 1987). Perinuclear clumps of microfilaments are thought to be a low molecular keratin or precursors of myofilaments although immunohistochemical staining did not supported.

The incidence of metastasis is higher than the Wilms' tumor and predilection sites are bone, lung, brain and regional lymph nodes. The actual 2 year survival rate is 39% to 49% in general (Beckwith and Palmer, 1978; Marsden and Lawler, 1980).

In summary, the microscopic and immunohistochemical features of CCSK are suggestive of pirmitive mesenchymal origin, distinguished from other primary renal tumors. Whatever its cell of origin or true nature might be, the CCSK seems to be unique renal tumor with histologic patterns and its prognosis.

REFERENCES

Beckwith JB: *Wilms' tumor and othere renal tumors of childhood: selective review from the National Wilms'tumor Study Pathology Center. Hum Pathol 14:481-492, 1983.*

Beckwith JB, Palmer NF: *Histopathology and prognosis of Wilms' tumor. Result from the first national Wilms'tumor study. Cancer 41:1937-1948, 1978.*

Carcassone M, Raybaud C, Bebreuil G: *Clear cell sarcoma of the kidney in children: a distinct entity. J Pediatr Surg 16:645-1981.*

Finegold M, Bennington JL: *Pathology of neoplasia in children and adolescents. Volume 18 in series MPP, W. B. Saunders Co., Philadelphia, P321-325, 1986.*

Haas JE, Bonadio JF, Beckwith JB: *Clear cell sarcoma of the kidney with emphasis on ultrastructural studies. Cancer 54:2978-2987, 1984.*

Kidd JK: *Exclusion of certain renal neoplasms from the catergory of Wilms' tumor. Am J Pathol 59:16a, 1970.*

Lawler W, Marsden HB: *Bone metastases in children presenting with renal tumors. J Clin Pathol 32:608-615, 1979.*

Marsden HB, Lawler W: *Bone metastasizing renal tumor of childhood. Cancer 42:1922-1928, 1978.*

Marsden HB, Lawler W: *Bone metastaizing renal tumor of childhood: Histopathological and clinical review of 38 cases. Virhows Arch A Path Anat and Histol 387:341-351, 1980.*

Morgan E, Kidd JM: *Undifferentiated sarcoma of the kidney. A tumor of childhood with histopathologic and clinical characteristics distinct from Wilms' tumor. Cancer 42:1916-1921, 1987.*

Okawa K, Nakashima Y, Kim YC, Yamabe H, Mizukami T, Kaneko S, Yamashima Y: *Unusual renal cell sarcoma in a young adult: Its simularities to clear cell sarcoma of the kidney. J Urol 137:730-731, 1987.*

Seo JW, Chi JG, Lee SK, Ahn HS: *Pathological Study on Wilms Tumor. Kor J Path 18:113-121, 1984.*