# Clear Cell Sarcoma of the Kidney—A Case Report

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Clear cell sarcoma of the kidney (CCSK) is a rare and highly malignant tumor which is usually confused with other kidney tumors. We experienced such a patient and present report this.

## Case Report

A 9-year old girl was admitted to a local hospital because of fever and a pain in her right abdomen. The computerized tomography showed: a mass in the lower pole of the right kidney. A laparotomy was conducted with the frozen tissue specimens showing necrosis and blood cells in addition to a few suspected cancer cells. The tumor was ablated and diagnosed as kidney cancer in the local hospital. But the consultation of the Department of Pathology in our hospital was CCSK. A CT after the operation showed that the tumor remained.

After a month, the patient received an operation in our hospital. The bone scan was normal before operation. This time, the right kidney was adhered to the surrounding tissue and the tumor had partly invaded the bowel. The right kidney and surrounding tissue were excised and the pathology after operation confirmed to be CCSK. Microscopic features showed the following: the tumor had nest or cord-like arrangement with distinct necrosis and ample vessels. The tumor cells were clear with abundant cytoplasm. Immunohistochemical studies provided these results: Vima(+), CK(+), EMA(-), S-100(-), desmin (-). We treated the patient with 4 cycles of chemotherapy every 3 weeks using vincristine and doxorubicin. She has been disease free for half a year.

## Discussion

Clear cell sarcoma of the kidney is a rare malignant kidney tumor which is distinguished from Wilms' tumor. It usually arises in male children and rarely occurs in adults. The tumor constitutes about 4% of all childhood renal tumors. Some patients have a family history of this tumor. The prognosis is poor with mortality being about 50%. Previously, CCSK was considered to be a type of Wilms' tumor, but these 2 types of tumors differ by their characteristic histologic features and a more aggressive clinical behavior of CCSK having a tendency to metastasize to bone.[1] CCSK is highly malignant, so a correct diagnosis should result in the application of appropriate treatment.

The typical clinical features of CCSK include a mass and pain in the abdomen and blood in the urine. There are no deformations that

occur in Wilms' tumor such as obesity, iris absence, etc. Since bone metastasis often are found in the skull (40~60%), it is also called bone-metastasizing renal tumor of childhood, while the typical Wilms' tumor often metastasizes to the liver and lung.

The typical gross features of CCSK include a large size, a mucoid texture, foci of necrosis, and prominent cyst formation. The margins of these tumors are sharply circumscribed. The classic microscopic appearance of CCSK is characterized by monomorpous proliferation of polygonal cells, which have oval to polygonal nuclei with tiny evenly granular chromatin and indistinct nucleoli, and poorly stained cytoplasm with indistinct cell boundaries. In addition a delicate fibrovascular stroma with evenly arborizing capillary networks is seen coursing through the nest of cord-like arrayed tumor cells.[2,3]

CCSK has a poor prognosis with almost no long-term survivors. Multimodal oncologic treatment includes surgery, chemotherapy and radiotherapy. Chemotherapy employing dixorubicin can improve the survival rate. Multivariate analysis reveals 4 independent prognostic factors for survival: treatment with doxorubicin, stage, age at diagnosis, and tumor necrosis. The ability to cure CCSK is poorer than that of Wilms' tumor with most patients dying of bone metastases. This characteristic indicates that a bone examination is of utmost importance, especially in follow-up.[4,5] This patient only received a tumor resection in her first hospitalization, consequently the tumor invaded the surrounding tissue prior to the second operation a month later which may affect the prognosis.

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