A Case of Clear Cell Sarcoma of the Kidney

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ABSTRACT

Clear cell renal sarcoma is a rare tumor and comprises 4% of primary pediatric malignant renal tumors. It is known as an aggressive tumor with poor prognosis. Clinically and radiographically, it resembles Wilms tumor. We present a case of a child with an abdominal mass that was diagnosed as clear cell sarcoma of her right kidney.

CASE REPORT

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A 3-year-old white female was found to have an abdominal mass by her primary care physician on routine visit. Initially, her abdominal mass was thought to be due to fecal impaction, and she was subsequently started on Miralax, Milk of Magnesia, and Glycolax. Consequently, she had several bowel movements, but the size of the abdominal mass did not decrease. She was then sent for further evaluation. Blood and urine tests were unremarkable. Abdominal sonography revealed a large heterogeneous right renal mass (Figure 1). Abdominal computerized tomography (CT) revealed a large right renal mass. The normal renal parenchyma encircled the mass producing a lobster claw deformity, called the radiographic claw sign. The mass had heterogeneous enhancement (Figure 2). The left renal vein was displaced by the mass medially. No thrombus was noted in the right renal vein, in the inferior vena cava, or in the right atrium. The patient underwent right nephrectomy. Initially post operatively, the mass was thought to represent Wilms tumor; however, the final diagnosis was renal clear cell sarcoma. The macroscopic nephrectomy specimen revealed a well-circumscribed tumor located in the inferior pole of the kidney (figure 3). Regional lymph node metastasis and nephrogenic rests were not identified. The tumor was diagnosed pathologically (Figures 4, 5) as renal clear cell sarcoma with focal microscopic penetration of the renal capsule but negative resection margins, consistent with stage II disease. Further evaluation by bone scintigraphy and chest CT revealed no distant metastases.

The patient underwent a chemotherapy regimen of cytoxan, doxorubicin, and vincristine and is doing well.

DISCUSSION

Malignant pediatric renal tumors include nephroblastoma, which can develop into Wilms tumor (85%), mesoblastic nephroma (5%), clear cell sarcoma (4%), rhabdoid tumor (2%), and other rare tumors (2%).

Wilms tumor, which accounts for 6-7% of all pediatric malignancies [1], is the most common renal tumor in children older than 6 months and younger than 12 years of age [2]. When a pediatric patient presents with an abdominal mass, rare childhood renal neoplasms should be considered along with Wilms tumor. Clear cell sarcoma of the kidney (CCSK) is one of these rare childhood renal neoplasms. CCSK represents one of the most common tumors with "unfavorable histology" listed by the National Wilms Tumor Study Group (NWTSG). Unlike Wilms tumor, CCSK is not associated with nephrogenic rests.

CCSK has been reported by the NWTSG to represent 4% of childhood renal tumors [3] and is seen mainly in young children with a peak incidence between 2 and 3 years of age [4] with a male predominance [5]. It is the most frequently misdiagnosed pediatric renal tumor, attributable to its infrequency, morphological diversity, and lack of specific diagnostic markers [3]. Approximately 5% of patients have metastatic disease at presentation. The most common site of metastasis at the time of presentation in patients with clear cell sarcoma of the kidney is the ipsilateral renal hilar lymph nodes. Skip metastases to periaortic lymph nodes have been reported as well. Treatment consists of nephrectomy and chemotherapy with current long-term survival rate of 60-70% [5].

One important distinguishing feature of CCSK is its 40-60% incidence of bone metastasis, which is much higher than the 2% incidence of bone metastasis found in Wilms patients [2]. The bone metastasis may also be both lytic and sclerotic. Bone is the most common site of distant metastases followed by lung, retroperitoneum, brain, and liver. Additionally, CCSK has been reported to metastasize to unusual sites such as the scalp, epidural space, nasopharynx, neck, paraspinal area, abdominal wall, axilla, and orbit [6]. Its aggressiveness and increased risk of bone metastases, along with its propensity for late relapse (up to 4 years from original diagnosis [6]) and relatively poor outcome compared to Wilms tumors, supports the importance of early and correct diagnosis [3].

Sonography is the initial modality to evaluate abdominal mass and helps demonstrate renal origin of the tumor. Sonography can distinguish between solid and cystic tumors and can diagnose other conditions such as hydronephrosis. In CCSK, the mass shows heterogenous echogenicity with cystic components and necrosis. Large fluid filled cystic spaces with echogenic septa may also be present. The pattern in computerized tomography (CT) is of inhomogeneous enhancement, with attenuation less than that of normal renal parenchyma. Furthermore, these tumors also contain areas of low attenuation corresponding to necrosis and cysts. The mass can cross the midline and displace vessels [7]. CT scans are invaluable in evaluating the size and extent of abdominal masses, any surrounding lymphadenopathy, and the resectability of tumors seen. However, these imaging studies are unfortunately nonspecific when it comes to renal neoplasms in the pediatric population. MRI has limited advantage in relation to CT. These days, CT coronal and sagittal reformats are nearly equivalent to multi-planar views of magnetic resonance images. The advantage of magnetic resonance imaging is the lack of exposure of ionizing radiation, which is highly desirable in the pediatric population. Radionuclide bone survey has an important role for detecting bone metastasis once the diagnosis is made, due to its high incidence.

Although clinical and imaging modality characteristics may support a particular diagnosis, the pretreatment gold standard should be final histopathologic determination. CCSK is consistently positive for vimentin and usually negative for cytokeratin [4]. Histopathologically, CCSK has been reported to have a distinctive complex vascular network, classically described as "chicken-wire" pattern [3]. CCSK is a malignant mesenchymal neoplasm that includes undifferentiated cells, cords and nests separated by fibrovascular septa, and abundant extracellular matrix. It has a variety of histologic patterns that includes classic pattern of nests or cords with arborizing vascular septa, myxoid, sclerosing, cellular, epithelioid, spindle cell, palisading, and sinusoidal (pericytomatous) pattern. Nevertheless, there are no tumor specific markers for CCSK, which makes the diagnosis difficult.

Treatment planning should involve a multidisciplinary team including pediatric surgeons or pediatric urologists, pediatric radiation oncologists, and pediatric oncologists. A pediatric radiation oncologist is needed for irradiation of the tumor bed and any other sites of the disease. A pediatric oncologist should be onboard to determine standard and investigational treatment protocols.

After a diagnosis of CCSK is made, radical nephrectomy is the initial treatment of choice if the lesion is resectable. Chemotherapy involves vincristine, doxorubicin, and dactinomycin for 15 months, which has shown an improved relapse free survival rate in contrast to shorter durations [8].

TEACHING POINT

Clear cell sarcoma of the kidney is a rare aggressive tumor with nonspecific imaging findings. Although Wilms tumor is the most common renal neoplasm in the pediatric population, other rare childhood renal neoplasms should be considered in a pediatric patient presenting with an abdominal mass. The practicing radiologist should be aware of clear cell sarcoma and include it in the differential diagnosis of pediatric renal tumors.

REFERENCES

1. Hadley GP, Sheik-Gafoor. Clear cell sarcoma of the kidney in children: experience in a developing country. Pediatric Surgery International. 26:345-348, 2010.

2. Miniati D. Gay AN, Parks KV, Naik-Mathuria BJ, Hicks J, Nuchtern JG, Cass DL, Olutoye OO. Imaging accuracy and incidence of Wilms and non-Wilms renal tumors in children. Journal of Pediatric Surgery. 43:1301-1307, 2008

3. Boo YJ, Fisher JC, Haley MJ, Cowles RA, Kandel JJ, Yamashiro DJ. Vascular characterization of clear cell sarcoma of the kidney in a child: a case report and review. Journal of Pediatric Surgery. 44:2031-2036, 2009.

4. Sebire NJ, Vujanic GM. Paediatric Renal Tumours: Recent Developments, New Entities, and Pathological Features. Histopathology. 54:516-528, 2009

5. Lowe LH, Isuani BH, Heller RM, Stein SM, Johnson JE, Navarro OM, Hernanz-Schulman M. Pediatric Renal Masses: Wilms Tumor and Beyond. Radiographics. 20:1585-1603, 2000.

6. Zhuge Y, Cheung MC, Yang R, Perez EA, Koniaris LG, Sola JE. Pediatric Non-Wilms Renal Tumors: Subtypes, Survival, and Prognostic Indicators. Journal of Surgical Research. 156(2): 257-263, 2010.

7. Glass RBJ, Davidson AJ, Fernbach SK. Clear Cell sarcoma of the Kidney: CT, Sonographic, and Pathologic Correlation. Radiology 180: 715-717, 1991.

8. Seibel Neil. Clear Cell Sarcoma of the Kidney. eMedicine Pediatrics. http://emedicine.medscape.com/article/993245, 2009

9. White KS, Grossman H. Wilms and associated renal tumors of childhood. Pediatr Radiol 21:81-88, 1991

FIGURES

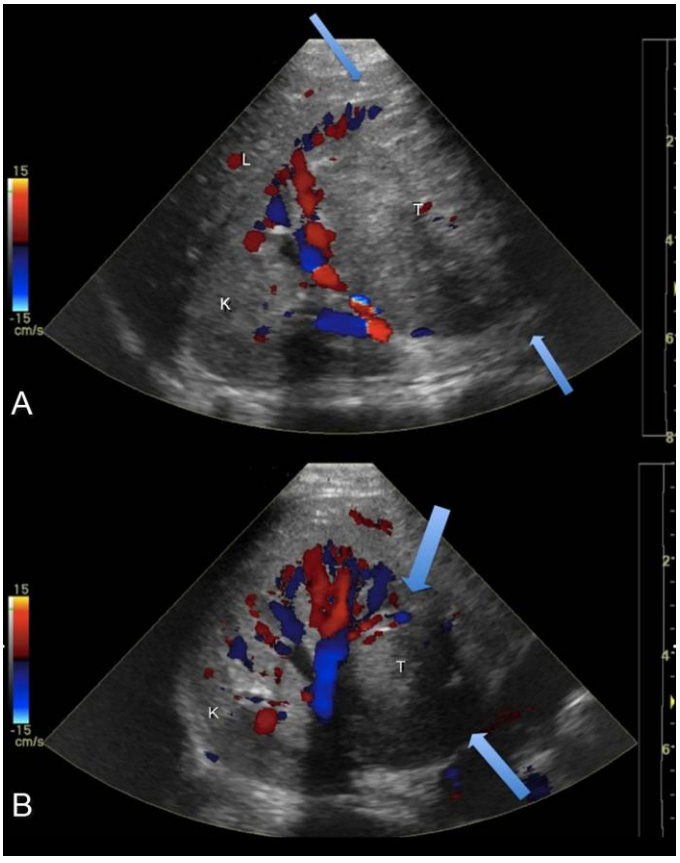

Figure 1: 3 year-old female with clear cell sarcoma of the right kidney. Sonogram of the right kidney performed with multi-frequency probe (GE sector 4-10) in longitudinal (a) and transverse (b) views demonstrates large hypoechoic and heterogeneous mass originating from the lower pole of the right kidney (arrows). Increased flow around the mass is demonstrated. (L - Liver; K - Right kidney; T - the mass)

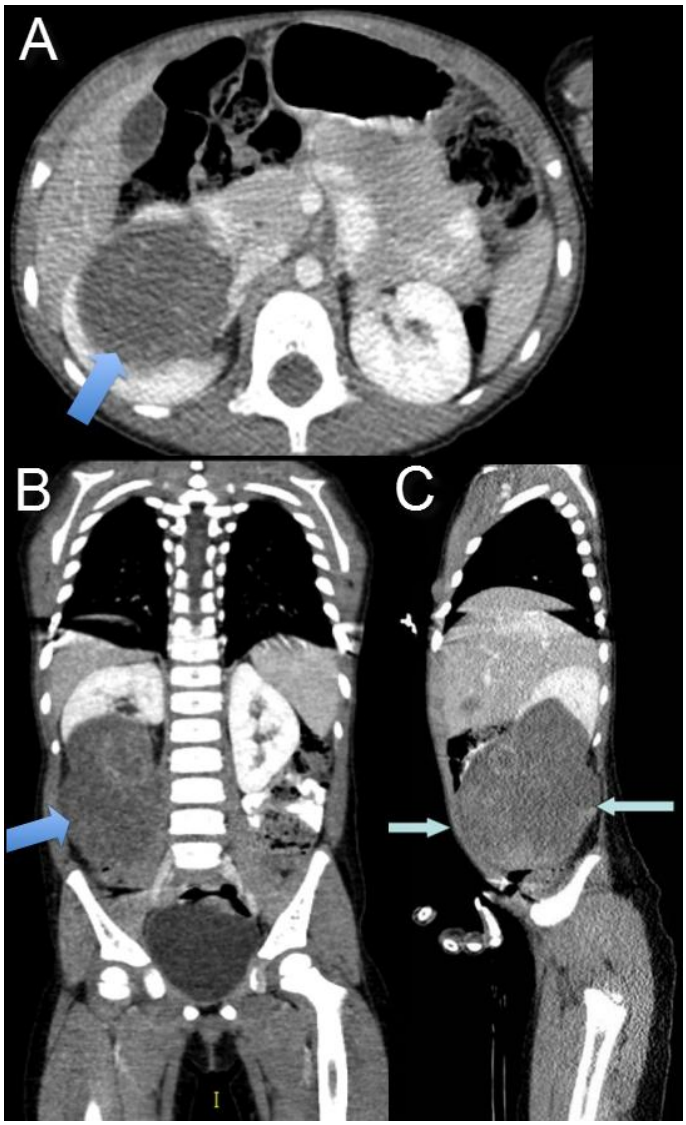

Figure 2: 3 year-old female with clear cell sarcoma of the right kidney. Axial (a), coronal (b), and sagittal (c) views of abdominal CT obtained following the intravenous administration of 30 cc of Omnipaque 350 and oral contrast. The CT setting was 100 kVp with modulated mAs. The images are 5mm slices at the level of the right kidney and demonstrate large renal mass. The mass measures 12.3 cm craniocaudally, 7.9 cm in the anteroposterior diameter, and 6.5 cm in the transverse diameter. The mass heterogeneously enhances (arrow). The mass originates from the lower pole of the right kidney. The kidney encircles the mass and forms the claw sign.

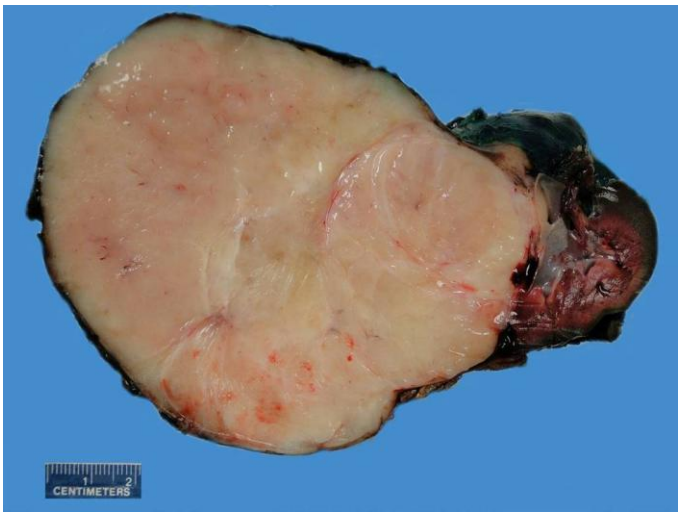

Figure 3: 3 year-old female with clear cell sarcoma of the right kidney. Gross photograph of tumor involving and extending from the lower pole of the kidney. The tumor is grossly contained by the renal capsule.

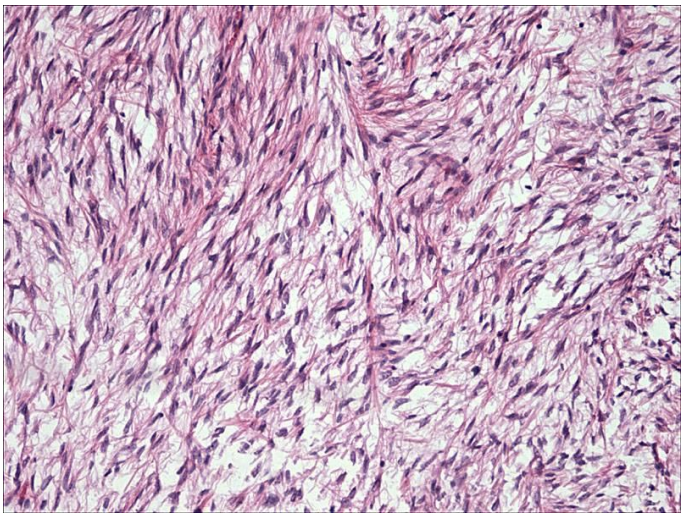

Figure 5: 3 year-old female with clear cell sarcoma of the right kidney. Photomicrograph showing an area with a less cellular spindle cell pattern. This looser, fibroblastic-like pattern was interspersed focally amidst the more cellular pattern shown in Fig. 4. H&E stain, 200x

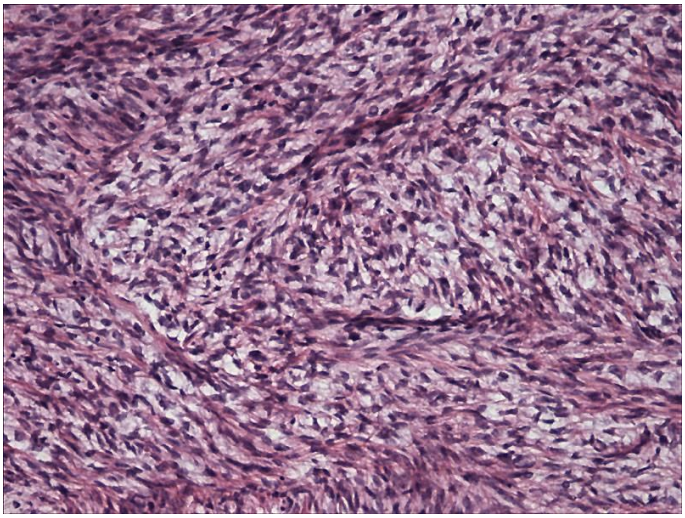

Figure 4 (left): 3 year-old female with clear cell sarcoma of the right kidney. Photomicrograph showing the spindle cell pattern that was seen throughout the vast majority of the tumor. H&E stain, 200x

Etiology	The tumor is composed of undifferentiated cells as illustrated by its relative lack of immunohistochemical reactivity
Incidence	4% of the malignant pediatric renal tumors. Approximately 20 new cases of CCSK are diagnosed each year in the United States
Gender Ratio	Males appear to be more commonly affected than females. Ratio 2:1
Age predilection	Peak incidence in the second year of life
Risk factors	Unknown
Treatment	Surgery, Chemotherapy, Radiation.
Prognosis	Poor outcome. 40 to 60% risk metastasizing to bone and other organs. Risk of late recurrence.
Findings on imaging	Nonspecific. Heterogeneous solid mass arising from the kidney on CT and Sonography. Claw sign on CT.

Table 1. Summary table of clear cell sarcoma of the kidney

Diagnosis	Ultrasound	CT	MRI
CCSK	A large renal mass indistinguishable from Wilms tumor [9]	A large renal mass indistinguishable from Wilms tumor [9]	A large renal mass indistinguishable from Wilms tumor [9]
Wilms tumor	Large mass of echogenicity slightly greater than the liver [9]	A large, well demarcated mass that has low attenuation as compared to adjacent renal parenchyma on enhanced scans [9]	Dark signal on T1weighted and bright signal on T2 weighted images. Inhomogeneity of the tumor is a common finding [9]
Malignant Rhabdoid tumor	The appearance is not universal and the masses may be identical in appearance to a typical Wilms tumor [9]	The appearance is not universal and the masses may be identical in appearance to a typical Wilms tumor [9]	The appearance is not universal and the masses may be identical in appearance to a typical Wilms tumor [9]
Multilocular cystic nephroma	Multiple cysts of varying size that do not communicate with each other or the renal pelvis.	Multiple cysts of varying size that do not communicate with each other or the renal pelvis.	Multiple cysts of varying size (low T1 and high T2) that do not communicate with each other or the renal pelvis.
Congenital Mesoblastic Nephroma	Solid mass that may show either a homogeneous or complex pattern [9]	Complex enhancing mass	Intermediate signal in T1WI and high signal in T2WI
Nephroblastomatosis (nephrogenic rests)	Generally hypoechoic [9]	Low attenuation on contrasted CT [9]	Intermediate signal in T1WI and high signal in T2WI

Table 2. Differential diagnosis table of clear cell sarcoma of the kidney

ABBREVIATIONS

CCSK = Clear cell sarcoma of the kidney
NWTSG = National Wilms Tumor Study Group
CT = Computerized tomography

KEYWORDS

Clear cell sarcoma; kidney; Wilms tumor