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

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 postoperatively, 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 NWTS 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 paraaortic 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 hydrenephrosis. In CCSK, the mass shows heterogeneous 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 (pericytomatosus) 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 dacitinomycin for 15 months, which has shown an improved relapse free survival rate in contrast to shorter durations [8].

REFERENCES


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

**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

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