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Large clear cell sarcoma of the kidney mistaken as Wilms' tumor

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ABSTRACT

Clear cell sarcoma of the kidney (CCSK) is a rare tumor. It has a wide histologic spectrum and often mimics other pediatric renal tumors, resulting in considerable diagnostic difficulty. We report the case of a two-year-old who presented with a large (14 cm) abdominal mass. Prior to neoadjuvant chemotherapy, a biopsy was performed, which revealed Wilms' tumor. The final pathology diagnosis at the time of resection revealed CCSK.

The differential diagnosis for an abdominal mass in a two-year-old child is broad and includes lesions of hepatic, renal, gastrointestinal, adrenal, and lymphatic origins. Of these, Wilms' tumor and neuroblastoma are the most common tumors [1], with Wilms' tumor representing 92% of renal masses in children [2]. Non-Wilms' renal tumors, rhabdoid tumors, and clear cell sarcoma of the kidney (CCSK) are uncommon. In fact, CCSK is particularly rare and represents only 17% of non Wilms' renal tumors in children with only 60 cases reported to the Surveillance Epidemiology and End Results (SEER) database between 1973 and 2005 [3]. In this report we present a child presenting with a huge renal mass consistent with Wilms' tumor on computed tomography (CT) and initial biopsy. However, the final pathologic diagnosis after resection revealed a CCSK.

1. Case report

A two-year-old boy presented with a one-year history of an enlarging abdominal mass. The patient was initially evaluated in Haiti and sent to the USNS Comfort Ship for further workup. CT at that time revealed a 14 cm heterogeneous mass arising from the right kidney, consistent with Wilms' tumor. He was referred to a non-governmental organization, Angel Mission, which arranged transfer to a large pediatric hospital in the United States for definitive care. No other pertinent medical or surgical history was reported at the time of transfer.

The patient presented to our institution with a very large abdominal mass that was associated with respiratory embarrassment, failure to thrive secondary to early satiety, and difficulty walking. There was no family history of malignancy. Physical exam revealed a malnourished, cachectic appearing child with a large palpable abdominal mass occupying the entire right upper, right lower and left lower quadrants. Marked superficial venous collateral circulation was noted on the anterior lower chest and upper mid-abdominal wall. He was hypertensive with a blood pressure of 125/86. Pertinent laboratory data included hemoglobin of 6.7 with a mean corpuscular volume of 58.1 and albumin of 2.6; he also was iron deficient. He was started on amlodipine, enalapril, and iron replacement therapy. Nasojejunal feeds were instituted to improve his nutritional status. CT of the chest, abdomen, and pelvis was performed and revealed a $17 \times 20 \times 17$ cm heterogeneous mass arising from the right kidney, consistent with Wilms' tumor (Fig. 1). The infrahepatic inferior vena cava (IVC) could not be visualized by CT or abdominal ultrasound. The intrahepatic vena cava, however, was patent by abdominal Doppler ultrasound. There was no evidence of metastatic diseases visualized.

Given the massive size of the tumor, the patient's poor nutritional status, and the possibility of tumor thrombus within the infrahepatic IVC the patient was taken to the operating room for open biopsy and

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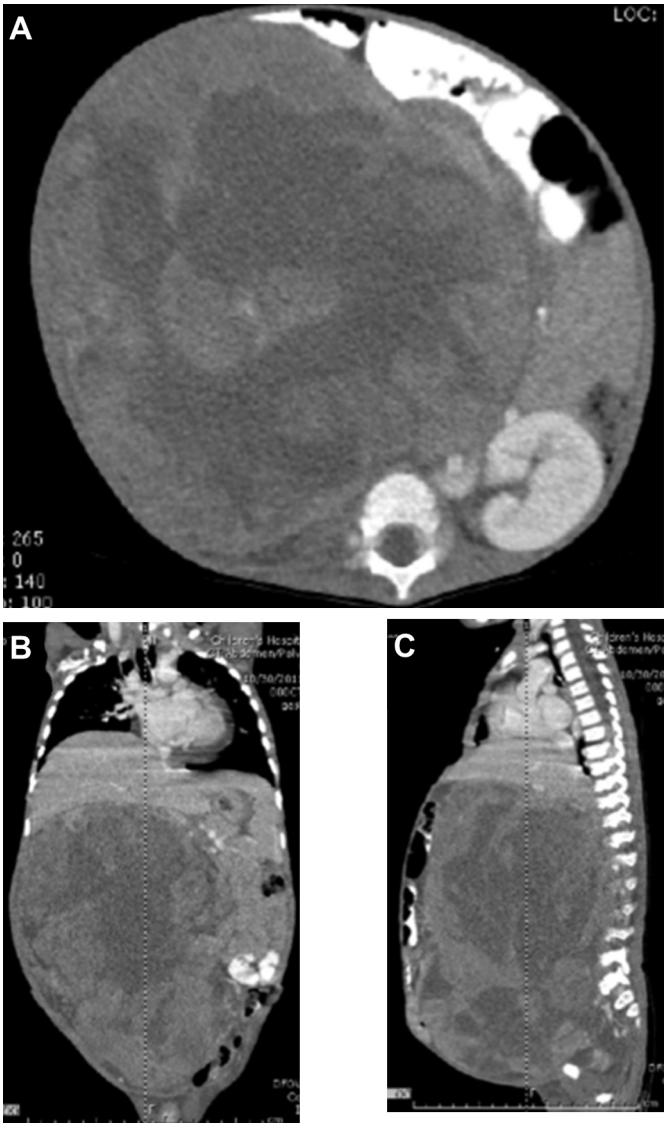

Fig. 1. Computed tomography of the abdomen demonstrating a large heterogeneous mass with mass effect, ascites, and possible invasion of the inferior vena cava. A, B, and C demonstrate displacement of the abdominal viscera in the axial, coronal and sagittal planes respectively.

port placement prior to neoadjuvant chemotherapy. Generous biopsies of the tumor were obtained and sent for pathology. Pathology revealed primitive tumor cells with hyperchromatic nuclei, small amount of cytoplasm and indistinctive cell borders consistent with a Wilms' tumor with favorable histology (Fig. 2A). FISH was negative for rearrangement of 22q12 EWSR1 and RT-PCR did not demonstrate translocations in EWS-FT1, EWS-EGR, PAX-FOX01, SYT-SSX, EWS-WT1, and ETV6-NTKR3. Further staining with vimentin, CD99 (Mic-2), CD56, and BCL-2 were positive. WT-1, cytokeratin, epithelial membrane antigen (EMA), desmin, S100, CD34, tyrosine hydroxylase, synaptophysin, and chromgranin A were negative.

The child underwent neoadjuvant chemotherapy consisting of vincristine, dactinomycin and adriamycin (regimen DD4A) for 6 weeks followed by repeat imaging. The CT revealed virtually no response to chemotherapy, and no new areas of progressive or metastatic disease. After multidisciplinary discussion it was felt that surgical resection was the best option for this child.

Exploratory laparotomy revealed a gigantic right-sided tumor with associated lymphadenopathy. Bilateral medial visceral rotation was performed to clearly identify tumor boundaries and define the vascular anatomy. The infrahepatic IVC was stretched over the tumor but patent. Great care was taken to preserve the left renal vein as well as the significantly enlarged left gonadal vein. A right radical nephrectomy was completed. The tumor measured $25.8 \times 19.2 \times 14.3$ cm and weighed 3.1 kg (Fig. 3). Multiple enlarged aorto-caval lymph nodes were resected and the ascending and descending colon were fixed to the abdominal wall. The child tolerated the procedure well and recovered uneventfully from his procedure without any complication.

The final pathologic diagnosis revealed CCSK as supported by the original immunohistochemical panel. A fine vascular pattern was appreciated on the gross specimen and review by a member of the Children's Oncology Group Renal Pathology Center confirmed the diagnosis (Fig. 2B). Focal penetration of the capsule and nine negative lymph nodes were identified. The patient was observed in the pediatric intensive care unit overnight, and transferred to the ward the following day. By postoperative day number 3 he was tolerating a regular diet. His superficial collateral venous pattern resolved within the first 24 h after surgery. Following diagnosis of clear cell sarcoma, a metastatic workup, including a bone scan and magnetic resonance imaging (MRI) of the brain, was performed and was negative for metastatic disease. He underwent and completed a course of radiation therapy to the abdomen as well as a course of adjuvant chemotherapy (regimen I), which consisted of alternating courses of vincristine, doxorubicin, and cyclophosphamide followed by the combination of cyclophosphamide and etoposide. No evidence of recurrent tumor has been noted 15 months from surgery.

2. Discussion

CCSK is a tumor that is frequently misdiagnosed due to its rarity and lack of specific features. Most patients present between two and three years of age [4] and show a 2:1 male to female predominance [5]. Clinical features are nonspecific making diagnosis of CCSK difficult. Workup should follow standard algorithms for an abdominal mass. CT and ultrasonography do not provide discriminatory findings that would differentiate between CCSK and Wilms' tumor [6]. Suspicion of CCSK should be heightened in children with evidence of bony metastases, which may develop in up to 60% of patients with CCSK compared to a 2% incidence of bony metastases in Wilms' tumor [4]. CCSK may also have a higher predilection to metastasize to liver, brain, and lung [7].

While not routine in the United States, preoperative percutaneous biopsy of suspected Wilms' tumors has been described and adopted by the United Kingdom Children's Cancer Study Group (UKCCSG). They recommend this approach for pathologic diagnosis prior to chemotherapy because they have found a high concordance between percutaneous biopsy and final surgical pathology in 94%–99% of cases [8]. Furthermore, use of pretreatment biopsy has not been associated with any increase in recurrence or complication [9].

Unfortunately, this patient fell into the small cohort of patients whose diagnosis on initial biopsy is discordant with the final pathologic diagnosis. CCSK is a malignant mesenchymal neoplasm characterized by undifferentiated cells with abundant extracellular matrix that are separated into cords and nests by a fine vascular network. Although the majority of CCSK have "classic" features on pathologic examination it is well known that CCSK is difficult to differentiate from other "renal blue round cell lesions" such as blastemal Wilms' tumor and primitive neuroectodermal tumor. The primary contribution of immunohistochemical analysis in the diagnosis of CCSK is the exclusion of other pediatric renal neoplasms, whereas immunohistochemical study has a limited role in the