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Case Report

Bilateral clear cell sarcoma of the kidney

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Abstract Clear cell sarcoma of the kidney (CCSK) accounts for 2–5% of all pediatric renal malignancies, and is known for its propensity to metastasize to bone and other sites. We are reporting two cases with bilateral CCSK that were diagnosed at our institution. One patient initially presented with bilateral renal masses, as well as pulmonary, hepatic and bone metastasis; while other present only with bilateral masses with no evident distant metastasis. Both patients received aggressive neo-adjuvant chemotherapy to decrease tumor size. One patient completed his designated treatment and initially showed complete remission (CR); eventually suffering from relapse. The other patient's tumor progressed during the course of chemotherapy. Both cases manifested brain dissemination at the time of relapse or progression. This emphasizes the importance of staging stratification in CCSK. This also illustrates CCSK's ability to metastasize to bone and other sites including the brain (a primary relapse site in our cases).

Introduction

Clear cell sarcoma of the kidney (CCSK) is an uncommon renal neoplasm of childhood [1]. It represents around 2–5% of pediatric renal tumors. Around 20 new cases of CCSK are diagnosed each year in North America [2,3]. It presents with bone metastasis in 40–60% of all cases [4]. The National

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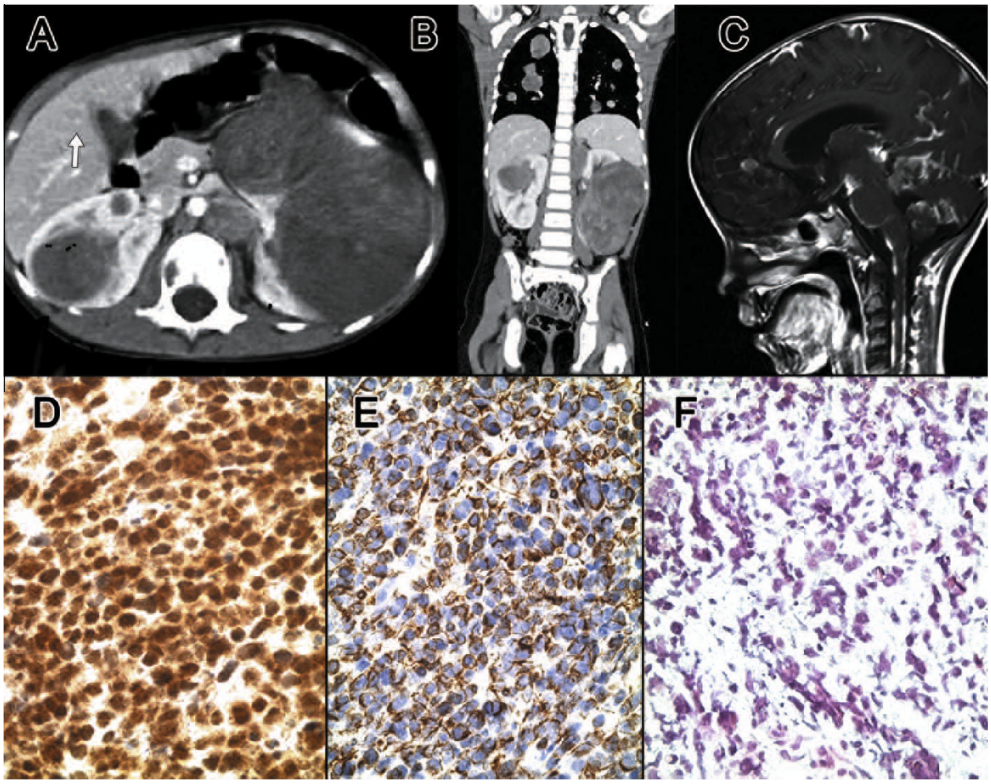

Figure 1 First patient. (A) Axial image showing a large lobulated left hypochondrial soft tissue mass; totally infiltrating the kidney with no residual parenchyma cells, and a right smaller soft tissue mass occupying the right kidney. (B) Coronal image of the same patient showing bilateral renal masses occupying both kidneys. (C) MRI revealing diffuse meningeal thickening and remarkable post contrast enhancement extending to the spinal meninges demonstrating diffuse metastases with diffuse brain edema. (D) Positive nuclear staining to INI-1. (E) Negative nuclear staining to WT-1. (F) H&E (20×) with anaplastic morphology.

Wilms' Tumor Study (NWTS) and the International Society of Pediatric Oncology (SIOP) indicated in 2004 that CCSK survival rates had increased to 87.5% and to 88% respectively [5]. Breslow and others revealed that intensive chemotherapy treatment including doxorubicin had increased overall survival of CCSK patients [6]. Staging based on the Children's Oncology Group (COG) qualifies stage I and stage II for upfront nephrectomy with lymph nodes sampling for pathological examination followed by a three-drug regimen. On the other hand, neoadjuvant chemotherapy is recommended for locally advanced stage III tumors along with assessment of the possibility of a postponed surgical excision; to be followed by adjuvant chemotherapy and flank irradiation. To our knowledge, the following two case reports represent the fourth and fifth reports of bilateral clear cell sarcoma of the kidney. Our aim is to report the clinical and pathological presentation and describe outcomes in both cases.

Case number 1

A 5.5 year old boy presented with jaw swelling, hematuria and abdominal swelling of 3 weeks duration and a progressive course. Computerized tomography (CT) scan of the abdomen revealed bilateral renal masses. The left kidney mass was huge ($11 \times 11.5 \times 9$ cm) and was lobulated, while the right kidney mass was smaller ($4.8 \times 4 \times 4.2$ cm). The CT also revealed para-aortic lymph node enlargement; the largest being about (4.5 cm) in diameter, a focal hepatic lesion in segment 7 measuring about (0.7 cm), and nodular pulmonary and bone metastatic lesions with no brain metastasis or inferior vena caval (IVC) thrombosis. Bone scan showed widespread multiple bony metastases. Initial bilateral renal tissue core biopsies revealed infiltration by malignant ovoid to spindle shaped cells having hyperchromatic nuclei and scanty cytoplasm. The cells showed evident mitosis and karyorrhexis, with loose stroma and focally myxoid wide areas of necrosis. Immunohistochemical stains revealed negative reactions to CD20, CD3, CD10, TdT, CP79a, LCA, CDS, synaptophysin, GD34, CD117, CD43, and WT-1, while staining for BCL-2 resulted in focally weak staining. There was a positive INI, myogenin and p53 with evidence of a high-grade mesenchymal sarcoma; possibly of the anaplastic subtype. The patient received 12 weeks of vincristine, doxorubicin, etoposide, carboplatin and cyclophosphamide. A regressive course of all masses ensued with collapse of involved C5, D1, and L1 vertebrae. After 12 weeks of treatment, the patient developed convulsions and deterioration of his conscious level, and was admitted to the ICU. Magnetic resonance imaging (MRI) of the brain revealed diffuse meningeal thickening and remarkable post contrast enhancement extending to the spinal meninges; demonstrating diffuse metastases with diffuse brain edema (see Fig. 1). The patient died after 5 months of initial diagnosis with progressive disease before undergoing any local control.