



Adult Clear Cell Sarcoma of the Kidney: Review of literature and a case report

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Abstract:

Introduction: Clear cell sarcoma of the kidney (CCSK) is a rare renal tumor which accounts for 3% of renal tumors. The tumor derives its name from the clear cytoplasm of the predominant cell type. Herein, we report a case of renal mass for which a radical nephrectomy was done and histologically proved to be CCSK. **Case Report:** A 34 year female presented with hematuria, dysuria, right flank pain and fever. Investigations revealed locally advanced right renal mass with right renal vein thrombosis. She underwent right radical nephrectomy followed by radiotherapy. Histopathology showed 10x11x11 cm of clear cell sarcoma tumor in upper and mid pole with multiple metastatic nodules in renal sinuses and in the lower pole. **Conclusion:** Clear cell sarcoma of the kidney is one of the rare renal tumor, we have added this case to the few cases published in the literature.

Key words: Hematuria, Kidney, Kidney Neoplasms, Nephrectomy, Sarcoma.

Introduction

Clear cell sarcoma of the kidney (CCSK) is a rare renal tumor which accounts for 3% of renal tumors. The tumor derives its name from the clear cytoplasm of the predominant cell type. CCSK is associated with bone and brain metastases. Patients with stage I had a 98% survival rate. We describe a case of clear cell sarcoma of kidney (NSCLC) who was treated by radical nephrectomy, and discuss treatments and outcomes.

Case Report

34 year female presented to our facility with recurrent attacks of hematuria and right flank

pain. Her past history was unremarkable. Clinical examination revealed fullness in right loin and palpable right kidney. Her hematological and biochemical tests were unremarkable. CT scan abdomen showed 12 x10x10 cm mass in the upper pole right kidney [Fig.1,2], involving the right adrenal and the retroperitoneum displacing the aorta and IVC. This was associated with multiple hilar, parahilar and intraaortocaval lymph nodes involvement. CT chest was unremarkable and isotope bone scan did not show any abnormal uptake in bones.

She underwent right radical nephrectomy and

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extensive lymph node dissection through midline incision. The operative finding was a huge right renal tumor which was mobile and not infiltrating the liver, with large paraaortic, interaortocaval and paracaval lymph nodes [Fig. 3,4,5].

Gross examination of the nephrectomy specimen [Fig.6] showed a large tumor measuring 10x11x11

cm located in upper and mid pole. Multiple metastatic nodules in renal sinuses and the lower pole were noted as well. The tumor invaded the renal capsule with gross tumor bulging from the capsule. There was non-contiguous extension of tumor into the ipsilateral adrenal gland. Renal vein margin and ureteric margin were positive for the tumor. Regional lymph nodes were involved.

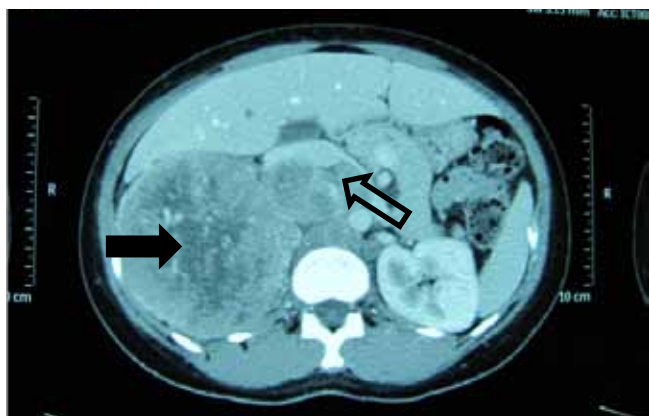


Fig.1: CT scan abdomen (cross section) showing the large right renal mass (filled arrow). Note the multiple hilar and para and intraaortocaval lymph nodes (empty arrow).

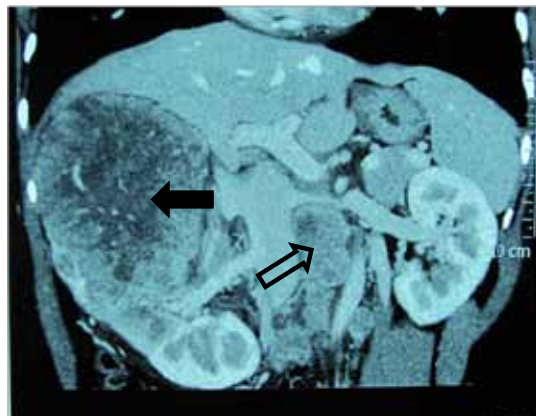


Fig.2: CT scan abdomen (coronal section) showing the large renal mass (filled arrow). Note the multiple hilar and para and intraaortocaval lymph nodes (empty arrow).

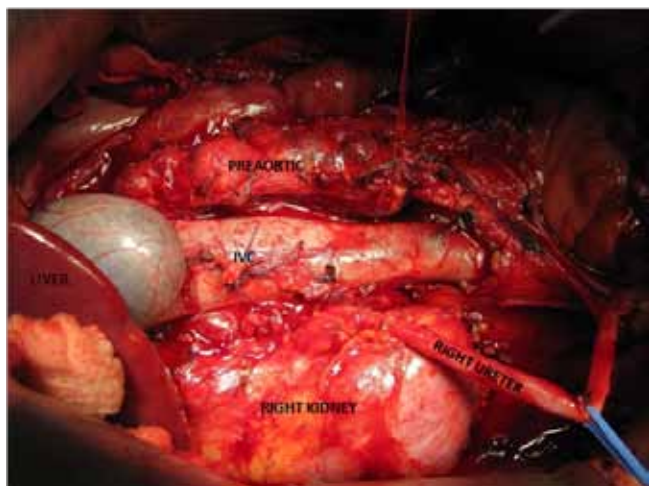


Fig.3: Intraoperative picture showing the large renal mass.



Fig.4: Intraoperative picture showing the large lymph nodes displacing the large vessels.

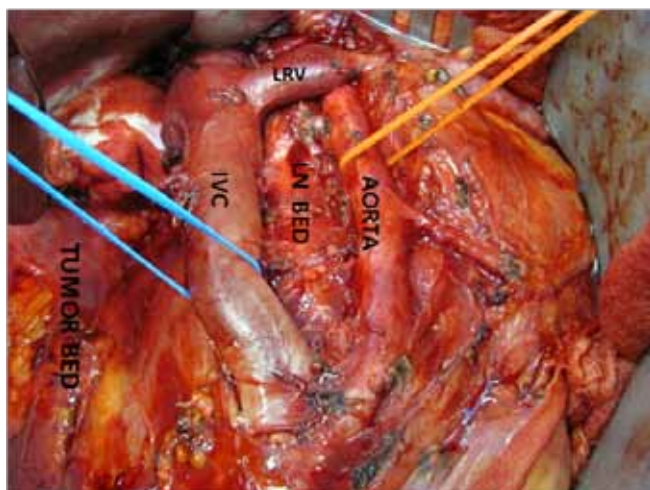


Fig.5: Intraoperative picture after tumor and lymph node beds.

Microscopic sections from the tumor revealed classic pattern of clear cell sarcoma of the kidney. The tumor cells were arranged in nests and separated by a fibrovascular stroma. The cells have fine nuclear chromatin, pale cytoplasm and indistinct cell borders. By immunohistochemistry, the tumor cells were diffusely positive for CD 10 and focally positive for vimentin, while negative for CKAE1/AE3, CK7, CK20, CEA, EMA, inhibin, melan A, hepar, synaptophysin, chromogranin and S-100.

Post-operative period was uneventful. The case was discussed in the tumor board and the decision was to give her radiotherapy followed by adjuvant chemotherapy. She received radiotherapy (50.4 Gy in 28 fractions using 3D conformal radiotherapy to the tumor bed, and completed within 2 months. Three months later, she started to complain of pain in the lower abdomen and in the left flank. CT scan was done and showed multiple hepatic cysts with suspicious metastasis, otherwise it was unremarkable. Case was discussed again in tumor board and decision was taken to proceed with 3 cycles of very aggressive chemotherapy using combined regimen of carboplatin, etoposide,

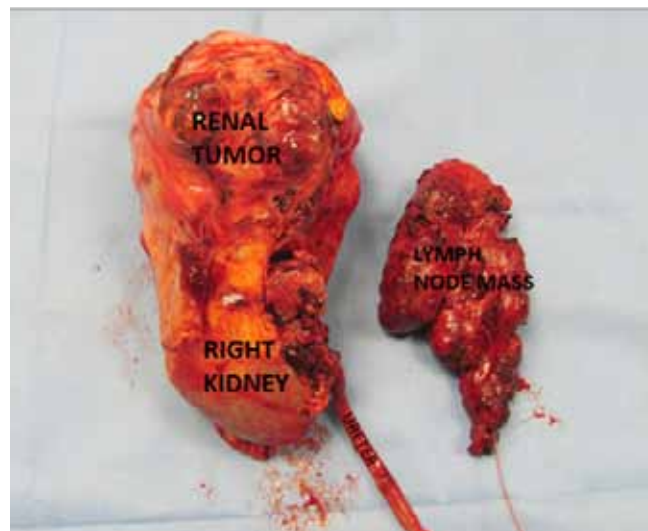


Fig.6: Macroscopic picture of the specimen.

cyclophosphamide, doxorubicin and vincristine.

Follow up CT scan after 2 months showed disease progression in abdomen, lungs and bones. Her condition deteriorated and she was sent back to her home country as a final stage, and she died one month later.

Discussion

Clear cell sarcoma of the kidney (CCSK) is a rare renal neoplasm of pediatrics, comprising about 3% of all pediatric renal tumors [1,2]. CCSK is distinct from Wilms tumor and also usually tends to metastasize to bones [1].

However, CCSK is extremely rare neoplasm in adults. Amin and colleagues reviewed around 12 cases of CCSK occurring in adult patients. They reported that clinical and pathologic features of CCSK did not differ significantly from pediatric patients [3]. They also noted that CCSK was negative for most of tumor markers except for vimentin and this finding was helpful to distinguish it from undifferentiated adult renal neoplasm including sarcomatoid renal cell carcinoma [3].

These tumors are usually unilateral and unicentric tumors arising from the medullary region of the kidney with mucoid texture, foci of necrosis, and cyst formations. It is called clear cell because of the presence of numerous intracytoplasmic vesicles. CCSK has a propensity to permeate through the renal and perirenal vascular system [5]. Most of these have a classic pattern either a predominant or a secondary morphology in 91% of cases [4]. Some published data suggest that clear cell sarcoma of the kidney in the young adult age group resembles its pediatric counterpart in ultrastructural and immunohistochemical characteristics, proclivity for skeletal and visceral metastasis, DNA diploid status with relatively low S-phase, and aggressive clinical course [3].

Management in children requires aggressive surgical approach followed by chemotherapy and radiotherapy as per National Wilms Tumor Study Group (NWTSG) protocols. Relapses, although late, are common even in stage one tumors. The overall survival is 69% [1]. While optimal treatment of adult patients with CCSK, still remains unclear. Surgery, radiotherapy and chemotherapy are combined or used separately. Benckroun *et al* reported a patient who underwent surgery followed by combination chemotherapy with cisplatin and doxorubicin and had no metastases 4 years later, but the metastases occurred within months in two patients who did not receive chemotherapy or radiotherapy after surgery [6]. In addition, Bhayani *et al* noted that a patient treated with surgery and combination chemotherapy (actinomycin, vincristine and doxorubicin) was disease-free at 12 months postoperatively [7]. In a report published by Rosso and colleagues, they treated a patient with surgery alone and stated that CCSK has a high resistance against radiation and chemotherapy [8].

In the case reported here, we report another rare adult type of CCSK. Our treatment plan reflects the non-standardized treatment plan for

the type of CCSK. The combination chemotherapy of carboplatin, etoposide, cyclophosphamide, doxorubicin and vincristine was adapted from a children protocol with some modification and substitution of cisplatin with carboplatin. However, with this very aggressive regimen and with the use of growth factors, we noted short durations of pancytopenia. Despite this aggressive combined chemotherapy regimen, disease progression was noted after 2 cycles and patient was terminally ill so comfort care was offered. As mentioned in Benckroun paper, this radio-chemotherapy did not change the aggressive prognosis of this tumor.

Conclusion

Clear cell sarcoma of the kidney in adult patients, although rare, must be differentiated from sarcomatoid carcinoma, sarcomas, and round cell tumors because of its unique characteristics in comparison to other renal neoplasms; thus, CCSK should be taken into consideration in the diagnosis of renal tumors. The malignant nature may relate not only to the biological features of these tumor cells, but also to the high resistance against radiation and chemotherapy. Being a rare malignant tumour with no standard treatment protocol, the management of such cases is still a subject of controversy and challenging.

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