



A Case of Clear Cell Sarcoma of Kidney in An Adult: An Extremely Rare Presentation

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ABSTRACT

Clear cell sarcoma of the kidney (CCSK) is a malignant renal tumor of childhood with a propensity to metastasize to bone and other organs. It is extremely rare in adults, must be differentiated from clear cell carcinoma of kidney which is more common in adults. We report a case of CCSK in a 45 year old female, confirmed by Immunohistochemistry (IHC) & managed with radical nephrectomy. The aim of this case report is to increase the awareness about this entity in adults with brief review of relevant literature.

Keywords: *Clear Cell Sarcoma, Immunohistochemistry, Kidney.*

Introduction

Clear cell sarcoma of kidney is a rare tumor (3-5% of childhood renal tumor), its mean age of diagnosis is 36 months with a range of 2 months -14 years [1]. It has propensity to metastasize to the bone & has unfavorable prognosis. Reports of these tumor diagnosed in adults is rare [2-5] & its treatment is controversial with combination of surgery & chemo radiotherapy. It is necessary to differentiate it from clear cell renal cell carcinoma which is most common in adults [2]. Immunohistochemistry is needed for confirmation of diagnosis, as this entity is not common in this age.

Case Report

A 47 year female presented with complaints of Left sided flank pain, fullness in abdomen & hematuria since 1 month. USG & CT scan with I.V. contrast revealed huge well defined heterogeneously enhancing mass lesion involving upper-mid poles of Left kidney with localized exophytic component with mass effect and without renal vein thrombosis. Radical nephrectomy was done under aseptic precaution in GA with Transabdominal approach. The patient was discharged post operative 5 days without complication.

Gross examination shows Left sided radical nephrectomy measuring 13x8x8 cm in size. Cut section shows grey-brown solid-cystic mass at upper-mid pole of kidney measuring 8x7x6 cm in size. Extensive hemorrhage & necrosis are seen [Figure 1].

Histo-pathological examination revealed diffuse monotonous proliferation of medium sized cells in nests

separated by thin fibro vascular septa. The cells show fine chromatin with indistinct cell border & clear cytoplasm [Figure 2]. Capsule, perinephric fat, fascia gerota, margin of vascular pedicle, renal sinus, ureter & adrenal gland were free from the tumor.

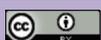
On Immunohistochemistry tumor cell expressed positivity for vimentin, bcl-2 & cyclin D1 which favors CCSK [Figure-3] and were immunonegative for Cytokeratin, EMA, Pax8 [Figure 4], Mic-2, Synaptophysin, Chromogranin A, Inhibin, Melan-A, CD-10, PgR, Desmin, SMA & c-kit (to rule out clear cell carcinoma of kidney, rhabdoid tumor, neuroendocrine tumor, mesoblastic nephroma).

Finally on the basis of histomorphology and Immunohistochemistry diagnosis of clear cell sarcoma was given. Patient was referred to onco-physician and chemo-radiotherapy was started.

Discussion

Clear cell sarcoma of kidney was first described in 1978 by three distinct groups: Beckwith and Palmer [6], Morgan and Kidd [7], and Marsden and coworkers [8] & it was initially named "bone metastasizing renal tumor of childhood" since it has predilection for skeletal metastasis. It is most frequently misdiagnosed renal tumor in children and it is rarely diagnosed in adults [2-3]. Males are slightly more affected (2:1).

Morphologically various patterns of clear cell sarcoma are noted with classical pattern of nests & cords separated by thin fibro vascular septae which is focally present in every case. The other patterns are myxoid (50%), sclerosing



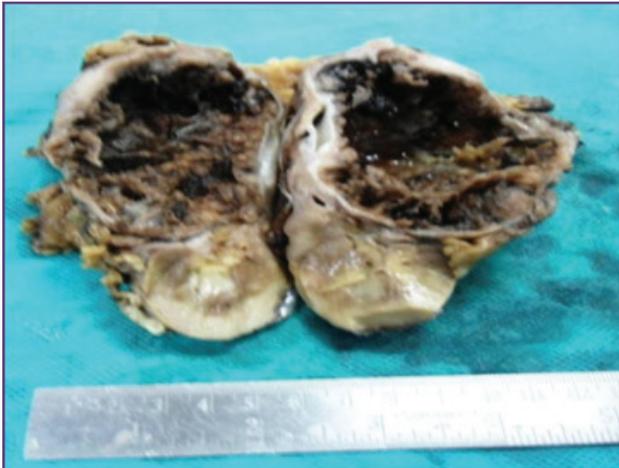


Fig. 1: Gross appearance of clear cell sarcoma of left kidney shows grey-brown solid-cystic mass at upper-mid pole of kidney measuring 8x7x6 cm in size with extensive hemorrhage & necrosis.

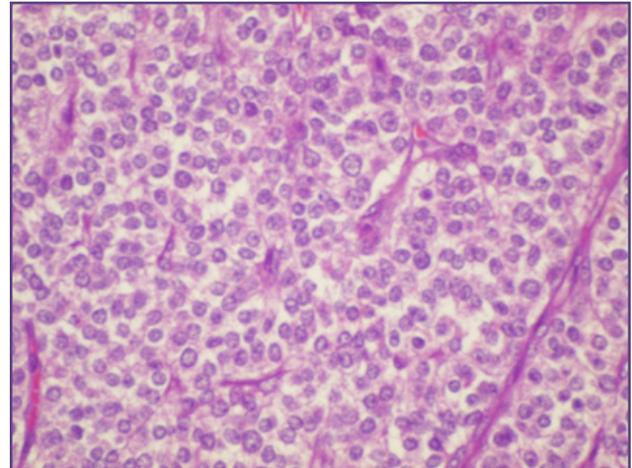


Fig. 2: H&E stain 40x, Clear cell sarcoma of kidney. Tumor cells showing clear cytoplasm and centrally placed nuclei arranged in nesting pattern separated by fibro-vascular stroma.

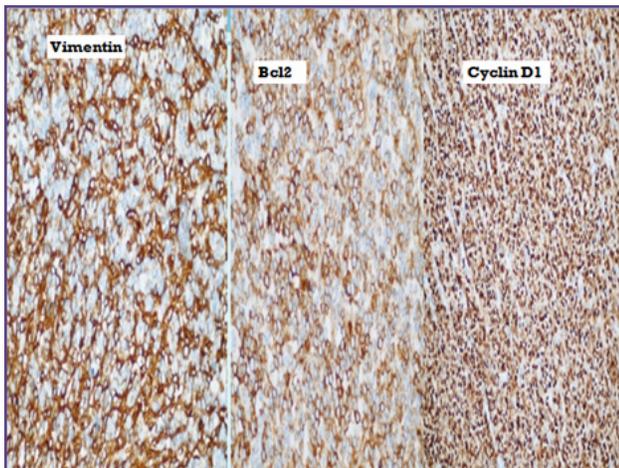


Fig. 3: (IHC stain- 20X) Clear cell sarcoma of kidney vimentin, cyclin D1 & BCL2 positive in tumor cells.

(35%), cellular (26%), epitheloid (13%), palisading verocay body (11%), spindle cell (7%), storiform (4%) and anaplastic (2.6%)^[1]. The nuclei of the CCSK are uniform with smooth chromatin and scant cytoplasm. On IHC tumor cells are consistently positive for vimentin & cyclin D1 with CD34 positivity in thin walled capillaries^[1,9]. Tumor cells are negative for Cytokeratin, EMA, WT1, CD99, Desmin and Synaptophysin.

Bone metastasis occurs in 17-40% of cases of CCSK tumor with the common site being of brain, regional lymph nodes, lungs, liver and soft tissue^[10].

Molecular genetics of CCSK is not fully elucidated. Internal tandem duplications in the BCOR gene (BCL6 co repressor) were identified in 100% (20 / 20) of CCSK of

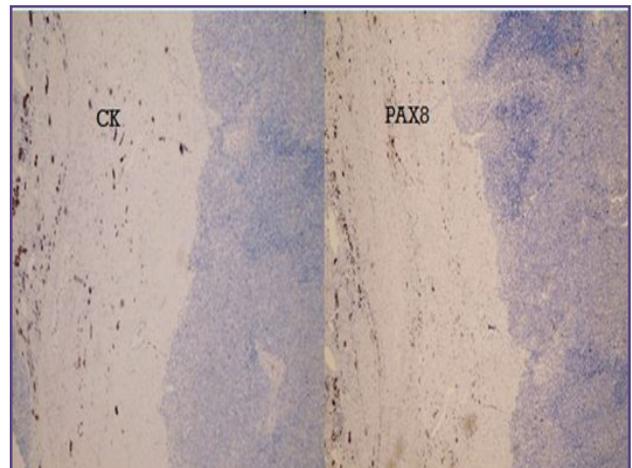


Fig. 4: (IHC stain 20x) Clear cell sarcoma of kidney CK & Pax8 negative in tumor cells.

pediatric renal tumors^[11] & 85% in another study done by Roy et al^[12].

Advanced clear cell renal cell carcinoma is highly resistant to cytotoxic chemotherapy drugs. Thus differentiation of CCSK from clear cell renal cell carcinoma of kidney and undifferentiated adult renal neoplasm including sarcomatoid renal cell carcinoma is especially important in adult patients.^[2] Prognosis of CCSK depends upon stage, age at diagnosis & tumor necrosis. Stage IV & young age are adverse prognostic factors for event free survival.

Total Radical nephrectomy followed by postoperative chemotherapy and flank radiation therapy is the treatment of choice of CCSK. The tumor is highly responsive to combination treatment with doxorubicin, vincristine and

actinomycin. The role of doxorubicin in therapy emphasizes the need for pathologists to accurately identify CCSK [1].

This case report highlights the importance in making accurate diagnosis in adult patients with CCSK suspected clear cell renal cell carcinoma. In particular, appropriate selection of IHC ensures timely diagnosis and improves patient management.

Conclusion

We concluded that clear cell sarcoma of kidney in adult is rare. Unfamiliar morphological patterns can cause difficulty in diagnosis. Awareness about its morphological patterns and diligent use of Immunohistochemistry will help in their identification. This is important as most of them can be cured and have good survival rate by adequate excision followed by chemo-radiotherapy. Its differentiation with clear cell renal cell carcinoma is necessary which is highly resistant to traditional cytotoxic chemotherapy.

Conflict of Interest

The authors declare that there is no conflict of interests regarding the publication of this paper.

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