

Case Report

Clear cell sarcoma of kidney in an adult: an extremely rare case

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ABSTRACT

Clear cell sarcoma of kidney is extremely rare in adults. It is a very malignant tumor, with a high tendency for relapse and a propensity for skeletal metastases, particularly skull. Treatment of clear cell sarcoma of kidney generally involves surgical intervention coupled with radiation and chemotherapy. This paper reports a case of adult clear cell sarcoma of kidney.

Keywords: Clear cell sarcoma, Kidney, Renal tumors

INTRODUCTION

Clear Cell Sarcoma, also known as bone metastasizing renal tumor, is a distinctive renal malignancy formerly regarded as a morphologic variant of Wilms tumor.¹ It comprises about 5% of childhood renal tumors; Its incidence peaks during the second year of life; However, However, Adnani et al. reported the case of a 58-year-old man² and Benchekroun et al. reported the case of a 65-year old female, who is the oldest patient with clear cell sarcoma of the kidney ever published in the literature.³

CASE REPORT

A female patient aged 38 years presented with complaints of left loin pain and hematuria to the department of urology, Gandhi hospital, Secunderabad. CT scan plain and contrast (Figure 1) of Whole abdomen revealed irregularly lobulated heterogeneously enhancing mass lesion measuring 5.3 x 4.8 cm in maximum dimension and 5 cm craniocaudally in central region of left kidney with necrotic areas suggestive of neoplastic lesion. No lymph node enlargement noticed. Right kidney appears normal. Under strict aseptic conditions, under general anesthesia left radical nephrectomy was done through

11th rib cutting incision. Specimen sent for histopathological examination. The patient was discharged from the hospital without post-operative complications. Grossly 5 cm diameter yellowish with grey white tinge tumor mass bulging from the surface in the central region of left kidney (Figure 2).



Figure 1: CT scan of Whole abdomen showing irregularly lobulated heterogeneously enhancing mass lesion in central region of left kidney.



Figure 2: Gross appearance of clear cell sarcoma of left kidney. 5 cm diameter yellowish to gray white bulging mass with areas of necrosis in the central region of the kidney.

Histopathological examination revealed classic pattern of clear cell sarcoma of the kidney composed of round to polygonal cells (Figure 3) with indistinct cell borders, abundant clear cytoplasm and vesicular to speckled nuclear chromatin forming nests separated by fibrovascular stroma (Figure 4). Immunohistochemistry with vimentin (Figure 4 inset), BCL₂ were positive (Figure 5) and with EMA negative (Figure 6).

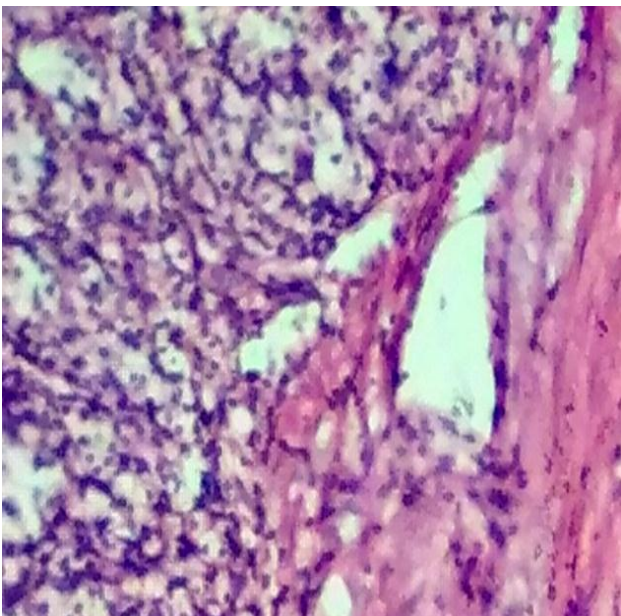


Figure 3: H&E x10, Tumor tissue with clear cells.

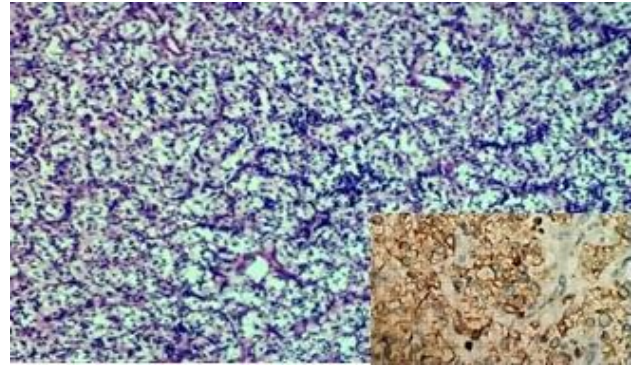


Figure 4: H&E x40, Clear cell sarcoma of kidney. Tumor cells showing clear cytoplasm and centrally placed nuclei arranged in nesting pattern separated by fibrovascular stroma. Inset: IHC marker vimentin positive.

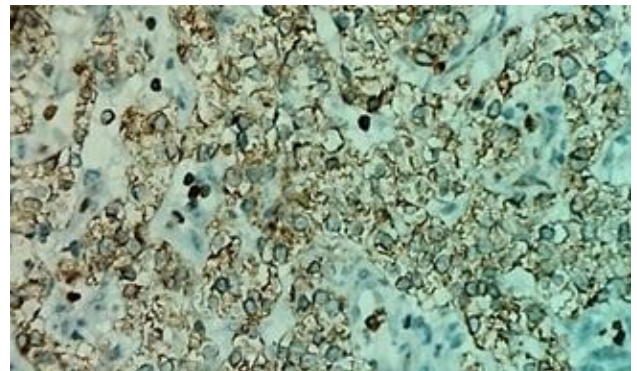


Figure 5: Clear cell sarcoma of kidney IHC marker BCL₂ positive.



Figure 6: Clear cell sarcoma of kidney IHC marker EMA negative in tumor cells.

DISCUSSION

Clear cell sarcoma of kidney is extremely rare in young adults. Clear cell sarcoma is a very malignant tumor with a high tendency for relapse and a propensity for skeletal metastases particularly skull.⁴ Metastases also occur to regional lymphnodes, brain, lung, and liver. These metastases tend to develop after long intervals following the removal of the primary tumor (5 years or more).

Treatment of clear cell sarcoma of kidney generally involves surgical intervention coupled with radiation and chemotherapy with cyclophosphamide, etoposide, vincristine and doxorubicin for 24 weeks. Optimal treatment for adult patients with Clear cell sarcoma of kidney still remains unclear. Surgery, Radiotherapy and chemotherapy are combined or used separately. Advanced renal cell carcinoma is highly resistant to cytotoxic chemotherapy drugs. Thus its differentiation from clear cell renal cell carcinoma and undifferentiated adult renal neoplasm including sarcomatoid renal cell carcinoma is especially important in adult patients.⁵ Nevertheless, there are no tumor specific markers for clear cell sarcoma of kidney, which makes the diagnosis difficult.⁶

However, Argani et al.⁷ reported that vimentin is readily demonstrable in nearly all specimens and BCL₂ is demonstrable in some but other markers EMA, CK, S-100, desmin, CD34 and CD99 are consistently negative. Our case showed vimentin and BCL₂ positive and EMA negative.

CONCLUSION

Clear cell sarcoma of kidney is rare in adults. We think that accurate diagnosis of Clear cell sarcoma of kidney and its differentiation from clear cell renal cell carcinoma and undifferentiated adult renal neoplasm including sarcomatoid renal cell carcinoma is especially important in adult patients.

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