University Journal of Pre and Para Clinical Sciences

ISSN 2455– 2879 Volume 2 Issue 3 2016

Adult clear cell sarcoma of kidney A case report

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Abstract :
Clear cell sarcoma of the kidney is an uncommon pediatric renal neoplasm which comprises approximately 4 Percent. Incidence peaks during second year of life and is extremely rare in adults. A 29 year old female came with a 4 month history of intermittent dragging pain over the right hypochondrial region. Abdominal pelvic ultrasonogram showed a heterogeneous mass originating from the mid region of left kidney. Right radical nephrectomy was performed. The histopathological diagnosis of the mass was Clear cell sarcoma of the kidney. This case is presented for its rarity of age group.

Keyword : Clear cell sarcoma of kidney, pediatric renal neoplasm, adult, Bone metastasizing renal tumour.

Introduction:

Clear cell sarcoma of kidney, originally called bone metastasizing renal tumour of childhood, is a highly malignant tumour of uncertain origin [1]. Clear cell sarcoma of kidney is usually diagnosed between 12 to 36 months of age and very rarely in adults. Identification of clear cell sarcoma of kidney in adults is very important because of its rarity and histomorphological resemblance with other variants of renal cell carcinoma. We report a case of a 29 year old female with clear cell sarcoma of kidney who presented with right hypochondrial pain.

Case report::

A 29 year old female was admitted to the department of urology with complaints of intermittent dragging pain over the right hypochondrial region for 4 months duration. Ultrasonography of abdomen and pelvis showed a heterogenous mass originating from the mid region of left kidney and there was no evidence of any metastasis. A right radical nephrectomy was performed. Specimen was sent for histopathological examination.

Pathological findings: Gross examination:

External surface of the kidney was nodular , about 10 x 6.5 x 5 cm in size. Cut surface showed a homogenous grayish white with focal brownish coloured vaguely circumscribed mass of size measuring about 5 x 4cm centered in the medullary region of the kidney. The mass was friable in the centre and soft to firm at the periphery (Figure1).

Microscopic examination: Microscopic examination showed a tumour composed of diffuse and solid sheets of monomorphous population of polygonal clear cells having pale cytoplasm with indistinct cell borders and uniform oval to round nuclei with fine granular chromatin. The tumour nests were separated by few bands of fibrosis with arborizing vessels (Figure 2,3). The tumour nest showed a pushing margin into the surrounding renal parenchyma with focal areas of necrosis. The tumour was confined to the renal parenchyma. Renal vein was free of tumour. Hence a histopathological diagnosis of stage I clear cell sarcoma of kidney was made. Further sections were stained with immunohistochemical markers cytokeratin, vimentin and epithelial membrane antigen. Immunostained sections showed positivity for vimentin (Figure 4) while stain for cytokeratin (figure 5) and epithelial membrane antigen (figure 6) were negative. The Immunohistochemical markers were supportive of the histopathological diagnosis of Renal clear cell sarcoma.

Discussion:

Clear cell sarcoma of kidney is a highly malignant tumor of childhood and is extremely rare in adults[2]. Male : female ratio is 1.7:1 with a male predominance[3]. The origin of this neoplasm remains uncertain and it may be histogenetically related to wilms tumour[4]. Clinical and pathological features of Clear cell sarcoma of kidney do not differ significantly between adults and pediatric patients[2,5].

Clear cell sarcoma of kidney exhibits nine histological patterns namely classical, myxoid, sclerosing, cellular, epithelioid, palisading, spindle, storiform and anaplastic[1]. Our case presented with classical histological pattern of monomorphic cells arranged in sheets supplied with a distinctive branching array of small blood vessels.

Immunohistochemically , Clear cell sarcoma of kidney are consistently positive for vimentin. Other markers are consistently negative. These include stain for epithelial markers (cytokeratins, epithelial membrane antigen),neural markers (**S-100** protein), neuroendocrine markers (chromogranin, synaptophysin) and muscle markers (desmin, myoglobin)[6].

Clear cell sarcoma of kidney is often confused with Wilms tumour in children. Clear cell sarcoma of kidney has to be differentiated from Wilms tumour by it's larger size ,areas of necrosis, myxoid changes and cyst formation. Clear cell sarcoma of kidney should also be differentiated from clear cell renal carcinoma and undifferentiated adult renal neoplasm including sarcomatoid renal cell carcinoma [2].The role of immunohistochemical markers cannot be underestimated in such cases. While Wilms tumour is positive for cytokeratin and clear cell carcinoma kidney is positive for epithelial membrane antigen, both are negative for vimentin [2,5].The present case was positive for vimentin and negative for cytokeratin and epithelial membrane antigen.The prognosis of clear cell sarcoma of kidney - stage I(5 year survival rate is 97%) is better than that of clear cell carcinoma kidney - stage I (5 year survival rate is 77.8%) and Wilms tumour- stage I (5 year survival rate is 90%)[7].

However Untreated clear cell sarcoma of kidney is a highly malignant tumour with high tendency for relapse and a propensity for metastasis to the skeletal system especially skull, regional lymph nodes, brain, lung and liver[6].

The occurrence of clear cell sarcoma of kidney in a non-pediatric age group is extremely rare. A thorough search of the available literature shows only very few documented cases. The present case showed no evidence of metastasis at the time of nephrectomy. A simple panel of immunomarkers are often helpful in supportive of a H/E diagnosis. Awareness of the existence of clear cell sarcoma of kidney in adults helps in the diagnosis and modifies the treatment regimens and prognosis.

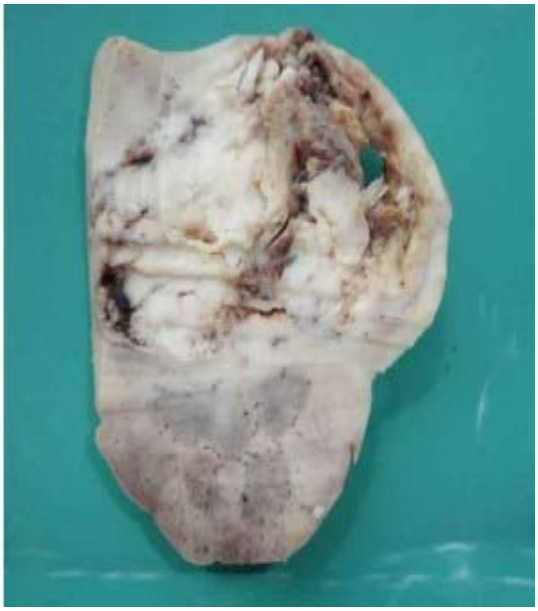


Figure 1: cut surface of kidney shows homogenous grayish white with focal brownish coloured vaguely circumscribed mass centered in the medullary region

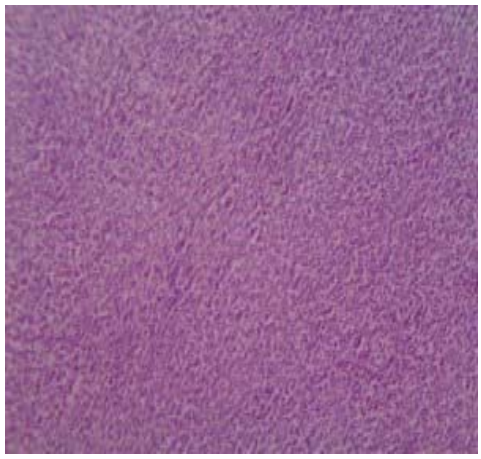
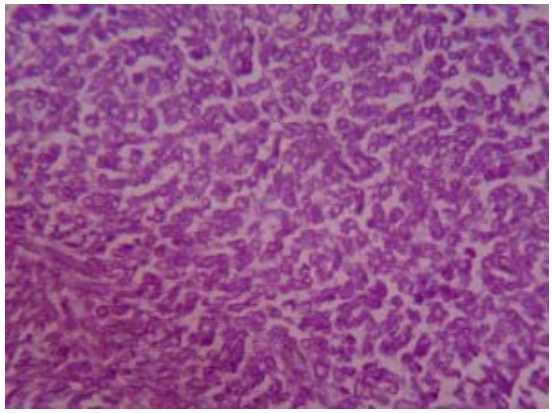


Figure 2: photomicrograph of tumour with classic pattern of polygonal cells with pale cytoplasm and round to oval nuclei (H&E 10x)



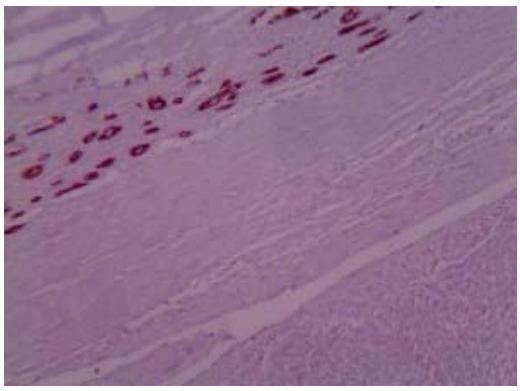
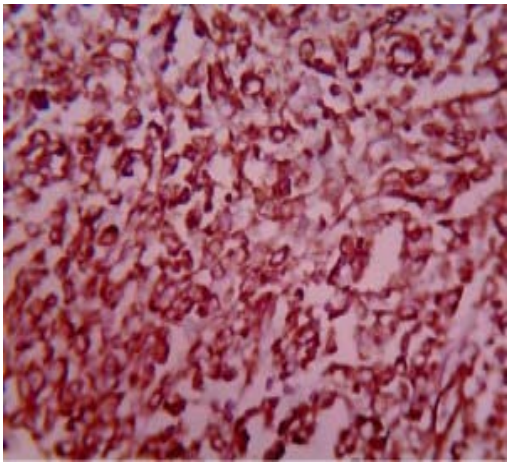


Figure 3: photomicrograph of tumour cells with clear cytoplasm and round to oval nuclei and inconspicuous nucleoli (H&E 40 x)

Figure 5: Photomicrograph shows tumour cells negative for cytokeratin while surrounding normal Renal tubules shows positivity for cytokeratin(10X)

Figure 4: photomicrograph shows tumour cells positive for vimentin (40x)



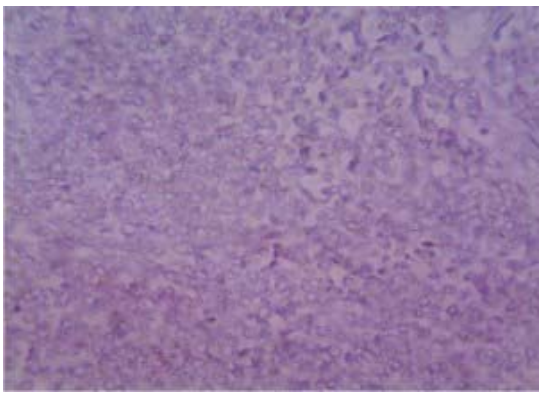


Figure 6: Photomicrograph shows tumour cells negative for epithelial membrane antigen (40x)

References:

1. Argani P, Perlman EJ, Breslow NE, Browning NG, Green DM, D'Angio GJ, Beckwith JB: Clear cell sarcoma of the kidney: a review of 351 cases from the National Wilms Tumor Study Group Pathology Center**.** Am J Surg Pathol 2000, **24:**4-18.

2 Ali R Kural, Bulent Onal, Hamdi Ozkara, Cansel Cakarir, Inci Ayan and Fulya Y Agaoglu Adult clear cell sarcoma of the kidney: A case report BMC Urology2006,6:11.

3 Joong seok seo, MD,Yong wook park MD,Kye yong song MD,Seon taek chang MD. Clear cell sarcoma of kidney –a case report.Journal of Korean medical science vol 4,No 3, P149-153 sep 1989.

4 Smith MA,Seibel NL,Altekruse SF,et al:Outcomes for children and adolescsnts with cancer: challenges for the twenty first century.J clin oncoll 28 (15):2625-34,2010.

5 Amin MB, de Peralta-Venturina MN, Ro JY, El-Naggar A, Mackay B, Ordonez N, Mani A, Ayala A: Clear cell sarcoma of kidney in an adolescent and in young adults. A report of four cases with ultrastructural, immunohistochemical and DNA flow cytometric analysis. Am J Surg Pathol 1999, 23:1455-1463.

6 Hideaki Oda, M.D., Junji Shiga, M.D., and Rikuo Machinami, M.D. Clear cell sarcoma of kidney two cases in adults oct 19,1992;71 (7):2286-2291.

7 Schmidt D,Harms D,Evers KG,Bliesener JA,Beckwith JB,Bone metastasizing renal tumour (clear cell sarcoma) of childhood with epithelioid elements.Cancer 1985,56:609-613.