HISTOPATHOLOGICAL AND IMMUNOHISTOCHEMISTRY BASED DIAGNOSIS OF CLEAR CELL SARCOMA IN A 5 MONTH OLD MALE CHILD: A CASE REPORT

M. Ramani¹, O. H. Radhka Krishna², K. Ramesh Reddy³, Ramsha Tahoor A⁴, C. Sandhya Rani⁵

1. Professor, Department of Pathology, Niloufer hospital.
2. Assistant professor, Department of Pathology, Niloufer hospital
3. Professor and HOD, Department of Paediatric Surgery, Niloufer Hospital.
4. IIIrd year under graduate, Osmania Medical College.
5. Post Graduate, Department of Pathology, Niloufer hospital

CORRESPONDING AUTHOR:
Dr. M. Ramani,
Niloufer Hospital for women and child Health, Red hills Hyderabad Andhra Pradesh.
E-mail: drmramani@sify.com

ABSTRACT: Clear cell sarcoma of the kidney is a rare paediatric malignant tumor having a high metastatic potential with predilection to involve the bone. It accounts for 4% of all primary childhood renal tumors. It represents one of the most common unfavourable tumors included in National Wilms’ Tumor Study Group clinical protocols. We here report a case of a 5 month old male child having this rare tumor. This case is presented because of the rarity of this tumor.

KEY WORDS: Clear cell sarcoma, bone metastatic renal tumor of childhood, 5 month male child.

INTRODUCTION: Clear cell sarcoma of the kidney also described as “Bone-metastasizing renal tumor of childhood” by Marsden et al. [1], is a rare paediatric malignant tumor. It comprises 4% of all primary childhood renal tumors [2]. Its incidence is between 1 and 4 years of age with a male: female ratio of 2:1[3]. It is the most frequently misdiagnosed paediatric renal tumor because of its infrequency, morphological diversity, and lack of specific diagnostic markers [4]. This rare tumor, owing to its aggressive nature, risk of metastasis to the bone and poor response to conventional therapy needs to be diagnosed early and correctly.

CASE REPORT: A 5 year old male child presented with history of a mass in the right upper abdomen since birth. Abdominal examination showed a firm, non-tender swelling occupying the right hypochondrium and lumbar region.

Plain and CECT examination of the abdomen showed a very large, solid, heterogenous tumor, arising exophytically from the middle and lower polar region of the right kidney measuring 11X9.5X8 cm with focal areas of necrosis and moderate enhancement on post contrast examination which was suggestive of Wilms’ tumor. Right kidney showed upper polar hydronephrosis. Retroperitoneal, prevertebral, paraaortic and retrocaval lymphadenopathy was observed.

With the provisional diagnosis of Wilms’ tumor, right Nephrectomy was done. We received a right renal mass measuring 13X10X7 cm. The surface was nodular and was irregular at the hilar area. The ureter was not identified. On cut section, rim of normal kidney was identified at the upper pole and the lateral margins. Tumor measured 11 X 8 X 7 cm and was grey white with areas of
haemorrhage, necrosis and few cystic and yellowish areas (Figure 1). The capsule was easily peelable.

On microscopy, multiple sections studied showed normal kidney architecture along with tumor. The tumor tissue, showed cells arranged in the form of solid sheets, cords, nests, and palisading pattern. The cells were polygonal with eosinophilic, vacuolated cytoplasm, with indistinct cell borders, finely granular chromatin, and inconspicuous nucleoli (Figure 2). Mitotic activity was not marked. Focal areas showed clear cells; extensive haemorrhage and necrosis. Sections from tumor kidney junction showed extension of tumor cells into the surrounding normal kidney, entrapping the nephrons.

Based on the above findings a differential diagnosis of Clear cell sarcoma of the kidney/monophasic Wilms’ tumor /small round cell tumor of kidney i.e -lymphoma was made.

IHC: IHC of the tumor showed strong positivity for vimentin (Figure 3) and was negative for CD34 and LCA.

In view of the above histological and immunohistochemical findings, diagnosis of Clear cell sarcoma stage I of kidney was confirmed. The patient had an uneventful post operative recovery.

DISCUSSION: Clear cell sarcoma of the kidney also described as “Bone-metastasizing renal tumor of childhood” by Marsden et al. is a rare paediatric malignant tumor. It comprises 4% of all primary childhood renal tumors. Its incidence is between 1 and 4 years of age with a male: female ratio of 2:1. It is a unilateral tumor 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 and has not shown any association with congenital anomalies. Most of the cases have a classic pattern, (91%) as a predominant or a secondary morphology. In the classic pattern, the tumor cells appear monomorphic with cords or nests of 6-10 cells separated by small, regularly spaced arborizing fibro vascular septa. While usually plump, the cells in the core uncommonly may assume a spindle shape. Nuclei are uniform in shape, with fine dusty chromatin without prominent nucleoli or coarse condensations. Empty appearing Orphan Annie eye nuclei are a frequent occurrence. Cytoplasm is sparse and borders indistinct. Sometimes the classical pattern can blend with variant patterns.

The histological variants recognised have been described in Table 1. Patients with clear cell sarcoma tumors without areas of necrosis have a more favourable prognosis. Twenty-nine percent of patients with clear cell sarcoma have lymph node metastases at the time of diagnosis, and bone metastasis is the most common form of relapse. Immunohistochemical staining shows reactivity for Vimentin in the form of perinuclear dot like pattern.

STAGING OF CLEAR CELL SARCOMA OF KIDNEY: Like other renal tumors of childhood, Clear cell sarcoma of kidney is staged by the National Wilms’ Tumor Study staging scheme, described in Table 2.

CONCLUSION: Clear cell sarcoma of the kidney is a rare paediatric malignant tumor. It is the most frequently misdiagnosed paediatric renal tumour. The current treatment for it is wide local tumour excision, with adjuvant radiation therapy for resection margins of less than 1 mm. It responds poorly to conventional chemotherapy and addition of doxorubicin to the treatment regimen has improved the overall outcome. Prognosis of Clear cell sarcoma of kidney is reported to be poor due to the great propensity to metastasise regionally and distantly. Relapses, although late, are common even in stage
one tumors. The overall survival is 69% [3]. It is of considerable therapeutic importance that clear cell sarcoma of kidney should not only correctly diagnosed but early too. This case has been presented in view of highlighting the salient features of clear cell sarcoma which is a very rare paediatric renal tumor.

REFERENCES:

TABLES AND FIGURES

Table 1: Histological variants of Clear cell sarcoma of kidney

| 1) Myxoid | 50% cases |
| 2) Sclerosing | 35% cases |
| 3) Cellular | 26% cases |
| 4) Epitheloid (trabecular/ acinar type) | 13% cases |
| 5) Palisading verocay body | 11% cases |
| 6) Spindle cell | 7% cases |
| 7) Storiform | 4% cases |
| 8) Anaplastic | 2.6% cases |
Table 2: Staging of Clear cell sarcoma of kidney

<table>
<thead>
<tr>
<th>Stage</th>
<th>Description</th>
<th>Percentage of Cases</th>
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<tbody>
<tr>
<td>Stage I</td>
<td>Tumor confined to the kidney and completely resected. Renal capsule or sinus vessels not involved.</td>
<td>25%</td>
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<tr>
<td>Stage II</td>
<td>Tumor extends beyond the kidney but is completely resected with negative margins. Any one of the following has occurred: (a) penetration of renal capsule (b) invasion of renal sinus vessels.</td>
<td>37%</td>
</tr>
<tr>
<td>Stage III</td>
<td>1 or more of the following criteria must be met: (a) Unresectable primary tumor. (b) Lymph node metastasis. (c) Positive surgical margins. (d) Tumor spillage involving peritoneal surfaces either before or during surgery, or transacted tumor thrombus.</td>
<td>34%</td>
</tr>
<tr>
<td>Stage IV</td>
<td>Metastasis outside the abdomen</td>
<td>4%</td>
</tr>
<tr>
<td>Stage V</td>
<td>Bilateral renal tumor</td>
<td>Not yet reported</td>
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Fig 1: Cut section of the tumor showing variegated appearance, normal kidney identified at the upper pole (arrow)

Fig 2: H&E (40x) showing tumor tissue consisting of polygonal cells with clear/vacuolated cytoplasm

Fig 3: Immunohistochemistry (40x) showing strong Vimentin positivity