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Pathology

CLEAR CELL SARCOMA OF KIDNEY – A RARE CASE REPORT

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ABSTRACT Clear cell sarcoma of kidney is a rare malignant tumor mostly seen in children less than five years of age with a slight male preponderance. Patients usually present with a palpable abdominal lump, abdominal distention, pain and gross hematuria. It is an aggressive tumor with about 17-40% of patients presenting with bone metastasis. It is one of the renal tumor in children which is most frequently misdiagnosed as it is rare, has varied histomorphology and lack of specific diagnostic markers. However, BCOR gene have emerged as a recent molecular marker. Management includes total nephrectomy followed by postoperative chemotherapy and radiotherapy. We describe a case of clear cell sarcoma of kidney diagnosed on histopathology in a 2.5 year old male child.

KEYWORDS : Children, Clear Cell Sarcoma, Renal

INTRODUCTION-

Clear cell sarcoma of kidney is a rare malignant renal tumor of children.It is second most common pediatric renal malignancy after Wilm's tumor,constituting around 4-5% of primary renal neoplasms. It is one of the aggressive tumor with a high tendency for recurrence and metastasis to bone.[1] First described in 1978 by three groups i.e Beckwith and Palmer, Morgan and Kidd, and Marsden and co-workers who named it "bone metastasizing renal tumor of childhood" as it has predilection for skeletal metastasis which occurs in 40% to 60% of patients.[2]The peak incidence is similar to Wilm's tumor seen between 2 and 5 years of age. It is very rare in infants less than 6 months old. Male to female ratio is 2:1.[3] Patients usually present with abdominal distension, a palpable abdominal mass, abdominal pain and gross hematuria.[4] Mostly patients are seen in early stage disease with only < 10% presenting with stage 4 disease. Management includes total nephrectomy followed by postoperative chemotherapy and radiotherapy.[5] Survival rate varies according to stage of the disease i.e 97% for stage I vs. 50% for stage IV disease.

We describe a case of clear cell sarcoma of kidney diagnosed on histopathology in a 2.5 year old male child.

CASE REPORT-

A 2.5 year old male child presented with gradually increasing abdominal distention and a palpable mass for 2 months. The mass was not associated with abdominal pain, constipation or urinary symptoms. On examination, a large lump was palpable in left lumbar region which was firm, non tender and ballotable. His routine hematological workup revealed mild anemia (Hb-10 gm%) however rest of the parameters were within normal limits. Serum urea (21.1mg/dl), serum creatinine (0.68mg/dl) and urine routine examination were within normal limits. No abnormality was detected on chest X-ray. USG abdomen revealed solid hypoechoic mass measuring 9x7cm at left upper pole of kidney. Liver, spleen, Right kidney were within normal limits. Abdominal CT scan revealed a large Heterogeneous enhancing mass involving superior pole of left kidney (Figure 1). Renal vein and ureter were free. There was no paraaortic lymphadenopathy.On radiology a provisional diagnosis of Wilm's tumor was rendered. Left total nephrectomy was performed and sent for histopathological examination. On gross examination, it was a globular encapsulated mass measuring 10x8x6 cm. On cut a solid, grey-white, homogeneous tumor was seen (Figure 2). No areas of hemorrhage and necrosis were identified. Renal vein and ureter were grossly uninvolved. Multiple sections examined on microscopy showed presence of tumor with sharp interface with surrounding kidney.Tumor was composed of sheets of medium sized monomorphic oval to elongated tumor cells separated by thin, incomplete fibrovascular septae.Tumor cells had indistinct cell borders, eosinophilic to clear vacuolated cytoplasm, round to oval nuclei showing mild pleomorphism and fine granular chromatin (Figure 3). Focal myxoid areas were also seen. Few mitosis were seen (0-2/hpf). No areas of necrosis, lymphovascular or capsular invasion were identified. Renal vessels and hilum were free of tumor. Attached perinephric fat and ureter were free of tumor. Immunohistochemistry showed strong diffuse positivity for Vimentin and Cyclin D1 (Figure 4) and negativity for EMA, Desmin, SMA,WT1 and BCL2. On the basis of histology and IHC, a final diagnosis of clear cell sarcoma, left kidney was given. Depending on histopathology report and clinical work up for metastasis, patient was found to have Stage 1 disease. Patient is currently undergoing chemotherapy according to regimen I of NWTS-5 protocol and has not yet shown any sign of metastasis.

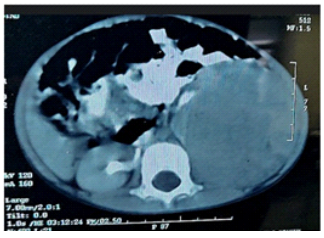

Figure 1: Abdominal CT of renal mass

Figure 2: Gross appearance of renal mass

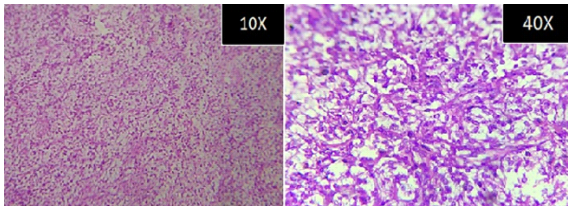

Figure 3: Microscopic appearance of renal mass showing fibrovascular septae and medium sized monomorphic oval to elongated tumor cells

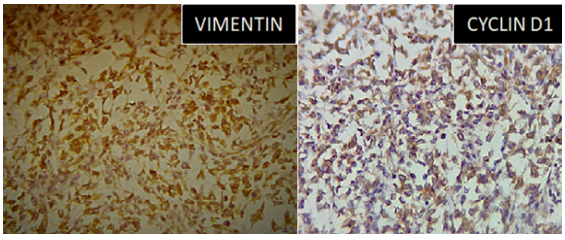

Figure 4 (40X): Tumor cells showing immunohistochemistry positivity

DISCUSSION-

Clear cell sarcoma of kidney is the second most common pediatric renal malignancy, constituting around 4-5% of primary renal neoplasms. It is one of the aggressive tumor have high tendency for recurrence and bone metastasis .[1] This tumor had been variously named as undifferentiated sarcoma of the kidney, bone metastasizing renal tumor of childhood and clear cell sarcoma and the name clear cell sarcoma of kidney was eventually adopted.[6] The peak incidence is similar to Wilm's tumor seen between 2 and 5 years of age. In our case also age of the patient was 2.5 years. Patients usually present with abdominal distension, palpable abdominal mass, abdominal pain and gross hematuria.[4] In this case also main complaints were abdominal mass and abdominal distension. Mostly patients have stage I, II, III disease at presentation with only < 10% presenting with stage 4 disease. This case too presented in stage I. The important prognostic factors are stage of the disease, age at the time of diagnosis, presence of tumor necrosis and treatment with doxorubicin.[7]Stage IV disease and young age are significant adverse prognostic factors. Clear cell sarcoma of kidney has a highly variable histologic appearance making histological diagnosis potentially challenging.Various histological patterns have been identified including spindle cell, sclerosing, epithelioid, myxoid, palisading, storiform and anaplastic variants, however the classic pattern is most frequently seen.[8] The classic pattern consists of monomorphic plump ovoid cells arranged in broad trabeculae or nests separated by arborizing fibrovascular septae with clear to vacuolated cytoplasm. Nuclei are round to oval and contain finely dispersed chromatin without any conspicuous nucleoli. In this case also classic pattern was seen on histopathology. Clinically as well as on radiology a provisional diagnosis of Wilm's tumor was given because of similar presentation and age. But, the characteristic histomorphology with delicate arborizing septae, monomorphous population and the immunohistochemical profile helps to distinguish it from Wilm's tumor.

CONCLUSION-

This interesting case is important as few cases have been reported in the literature. Clear cell sarcoma of kidney is one of the most frequently misdiagnosed renal tumor in children as it is uncommon, has varied morphology and there are no specific diagnostic markers. However, BCOR gene have emerged as a recent molecular marker. The need of clear cut distinction of clear cell sarcoma of kidney from other primary renal tumors of childhood such as wilms tumor, congenital mesoblastic nephroma, rhabdoid tumor and neuroblastomas is of utmost importance because of its aggressive nature, poor prognosis and treatment protocol will ultimately depend on the final diagnosis.

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