Primary Ewing Sarcoma of Kidney; Case Report of A 20 Year Boy

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ABSTRACT

Background: Primary Ewing sarcoma (ES)/ Primitive Neuroectodermal tumor (PNET) is a very rare tumor occurring mostly in young adults. It presents with vague flank pain. It is an aggressive tumor with high risk of metastasis and poor prognosis.

Case Presentation: We report a case of a 20 year old boy who presented with mass abdomen. CT scan revealed left renal mass. His nephrectomy followed by histopathology and immunohistochemistry showed primary renal Ewing sarcoma. It was limited to kidney without any metastasis.

Conclusion: Although a rare tumor early diagnosis is important to prevent metastasis and better overall prognosis.

Key words: Ewing Sarcoma, Primitive Neuroectodermal tumor, Central nervous system

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INTRODUCTION

Primitive Neuroectodermal tumor (PNET) is common in Central nervous system (CNS) but peripherally occurring PNETs belong to Ewing family of tumors.1 Previously Ewing sarcoma and PNET were considered separate conditions, but molecular studies have demonstrated that they belong to same family of tumors2. Now the term Ewing sarcoma is used for both these conditions.

Primary Ewing sarcoma of kidney is a rare tumor. Very few cases have been reported in literature. They are usually asymptomatic until they reach a large size and present with flank pain, hematuria, and palpable mass in abdomen.3 These are aggressive tumors with tendency to metastasize and with poor prognosis. 4

We present a case of 20 year old male who presented with primary renal Ewing sarcoma.

CASE REPORT

A 20 year old male presented with pain lower abdomen followed by hematuria. His CT scan abdomen revealed a large mass involoving the left kidney. His nephrectomy was performed followed by histopathological examination. Gross examination of the tumor revealed a renal mass measuring 10x10x8 cm. Cut surface showed cystic and haemorrhagic areas. On microscopic examination the tumor was composed of small round to oval cells with hyperchromatic nuclei and scanty cytoplasm forming rosettes as shown in figure 1. Immunohistochemically the tumor was positive for CD99, FLI1 as shown in figure 2. LCA, Desmin and WT1 were negative. Final diagnosis of Ewing sarcoma was made. FISH was performed which showed EWSR1 gene rearrangement. His Photon emission tomography-computed tomography (PET-CT) Scan ruled out any systemic skeletal metastasis. The patient was put on chemotherapy regimen VAC-IE (Vincristine, Adriablastina, Cyclophosphamide, Ifosfamide, Etoposide). So far 6 cycles of chemotherapy are completed. No signs of recurrence has been seen after the operation so far.

Figure1. Showing small round blue cells with scant cytoplasm)

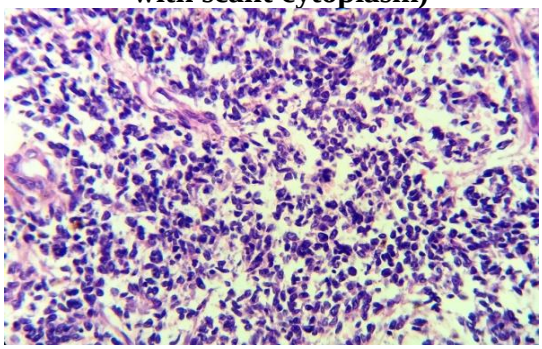
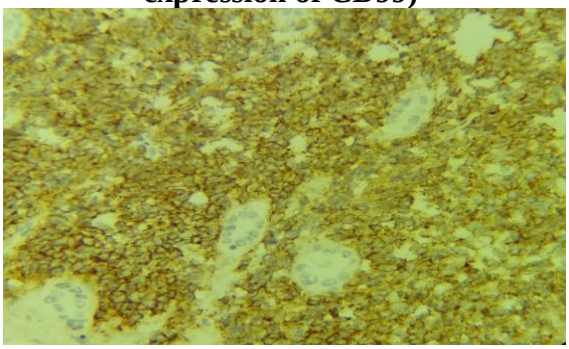


Figure 2. Immunohistochemical expression of CD99)



DISCUSSION

Renal cell carcinomas constitute 90% of kidney tumors. Renal sarcomas account for less than 1 % of all tumors.5,6 Ewing sarcoma of kidney is very rare tumor which mostly affect young males with median age of 28-34 years.7 In our case the patient was 20 years of age. These patients are usually asymptomatic until their size increase to cause local pressure symptoms. Average size at time of diagnosis is 5.5 cm to 23 cm.5,7 In our case the tumor was 10 cm in maximum dimension. Common symptoms in these patients are pain (85%), palpable mass (60%), and hematuria (37%). 7,8 As in our case patient presented with flank pain and later hematuria.

Radiological findings of Ewing sarcoma on kidney are non specific and show overlapping with other tumors of kidney like renal cell carcinoma, Wilm's tumor, neuroblastoma, lymphoma, desmoplastic small round cell tumor, renal cell sarcoma and metastatic carcinoma.5 Ultrasonography, CT scan and MRI have variable role in demonstrating the tumor size and its relationship with the kidney and surrounding structures.

Histopathological examination with immunohistochemistry has vital role in final diagnosis of tumor and further workup. On histopathological examination these tumors are composed of small round blue cells forming Homer wright rosettes.

Immunohistochemically these tumors are positive for CD99, and FLi1. Negativity for other markers like CK, LCA, Desmin, WT1, GFAP, PAX2 will rule out other small round blue cell tumors.4 Table 1. Show comparison of age, tumor size, metastasis, immunohistochemical markers and EWSR1 gene rearrangement of our case with other regional and international studies.

Table1. Comparison with regional and international studies

	Katkar AS et al(2014)	Badar Q et al (2010)	Almeida MF et al (2014)	Nam JK et al(2015)	Zubair et al (2016)
Age	46F	13F	19M	30M	20 M
Tumor size	15.7 cm	9x6 cm	-	30x20cm	10x10 cm
Metastasis	Not seen	Lung Mets	Lung mets	Not seen	Not seen
IHC	CD99, FLI 1	CD99	Not given	CD99	C99, FLI 1
EWSR1 Rearrangement	Seen	-	Seen	-	Seen

After establishing diagnosis appropriate treatment strategies like surgery, chemotherapy and radiotherapy are formulated. 9 Current studies suggest that the addition of ifosfamide and etoposide to a regimen containing doxorubicin, vincristine, dactinomycin, and cyclophosphamide, improves the outcome for patients with both non metastatic and metastatic disease 10. Despite aggressive therapy, however, the prognosis for this tumor is poor, with a 20% as overall cure rate 11.

CONCLUSION

Ewing sarcoma is rare differential diagnosis in renal tumors. It occurs mostly in young adults with aggressive course. Patients have vague, nonspecific symptoms. Diagnosis is based on histologic examination, immunohistochemistry, and molecular findings. Despite the multimodal treatment the tumor has poor prognosis.3

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