Primary Ewing's Sarcoma/Primitive Neuroectodermal Tumor of Kidney: A Rare Case Report

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Abstract

Ewing's sarcoma/primitive neuroectodermal tumor (ES/PNET) is a member of small round cell "blue tumor" family, occurring primarily in kidney is quite rare. Hence, detail appraisal is needed using histopathology, immunohistochemistry, and cytogenetic studies to attest the final diagnosis of ES/PNET, as this has an aggressive course and poor outcome. Here, we present a case of primary ES/PNET of kidney in 22-year-old male with locally invasive disease. Tumor cells were strongly positive for CD99 and FLI-1 on immunohistochemistry, thus confirming the final diagnosis.

Keywords: *CD99, Ewing's sarcoma/primitive neuroectodermal tumor, FLI-1, kidney, primary*

Introduction

Ewing's sarcoma/primitive neuroectodermal tumor (ES/PNET) is a member of small round cell "blue tumor" family. The overall incidence of ES and PNET is <1%. ES/PNET, both are equivalent tumors microscopically, immunohistochemically, and genetically with bone is the most common site affected.[1,2] Rarely, it can present as a primary renal mass with <100 cases have been reported in literature till date.[3,4] Primary renal PNET has highly aggressive clinical course than other sites with a tendency for early metastasis and death.[5] Differential diagnosis includes the other members of small round cell "blue tumor" family which can impose diagnostic difficulties due to frequent overlapping features between the entities. Histopathology, immunohistochemistry along with cytogenetic analysis are essential to arrive at the final diagnosis of ES/PNET of kidney. The presence of small round cells with rosettoid arrangement on histopathology and identification of CD99 and FLI-1 expression is critical evidences for the diagnosis. In addition, cytogenetic detection of EWL/FLI-1 fusion protein is strongly supportive for final diagnosis.[6]

Here, we report a rare case of ES/PNET in a young adult.

Case Report

A 22-year-old male patient presented with a history of hematuria and pain in right lumbar region of 2 weeks duration. For these complaints, he was subjected for ultrasonography and computerized tomography scan of abdomen which showed a solid lesion with necrotic areas in the middle part of right kidney measuring 6 cm × 5.2 cm × 4 cm [Figure 1]. There was no other lesion at any other site. Viewing the clinical and radiological profile, right radical nephrectomy was done. On gross examination, the specimen consisted of kidney with ureter. On cut section – a gray-brown mass of size 4 cm × 4 cm seen in mid kidney destroying medulla and cortex. Another cystic lesion of size 1.5 cm diameter noted in mid cortex of the kidney [Figure 2]. Histologically, tumor composed of small uniform, dark, round cells arranged in sheets, and rosettoid pattern. Cells had round to oval nuclei, dark clumped chromatin, inconspicuous nucleoli, and a small amount of vacuolated cytoplasm [Figures 3 and 4]. Both the masses in kidney showed similar histology. Tumor cell infiltration was noted in renal capsule, perirenal fat, renal sinus fat, and Gerota's fascia. A wide panel of immunohistochemistry (IHC) markers was applied to rule out the other members of small round cell "blue tumor" family. Tumor cells show strong diffuse membranous positivity for CD99 [Figure 5], moderate positivity for FLI-1 [Figure 6] and neuron-specific enolase (NSE). However, tumor cells were negative for cytokeratin (CK), EMA, WT1,

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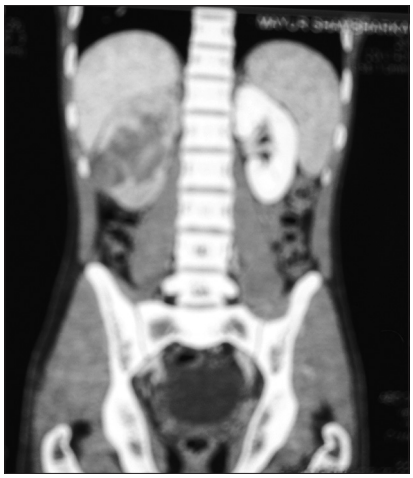

Figure 1: Computerized tomography scan showing solid lesion with necrotic areas in the middle part of right kidney

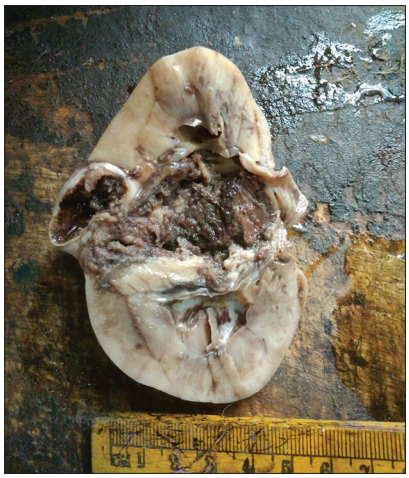

Figure 2: Gross photograph showing two mass lesions in the middle part of kidney

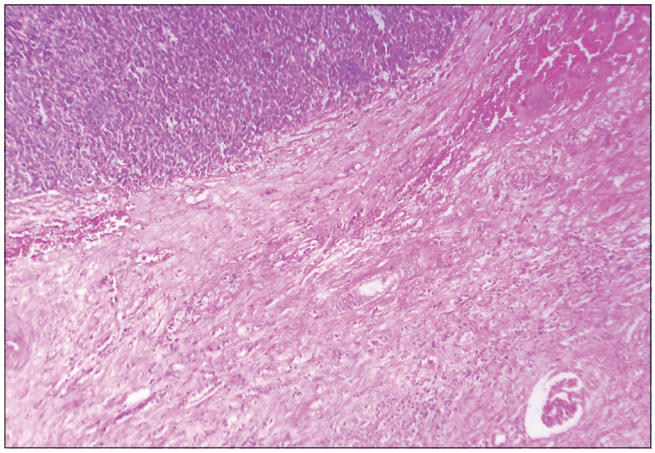

Figure 3: Microscopic photograph showing glomeruli one side and sheets of blue cells on other side (×100)

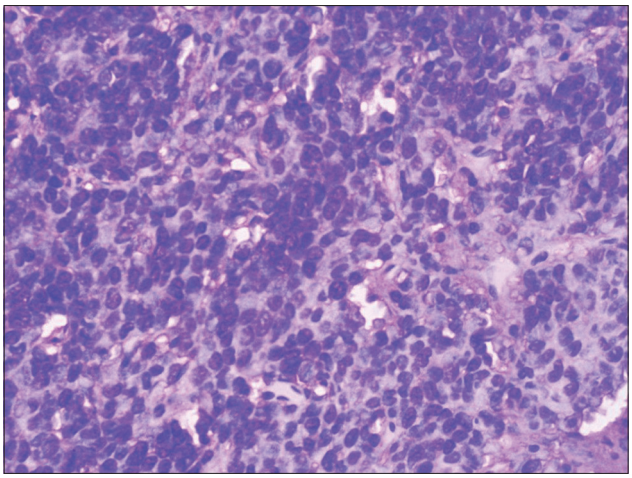

Figure 4: High power showing cells with nuclei round to ovoid, dark clumped chromatin &scanty cytoplasm (×200)

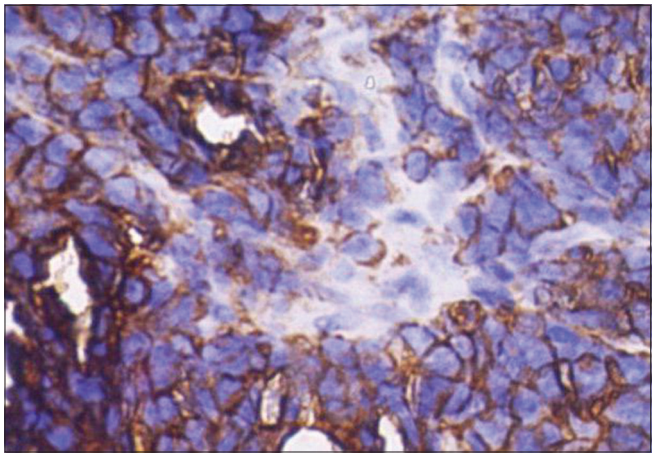

Figure 5: Immunohistochemical image of CD99 showing strong diffuse membranous positivity

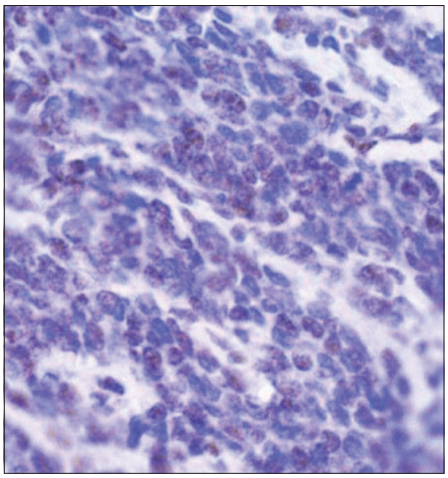

Figure 6: Immunohistochemical image of FLI-1 showing moderate positivity

LCA, TdT, synaptophysin, desmin, and vimentin. Tumor was mitotically active with ki-67 – 25%.

Hence, with these morphological and IHC findings, diagnosis of primary ES/PNET of kidney was given.

Discussion

ES/PNET is a member of small round cell "blue tumor" family with bone and soft tissues are the most common

sites affected. Occurring primarily in kidney is a rare with first case of renal PNET was reported by Mor *et al*. in 1994.[7] Majority of the patients affected by ES/PNET of Kidney are in their second and third decades of life with slight male predominance.[8,9] The presenting symptoms and radiological findings of PNET kidney are nonspecific and can mimic other renal mass lesions such as RCC and nephroblastoma.[10,11] The patient was 22-year-old male patient presented with right-sided lumbar pain and hematuria of short duration with radiological diagnosis offered were? RCC? nephroblastoma, our findings matching with other authors. Grossly, renal PNET frequently presents with areas of necrosis and hemorrhage along with cystic degeneration.[6,12] In our case, we had also similar gross findings.

Primary renal PNET behaves more aggressively than PNET arising at other sites with tendency for has with tendency for early local recurrence and remote metastasis to lymph nodes, lung, liver, and bone which entail the worse prognosis.[13,14] Moreover, postoperative radiotherapy is recommended in advanced local invasion to perineprhic tissue and Gerota's fascia.[15]

In our case, secondary cystic tumor nodule was present in kidney itself along with tumor cell infiltrates into Gerota's fascia, perirenal fat, and renal sinus fat indicating advanced local spread of the tumor despite of small size of the primary lesion.

The differential diagnosis of primary renal PNET includes other members of small round cell "blue tumor" family-like neuroblastoma, blastema-rich nephroblastoma, malignant lymphoma, monophasic synovial sarcoma, and rhabdomyosarcoma and desmoplastic small round cell tumor. Hence, for the complete appraisal of small round cell "blue tumor" family, expanded IHC panel was applied which showed strong positivity for CD99, moderate positivity for FLI-1 and NSE. However, CK, EMA, WT1, LCA, TdT, synaptophysin, desmin, and vimentin were negative.[15,16] We had NSE positive. However, literature says, PNET cells can also show NSE positivity depending on the differentiation, but neuroblastoma cells are always CD99 negative. CD99 is also positive in synovial sarcoma, but it will also show positivity for bcl-2 and CK which was negative in our case.[4] Most of the ES/PNET of kidney shows the translocation of t (11:22) as a result of fusion of EWS gene on chromosome 22 to the FLI-1 gene on chromosome 11.[12]

This multimodality diagnostic approach is required for confirmation of the diagnosis and further management of the ES/PNET of kidney.

Conclusion

Primary ES/PNET of kidney is a rare diagnosis, needs detail appraisal from other members of small round cell "blue tumor" family, using histopathology, immunohistochemistry, and cytogenetic studies. These diagnostic modalities are crucial to attest the final diagnosis and to adapt appropriate management protocol; as renal ES/PNET has an aggressive course and poor outcome.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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