Case Report

Primitive neuroectodermal tumor of the kidney

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ABSTRACT

Primitive neuroectodermal tumor of the kidney is a rare entity. Only a few cases have been reported in the literature and they revealed a variable presentation and an aggressive behavior. Most commonly it is seen in the relatively young population. The diagnosis is usually made at histopathology.

Key words: Fluorodeoxyglucose (^{18}F-FDG), Ewing's sarcoma, primitive neuroectodermal tumor

INTRODUCTION

The primitive neuroectodermal tumors (PNETs)/Ewing's sarcomas (ESs) are small-round-cell tumors of neural crest origin. The commonest site is the central nervous system. Peripheral localization is rare. Only a few cases of primary renal PNET have been reported.[1]

The mean age for renal PNET is 28 but it can be seen in a wide range between 4 and 69 years.[2] As it is a rare localization, it can be confused with other renal tumors like Wilms' tumor. Due to its distinctive prognostic and therapeutic features, differential diagnosis is very important. Renal PNET is more aggressive than in other sites. It has a high risk of local recurrence and distant metastasis.[2] The five-year disease-free survival rate for localized extraskeletal PNET is around 45–55%, and cases with advance disease at presentation have a median relapse-free survival of only two years.[3]

CASE REPORT

An 18-year-old female presented at our outpatient department (OPD) with complaints of pain in the abdomen and hematuria. There was no history of fever, vomiting, or burning micturition. On examination, a lump was found in the left flank. Ultrasonography (USG) indentified an enlarged left kidney showing 93 × 85 mm mass of mixed echopattern in relation to the lower pole. The mass showed central anechoic areas, and no calcification was seen. Computed tomography (CT) scan revealed left renal mass with inferior vena caval thrombus. She underwent left radical nephrectomy with inferior vena caval thrombectomy. Intraoperatively, the left renal mass with left renal vein thrombosis extending into the inferior vena cava for about 2 cm was found. Multiple enlarged and hard lymph nodes in para-aortic region inferior to the caval region were found. A cut section of the left kidney revealed a large white tumor occupying the middle and lower portions of the kidney, measuring 13 × 11 × 10 cm. The cut section of the tumor was soft and friable with extensive areas of necrosis and hemorrhage. The tumor was grossly limited to the kidney, and invaded the hilum, with the presence of tumor emboli in the branches of major vessels. The renal vein was filled with tumor embolus extending up to the cut surface. Multiple coalesced lymph nodes with some grossly involved with the tumor were identified, with the largest measuring 3 cm in diameter.

The surgical specimens were formalin fixed and paraffin embedded. Microscopy revealed a high-grade malignant neoplasm with pushing borders stretching the renal capsule and confined within the kidney. The tumor was found to be highly cellular and showed large geographic areas of coagulative necrosis, with variable tumor cells condensed around blood vessels (perivascular survival pattern). The tumor was composed of diffuse sheets of monomorphic-appearing malignant small round cells showing brisk mitosis, apoptotic activity,

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high nucleocytoplasmic ratio, scant pale cytoplasm, and round-to-ovoid hyperchromatic nuclei with finely granular chromatin, inconspicuous nucleoli, and slightly contorted nuclear outlines at places [Figure 1a]. Immunohistochemistry was performed using the UltraVision polymer detection system and the antibodies used included cytokeratin, vimentin, WT-1 (WT: Wilms' tumor protein), CD99 (CD: Cluster of differentiation), chromogranin, and desmin. Immunohistochemical evaluation revealed a diffuse CD99 positivity in the cytoplasam of the neoplastic cells. Cytokeratin, WT-1, and vimentin positivity was focal. Chromogranin and desmin were negative. These findings along with the clinical presentation, the microscopic aspect, and cytological characteristics were in favor of a diagnosis of primary neuroectodermal tumor of the kidney [Figure 2a-c].

A whole-body ^{18}F-FDG PET (PET: Positron emission tomography) study was done which revealed multiple pulmonary nodules in both lungs and L1-2 vertebrae showing increased FDG uptake, along with evidence of left pleural effusion [Figure 1b]. Four cycles of chemotherapy with vincristine, adriamycin, and cyclophosphamide were performed.The first cycle of vincristine, adriamycin, and cyclophosphamide was alternated with ifosamide, carboplatin, and etoposide.

The subsequent three cycles of vincristine, adriamycin, and cyclophosphamide were alternated with three cycles of ifosamide and carboplatin. Palliative radiotherapy, 8 Gray in 1 fraction was given to the D12-L3 vertebrae.The patient is to receive further cycles of chemotherapy of the above regimen.

DISCUSSION

The peripheral PNET was first recognized by Arthur Purdy Stout in 1918.[4] The first report of renal PNET was by Seemayer *et al*. in 1975.[5] The rarity of these tumors preclude a meaningful analysis of the clinical outcome. The diagnosis of renal PNET/Ewing's tumor is very rare and usually involves several different diagnostic techniques. The most challenging part is the differential diagnosis which is broad with morphological and immunohistochemical overlapping features between the entities. Therefore, mostly frequently, molecular biology techniques need to be applied for a definitive diagnosis. The differential diagnosis of renal PNET includes rhabdomyosarcoma, Wilms' tumor, carcinoid neuroblastoma, clear-cell

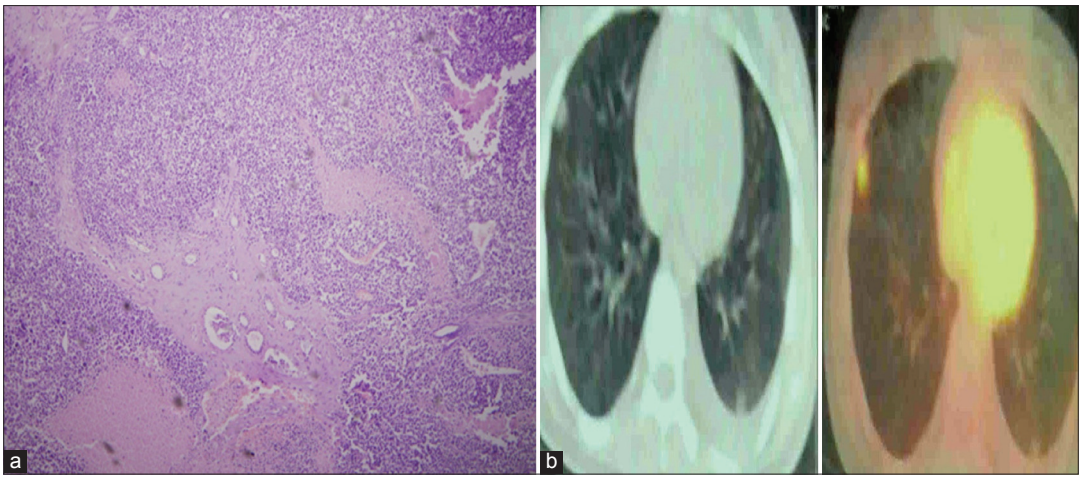

Figure 1: (**a**) A×40 showing a malignant small-round-cell tumor of kidney, (b) Fluorodeoxyglucose (^{18}F) positron emission tomography scan image revealing metastatic nodules in lungs

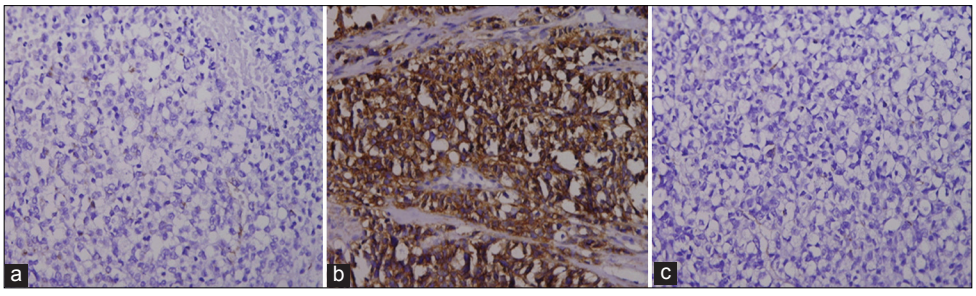

Figure 2: (a) ×200 showing negative staining for WT-1, (b) 2×200 immunohistochemistry showing positive membranous staining for mic-2, (c) 2×200 immunohistochemistry showing negative staining for synaptophysin

sarcoma of the kidney, lymphoma, small-cell variant of osteosarcoma, desmoplastic small-round-cell tumor, small-cell anaplastic neuroendocrine carcinoma, and nephroblastoma.[6] These tumors typically manifest in adolescents and young adults and have aggressive behavior. Despite aggressive treatment of these tumors by combination therapy with surgery, chemotherapy, and radiotherapy, the prognosis remains poor and overall five-year survival rates have been reported at 45% to 55%.[7] Our patient is currently alive and scheduled for the next cycle of chemotherapy.

CONCLUSION

PNET of the kidney is a rare tumor with very aggressive behavior. Unusual presentation usually delays diagnosis. Due to the aggressive nature of the tumor, multimodality treatment is recommended. Complete resection of the tumor with nodes should be done in all cases wherever feasible. Combination chemotherapy gives best results. The role of radiotherapy is not clear but indicated in unresectable disease or in case of gross residual disease. Despite all modalities, overall survival is dismal in these patients.

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