Primitive neuroectodermal tumor of the kidney: A case report and review of the literature

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Background: Primitive neuroectodermal tumors (PNETs) represent a family of neoplasms, presumed to be of neuroectodermal origin and mostly presenting as bone or soft tissue masses in the trunk or axial skeleton in adolescents and young adults.

Case: The patient was a 32-year-old man. On gross examination, the tumor measured 16x14 cm in dimensions, with cystic areas and necrosis. Microscopically, the neoplasm was highly cellular consisting of small-round-to-oval cells with irregular nuclei. Immunohistochemical stains were positive for vimentin and CD99. Microscopic and immunohistochemical findings were compatible with PNET.

Conclusion: Although rare, PNET must be included in the differential diagnosis of renal tumors especially in children and young adults. We present a case of renal PNET as an unusual case, discuss the differential diagnosis of small round-cell tumors in this location and review the literature.

Keywords: primitive neuroectodermal tumor, kidney

Introduction

Primitive neuroectodermal tumor (PNET) is thought to arise from primitive cells of neural crest and commonly involves the bone and soft tissue in adolescents. It was first described by Stout in 1918. Renal PNET is an extremely rare entity. It had been documented as isolated case reports. There are only two series of renal PNET that was published by Parham and Jimenez in the literature. Here we report a rare case of renal PNET and review the literature.

Case report

A 32 year-old-man was admitted to a hospital with abdominal pain. Physical examination revealed a palpable mass in the right upper abdomen. Computerized tomography (CT) of the abdomen demonstrated a large tumor which was confined to the right renal parenchyma. The results of laboratory studies were within normal limits. Chest X-ray and bone scan showed no evidence of a metastatic disease. CT investigations ruled out any other primary tumor, except kidney. The patient underwent a right nephrectomy and was introduced to our hospital for further evaluation. Gross examination of the resected kidney showed extensive replacement of the renal parenchyma by grayish-yellow, 16 x 14 x 10 cm tumor with focal cystic areas and irregular areas of necrosis (Figure 1). Macroscopically, renal parenchyma, perirenal space and renal vein were invaded by tumor. Microscopically tumor consisted of small-sized, round-oval shaped cells with irregular nuclei and ill defined cytoplasmic borders. Poorly formed rosette-like structures were rarely observed (Figure 2). There were extensive areas of hemorrhage and necrosis. Paraffin-embedded preparation was stained immunohistochemically using streptavidin biotin.
Immunohistochemical stains were positive for vimentin and CD99 (MIC-2 gene product, 12E7) (cytoplasmic surface membrane staining) whereas negative for cytokeratin, LCA, desmin, S-100, Tdt (Terminal deoxynucleotidyl transferase), NSE, and synaptophysin (Figure 3). There was also focal positivity with chromogranin. Based upon the microscopic appearance and immunohistochemical features, the diagnosis of PNET of kidney was established. Unfortunately, we couldn’t perform cytogenetic analysis. The pathologic stage of tumor was pT3b. Despite additional chemotherapy the patient died two months after the initial diagnosis.

**Discussion**

The small cell tumors of the kidney are a heterogeneous group of neoplasms with overlapping morphologic features and different prognostic/therapeutic implications. This group of tumors usually include blasteme-predominant Wilms’ tumor (WT), PNET, neuroblastoma, rhabdomyosarcoma, lymphoma and desmoplastic round cell tumor.4–6 Among these, PNET of the kidney is a rare entity that classically occurs in children and young adults.7–10

Several theories have been proposed for a reasonable explanation for the genesis of PNETs which arise at peripheral sites.5,11 One of them is the presence of aberrant neural crest cells in the kidney. Another explanation for genesis is arising from the neural ramifications of the celiac plexus that intervates the kidney.5,11

Renal PNET was first reported by Mor et al.5 Between 1994-2004, approximately 40 renal PNET cases have been reported.1–17 However, Parham et al. have reported a large series of malignant neuroepithelial tumors of the kidney including 79 cases considered to be PNET on the basis of histology and immunohistochemistry.11 Recently, Jimenez et al. have cited additional 11 cases including 8 cases with follow-up information.12 Among these, one patient was alive without evidence of disease with a survival of 64 months which seems to be the longest survival in the literature.12 In other reports, survival time ranged from
1 to 24 months with distinctively poor prognosis among patients with distant metastases at diagnosis. The most common metastatic sites were lymph nodes, lung, liver and bone. Most of these previously reported cases have occurred in young male adults. All of the cited cases exhibited the similar histologic features with small cells arranged in cords, nests or clusters with or without rosettes and pseudorosettes. A summary of cases cited in the literature appears in Table 1.

All other small round cell tumors of the kidney must be included in the differential diagnosis of renal PNET. Immunohistochemically CD99, NSE and vimentin are expressed by PNET cells. S-100 protein,
lymphoblastic lymphoma. Focal areas of neural and Tdt excluded Non-Hodgkin lymphomas especially with antibody to CD99. Negative staining with LCA largely confined to the kidney and reacted positively exhibited features of the so-called small round cell examinations. Histologically, the present case histopathological and immunohistochemical diagnosis of PNET of the kidney based on shows no reactivity for WT-1 gene product and can be occurs in approximately 90% of these neoplasms.4–11

Despite their genetic and antigenic similarity, most authors recognize PNET and extraskelatal ES as separate entities. Distinction is based primarily on the more neural differentiation of PNET.7 Several recent cytogenetic analyses of PNET have demonstrated a reciprocal translocation t(11;22)(q24;q12). This translocation which appears to be unique to the PNET and Ewing’s sarcoma (ES), occurs in approximately 90% of these neoplasms.4,6,7,9

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Renal PNETs are frequently aggressive and almost 30% of all newly diagnosed cases present with distant metastases. Surgical intervention, intensive chemotherapeutic drugs and radiation therapy are the best choices for the management.8,11

The features of our case strongly support the diagnosis of PNET of the kidney based on histopathological and immunohistochemical examinations. Histologically, the present case exhibited features of the so-called small round cell tumor. The neoplasm arose in a young male adult, was largely confined to the kidney and reacted positively with antibody to CD90. Negative staining with LCA and Tdt excluded Non-Hodgkin lymphomas especially lymphoblastic lymphoma. Focal areas of neural differentiation and positive reaction with antibody to chromogranin tend to favor PNET over ES. Immunohistochemically positive reaction with CD99 and negative reaction with cytokeratin, desmin, NSE and synaptophysin widely helped to differentiate most other small cell tumors like renal carcinoid, neuroblastoma and small cell carcinoma. In spite of an aggressive combined treatment (surgery and chemotherapy), the patient developed liver metastases and died from tumor progression after 2 months.

In conclusion, renal PNET is a rare neoplasm and should be differentiated from the other small cell tumors of the kidney. Immunohistochemistry for CD99 and if possible cytogenetic studies should be performed in all cases where PNET is considered.

References