

Case Report

Middle East Journal of Cancer; October 2014; 5(4): 233-236

Primitive Neuroectodermal Tumor of the Kidney in a 22-Year-Old Male: A Rare Entity

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Abstract

Primitive neuroectodermal tumors of the kidney are rare tumors representing about 1% of all sarcomas. Extraskelatal primitive neuroectodermal tumors may be found in the genitourinary tract, the testes, ovaries, uterus, or pancreas. Renal primitive neuroectodermal tumors 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 management.

We discuss the case of a 22-year-old male who presented with a left renal mass. He underwent nephrectomy followed by histopathological examination. Microscopic and immunohistochemical examination revealed a primitive neuroectodermal tumor.

Keywords: Primitive neuroectodermal tumor (PNET), Kidney, Rare

Introduction

Primitive neuroectodermal tumor (PNET) is an uncommon malignancy of bones and soft tissue which rarely occurs in the kidneys.¹ In the kidneys most cases occur in the medullary/pelvic region.² It is a small round cell tumor that has neuroepithelial differentiation, which typically occurs in young children and adolescents.¹ This tumor is a rare entity and its diagnosis is usually made at histopathology. A few cases that have been reported in the literature reveal a variable presentation and aggressive behavior.³

Case Report

A 22-year-old male presented with a tender mass in his left loin. Past medical history was unremarkable. Routine hematological, biochemical and urine examinations were within normal limits. Computerized tomography (CT scan) of the abdomen revealed a mass that involved the upper and middle poles of the left kidney without any capsular invasion or renal vascular involvement. There was no evidence of intra-abdominal or retroperitoneal lymphadenopathy or any abdominal organ involvement. Chest X ray, CT scan of the thorax and bone scan did not reveal any evidence of lung or

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Received: March 17, 2014; Accepted: June 1, 2014

bone involvement. The patient subsequently underwent left radical nephrectomy.

Gross inspection revealed a mass that measured 11×9 cm. The cut surface was variegated with areas of hemorrhage and necrosis (Figure 1). Microscopic examination revealed a malignant small round cell tumor (Figure 2) with rosetting in places (Figure 3a). The tumor cells expressed CD99 (Figure 3b) and vimentin (Figure 3c), but were negative for CD10 (Figure 3d), cytokeratin and leucocyte common antigen (LCA), which excluded the diagnoses of renal cell carcinoma, Wilms' tumor (WT) and lymphoma, respectively. Tumor cells also lacked expressions of desmin, neuron specific enolase (NSE), synaptophysin and chromogranin, which thereby eliminated the possibilities of rhabdomyosarcoma, desmoplastic small round cell tumor and neuroblastoma. The diagnosis of primitive neuroectodermal tumor (PNET) was rendered. The patient was started on three cycles of cyclophosphamide, vincristine, adriamycin and three cycles of vincristine, etoposide and ifosfamide over a period of ten months. He achieved good clinical response with no signs of local recurrence or distant metastasis during one year of follow-up.

Discussion

The most common renal tumor is renal cell carcinoma which has an accuracy of more than 85%.¹ Renal PNET is an extremely rare entity.⁴ The histogenetic origin of PNET remains

hypothetical. Because of their morphological resemblance to neuroblastomas, PNET have been considered to be neural crest derivatives.⁵ Theories explaining the genesis of PNETs which arise at peripheral sites include the presence of aberrant neural crest cells in the kidney or genesis from the neural ramifications of the celiac plexus that innervate the kidney.⁴

About 75% of cases occur before the age of 35 years and the median age is 20 years. The initial signs and symptoms are similar to those of other renal tumors. Radiographic features of PNETs are large size, lack of extensive parenchyma infiltration, lack of renal vein invasion, diffuse large calcification, areas of internal hemorrhage and necrosis, and peripheral hypervascularity.⁵ Our patient presented with pain in his left loin and the CT scan revealed a circumscribed renal mass that lacked involvement of renal vasculature or other abdominal structures.

Histologically, PNET consist of a sheet-like proliferation of small round cells with small amounts of clear to eosinophilic cytoplasm that form rosette-like structures in some areas. The tumor cells are usually uniform and small with finely dispersed chromatin and small nucleoli. Geographic necrosis and mitotic figures are frequently present.¹

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



Figure 1. Gross specimen of kidney showing a grey-white tumor with areas of necrosis and hemorrhage.

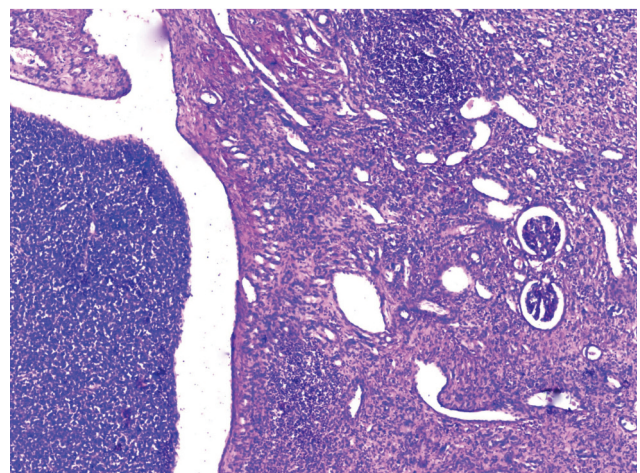


Figure 2. Microphotograph of a malignant round cell tumor of the kidney with normal kidney on the right (40×; H&E).

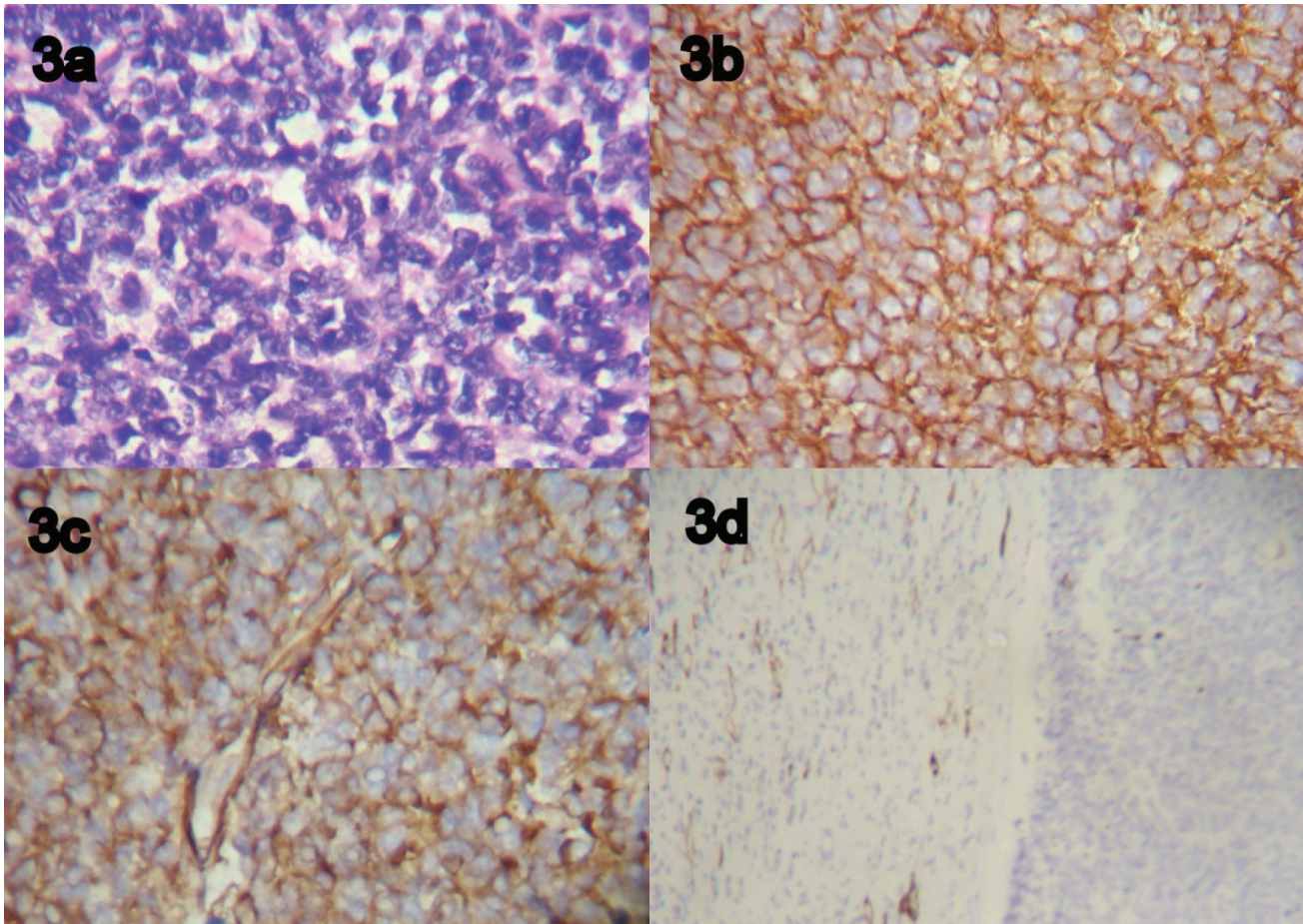


Figure 3. a) Microphotograph of a tumor composed of small round cells with round nuclei and scant cytoplasm arranged in sheets and rosettes in places (400 \times ; H&E). b) The tumor cells were immunoreactive for CD99 (400 \times ; IHC), and c) vimentin (400 \times ; IHC). d) CD10 was positive in adjacent normal renal tubules in contrast to its absence in the tumor cells (100 \times ; IHC).

tumors usually include blastemal predominant WT, PNET, neuroblastoma, rhabdomyosarcoma, lymphoma and desmoplastic round cell tumor.⁴ The cells of PNET express CD99, vimentin, NSE, and FL1 but do not express CK and WT1, LCA, synaptophysin and chromogranin which differentiates it from other light microscopic differentials including monophasic WT, lymphoma, carcinoid and neuroblastomas.^{1,5} The diagnosis of renal PNET must be considered in young patients who present with renal mass.³

PNETs are usually highly aggressive, with 25%-50% of patients presenting with metastatic disease that most commonly occurs in the lungs, bones, and liver.⁵ The five year disease-free survival rate is around 45%-55% in well confined cases, whereas cases with advanced stage at presentation have a median relapse-free survival of only two years.² The tumor in our case was

limited to the kidneys without any evidence of metastasis.

Standard therapy consists of a combination of surgical resection, postoperative irradiation and chemotherapy. The chemotherapy regimen used is either the round cell tumor (RCT II) protocol or Ewing's family of tumors (EFT 2001) protocol. However, further studies are required to validate the appropriate chemotherapy protocol.³

Conclusion

PNET of the kidneys, although rare, should always be considered in the diagnosis of round cell tumors that involve the kidneys. Immunohistochemistry and genetic studies (if available) should be employed to differentiate PNET from other tumors because of its poor prognosis and need for an adjuvant chemoradiotherapy.

Conflict of Interest

No conflict of interest is declared.

References

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