Case Report

Primitive neuroectodermal tumor of the kidney in a young male: Case report and review of literature

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Abstract

Primitive neuroectodermal tumor of the kidney is a rare tumor. A total of approximately 79 primary renal cases have been reported to date. Primitive neuroectodermal tumors occur preferentially in the soft-tissues of the paravertebral region and chest wall, less frequently in extremities, with a slight male predominance. We report a case of primitive neuroectodermal tumor of the kidney in a 17-year-old male with a pre-operative diagnosis of renal cell carcinoma-stage 4. The patient underwent radical nephrectomy and histopathological examination revealed a highly aggressive tumor of monotonous sheets of round cells with focal areas of rosette formations and high mitotic rate with Ki67 index of 25-30%. Tumor cells were positive for CD 99 confirming the diagnosis of primitive neuroectodermal tumor. Primitive neuroectodermal tumor of the kidney needs to be kept in mind as a differential diagnosis in young adults presenting with a large kidney mass.

Key Words: Blastemal wilms, immunohistochemistry, kidney, primitive neuroectodermal tumor

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INTRODUCTION

Primitive neuroectodermal tumor (PNET) of the kidney is a rare malignancy. In the literature reviewed approximately 79 primary renal cases have been reported.[1] Renal PNET is highly aggressive, presenting at an advanced stage, with distant metastasis and subsequent poor prognosis. It affects young adults with significant mortality.[2] We report a case of primary Renal PNET in a young male.

CASE REPORT

A 17-year-old male presented with complaints of hematuria and heaviness in left flank since 1 month. Physical examination revealed a soft lump in left hypochondrium. Ultrasound examination of the abdomen showed a mass in the left kidney. CT scan of the abdomen demonstrated a large significantly enhancing left renal upper mid polar mass with necrosis and a possible thrombus in left renal vein [Figure 1]. Left radical nephrectomy was performed. Gross examination revealed a friable, grayish white, lobulated mass ($15 \times 13 \times 7$ cm^3), with multiple foci of hemorrhage and necrosis [Figure 2]. The renal vein and ureter were free of the tumor. Four lymph nodes were dissected from perihilar region.

Microscopically, the tumor was composed of monotonous sheets of round cells divided by fibro vascular septae into lobules along with large areas of necrosis [Figure 3]. There were focal areas of rosette formations [Figure 4]. The individual cells had round to ovoid nucleus with a distinct nuclear membrane, fine powdery chromatin and 1 or 2 small nucleoli with 5-10 mitosis per high power field [Figure 5]. The cytoplasm was ill defined, scanty, and pale staining. Three of the four lymph nodes were involved. Immunohistochemically, the tumor cells were positive for

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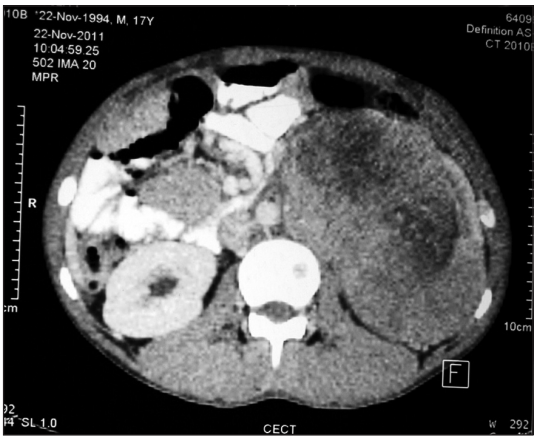

Figure 1: CT abdomen showing large significantly enhancing left renal upper mid polar mass with necrosis and a possible thrombus in left renal vein. No renal hilar lymphadenopathy

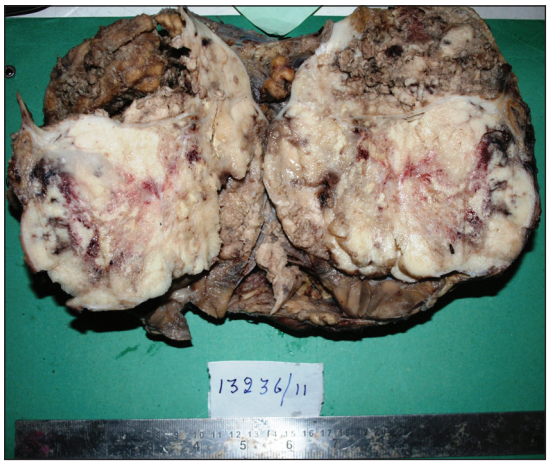

Figure 2: A friable, greyish white, lobulated mass, with multiple foci of hemorrhage and necrosis

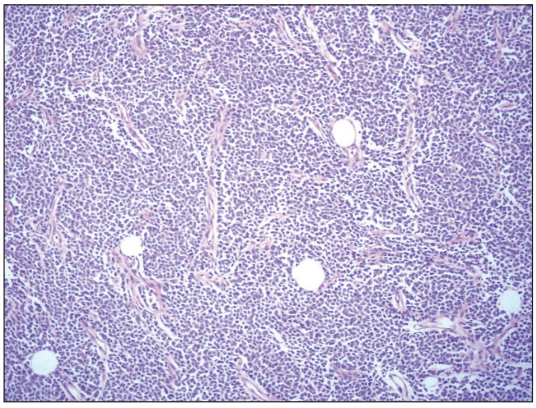

Figure 3: Tumor composed of monotonous sheets of round cells divided by fibro vascular septae

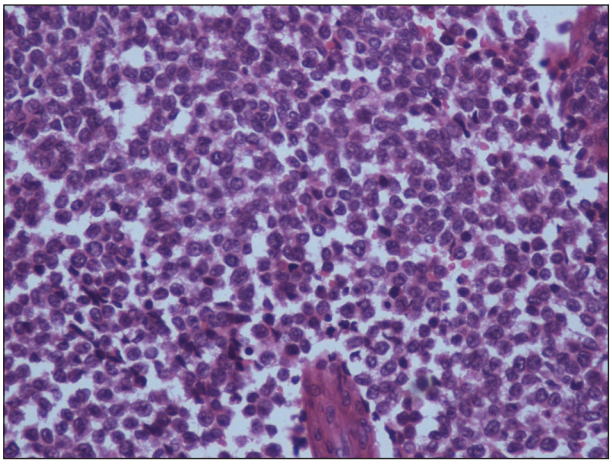

Figure 4: Focal areas of rosette formations

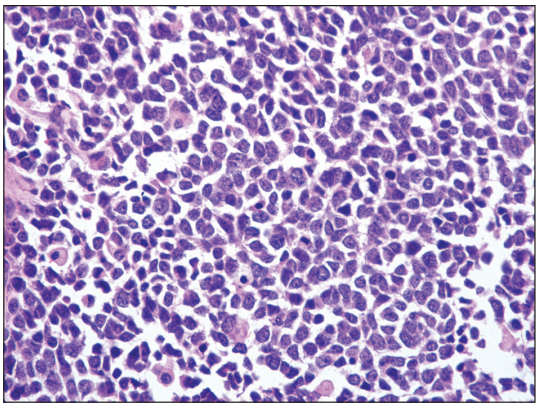

Figure 5: The individual cells with round to ovoid nucleus with distinct nuclear membrane, fine powdery chromatin 1 or 2 small nucleoli and ill-defined and scanty cytoplasm

CD99 [Figure 6] weakly positive for NSE, focally positive for WT1 but negative for chromogranin. The tumor had a high proliferation rate with labeling index of 25-30 % on Ki67 immunostaining [Figure 7]. A diagnosis of PNET was made based on the above findings and patient was referred to oncology center for further management.

DISCUSSION

The PNET, first recognized by Arthur Purdy Stout in 1918, is a member of the family of "small round-cell tumors."[3] PNET arising in the kidney was first reported by Mor in 1994.[4]

PNET is an aggressive tumor that tends to recur locally and to metastasize to lymph nodes, lung, liver, bone, and bone marrow which entail a worse prognosis.[1] Patients show a male predominance (58%) with 85% cases being diagnosed during the second to fourth decades.

Grossly, PNET's of the kidney are typically large with 65% measuring greater than 10 cm in diameter with replacement of the kidney.[2] They tend to be grayish in color, encapsulated and contain focal areas of hemorrhage or necrosis. Microscopically, the tumor is composed of lobular

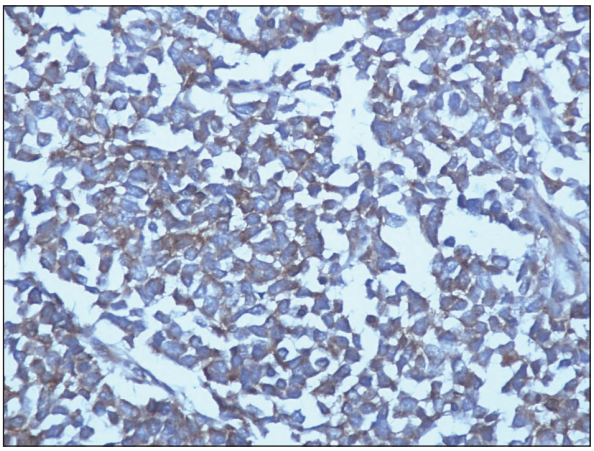

Figure 6: Tumor cells positive for CD99

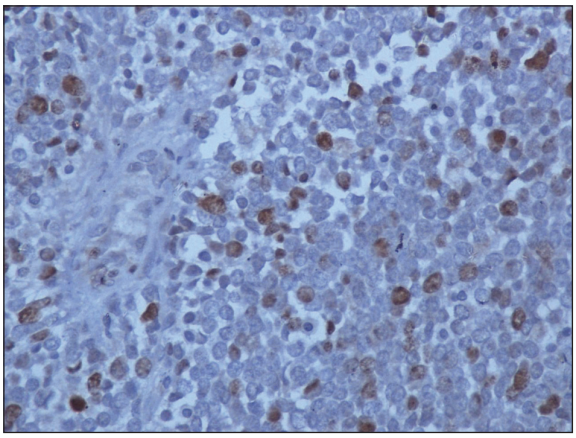

Figure 7: Tumor cells positive for Ki67 immunostaining

pattern of strikingly uniform round cells; individual cells have a round to ovoid nucleus with a distinct nuclear membrane, fine powdery chromatin, and 1 or 2 small nucleoli. The cytoplasm is ill defined, scanty, usually PAS positive and mitosis are variable. Perivascular pseudo rosettes and Homer-Wright rosettes are common.

To better address the diagnosis, an immunohistochemical analysis is necessary. CD99 (the product of MIC2 gene) greatly aids in recognizing the ES/PNET family of tumors.[5] Many PNET also stain for neural markers, including NSE, Leu-7, S-100 protein, synaptophysin, and PGP9.5.[6]

The most frequent translocation in PNET is t(11;22) (q24;q12), detected in approximately 90% of cases, resulting in EWS-FLI fusion gene. Monoclonal antibodies to FLI1 can be used to detect these tumors.[7]

The differential diagnosis includes Blastemal Wilms, carcinoid, neuroblastoma, lymphoma, rhabdomyosarcoma, clear cell sarcoma of the kidney, the small cell variant of osteosarcoma, and desmoplastic small round cell tumor.[8]

The most important differential is Blastemal Wilms which presents with lump abdomen, hematuria. The common age of presentation is 0-5 years. On gross, it may be difficult to differentiate. Microscopically, both are small round cell tumors with high N/C ratio and both may show rosettes. The presence of nephrogenic rests in approximately 25-40% cases in Wilms is helpful in distinguishing between Wilms tumor from PNET. In the remaining cases immunohistochemistry and molecular biology studies are essential to differentiate.

Immunohistochemically, Wilms tumor is WT1 positive and rarely positive for CD99 while diffuse and strong CD99 expression with rare WT1 expression was detected in PNET.[9] Ellison *et al*. reported both CD99 and WT1 expression in seven of 30 primary renal PNETs.[10] When both CD99 and WT1 expression are positive, the distinction of PNET and Wilms tumor may be problematic. Nevertheless, age of the patient, morphological features and aggressiveness of the tumor with diffuse strong membranous CD99 and positive NSE have supported to PNET rather than Wilms tumor. Diffuse nuclear WT1 and focal CD99 expression may be interpreted as Wilms tumor rather than PNET.[9]

Immunohistochemical markers like neuron-specific enolase (NSE), chromogranin, synaptophysin, and vimentin may be useful to differentiate PNET from other differential diagnosis.

Survival rate of renal PNET is poor despite multidisciplinary approach for treatment and most patients die within 1 year of the diagnosis. On the other hand, with current therapies, more than 90% of children with Wilms' tumor are expected to survive 5 years after initial diagnosis.

Hence, PNET of the kidney needs to be kept in mind as a differential diagnosis in young adults presenting with a large kidney mass because it is a highly aggressive tumor and needs multidisciplinary approach for treatment.

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