Primitive Neuroectodermal Tumor of the Kidney: Case Report in a 13-Year-Old Boy

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ABSTRACT
A 13-year-old boy presented with acute right loin pain. Computed tomography scan showed a huge mass originating from the upper pole of the right kidney. Right radical nephrectomy was performed. Histopathological examination revealed a neuroectodermal tumor of the kidney. There was local and lymph node invasion as well as distant metastasis to the lungs, liver and bone. The patient died 18 months from the time of initial diagnosis. Nearly 94% of primitive neuroectodermal tumor shows the complete response to combined surgery, radiation and chemotherapy. However, in the presence of metastases, the 5-year survival drops to 29%.

Key words: Extra osseous, kidney, primitive neuroectodermal tumor, Wilms

INTRODUCTION
Primitive neuroectodermal tumor (PNET) is a small, round-cell neoplasm that represents about 1% of all sarcomas. It affects cerebral and cerebellar tissues and is considered to be a neural crest derivative.[1] PNET was first described by Stout in 1918.[2] In 1975, Seemayer was the first to report PNET in the kidney.[3] It closely resembles Ewing’s Sarcoma and is difficult to be differentiated from it.[4] Renal PNET occurs more frequently during the childhood or adolescence with an aggressive behavior. It often recurs locally and metastasizes early to regional lymph nodes, lungs, liver, bone and bone marrow with poor prognosis.[5] The diagnosis of PNET is based on a combination of electron microscopy, immunohistochemistry, chromosomal analysis, FISH, and light microscopy.[6]

CASE REPORT
This was a case report of a 13-year-old boy who was transferred to the “Emergency Room” with severe acute right loin pain for a few hours. There was no history of a similar attack or any comorbid diseases. The surgical team made a diagnosis of acute appendicitis based on the right iliac fossa pain with rebound tenderness. He was scheduled for appendectomy. Through a Mcburny incision, a normal appendix with hemoperitonium was noticed. A right paramedian incision with hemoperitonium was noticed. A right paramedian incision was performed to detect the source of bleeding. A bleeding right retroperitoneal mass related to the kidney was detected. Evaluation of the retroperitoneal mass by urology team revealed an upper pole right renal mass with infiltration of the posterior peritoneum. A biopsy was taken from the mass, bleeding was controlled and the abdomen was closed. Post-operative computed tomography (CT) scan revealed a huge mass originating from the upper
pole of the right kidney. The mass was composed of two components, a superior large cystic component with a thick capsule and an inferior solid enhancing component. The para-aortic and the retrocaval lymph nodes were enlarged. There was no involvement of the renal vein or inferior vena cava (IVC) [Figure 1]. Magnetic resonance imaging findings were consistent with those of the CT scan [Figures 2 and 3]. Histopathological examination of the mass biopsy revealed malignant changes of unspecified nature. At 2 days later, right radical nephrectomy was performed and the specimen was sent for pathological evaluation. Grossly, there was an upper polar renal 17 cm × 15 cm × 11 cm mass. It was solid, nodular, with areas of necrosis [Figures 4 and 5]. Microscopically, sheets, groups, and cords of neoplastic cells were noticed arranged around blood vessels. The cells had plumped vesicular nuclei, mild pleomorphic inconspicuous nucleoli and frequent mitoses. Some cells had clear and others had eosinophilic cytoplasm with indistinct cell borders. The lymph nodes were invaded by tumor cells. This picture was consistent with a malignant high grade round cell tumor (high monomorphic blast cell Wilms’ versus clear cell sarcoma of the kidney). The tumor was stage III, T2N1M0 tumor). Immunohistochemical staining (CD99) confirmed sarcoma of the kidney. It had features that suggested a PNET. Due to a local and lymph node invasion, the patient was referred to Medical Oncology for chemo-radiotherapy. One year later, the patient had surgical excision of a local recurrence. It was followed by a second course of chemotherapy and radiotherapy. The patient developed local as well as distant bone, lung and liver metastases, and died 18 months from the initial diagnosis.
Primary PNET of the urinary tract is a rare disease with an aggressive behavior and poor prognosis.\(^7\) Due to its morphological resemblance to neuroblastoma, it has been considered to be neural crest derivative.\(^4\) Extracranial PNET can occur at any location in the body including the chest wall (Askin’s tumor), genital tract, retroperitoneal cavity and soft-tissue of the head and neck.\(^8\) It usually occurs during childhood or adolescence and less commonly in young adults, the average age at diagnosis being 27.7 years.\(^9,10\) The symptoms and signs are similar to those of other renal tumors and take an aggressive course.

Radiographic features of PNETs include a large size, lack of extensive parenchyma infiltration, lack of renal vein invasion, diffuse calcification, areas of internal hemorrhage and necrosis, and peripheral hyper-vascularity.\(^11\)

Renal PNET needs to be differentiated from other primary and metastatic renal tumors. The distinction is crucial for determining prognosis. Differential diagnosis includes extra osseous Ewing’s sarcoma, rhabdomyosarcoma, Wilms’ tumor, carcinoid, neuroblastoma, clear cell sarcoma of the kidney, lymphoma, the small cell variant of osteosarcoma, desmoplastic small round cell tumor and nephroblastoma.\(^12\)

Light microscopy shows a tumor of small, round blue cells that lacks markers for lymphoma, neuroblastoma and rhabdomyosarcoma. Cytogenetically 90-95% of the Ewing’s family of tumors have a translocation between the EWS gene on chromosome 22 and the FLI1 gene on chromosome 11 [t(11;22)(q24;q12)] or the ERG gene on chromosome 21 [t(21;22)(q22;q12)].\(^12\)

Treatment of PNET includes a combination of surgery, irradiation, and chemotherapy. Current standard chemotherapy includes doxorubicin, vincristine, and cyclophosphamide alternating with ifosfamide and etoposide. Post-operative radiotherapy may be added when regional lymph nodes are involved.\(^13\)

The few reported cases described variable presentations with aggressive behavior. About 20% of patients presented with metastatic disease. Among those, 44% presented with lung metastases, and 51% had bone or bone marrow involvement (with or without lung metastases). The remaining 5% presented with metastases to other organs.\(^14\) Direct invasion of the renal vein and IVC has also been reported.\(^10\)

PNET is rare and presents an aggressive clinical behavior with worse prognosis.\(^15\) Prognosis is poor if distant metastases are present. Furthermore, palpable tumor masses and synaptophysin expression are associated with a shorter survival.\(^16\) The 5-year relapse free survival for patients with lung metastases was 29%. Major improvements in the long term survival rates will probably not occur until development of better systemic agents or genetic therapy targeted at the unique translocation that produce these malignancies.\(^17\)

REFERENCES


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