CASE REPORT

Primitive neuroectodermal tumor of the kidney in children

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Abstract

Objectives: to characterize primitive neuroectodermal tumor of the kidney as a differential diagnosis for Wilms tumor, and to emphasize the severity of the disease.

Methods: we report the case of a patient with a diagnosis of primitive neuroectodermal tumor of the kidney who underwent nephrectomy and complete tumor resection combined with chemotherapy.

Results: initially, the response of the patient to treatment was favorable, with a few events associated with agranulocytosis resulting from the chemotherapy. Ten months after the end of treatment, the tumor recurred in the paraspinal cervical region.

Conclusions: primitive neuroectodermal tumors involving the kidney are very rare and extremely aggressive. In such cases, the pathologist has a major role in establishing a final diagnosis through histological and immunohistochemical methods.


Introduction

Extracranial primitive neuroectodermal tumors (PNET) are described as small round cell, malignant tumors that have the neural crest regarded as the most likely progenitor and that arise from outside the central nervous system.1 Other terms used for PNET are peripheral neuroepithelioma, adult neuroblastoma, and Askin tumor; the latter involving the chest wall and lung. These tumors are part of the Ewing’s sarcoma family of tumors.2

The peak incidence of PNET occurs within the second decade of life. Recent studies, however, have described cases of PNET within an age range of newborn patients to 14-year old patients and for a median of 1 year of age in a population of 26 patients.2,3

The primary site for PNET is frequently the thorax, either in the intrathoracic or chest-wall areas.3 Other less frequent primary sites are pelvis, retroperitoneum, limbs, neck, and the paraspinal region.4 There are few cases of PNET of the kidney reported in the literature. Moreover, these studies have described PNET of the kidney as a rare entity.5,6 Cases of PNET usually present metastases to the bones, bone marrow, and lungs.4

PNET has been diagnosed with increased precision by immunohistochemical technique employing the MIC2 monoclonal antibody.7 PNET can be confounded with sarcomas and neuroblastomas negative for urinary catecholamine due to the histopathologic, immunohistochemical, and cytogenetic similarities found in Ewing’s sarcomas.8

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VDK, a 9-year old male patient presented with history of abdominal pain and increase in abdominal mass around 25 days prior to presentation and with progressive characteristics. In addition, patient also presented a loss of weight of 5 kg during this period. VDK was negative for family history of neoplasia and of morbid events without presenting any differentiating characteristics.

At physical examination, patient presented with regular overall clinical status and with pale skin; patient was also eutrophic and afebrile. Patient abdominal mass was painful to the touch, in flank topography in the right hypochondrium, firm, and attached to the right kidney. No other alterations were verified during physical examinations.

We carried out laboratory and X-ray examinations. Ultrasonography and tomography exams of the abdomen indicated voluminous tumoral mass in the right hypochondrium that was adherent to the kidney, had heterogeneous echogenicity, and exhibited rough calcification with approximately 15 x 13 x 14 cm in diameter, thus exceeding the medial line. Patient presented normal for other additional examinations.

Our patient was submitted to surgical procedure of nephrectomy and complete tumor resection. The tumor presented the macroscopic features of a Wilms tumor.

Initial anatomicopathological diagnosis was of a small-cell, malignant neoplasia infiltrating the kidney (Figure 1) with a focus of capsular invasion, tumor-free vascular and urethral margins, infiltration of hilar fat by neoplasia, and tumor-free paraaortic lymph nodes.

We carried out an immunohistochemical study using the avidin-biotin peroxidase method. The study showed expansile infiltration into the kidney by neoplasia constituted of trabeculae and small-cell colonies, undifferentiated cytoplasm, oval and hyperchromatic nuclei with rough chromatin, and mitosis. Neoplastic cells were negative for vimentin, cytokeratin, enolase, and leukocytic antigens but positive for MCI2 (Figure 2). A final diagnosis of PNET was established.

Patient was treated according to the P6 protocol suggested by the Memorial Sloan-Kettering Cancer Center, which consists of seven courses of chemotherapy with vincristine, doxorubicin, high doses of cyclophosphamide, ifosfamide, and etoposide.9

Patient presented clinical development that was favorable to this type of treatment despite episodes of intercurrence related to agranulocytosis as a result of high doses of chemotherapy.

After then months of treatment, patient presented relapse in the paraspinal cervical region and abandoned treatment.

Discussion

PNET is a malignant neoplasia uncommonly found in children, it is highly aggressive and presents high rates of relapse. PNET of the kidney should be included in the differential diagnosis of renal tumors, particularly in patients who present with advanced disease at diagnosis. It is important to distinguish PNET of the kidney from Wilms tumor since they require different therapeutic conduct and present different treatment outcomes.6

As to what concerns the histology of the tumor, it is also important to include small-cell renal carcinoma in differential diagnosis of neuroblastoma. PNET of the kidney presents as a small-round-cell tumor without any tubular or glomerular differentiation and with possibility of presenting areas with small cell aggregate formation. In PNET, electron microscopy examination can present neurosecretory granules and immunohistochemical reactions for neuro-specific enolase, chromogranin A, synaptophysin, and MIC2 are positive.10-13
Cytogenetic alterations observed for PNET have been described as identical to those observed for Ewing’s sarcoma, sharing a unique and specific t(11;22)(q24;q12) chromosomal translocation. This alteration was observed in approximately 90% of patients.14,15

Poor-risk for PNET is indicated by tumor volume more than 100 cm³ and over 5 to 10 cm in extension; metastatic spread to the lungs or bone marrow; high levels of lactic acid dehydrogenase; and tumors in adolescents and young adults.9

The recommended treatment for PNET is surgical resection of the tumor associated with chemotherapy and radiotherapy treatment. Current protocols of chemotherapy indicate the use of courses of high-doses of cyclophosphamide associated with vincristine and doxorubicin, alternating with courses of ifosfamide and etoposide.9 Centers of reference for oncologic treatment have described the use of high-dose chemotherapy together with autologous bone marrow transplantation for improved effectiveness of results.16

It is understood that the PNET of the kidney may have a different presentation from other PNETs. The PNET of the kidney is characterized by aggressive clinical behavior, which in turn makes it similar to malignant rhabdoid tumor. Though uncommon, PNET of the kidney should be included in differential diagnosis of childhood and adolescence kidney tumors. Patients with PNET of the kidney usually present with advanced disease and have poor response to treatment.6

References


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