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Letter to the Editor: Primitive Neuroectodermal Kidney Tumor

To the Editor: Primitive neuroectodermal tumors (PNET) and Ewing sarcoma (ES) belong to a group of neoplasms defined by neuroectodermal differentiation and a characteristic cytogenetic translocation, t(11;22)(q24;q12) or gene rearrangements between chromosomes 21 and 22 [1]. They are generally aggressive tumors that present as metastatic disease in nearly 50% of the cases. ES is frequently a bone disease, whereas PNET can occur in bones, soft tissues, or any other site. Renal PNETs are extremely rare, with only a few cases reported [2]. We here record an adult with renal PNET and bone metastases at diagnosis. Because these tumor can also be found in children [3] our experience may therefore be helpful to pediatric oncologists.

A 38-year-old male with no medical antecedents consulted in November 1997 with a 6-month abdominal history of pain in the upper and mid-left quadrants. Physical examination showed what appeared to be a painful, enlarged spleen. Computed tomography (CT) and magnetic resonance imaging (MRI) revealed a 120×120-mm tumor mass involving the left kidney. A technetium bone scan showed an enhanced, infiltrative vertebral lesion (T-10). A radical left nephrectomy was performed on 12/17/97; pathologic examination discovered a renal PNET, confirmed by the detection of the translocation t(11;22) in tumor cells.

After surgery, external beam irradiation was delivered to the tumor bed and bone lesion (50 Gy, 180 cGy five days each week). Simultaneously, systemic EVAIA chemotherapy was started (etoposide 120 mg/m^2 on days 1–3, vincristine 2 mg total dose on day 1, doxorubicin 20 mg/m^2 on days 1–3 alternating in cycles with dactinomycin 0.5 mg/m^2 on days 1–3, and ifosfamide 3,000 mg/m^2 on days 1–3). The patient received 9 cycles. On November 1998, he underwent high-dose chemotherapy (carboplatin, etoposide, and cyclophosphamide) and autologous peripheral blood stem-cell transplantation as consolidation of a complete response.

On March 1999, the patient was readmitted with left shoulder pain and multiple bone lesions were found. He then received palliative irradiation and salvage chemotherapy with high-dose ifosfamide and doxorubicin. The treatment was stopped after 4 cycles due to myelotoxicity and absence of clinical response; three months later he developed dyspnea. A CT scan revealed multiple lung and liver metastases, with pleural effusions. Shortly after, he developed spinal cord compression with paraplegia and died from tumor progression.

Genito-urinary tract PNETs are rare, with isolated cases reported in children as well as adults of renal, uterine, and epididymal involvement [2,3]. Almost 30% of all newly diagnosed cases present with distant metastases, thus showing the biologic aggressiveness of the disease. The most frequent metastatic sites are the lung, bone, and bone marrow.

This case is representative of the clinical course of metastatic PNET with poor prognostic factors (bulky primary disease and bone lesions). With aggressive, combined modality treatment (surgery, irradiation, chemotherapy, and autotransplant), a complete response was achieved. However, the patient soon developed a systemic relapse with a poor response to salvage therapy. We conclude that patients with the Ewing family of tumors and poor prognostic factors must be treated with curative intent, even when the primary site is atypical.

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