Primitive Neuroectodermal Tumor Arising From the Kidney

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Primitive neuroectodermal tumor is a type of sarcoma that occurs in the first two decades of life.[1] It usually appears as a soft-tissue mass in the chest wall and paraspinal region. This tumor has been said to be the soft-tissue equivalent of the Ewing sarcoma and malignant small cell tumor of the thoracopulmonary region.[2] It usually arises from tissue of the chest wall and thoracopulmonary paraspinous musculature. Such a tumor arising from the kidney is quite rare, and few cases have been reported. The treatment is surgical removal and combined radiation and chemotherapy. The case described here is that of primitive neuroectodermal tumor arising from the kidney in a 25-year-old woman who had abdominal pain and a mass. This case is a rare presentation of this tumor as an organ-based neoplasm.

Case Report

A previously healthy 25-year-old woman came to the emergency department complaining of abdominal pain of 2 weeks' duration. The pain had worsened during the preceding 2 days. She had no fever, chills, nausea, vomiting, diarrhea, or change in bowel habits. The pain was described as constant, with no change related to eating. The patient described the pain as initially a sharp, stabbing pain, with some radiation to the back. As the course of the illness progressed, the pain became dull and constant. For the past 2 to 3 days before her visit, the pain has been constant with no relieving or exacerbating factors. This patient sought treatment because of worsening pain, progressive discomfort, and inability to obtain relief by such over-the-counter medications as antacids and acetaminophen. The patient had no history of any intraabdominal disease. She denied any hematuria, dysuria, pain, or burning on urination; bloody stools; or melenic stools. She denied any possibility of pregnancy.

Her medical history was notable for two abnormal Papanicolaou smear results within the last year; however, the patient stated that she had not had any follow-up care or evaluation. She admitted to smoking one-half pack of cigarettes per day (7.5 pack years) and admitted to alcohol intake of five to six beers a day. She denied use of recreational drugs. She had history of Legg-Calvé-Perthes disease as a teenager (bacterial disease caused by infection with *Legionella pneumophila*). She had no previous history of an operation.

On physical examination her blood pressure was 130/59 mm Hg, pulse was 92 beats per minute, respirations were 18/min, and temperature was 37.3°C. Findings on examination of her head, eyes, ears, nose and throat were unremarkable. Her neck was supple and without adenopathy. Her heart was regular and without murmurs, and a pulmonary examination showed equal breath sounds bilaterally. Her abdomen was soft. A mass in the left upper quadrant filled the entire left side of the abdomen and was palpable to the pelvic rim. It was fixed, tender, and without any associated peritoneal signs. Initially the mass was estimated to be approximately 10 cm long, extending from the left subcostal margin to the anterior superior iliac crest on the left side. The mass did not cross the midline, but extended laterally to the anterior axillary line. As the mass was palpated inferiorly, it became more difficult to follow, presumably because it became a deeper structure. There was no well-defined inferior border. Bowel sounds were normal. There were no inguinal nodes. Findings of a pelvic examination were normal, with no cervical discharge. Normal menstrual flow was observed. The uterus was not enlarged, and there were no adnexal masses or tenderness.

Her laboratory values were as follows: sodium 141 mEq/L, potassium 3.9 mEq/l, chloride 108 mEq/L, carbon dioxide content 24 mEq/L, glucose 90 mg/dL, blood urea nitrogen 10 mg/dL, and

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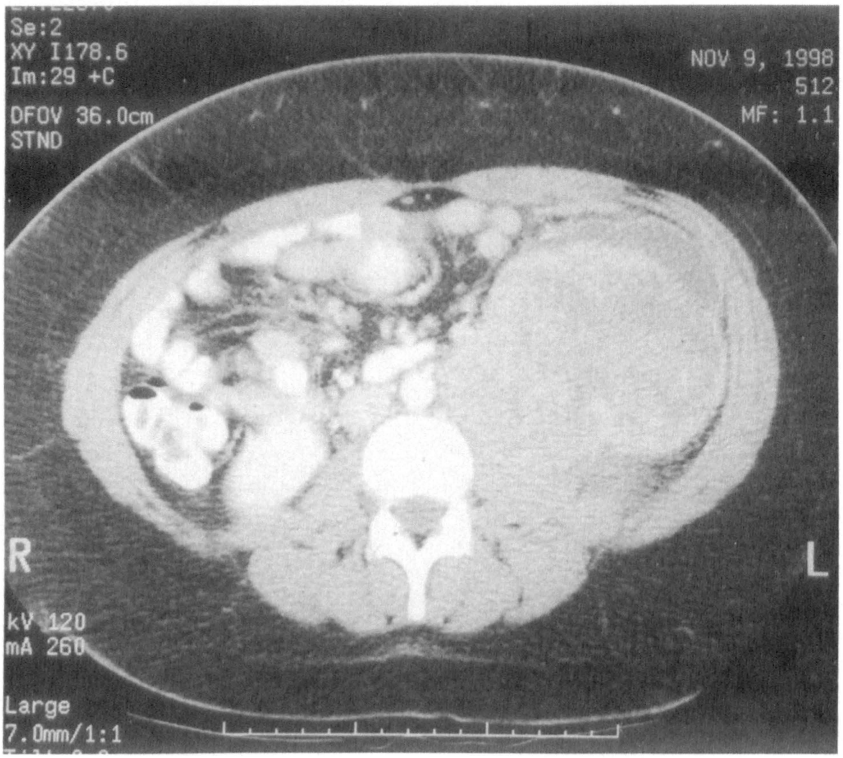

Figure 1. Computed tomogram of the abdomen showing a huge tumor involving the left kidney, filling almost the entire left side of the abdomen with posterior involvement of the psoas muscles.

creatinine 0.7 mg/dL. Her white blood cell count was 9,400/μL, with a hemoglobin of 12.7 g/dL, and a hematocrit of 37.7%. The platelet count was 286,000/μL. A pregnancy test was negative. The urinalysis was negative for any red blood cells, white blood cells, bacteria, glucose, or protein. Urine specific gravity was 1.012. Abdominal films showed a soft-tissue mass in the left side of the abdomen, with no free air, obstruction, or ileus. Computed tomography (CT) of the abdomen with contrast showed a large heterogenous renal 12×12×14-cm mass extending caudally from the mid pole of the left kidney (Figure 1). Given the age of the patient, the mass was strongly suspected to be renal cell carcinoma. The patient was examined by an urologist and scheduled for surgery.

During the operation a 13.5×12.0×9.5-cm tumor was found that had invaded the perirenal fat and involved the margin of resection of Gerota fascia. The lower pole of the kidney was tumor, which also involved the capsule. Extensive necrosis and hemorrhage were found. Histologically the tumor was composed of sheets of small round cells with frequent Homer-Wright rosettes (Figure 2). This pattern and appearance on the initial evaluation raised the possibility of neuroectodermal tumor, because they are characteristic of this tumor. Accordingly, specific immunohistochemical staining was used to confirm the diagnosis. Results were positive using stains and techniques specific for neuroectodermal tumors, specifically cytokeratin and neuron-specific enolase. Stains for synaptophysin and vimentin, which also correlate highly with neuroectodermal tumor, showed weak reactivity. Electron microscopy showed poor ultrastructural preservation.

The diagnosis of primitive neuroectodermal tumor was made based on the strong positive correlation of the microscopic staining and immunohistochemical staining. Three consulting pathologists also came to the same diagnosis in light of the characteristic Homer-Wright rosette pattern and high correlation with confirmation by histochemical staining.

Tumor cells were also found in 2 of 23 lymph nodes submitted, including one para-aortic node. Radiation and chemotherapy were recommended, and further evaluation consisted of a bone scan, which was negative, and a postoperative CT scan of her chest, abdomen, and pelvis, which was negative. A bone marrow evaluation was negative for malignant cells. The patient started radiation and che-

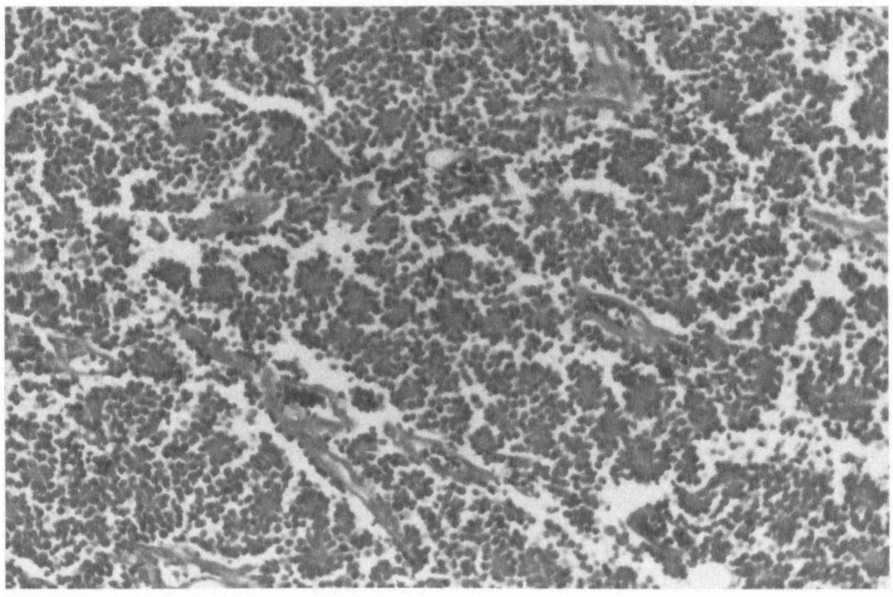

Figure 2. Microscopic section of the tumor showing Homer-Wright rosette formation.

motherapy and was discharged from the hospital. She has subsequently been readmitted to the hospital on two other occasions for complications related to her treatment, including radiation-induced colitis and profound granulocytopenia.

Discussion

Primitive neuroectodermal tumor has been classified as belonging to the Ewing family of sarcomas.[2–4] These tumors were first described by Stout in association with peripheral nerves.[5] Children and adolescents are most frequently affected, with occurrences in the adult years rare. The most common locations are the head, neck, trunk, and extremities.[5,6] These tumors have a poor prognosis, with a disease-free survival of 45% at 7.5 years.[7]

The differential diagnosis of renal tumors in this age-group includes renal cell tumor, Wilms tumor, and lymphoma.[7,8] The diagnosis of primitive neuroectodermal tumor was made in view of the cytologic picture of the surgical specimen, the Homer-Wright rosette formations, and the high degree of correlation with immunohistochemical staining to confirm this particular tumor. The distinguishing genetic factor in primitive neuroectodermal tumor is the association with a translocation between chromosomes 11 and 22, the t(11;22)(q24;q12).[7–10] Poorly differentiated rosettes are found on Homer-Wright staining.[7,10,11] Immunohistochemical staining for cytokeratin and neuron-specific enolase are usually positive, as are stains for vimentin and synaptophysin.[11] All these markers point to a diagnosis of primitive neuroectodermal tumor. It is currently thought that the classification of primitive neuroectodermal tumor should be separate from the Ewing sarcoma, as the prognosis is worse for Ewing sarcoma because it is more invasive.[10]

The treatment of primitive neuroectodermal tumor is surgical resection and a chemotherapeutic regimen consisting of combinations of doxorubicin, cyclophosphamide, vincristine, and dactinomycin. In some cases chemotherapy is started before surgery.[8] Radiation therapy is also used.

Described is a case of a rare tumor of which there have been few previously reported cases.[1,7,10,11] The pathologic picture made a strong case for the diagnosis of primitive neuroectodermal tumor. A review of the literature indicates fewer than 10 previously reported cases of primitive neuroectodermal tumor. This diagnosis should be considered for primary kidney tumor in patients of this age-group. It is interesting to note that this patient had two Papanicolaou smears during the time immediately before the diagnosis of this tumor. It is likely that the tumor might have been palpable during those examinations. Often family physicians have the first opportunity to make the diagnosis of a rare disease by performing careful and thorough examinations during office visits.

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