

Primary Ewing Sarcoma of Kidney in an Elderly

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Primary Ewing sarcoma of the kidney is a very rare neoplasm that generally occurs in young adults. We report a 69-year-old woman with a history of breast cancer who on a routine annual bone scan was found to have a suspicious area over the right kidney. Computed tomography revealed a 16 by 15-cm mass that was removed by a radical nephrectomy. Pathologic examination confirmed the diagnosis of Ewing sarcoma. Three months later, the patient developed recurrence of the tumor at the nephrectomy site that significantly regressed after chemotherapy.

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INTRODUCTION

Ewing sarcoma is the second most common malignant bone cancer, comprising approximately 10% to 15% of all bone sarcomas. Eighty percent of patients are diagnosed before the age of 20 years. Men are more commonly affected than women. Ewing sarcoma is most commonly found in the trunk and long bones, especially in the pelvis and femur.^{1,2} Primary Ewing sarcoma/primitive neuroectodermal tumor (ES/PNET) of the kidney is a very rare and aggressive neoplasm with only a small number of cases reported and generally in young adults. Hereby, we present a case of primary Ewing sarcoma of the kidney that was incidentally found in a 69-year-old woman who was undergoing routine bone scan for her previously diagnosed breast cancer.

CASE REPORT

The patient was a 69-year-old white woman with a history of moderately differentiated infiltrating ductal carcinoma of the breast diagnosed in 2001, which was treated with lumpectomy, radiation, and chemotherapy, and 5 years of tamoxifen followed by exemestane. The patient had remained in remission. She also had history of hypothyroidism, diabetes mellitus, hypertension, asthma, and sleep apnea. She reported no weight loss, fever, abdominal pain

or fullness, or gross hematuria. Her family history was only positive for heart disease and prostate carcinoma. She denied any tobacco, alcohol, or intravenous drug use.

In January 2008, her routine annual bone scan showed a suspicious area in her right kidney (Figure 1). A subsequent computed tomography revealed a 16 × 15-cm heterogeneous multilobulated right renal mass (Figure 2). At this time, her examination revealed a morbidly obese female with blood pressure of 165/70 mm Hg, heart rate of 86 beats/min, body temperature of 37.4°C, and body weight of 94.5 kg. The examination of the heart, lungs, abdomen and extremities were unremarkable. Laboratory examination revealed a blood urea nitrogen of 18 mg/dL; serum creatinine, 1.5 mg/dL; hemoglobin, 11.5 g/dL; and normal mean cell volume. Urinalysis showed no microscopic hematuria.

In February 2008, the patient underwent right radical nephrectomy. The gross overall specimen weighed 2425 g and measured 21.0 × 17.0 × 13.0 cm. A variegated pink-tan to brown-tan irregular lobular tumor, measuring 18.0 × 16.5 × 13.0 cm, with areas of necrosis arose in the mid portion of the kidney and extended past the capsule into the perinephric fat. It did involve the renal vein. Microscopic examination showed monomorphic population

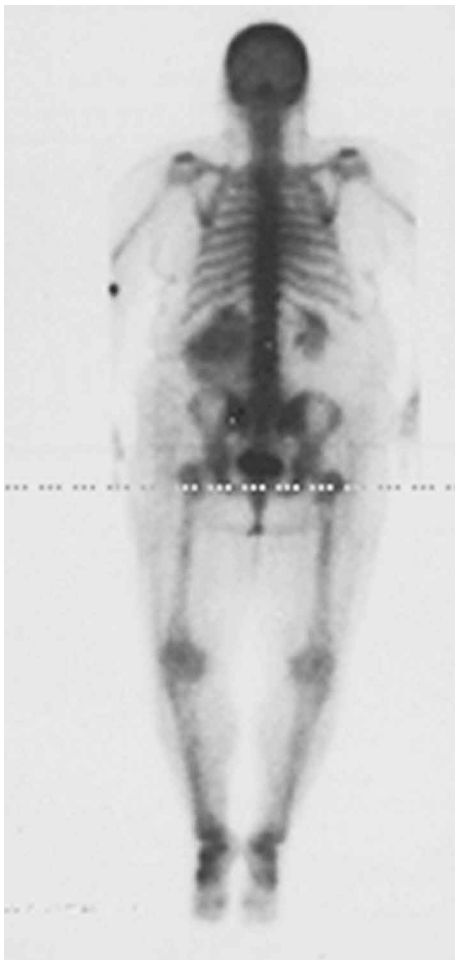


Figure 1. Abnormal soft tissue uptake in the inferior lateral aspect of the right kidney highly suspicious for malignancy.

of small blue cells with variably conspicuous nucleoli and scant cytoplasm. Abundant Homer-Wright rosette formation and tumor necrosis were identified (Figure 3). No lymphatic invasion was found. Immunohistochemistry was strongly positive for CD99, INI-1 (BAF-47), and vimentin (Figure 3).

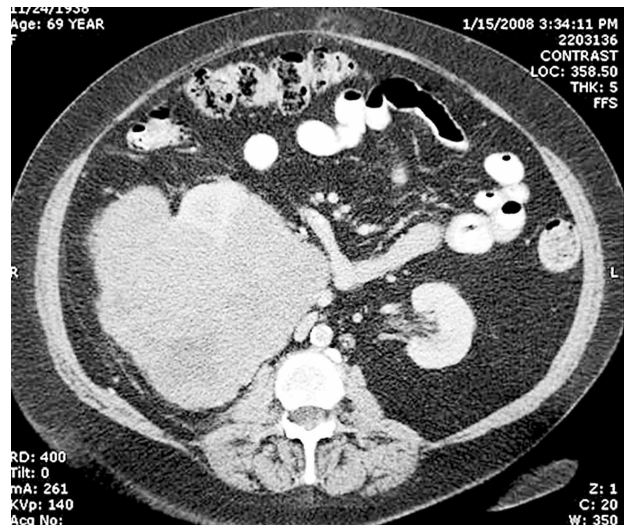


Figure 2. Pre-operative computed tomography of the abdomen showing a 16 × 15-cm heterogeneous multilobulated right renal mass.

The tumor cells were negative for chromogranin, myogenin, MYO-D1, WT-1, cytokeratin, AE1 / AE3 keratin, and CD45. The final diagnosis was ES/PNET pT3aNxMx, AJCC stage 3. A subsequent FISH analysis showed one splitting signal indicative of a rearrangement involving the *EWSR1* gene in 80% of cells. The remaining 20% showed normal signal patterns.

In March 2008, the patient had a follow up positron emission tomography of the abdomen, which was unremarkable. In May 2008, she presented with a 10-day history of right lower abdominal and flank pain. Positron emission scan showed a recurrence of the tumor in the right nephrectomy site with peritoneal metastases (Figure 4). Chemotherapy with ifosfamide, doxorubicin, cyclophosphamide, vincristine, and etoposide resulted in significant regression of the tumor (Figure 5).

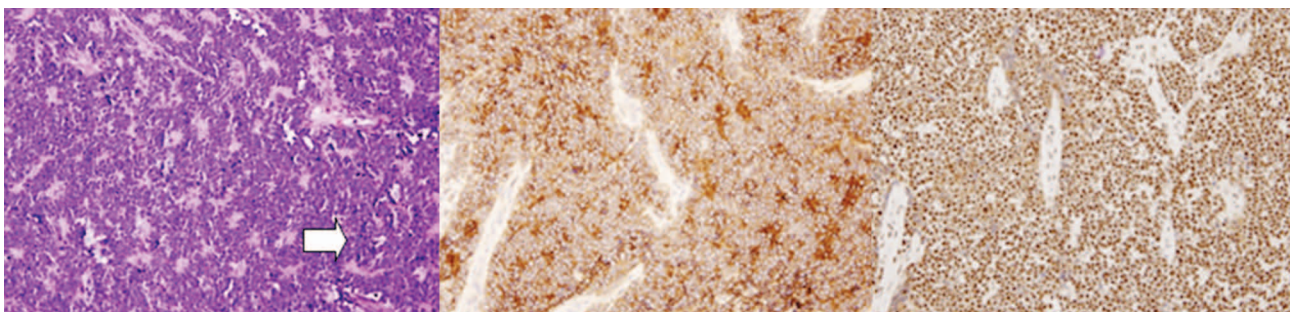


Figure 3. **Left,** A Monomorphic small round blue cell tumor with many Homer-Wright rosettes (arrow) and tumor necrosis (mid bottom; hematoxylin-eosin). **Middle,** On Immunohistochemistry, the tumor cells show strong membranous staining for CD99. **Right,** nuclear staining for INI-1 (BAF-47).

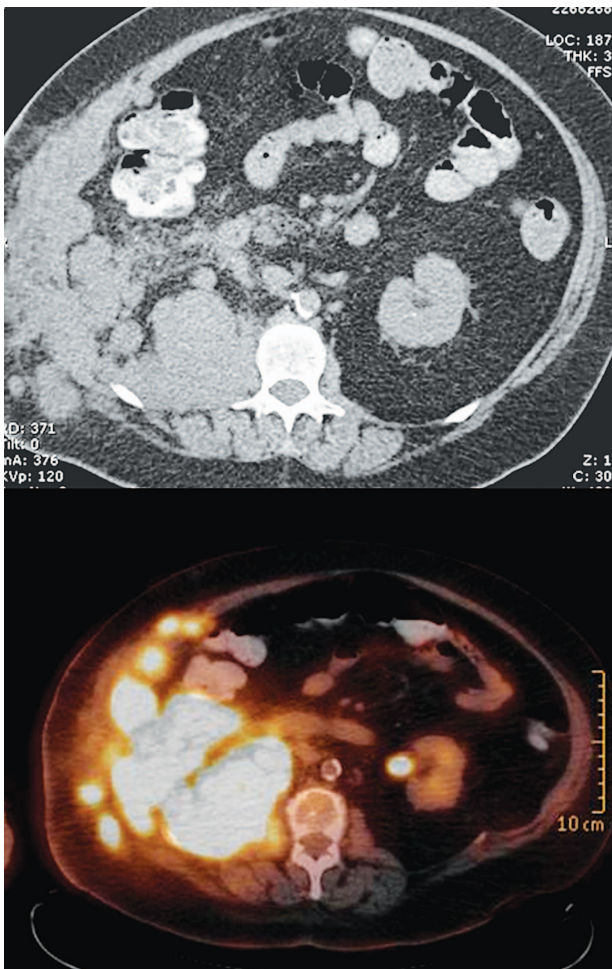


Figure 4. Top, Three months after a right radical nephrectomy, a computed tomography of the abdomen shows multiple nodular masses of various sizes in the surgical bed with involvement of the peritoneum, muscle and subcutaneous tissue of the right lateral abdominal wall. Bottom, positron emission scan shows fludeoxyglucose-avid masses in the right nephrectomy bed, anterior abdominal wall and peritoneum highly suggestive of recurrence of the tumor and metastatic disease.

DISCUSSION

Ewing sarcoma is a high-grade malignant tumor that belongs to the family of small-round cell tumors, “the Ewing family of tumors,” that are derived from neuroectodermal cells. They are localized in bone, soft tissue, and visceral (intra-abdominal, intrathoracic, and urogenital) organs. Primary Ewing sarcoma of the kidney is a very rare neoplasm with a mean age at presentation between 28 and 36 years (range, 4 to 73 years).³⁻⁵ Patients usually present with abdominal and flank pain or hematuria.⁶ Computed tomography confirms the renal masses and usually suggests areas of necrosis and hemorrhage. The diagnosis can be made based

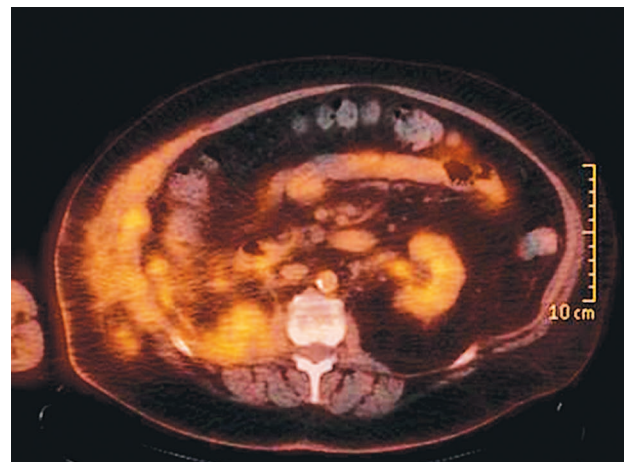


Figure 5. Positron emission scan after chemotherapy shows persistent but decreased fludeoxyglucose uptake in the multiple retroperitoneal and peritoneal right abdominal masses, which have decreased in size.

on positive staining with immunohistochemical markers CD99 and FLI-1. The CD99, FLI-1, and WT-1 markers are important to distinguish ES/PNET tumors from other small blue round cell tumors. Primary ES/PNET of the kidney is an aggressive neoplasm that typically shows poor response to therapy, with a median survival of 16.8 months,⁶ and a 5-year disease-free survival of around 45% to 55%.⁷⁻⁹

In summary, the presented case is an unusual presentation of primary Ewing sarcoma of the kidney in a totally asymptomatic elderly female with normal urinalysis and physical examination.

CONFLICT OF INTEREST

None declared.

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