CLINICAL VIGNETTE

Ewing Sarcoma of the Kidney: Rare Tumor Found on Physical Exam

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Case Report

A 49-year-old female presents to her primary care doctor for her annual physical exam without significant complaints. Her past history is significant only for hypothyroidism on levothyroxine and cosmetic rhinoplasty. She does not smoke, drink alcohol or use illicit drugs. In the prior year she reported blood in the urine and went to a local urgent care and was prescribed ciprofloxacin for a presumed urinary tract infection. She denies any flank pain or hematuria at the time of her visit. On physical exam, her vitals are within normal limits. Physical exam of the abdomen reveals a large firm mass in the right upper quadrant that is nontender to palpation. An ultrasound revealed a "large cystic lesion in the right kidney with irregular mural nodules and associated vascularity, suspicious for cystic renal cell cancer." Follow up CT scan revealed "a 12 x 8 cm cystic lesion within the right kidney with internal soft tissue components along the cyst periphery and minimal enhancement concerning for cystic renal cell carcinoma. No definite renal vein invasion. No definite metastatic disease." Laboratories at the time of her visit included normal CBC, CMP and urinalysis. She is referred to urology for further management and undergoes a right radical nephrectomy without any complications. The final pathology report shows a large 12.5 cm Ewing sarcoma (solid and prominent cystic components with focal infiltration of the renal sinus fat but no necrosis or lymphovascular invasion otherwise). The surgical margins are negative for cancer. She is seen by hematology-oncology and a staging PET CT shows no local recurrence or definite metastatic disease. The patient is started on adjuvant chemotherapy with alternating cycles of VAC (vincristine, adriamycin, cytoxan, mesna) with temodar and Irinotecan. Unfortunately, she only receives 1 cycle of adjuvant chemotherapy VAC regimen due to very poor tolerance with significant weight loss and deterioration of her performance status. She subsequently declined to continue further chemotherapy despite multiple discussions to modify her regimen. The patient also declined surveillance PET CTs and wished to be followed clinically.

Five years after initial diagnosis, she presented back to her primary care doctor for her annual exam. She is doing remarkedly well clinically without any symptoms. Laboratories and urinalysis continue to be normal. The patient continues to decline surveillance imaging.

Discussion

Ewing sarcoma (ES) and peripheral primitive neuroectodermal tumors (PNETs) comprise the same spectrum of neoplastic diseases known as the Ewing sarcoma family of tumors (EFT), which also includes malignant small-cell tumors of the chest wall (Askin tumor). Because of their similar histologic and immunohistochemical characteristics and shared nonrandom chromosomal translocations, these tumors are considered to be derived from a common cell of origin. The Ewing sarcoma family of tumors (EFT) are characterized by distinct nonrandom chromosomal translocations, which all involve the Ewing sarcoma gene (EWS) on chromosome 22. These translocations result in the fusion of distinct genes on different chromosomes, and these fused genes then encode hybrid proteins, which are thought to be involved in tumorigenesis. At least 18 different structural possibilities for gene fusions have been reported in these tumors.[1]

Ewing sarcoma family of tumors most often arises in the long bones of the extremities (predominantly the femur, but also the tibia, fibula, and humerus) and the bones of the pelvis. Ewing sarcomas/primitive neuroectodermal tumors (ES/PNET) of the kidney are very rare and are often high-grade malignant tumors with poor prognosis. Although established treatment guidelines for Ewing sarcoma of kidney are scarce, a multi-modality treatment approach is typically implemented,[2] which includes surgery, adjuvant chemotherapy and sometimes radiation.

In a study of 30 patients between the ages of 8-69 with Ewing sarcomas/primitive neuroectodermal tumors (ES/PNET) of the kidney from 1990-2013, presented event-free and overall survival. Six patients (20%) had tumors confined to the kidney, seven (23.3%) had local tumor extension beyond the kidney, and 17 (56.7%) had distant metastasis at diagnosis. Twenty-five (83.3%) patients underwent radical (19 upfront, 5 delayed) or partial (1 upfront) nephrectomy, 25 (83.3%) chemotherapy and 4 (13.3%) radiotherapy. The 4-year event-free survival and overall survival were 43% (95% CI, 26-61%) and 63% (95% CI, 46-81%), respectively. It reported patients with disease confined to the kidney treated with nephrectomy and adjuvant chemotherapy had favorable outcomes. Local tumor extension beyond the kidney, tumor thrombus, and distant metastasis were unfavorable factors that warrant intensification or novel approaches of therapy. The presence of tumor thrombus in renal vein and/or inferior vena cava was associated with worse event-free survival ($p = 0.053$).[3]

This case illustrates the importance of the physical exam. While the patient did allude to having hematuria the year prior, no imaging was performed for an isolated incident of hematuria. Interestingly at the time of her diagnosis, her urinalysis was completely normal despite her tumor reaching a size of over 12cm. While our patient had disease confined to the kidney, she unfortunately did not complete chemotherapy due to intolerance. She is five years out from her diagnosis with subsequent nephrectomy and continues to do well clinically.

REFERENCES

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