Primitive neuroectodermal tumor of the kidney with level II inferior vena cava involvement

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Primitive neuroectodermal tumor of renal origin, PNET, is extraordinarily rare and often lethal. Here we present a case of renal PNET managed successfully by radical nephrectomy, caval thrombectomy, and retroperitoneal lymph node dissection.

Key Words: primitive neuroectodermal tumors, kidney neoplasms, lymphatic metastasis, nephrectomy, inferior vena cava

Introduction

Primitive neuroectodermal tumor is an extremely aggressive neoplasm which belongs to the Ewing family of tumors.[1] Despite multimodal therapeutic approaches for this neoplasm, prognosis remains poor. The literature on renal PNET with inferior vena cava involvement is particularly limited.

Case report

A 33-year-old Hispanic female presented with a 1 month history of gross hematuria. Medical history, physical examination, and preoperative laboratory values were all unremarkable. She underwent a computerized tomography (CT) demonstrating a 9.5 cm x 5.5 cm x 7 cm right upper pole renal mass with extension into the renal vein and into the inferior vena cava. Figure 1. Magnetic resonance imaging confirmed the 2 cm involvement of the inferior vena cava. Chest CT demonstrated a 6 mm x 8 mm right lower lobe nodule of uncertain significance.

She underwent a right radical nephrectomy, level II inferior vena caval thrombectomy, and interaortocaval / paracaval retroperitoneal lymphadenectomy. Gross and microscopic examination revealed a 10 cm tan focally necrotic mass with extension into the renal capsule. The largest interaortocaval node measured 2.5 cm with a total of 2 out of 20 nodes positive for metastatic primitive neuroectodermal tumor. Immunohistochemical staining for CD99 was strongly positive and focally positive for FLI-1. Immunostaining for WT-1, S-100 protein, chromogranin, etc, were negative strongly supporting the diagnosis of PNET.

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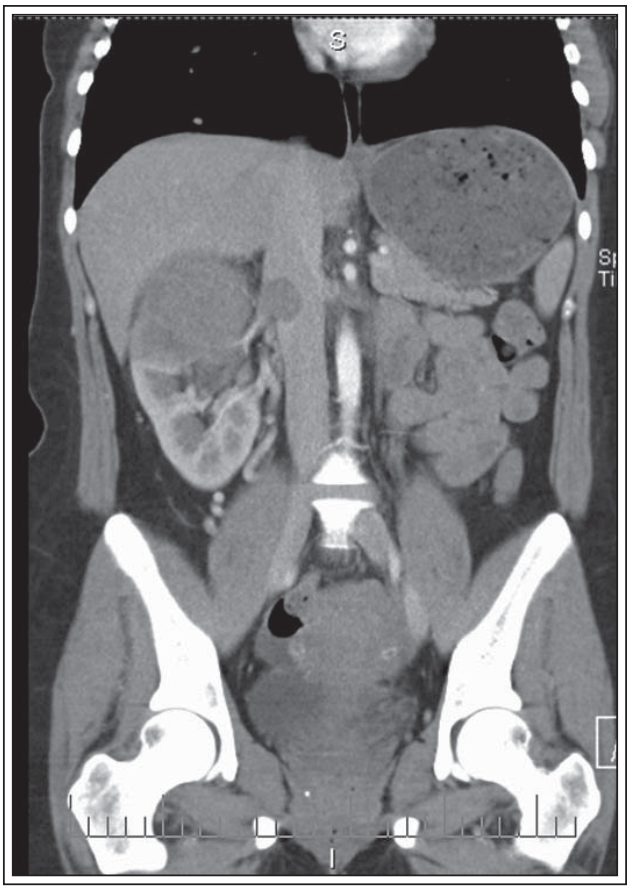

Figure 1. Computerized tomography demonstrating inferior vena cava involvement.

She was started on an alternating chemotherapy regimen consisting of cyclophosphamide/adriamycin/vincristine, and ifosfamide/VP-16, and platinum/ifosfamide. She underwent a PET scan showing no evidence of metastatic disease prior to beginning chemotherapy. The 8 mm lung nodule of uncertain significance had nearly resolved on subsequent CT imaging following one course of CAV. Currently six courses of chemotherapy are complete and she remains disease free at a limited 9 month postoperative follow-up.

Discussion

To our knowledge this case is one of only a handful of renal primitive neuroectodermal tumors in the literature with involvement of the inferior vena cava. Primitive neuroectodermal tumor was first described in the literature in 1918 by Stout and is a member of the small round cell tumors. It generally exhibits neuroepithelial components and is most commonly found in the axial skeleton of adolescents and young adults.[2] PNET of renal origin is exceedingly rare.

In many published reports the average age at diagnosis ranges from 27-33 years of age. Because it belongs to the family of round cell tumors, the immunohistochemical staining plays a pivotal role in diagnosis. Jimenez et al found that the staining for the carboxy-terminus of FLI-1 is sensitive and highly specific for PNET. In addition to this, the surface membrane protein CD-99 is overexpressed in the Ewing family tumors which can assist in confirming the diagnosis.

Renal primitive neuroectodermal tumor is a very aggressive tumor with an overall 5-year disease free survival of 45%-55%.[4] The outcomes are strikingly different when broken down by pathologic stage. Patients with localized disease have an overall survival of 60 months when compared with patients who have disease at regional and/or distant sites in whom overall survival drops to only 15 months.[2] The mainstay of therapy is multiagent chemotherapy and radical surgery. Retroperitoneal lymph node dissection not only plays a pivotal role in pathologic staging, but may also provide therapeutic eradication of all loco-regional neoplasm. □

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