Urology Case Reports 3 (2015) 12–14

Contents lists available at ScienceDirect

Urology Case Reports

journal homepage: www.elsevier.com/locate/eucr

Oncology

Rare Renal Incidentaloma in Pregnancy: An Unusual Primitive Neuroectodermal Tumor Presentation

Gonzalo Torga [a,*], Willis Barrow [b], Misop Han [a]

[a] The James Buchanan Brady Urological Institute, Department of Urology, Johns Hopkins School of Medicine, 600 N. Wolfe Street, Baltimore, MD 21287, USA
[b] Department of Pathology, Johns Hopkins School of Medicine, 401 N. Broadway Street, Baltimore, MD 21287, USA

ARTICLE INFO

Article history:
Received 5 December 2014
Accepted 9 December 2014
Available online 23 January 2015

Keywords:
Primitive Neuroectodermal Tumor (PNET)
Kidney
Renal
Pregnancy
Hodgkin lymphoma

ABSTRACT

Peripheral Primitive Neuroectodermal Tumors (PNETs) are rare lesions that arise from outside the central nervous system and normally do not affect the genitourinary system. Primary renal presentations are extremely rare but given their aggressive behavior and characteristic cytomorphologic and genetic features should be considered well-defined distinct clinical entities in order to distinguish them from other primary tumors featuring round cells in the kidney. We report one case of PNET involving the kidney and associated with pregnancy.

Introduction

Ewing Sarcoma/Primitive Neuroectodermal Tumor family represents 1% of all sarcomas and consists of Ewing Sarcoma (ES), Primitive Neuroectodermal Tumor (PNET) and Askin Tumor. These three distinct entities are characterized by common cytomorphologic, immunohistochemical and molecular pathologic features.[1] PNETs are presumed to arise from the cells of the primitive neuroectoderm located in the neural crest.[2] Tumors located outside the central nervous system, whose most common presentation is as a bone or soft tissue mass in the chest wall or paraspinal regions in adolescents and young adults,[1–3] are categorized as peripheral PNETs (pPNETs). Although very unusual, several genitourinary locations including uterus, cervix, ovary, testis, epididymis, bladder and kidney have been documented.[2] Renal Primitive Neuroectodermal Tumors (rPNETs) have an extremely low incidence and thus most of the literature consists of isolated case reports. The few cases reported described a variable presentation, more aggressive behavior and association with a higher mortality than in any other location.[2] Here we present the case of a 33-year-old pregnant woman with rPNET.

Case presentation

A primiparous 33-year-old woman was in her usual state of health until she presented during the 24th week of her second pregnancy complaining of left groin pain in June of 2014. An ultrasonography examination revealed a large left renal mass, and subsequent MRI demonstrated a 3.5 × 5.2 cm in the mid and lower pole of the left kidney located in the medial and posterior position that exerted a mass effect on the renal sinus without venous involvement (Fig. 1). Past medical history was remarkable for successfully treatment for Hodgkin lymphoma with 6 cycles of ABVD chemotherapy regimen, consisting of Adriamycin, Bleomycin, Vinblastine and Dacarbazine, in 2004 and a follow-up abdominal CT scan from November 2012 could be compared proving the renal mass was not present at that time. A family history of olfactory neuroblastoma was also reported. The patient was referred to our Urology department, and after having thoroughly discussed the likelihood of malignancy, evaluated the options available as well as the associated risks, decided to schedule an elective delivery then undergo radical nephrectomy. A presurgical MRI in August 2014 demonstrated a significant increase in tumor size to 4.4 × 7.2 cm within 42 days (Fig. 1). Hence, at 32 weeks of gestation the baby was delivered and 1 week later the patient underwent robot assisted

* Corresponding author. The James Buchanan Brady Urological Institute, Department of Urology, Johns Hopkins School of Medicine, 600 N. Wolfe Street, Baltimore, MD 21287, USA. Tel.: +1 410 614 4974.
E-mail address: gtorga@jhmi.edu (G. Torga).

2214-4420/
http://dx.doi.org/10.1016/j.eucr.2014.12.004

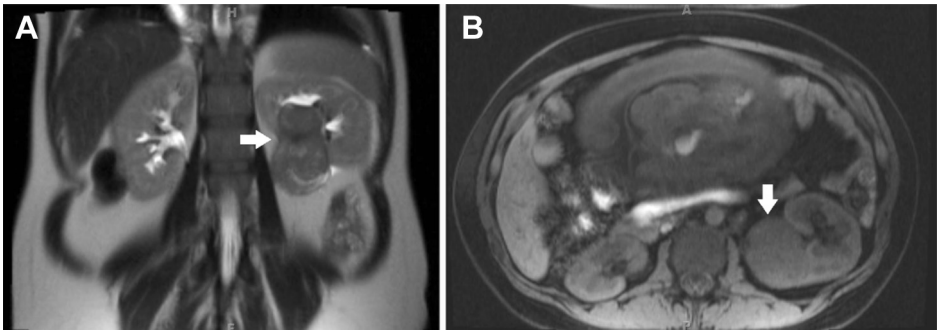

Figure 1. Coronal T2-weighted abdominal magnetic resonance imaging (MRI) demonstrated a large mass (arrows) in the mid and lower pole of the left kidney (A). Axial T1-weighted abdominal MRI revealed the tumor was located in the medial and posterior position and exerted a mass effect on the renal sinus without venous involvement (B).

laparoscopic left radical nephrectomy without lymphadenectomy. Surgical pathological report revealed a small round blue cells tumor of 8 cm consistent with PNET in which the bulk of the lesion was contained within the renal hilum and no extracapsular extension (Fig. 2). Immunochemical studies were performed supporting the previous diagnosis (Fig. 3). At her most recent follow-up in September 2014, 4 weeks after surgery, CT/PET scan results demonstrated no abnormal FDG activity in the remainder of the soft tissue or osseous structures to suggest metastatic disease. The patient was scheduled to initiate adjuvant chemotherapy.

Discussion

Primary renal PNETs are a rare and aggressive entity, usually characterized by an advanced stage at the time of diagnosis.[1] More than 50% of the rPNET cases either develop local recurrence or metastatic disease, with lymph node, liver and lung the most frequent sites.[3] Mean age at diagnosis is 26 years and the 5-year disease-free survival has been reported to be 45–55%.[3] The existence of other primary malignancies featuring round cells, associated with the uncommon nature of these malignancies, can lead to misdiagnosis.[1] rPNETs require a more extensive therapeutic approach as compared to other comparable tumors of the kidney such as Wilms tumor, monophasic synovial sarcoma, lymphoma, clear cell sarcoma of the kidney, small cell carcinoma, neuroblastoma or desmoplastic small round cell tumor.[1] Definitive diagnosis of rPNET should be confirmed based on histologic features that include small uniform round cell morphology associated to a variable level of rosette formation,[3] molecular markers for translocations resulting in a fusion transcript of the EWS gene and the ETS-related family of oncogenes,[1,3] as well as immunohistochemistry staining.[2] This tumor was partially positive for S-100, focally positive for NSE, strongly positive for CD99 and negative for synaptophysin, chromogranin, WT-1, desmin, myogenin, CD45, AE1/AE3 and CAM5.2. Clinical management includes resection associated with adjuvant chemotherapy.[2] The current standard of care remains the intensive multidrug chemotherapy regimen published by Grier et al in NEJM in 2003, which includes 5 cycles of vincristine, doxorubicin and Cytoxan, followed by 7 cycles of ifosfamide and etoposide and 2 cycles of vincristine and Cytoxan.[4] Despite the limited evidence regarding pPNETs available in the literature, several cases associated with pregnancy have been reported in different locations including ovary, cervix, brain and pancreas.[5] One case of rPNET has been previously described in a pregnant woman.[5] This association might suggest a possible effect of hormones on PNETs during pregnancy, but this association still remains unclear. This patient presented with a personal history of Hodgkin

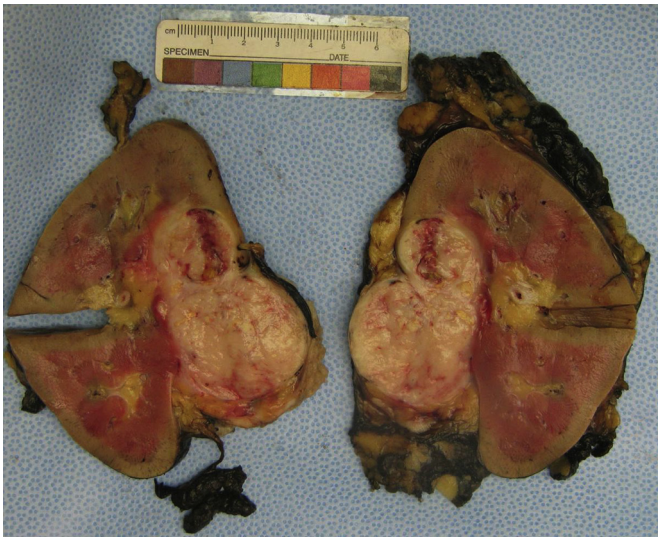

Figure 2. Macroscopic examination of the specimen.

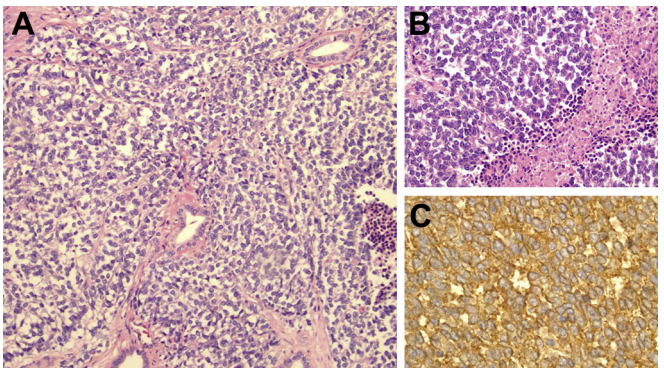

Figure 3. Microscopic examination of tumor tissue showed sheets of tumor cells entrapping tubules (A) composed of monotonous cells with round, vesicular nuclei with fine chromatin, small nucleoli, and indistinct cell borders. Tumor cells in a pseudorosette pattern and areas of necrosis were common within the tumor. Cytoplasmic clearing was also present due to the presence of glycogen (B). Tumor cells exhibited diffuse, membranous positivity for CD99 supporting the diagnosis (C).

Lymphoma as well as family history for olfactory neuroblastoma, another rare entity whose origin is located in the neural crest.

Conclusion

Renal PNETs should be considered in the differential diagnosis of rapidly growing renal mass with aggressive presentation, especially during pregnancy.

Conflict of interest

The authors declare no conflicts of interest.

References

1. Karpate A, Menon S, Basak R, et al. Ewing sarcoma/primitive neuroectodermal tumor of the kidney: clinicopathologic analysis of 34 cases. *Ann Diagn Pathol*. 2012;16:267–274.
2. Thyavihally YB, Tongaonkar HB, Gupta S, et al. Primitive neuroectodermal tumor of the kidney: a single institute series of 16 patients. *Urology*. 2008;71:292–296.
3. Sun C, Du Z, Tong S, et al. Primitive neuroectodermal tumor of the kidney: case report and review of literature. *World J Surg Oncol*. 2012;10:279.
4. Grier HE, Krailo MD, Tarbell NJ, et al. Addition of ifosfamide and etoposide to standard chemotherapy for Ewing's sarcoma and primitive neuroectodermal tumor of bone. *N Engl J Med*. 2003;348:694–701.
5. Wu YJ, Zhu YC, Chen H, et al. Primitive neuroectodermal tumor of the kidney with inferior vena cava tumor thrombus during pregnancy response to sorafenib. *Chin Med J*. 2010;123:2155–2158.