Case Report

Metastatic Primitive Neuroectodermal Tumor of the Prostate: A Case Report and Review of the Literature

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Clinical Practice Points

- Peripheral primitive neuroectodermal tumor (PNET) is a rare malignant neural crest tumor that is now included in the Ewing family of tumors. PNET is specifically thought to be related to Ewing sarcoma, sharing overlapping cytogenetic features and translocation. Although Ewing sarcoma often occurs in the soft tissue and bones of children, very rare cases of PNET in the bladder, prostate, kidney, and adrenal gland have been reported.
- A 38-year-old male presented with a lower abdominal pain and hematuria for 2 months. Computed tomography and magnetic resonance imaging showed a large pelvic mass, suspicious of malignancy with evidence of local invasion and distant metastasis. Biopsy confirmed PNET of the prostate, and the patient underwent chemotherapy and surgical resection with a survival of 14 months.
- Review of the previously reported cases revealed a young age at presentation with a mean ± SD of 28.3 ± 8.0 years (N = 10; range, 20-49 years). Forty percent of the patients presented with metastatic disease, with the lung being the most common site for the metastasis. Ninety percent of patients received chemotherapy, whereas 3 out of 8 patients received radiotherapy, and 50% (4 of 8 patients) underwent surgery. The survival was 10.4 ± 8.6 years (N = 5; range, 2-24 months; median, 10 months).
- PNET of the prostate involves young male adults, may present as metastatic disease, and has poor outcomes, with a median survival of less than 1 year despite multimodal treatment strategy of the combination of chemotherapy, radiotherapy, and surgical resection.

Clinical Genitourinary Cancer, Vol. ■, No. ■, ■-■
Keywords: Ewing family of tumors, Ewing sarcoma, Malignant neural crest tumor, Magnetic resonance imaging, Staging

Introduction

Peripheral primitive neuroectodermal tumor (PNET) is a rare malignant neural crest tumor that is now included in the Ewing family of tumors. PNET is specifically thought to be related to Ewing sarcoma (ES) as they have been found to share overlapping cytogenetic features and translocation of t(11;22)(q24;q12).[1] Although ES often occurs in the soft tissue and bones of children, very rare cases of PNET in the bladder, prostate, kidney, and adrenal gland have been reported.[2-5] To our knowledge, there have been 10 reported cases of PNET of the prostate.[4,6-14] Patients with PNET of the prostate typically undergo a combination of chemotherapy, radiotherapy, and surgery but often suffer poor outcomes. In this study, we present an extremely rare case of metastatic PNET of the prostate with a brief review of the current literature.

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Submitted: Oct 20, 2017; Accepted: Oct 30, 2017

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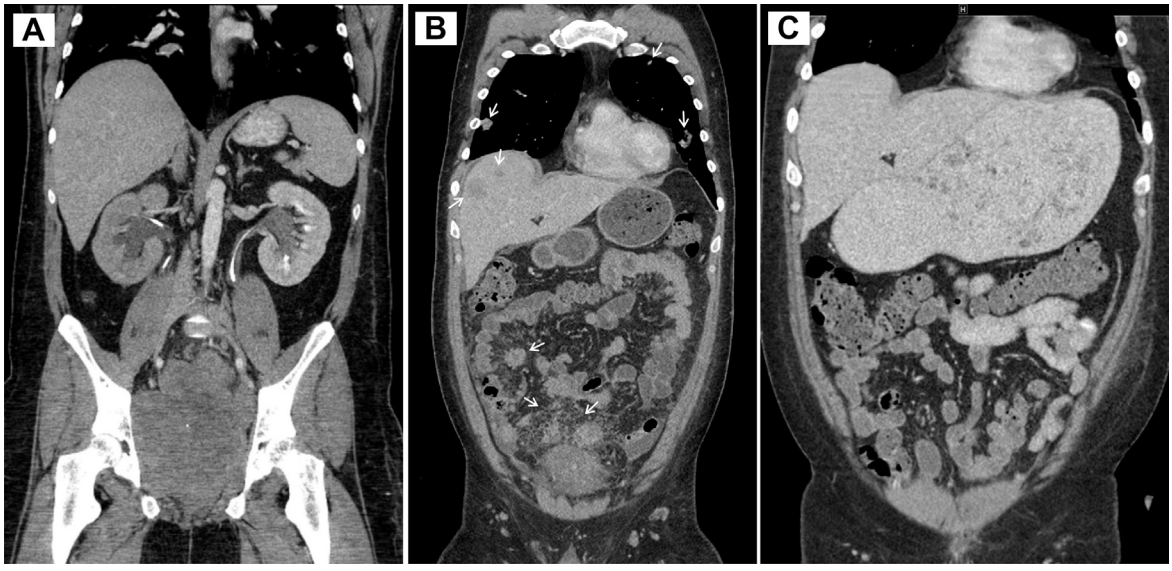
Case Report

A 38-year-old male with no previous history of malignancy presented to the urology clinic at the University of Illinois at Chicago with complaint of lower abdominal pain, constipation, pain with defecation, and hematuria for about 2 months. There was no reported weight loss. Physical examination revealed a soft,

1558-7673/$ - see frontmatter
https://doi.org/10.1016/j.clgc.2017.10.023

Figure 1 Coronal Computed Tomography (CT) of the Abdomen and Pelvis With Contrast at Presentation (A) Demonstrates a Heterogeneous Mass Within the Pelvis Abutting the Urinary Bladder With Bilateral Mild to Moderate Hydronephrosis. Bilateral Ureteral Stents Are Partially Visualized. Coronal Contrast-enhanced CT of the Chest, Abdomen, and Pelvis (B) Demonstrates Multiple Metastases (White Arrows) Within the Lungs, Liver, and Peritoneum. Post-chemotherapy CT Imaging Shows Favorable Response to the Treatment With Improvement of the Pulmonary (Partially Shown), Hepatic and Peritoneal Metastases



nondistended abdomen and no inguinal lymphadenopathy. Digital rectal examination revealed a firm mass fixed anteriorly with distal extension to the superior portion of the ano-rectal ring. Biochemical investigations demonstrated slight elevation of the transaminases and white blood cell counts.

A computed tomography (CT) scan of the abdomen and pelvis with intravenous contrast revealed a heterogeneous mass measuring 11.1 × 11.3 × 14.4 cm arising from the prostate and abutting the bladder. Nodular implants suspicious for metastases were noted on the omentum, peritoneum, and bladder (Figure 1). Magnetic resonance imaging (MRI) of the pelvis confirmed a lobulated mass replacing the prostate and seminal vesicles, occupying a large portion of the pelvis. The mass appeared hypointense on T1-weighted images and heterogeneously isointense to hyperintense on T2-weighted images. Tumor invasion was noted on the posterior bladder wall and right wall of the rectosigmoid colon (Figure 2).

A transrectal biopsy of the mass was performed. Microscopic examination revealed a poorly differentiated neoplasm demonstrating focal neuro-ectodermal morphology (Homer Wright rosettes) as well as focal membranous staining for CD99 by immunohistochemical studies (Figure 3). Molecular pathology analysis revealed an EWSR1/FLI1 fusion transcript confirming a final diagnosis of PNET.

The patient was scheduled for resection of the prostatic mass. However, a colonic perforation occurred while the patient was awaiting the procedure, necessitating a bowel resection with colostomy. The patient subsequently presented to the emergency room with worsening pain, and an abdominal CT and chest radiograph revealed a rapidly growing pelvic tumor with metastases to the liver and lung. Bilateral hydronephrosis was also noted secondary to mass effect causing ureteral obstruction, requiring bilateral nephrostomy. The medical team arrived at a multidisciplinary decision to initiate chemotherapy prior to surgery. Pelvic exenteration was performed after several rounds of chemotherapy with cyclophosphamide, doxorubicin, and vincristine, which was complicated by fever, neutropenia, and recurrent urinary tract infection. Pathology studies of the surgical specimens demonstrated extraprostatic tumor extension to the muscular layer of the bladder, with negative tumor involvement in the peritoneum, small bowel, colon, and ureters.

An aggressive treatment approach was adopted, stem cell harvest was performed, and chemotherapy was reinitiated prior to planned radiotherapy. However, after 6 rounds of chemotherapy, the patient subsequently developed sepsis and ultimately expired after hemodynamic deterioration.

Previous Cases of PNET of the Prostate

Table 1 summarizes the demographics and clinical data of previously reported cases of PNET of the prostate. The mean ± standard deviation (SD) age of the reported cases was 28.3 ± 8.0 years (N = 10; range, 20-49 years). Forty percent of the patients presented with imaging evidences of metastatic disease, with lung being the most common site for the metastasis. The pathologic evaluation was available in 6 cases and demonstrated stage T2b in all cases, defined as having a tumor > 5 cm in largest dimension in the deep tissue. Four of the 10 cases demonstrated findings suspicious for metastases. The pathologic staging in the other 4 cases could not be determined from the reports alone.