

Primary Primitive Neuroectodermal Tumor of the Urinary Bladder: A Case Report of a Rare Pathological Entity with a Rapidly Progressing Clinical Course

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Abstract

Primary primitive neuroectodermal tumors (PNET) are a group of malignant neoplasms composed of sheets of uniform small round cells that exhibit neuroblastic differentiation outside the nervous system. They are closely related to Ewing's sarcoma and predominantly affect children and adolescents. There are few reports of primary visceral PNET's in the literature, most of them in the urogenital organs, and even fewer reports of PNET's of the urinary bladder.

Thus, every case counts in increasing the pool of knowledge as far as the symptoms, risk factors, histopathology and treatment of this rare and aggressive entity. We present such a case.

Keywords: Bladder; Ewing; Neuroectodermal; PNET; Primitive

Case Report

A 76 year old male, with past medical history significant for diabetes, chronic renal failure hypertension and morbid obesity presented to the emergency room with urinary complaints. The patient, who had no urological history thus far, complained of dysuria, dribbling and difficulty to void for the past two weeks. Physical examination revealed a rigid mass palpated in the lower abdomen and the digital rectal exam was normal. Complete blood count showed hemoglobin of 12.6 g/dl, white count of 9.06 k/ul with normal differential. Blood chemistry revealed acute on chronic renal failure with a creatinine value of 4.7 mg% and urea of 217 mg%. Total PSA was 0.03 ng/dl. An abdominal ultrasound revealed bilateral moderate hydronephrosis (Figure 1a and 1b), a urinary bladder with thickened walls hyperechoic fullness and no residual urine (Figure 1c). ANCCCT confirmed the same findings without lymphadenopathy (Figure 1d).

After stabilizing the patient in the following days, a chest CT and bone scan showed no evidence of systemic metastasis. Cystoscopy demonstrated a normal urethra, enlarged but otherwise normal prostate lobes with a small capacity urinary bladder. The bladder walls were filled with solid lobular tumors of different sizes. Numerous biopsies were taken. The patient was discharged 2 days after exhibiting clinical and laboratory improvement.

Pathologic Findings

Microscopic examination revealed fragments of urothelial mucosa extensively infiltrated by a malignant undifferentiated tumor composed of small round blue cells with irregular appearing nuclei, increased mitotic activity and karyorrhexis. By immunohistochemistry, tumor cells showed positive staining for CD99 (O13), CD56, C-Kit (CD117) and negative for vimentin, CLA, CD20, CD3, M-POX, CD34, pankeratin, CK20, CK7, EMA, CEA, S100, panmelanoma, CK5/6, P63, synaptophysin (twice performed) and TTF1. Proliferation index (Ki-67) of the tumor cell nuclei was up to 90% positive. Findings were consistent with Primitive Neuroectodermal Tumor (PNET) (Figure 2).

Clinical Course

The patient returned 10 days later with significant worsening edema of his lower limbs. A cardiac ultrasound showed normal function and there was no evidence of deep vein thrombosis on a lower limb duplex. A CT scan that was performed only 3 weeks after his initial presentation revealed further spread of the mass to the retroperitoneum (Figure 3).



Figure 1: A and B: Abdominal ultrasound showing kidneys of normal size and texture with bilateral hydronephrosis. C: The urinary bladder demonstrates a thickened wall and hyperechoic fullness. D: CT urography confirmed hyperechoic fullness of the urinary bladder measuring 40 HU and bilateral hydronephrosis.



Figure 2: An antegrade pyelography demonstrating both mid ureters blocked.

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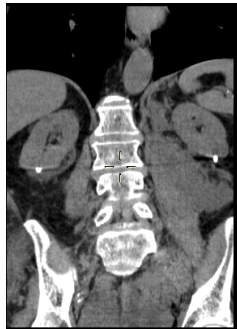


Figure 3: Abdominal non contrast CT demonstrating the tumor's spread to the retroperitoneum, especially on the left.

Due to the rapid deterioration and the pathological diagnosis of PNET that was known at this point the patient was transferred to a tertiary hospital to receive chemotherapy. The treatment included a combination of Doxorubicin Cyclophosphamide and Vincristine. The patient only managed to receive one course of the treatment. In the following few days the patient was oliguric and uremic and underwent several sessions of urgent dialysis. The abdominal mass grew further engulfing the rectum and sigmoidcolon up to S1 and in the retroperitoneum up to the level of both kidneys and intraperitoneally along the mesentery, causing large bowel obstruction. Eventually, the patient died of septic complications only 36 days after his initial presentation to our service.

Discussion

Due to scarce descriptions in the literature it is difficult to establish the way of conduct with this entity as far as diagnosis and treatment, and in this lies the importance of reporting each case. Our patient presented with urinary complaints and bilateral hydronephrosis unlike the majority of the cases described thus far in which the presenting symptom was hematuria [1,2]. He had no recognizable risk factor such as immunosuppression suggested in other cases [2].

As in the other cases the diagnosis was suggested by a poorly defined and aggressive tumor that fills the lumen of the urinary bladder and usually spreads later on to adjacent organs of the pelvis. The diagnosis was established on a typical pathological examination complemented by a typical immunohistochemical staining. As in prior cases, strong staining with CD99 and negative staining for markers excluding other types of small, blue and round cell tumors such as lymphoma and neuroendocrine cancers was very characteristic [3,4]. The patient died before the presence of EWS-FLI1 translocation could be detected by fluorescent in situ hybridization or reverse transcriptase-polymerase chain reaction as occurs in about 95% of the cases.

Due to the close relation to Ewing sarcoma the treatment regimen is combined of radical resection and adjuvant radiotherapy and chemotherapy [5]. As in most cases, our patient most probably already presented with a subclinical metastatic disease and unfortunately he was not fit enough and his disease was too aggressive to allow a comprehensive treatment.

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