Case Report

Primary primitive neuroectodermal tumor of the urinary bladder: A rare case report and review of literature

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ABSTRACT

We report a rare case of primary primitive neuroectodermal tumor (PNET) of the urinary bladder. Primary PNETs of the bladder are very aggressive neoplasms. They are extremely rare and only few cases have been reported in the literature. A 26-year-old female patient who presented to us after excision biopsy is reported here. Initial histopathology examination report said the case to be small cell variant of malignant mesothelioma. Review and immunohistochemistry report confirmed the case as urinary bladder small round cell tumor with tumor cells expressing MIC-2. After excision of the tumor, bladder cystoscopy reveals normal study and patient is undergoing chemotherapy now.

Key words: Primitive neuroectodermal tumor, urinary bladder, PNETUB

INTRODUCTION

A primitive neuroectodermal tumor (PNET) is a malignant small round blue-cell tumor exhibiting a variable degree of neural differentiation, which arises outside the brain, spinal cord and sympathetic nervous system.[1] These tumors are closely related to osseous or extraosseous Ewing's sarcomas, with which they share the same chromosomal abnormality: t (11;22)(q12;q24).[2] In general, a PNET is a very aggressive tumor with rapid local infiltration combined with widespread metastasis.[3,4] Its aggressive behavior is reflected by a low 5-year survival rate ranging from 60% to 90% when treated with surgical resection in combination with radio-chemotherapy.[4] Recently such rare tumor with a highly malignant potential have been increasingly reported in various organs, including kidney[4] and prostate[5] in the field of urology. PNET of the urinary bladder is extremely rare, with only 11 articles [Table 1] previously reported to the best of our knowledge. Herein, we present an additional case of a patient and review of the literature.

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CASE REPORT

The present case report is about a 26-year-old female patient who presented with a history of lower abdominal pain and distension for the duration of 6-7 months. Magnetic resonance imaging lower abdomen showed evidence of cystic (hypointense on T1WI, hyperintense on T2WI) lesion with thin wall except some nodular and cystic component in the antero-inferior wall of the lesion; no fluid level/obvious calcifi cation/bleeding was seen [Figure 1]. It measured 10 cm^3 × 18 cm^3 × 17 cm^3 in dimensions. Mild fluid was seen in the pelvic cavity. Impression was (?) cystic mesothelioma. Patient then underwent exploratory laparotomy followed by excision biopsy. As per operative note it was completely removed and no vascular or adnexal invasion was found. Initial histopathology examination report revealed tumor composed of small round cells with fine chromatin, inconspicuous nucleoli and scanty cytoplasm. Large areas of necrosis and a peritheliomatous pattern of growth around blood vessels were seen [Figure 2]. Also pseudorosette can be seen in the background of small round ablue cells [Figure 3]. Possibility of small cellvariant of malignant mesothelioma was suggested. Further review of the report and immunohistochemistry

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Table 1: Reported cases of primary neuroectodermal tumor of the bladder

Author (journal)	Age/ sex	Chief complaint	Therapy	Follow up	Immunohistochemical study CD99 S100 PSA NSE CK
Banerjee SS (Histopathology, 1997)	21/M	Microscopic hematuria	Total Cystectomy chemotherapy	NED: 18 months	(++ (-) (+) (+/-) (-)
Angelo E (J Urol, 1997)	15/F	Gross hematuria	Partial Cystectomy Chemotherapy	NA	(++) (-) (+) +/-) NA
Mentzel T (Pathology, 1998)	62/M	High fever attack	TURBT	DOC: 3 weeks	(++) (+/-) (+) (+/-) (-)
DesaiS (Histopathology, 1998)	38/F	Gross hematuria	Total cystectomy	NA	(++) (-) (+) (++) (-)
Colecchia M (Histopathology, 2002)	61/F	NA	No treatment	NA	(++) (-) (+) NA (-)
Kr ger S (Pathology, 2003)	81/M	Urge incontinence	TURBT	DOD: 2 weeks	(++) (-) (+) (++) (-)
Lopez-Beltran A (J Clin Pathol, 2006)	21/F	Gross hematuria	Total Cystectomy chemotherapy	NED: 3 years	(++) (+/-) (+) (-) (+/-)
Elinger J (Urology, 2006)	72/M	Gross hematuria	TURBT	NA	(++) NA (+) NA NA
Okada.Y Int J clin oncol 2011	65/M	Hematuria dysuria	TURBT	DOD: 8months	(+) NA NA NA NA
Zheng Y Medical oncology, 2011	74/M	Gross hematuria Dysuria	Palliative chemotherapy	DOD: 4months	(++) NA NA NA (-)
Y.Kobayashi Journal of Solid Tumors, 2012	85/M	Gross hematuria	TURBT	DOD:3months	(++) (+) (+) (+) (-)
Our case	26/F	Lower abdominal discomfort	Total cystectomy Chemotherapy	Treatmaent going on	(++) (-) NA NA (-)

TURBT: Transurethral resection of bladder tumor, DOC: Died of other cause, NA: Not available, NED: No evidence of disease, DOD: Died of disease, PSA: Periodic acid-Schiff, NSE: Neurone specific enolase, CK: Cytokeratin

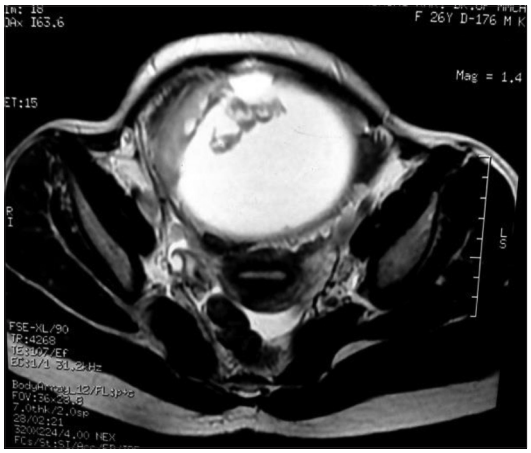

Figure 1: Magnetic resonance imaging showing the bladder cyst

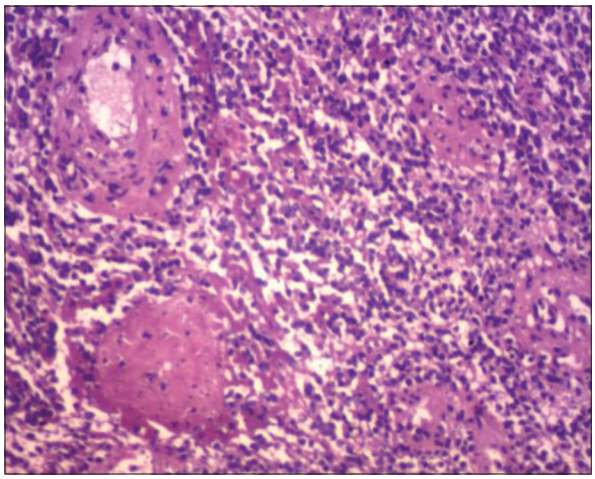

Figure 2: Peritheliomatous pattern of growth around blood vessels

(IHC) study showed it as a case of small round cell tumor with tumor cells expressing MIC-2 and was negative for chromogranin-A, cytokeratin, epithelial membrane antigen, S-100 protein, desmin and leukocyte common antigen. Present ultrasonography whole abdomen and chest X-ray study are absolutely normal. The patient is undergoing course of chemotherapy now.

DISCUSSION

A PNET of the urinary bladder is extremely rare, with only 11 cases previously reported, to our knowledge. A review of those previous reports including the present case is shown in Table 1. The affected patients were seven men and five women, with a mean age of 50.7 years old (range: 15-85). Surgical treatment was performed in 10 cases (total cystectomy in 4, partial cystectomy in 1, transurethral resection of bladder tumor [TURBT] only in 5). Adjuvant chemotherapy was given in four cases. Two patients remained free of disease at follow-up examinations performed at 18 months and 3 years, respectively. In contrast, four patients treated by TURBT showed rapid recurrence and thereafter died of tumor. Therefore, a total cystectomy with adjuvant chemotherapy seems to provide long-term survival for PNET of the urinary bladder. The recommended chemotherapy regimen consists of vincristine, doxorubicin and cyclophosphamide, alternating

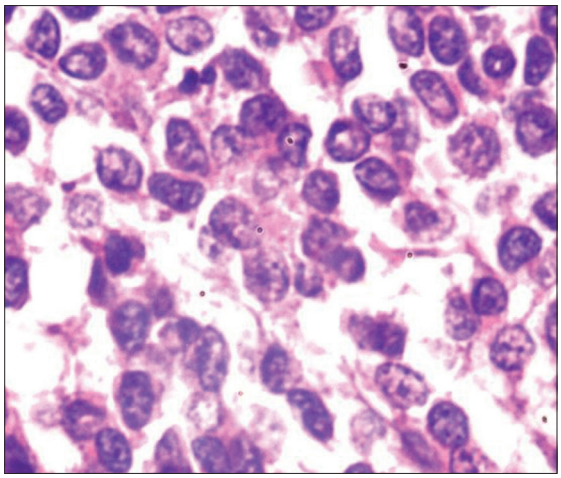

Figure 3: Small round blue-cells and rosettes

with ifosphamide and etoposide.[6,7] Histologically, PNET is a malignant small round blue-cell tumor that exhibits a variable degree of neural differentiation, though all points on the spectrum share a predominantly lobular growth pattern.[8] For IHC analysis, the most useful neural markers are neuron specific enolase, vimentin, S-100 and synaptophysin, with staining detectable in up to 60% of the cases.[9] Further, the tumor cells show a strong expression of MIC-2 protein (CD99).[10] In order to qualify for the designation of PNET, it is suggested that the tumor must show rosettes and be positive for at least two of the neural markers.[8] The detection of EWS-FLI1 type I fusion also allows us to exclude difficult differential diagnoses that cannot be separated immunohistochemically.[11] In the present case, positive staining for CD99 was helpful for the diagnosis of PNET.

CONCLUSION

We report the 12th known case of PNET of the urinary bladder. Diagnosis of this type of tumor and effective therapy must be conducted as soon as possible because of its highly aggressive behavior. Our patient underwent adjuvant chemotherapy with vincristine, doxorubicin and cyclophosphamide, alternating with ifosphamide and etoposide and at 12 months follow-up period, she is disease free.

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