Letter to the Editor

Ewing's sarcoma/primitive neuroectodermal tumor (ES/PNET) of the bladder in an adolescent: a case description

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Introduction

Extraosseous Ewing's sarcoma/primitive neuroectodermal tumors (ES/PNET) are a relatively rare group of malignant tumors (1,2) that occur in children and adolescents aged between 10–20 years (3). ES/PNET are highly aggressive and generally portend poor prognoses (4) because of the lack of clear clinical symptoms during the early stages. These tumors frequently occur in the extremities and retroperitoneum (5,6), and are seldom in the internal organs. ES/PNET of the bladder is rare, with only a few reports available at present (7,8). We report a case of primary ES of the bladder that occurred in a 19-year-old female.

Case presentation

A 19-year-old female patient was admitted to our hospital after experiencing gross hematuria without concomitant urinary frequency, urgency, painful urination, and blood clots for 1 week. Urine samples were routinely collected on admission, and the red blood cell count was 7,796/μL. The patient had been smoking for the previous year and had no family history of tumors.

Outpatient ultrasound suggested a left ureteral end cyst with abnormal internal echogenicity and clot but did not exclude the possibility of a tumor. Contrast-enhanced ultrasound (CEUS) suggested mixed (predominantly solid component) left end ureteral occupancy (*Figure 1A*). Computed tomography (CT) enhancement showed a cystic and solid mass in the lower left ureter, indicating the presence of tumor lesions. Magnetic resonance imaging (MRI) suggested that the left ureteral bladder wall was occupied, suggesting the possibility of a ureteral cyst combined with hemorrhage (*Figure 1B*,*1C*).

The patient then underwent cystoscopy and transurethral resection of bladder tumor (TURBT). Intraoperatively, the lesioned tissue had old hemorrhage and hematoma. Pathological examinations indicated microscopic infiltration and growth of small round tumor cells with higher nuclear plasma and darker nuclear chromatin (*Figure 1D*). Immunohistochemical staining of the paraffin tissue block A8 indicated that the lesion was positive for CD99, FLI1, Vimentin (+), SYN (focal weak), NSE (focal weak), β-catenin (cytosolic and cytoplasmic), INI-1, CD117, and Ki-67 (~40%), and negative for CgA, CK7, CK20, CK-pan, EMA, P63, GATA3, S100p, HCG, CD56, S100, Desmin, SMA, Myo-D1, Myogenin, and LCA. These findings were typical of extraosseous ES/PNET. The patient received gemcitabine infusion chemotherapy after surgery and was not subjected to further treatment. Notably, there was no sign of recurrence during the two years of follow-up.

All procedures performed in this study were in accordance with the ethical standards of the institutional and national research committee(s), and the Helsinki Declaration (as revised in 2013). Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the editorial office of this journal.

Discussion

ES/PNET is a rare primary small round cell malignancy. It

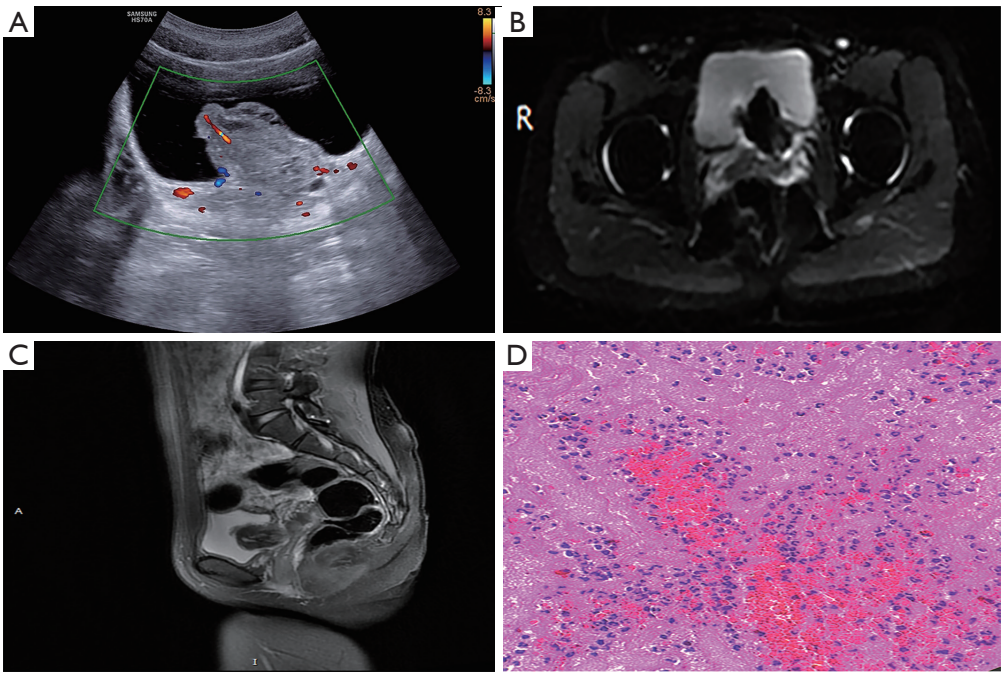

Figure 1 The imaging findings of the lesion. (A) Mixed echogenic mass at the left ureteral orifice in the bladder triangle with CDFI showing a few dotted streaks of blood flow signals in and around it. (B,C) T2WI with predominantly low signal and a thin strip of slightly high signal area. The bladder wall signal is at the edge of the lesion, with low DWI signals. (D) Microscopic infiltration and growth of small round tumor cells with higher nuclear plasma and darker nuclear chromatin (HE ×40). CDFI, Color Doppler flow imaging; T2WI, T2 weighted imaging; DWI, diffusion-weighted imaging; HE, hematoxylin-eosin staining.

is difficult to establish clear guidelines for its management and treatment because of the minuscule number of reported cases. Its preoperative diagnosis is further complicated by the lack of specific imaging characteristics. Definitive diagnosis relies on postoperative pathology, immunohistochemistry, and genetic analysis. Early diagnosis and treatment of ES/PNET patients are thus highly challenging.

The case described in this report is of a 19-year-old female with clinical hematuria and hydronephrosis. The patient had a large lesion of approximately 55 mm ×36 mm, and had clear borders, irregular morphology, and convex growth towards the internal bladder. The lesion was close to the left ureteral bladder entrance, causing continuous compression to the left kidney and left ureteral fluid. The appearance of the lesion was very confusing on MRI, with significant low signals in T2WI and the center of DWI. The lesion had circumferential high signal at the edges, and T1WI showed equally high signal, slightly higher than the surrounding muscle signal. The internal signal of the lesion was more like a hematoma, and was also reflected in the surgical records. The pattern had a mild enhancement during the arterial phase, peaking during the venous phase, and decreasing during the delayed phase, with a net CT increase of approximately 8–29 HU. MRI showed discontinuous circumferential enhancement of the lesion with a significant visible gap in the right anterior.

In the previously reported cases (*Table 1*), the youngest patient was 10 years old, the oldest was 81 years old, bringing the mean age to 45 years. Notably, only one patient reported massive bleeding (25). Many patients had tumors on the right side of the bladder (7/21). In particular, there was extrinsic bladder protrusion towards the pelvis and the lesions were larger in most cases. However, the previous reports did not elaborate more on the imaging presentation. The imaging presentation of extraosseous Ewing sarcoma is not specific and may vary depending on the site. Generally, cystic necrosis is more common, calcification is rare, and pseudo-envelope may be present. Reinforcement varies depending on the site of occurrence and is mostly heterogeneous and variable (28,29). Ewing sarcoma of the bladder is rare but is more malignant and

Table 1 Reference cases of ES/PNET of the bladder

References	Sex/ age	Symptoms	Diagnostic	Tumor size (mm)	Local	Surgery
Present case	F/19	Gross hematuria	US, CT, MRI	55×36	Left posterior	TURBT
Sueyoshi *et al.* (9)	M/10	Polyuria, lower-abdominal swelling	US, CT	135×131×129	Right side	Double J tube + partial cystectomy
Osone *et al.* (10)	M/10	Dysuria, hematuria, hematuria, fever	US, CT, cystoscope	10	Base	TURBT
Rao *et al.* (11)	F/14	Dull pain, lower-abdominal lump	US, CT, needle biopsy	150×120×75	Posterior	Partial cystectomy
Gousse *et al.* (12)	F/15	Hematuria	IVP, cystoscopy	30×20×20	Right anterior lateral	TURBT
Lopez-Beltran *et al.* (13)	F/21	Frequency, dysuria, hematuria	US, cystoscope biopsy	90×80×60	The posterior and right and left sides	Cystectomy + TH + BSO
Banerjee *et al.* (14)	M/21	Frequency, dysuria, hematuria	IVP, cystoscopy	80×60×40	Right side	Cystectomy
Vallonthaiel *et al.* (15)	F/27	Frequency, hematuria	US, CT	103×98×47	Left anterior and lateral	TURBT
Lam *et al.* (16)	F/30	Polyuria, hematuria	US, MRI	64×94×77	Right side	TURBT + cystectomy + Indiana pouch
Tonyalı *et al.* (17)	F/38	Hematuria	CT	40×26×25	Right side	TURBT + cystectomy + TH + BSO + ileal conduit
Desai (18)	F/38	Hematuria	Cystoscope	120×70×35	Posterior and right and left	Cystectomy + TH + BSO
Gao *et al.* (8)	F/45	Frequency, urgency, dysuria	US, CT, cystoscope	30	Right neck	TURBT + cystectomy + TH + ileal conduit
Busato *et al.* (19)	F/52	Frequency, dysuria, pelvic pain, hematuria	US, cystoscope	33×15×22	Right base	TURBT
Colecchia *et al.* (20)	F/61	Hydronephrosis, renal failure	CT, cystoscope biopsy	–	–	–
Mentzel *et al.* (21)	M/62	Dark urine, fever, backache, AUR	MRI	140×100×100	–	TURBT + nephrostomy
Liu *et al.* (7)	M/64	Abdomen dull pain	CT	60×50	Left lateral out of bladder	–
Okada *et al.* (22)	M/65	Hematuria, dysuria	US, CT, cystoscope	50	Left posterior	TURBT + cystectomy
Al Meshaan *et al.* (23)	F/67	Hematuria, fever, hydronephrosis	US, CT, cystoscope	30×25×10	Posterior	TURBT + partial cystectomy
Ellinger *et al.* (24)	M/72	Hematuria, oliguria	MRI	–	–	TURBT
Zheng *et al.* (25)	M/74	Frequency, dysuria, hematuria	CT	–	Neck	TURBT
Zhang *et al.* (26)	F/78	Gross hematuria, blood clot, urinary frequency, urgency	CTU	63×44	Right posterior	TURBT
Krüger *et al.* (27)	M/81	Lymphedema, fatigue, urge, incontinence	US, CT	–	–	TURBT + nephrostomy

ES/PNET, Ewing's sarcoma/primitive neuroectodermal tumor; F, female; M, male; US, ultrasound; CT, computed tomography; MRI, magnetic resonance imaging; TURBT, transurethral resecting of bladder tumour; IVP, intravenous urography; TH, total hysterectomy; BSO, bilateral salpingo-oophorectomy; AUR, acute urinary retention; CTU, computed tomography urography.

needs to be differentiated from the common uroepithelial carcinoma of the bladder. Based on the documented cases, Ewing sarcoma in the bladder seems to be common among middle-aged persons and young adults, while general uroepithelial carcinoma is common among middle-aged and older men. Notably, smoking is an important risk factor in both. CT enhancement demonstrates that there is progressive strengthening during uniform strengthening, with the most significant strengthening amplitude happening during the arterial phase (30). Of note, the final diagnosis of this disease relies on pathology because there are cases of ES combined with epithelial carcinoma. The imaging presentation of ES combined with epithelial carcinoma is somewhat different from that of most extraosseous ES because of the interference of massive bleeding. The descriptions of more new cases are thus necessary to improve the existing knowledge and experience.

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Footnote

Conflicts of Interest: All authors have completed the ICMJE uniform disclosure form (available at https://qims.amegroups.com/article/view/10.21037/qims-22-867/coif). The authors have no conflicts of interest to declare.

Ethical Statement: The authors are accountable for all aspects of the work in ensuring that questions related to the accuracy and integrity of any part of the work are appropriately investigated and resolved. All procedures performed in this study were in accordance with the ethical standards of the institutional and national research committee(s), and the Helsinki Declaration (as revised in 2013). Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the editorial office of this journal.

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