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Case Report

Primary primitive neuroectodermal tumor in pelvic cavity: an unusual case and literature review

Fan Wang, Xiaoman Zhang, Yanwei Shen, Shuting Li, Meng Lv, Chunli Li, Jiao Yang, Zheling Chen, Jin Yang

Department of Medical Oncology, The First Affiliated Hospital of Xi'an Jiaotong University, Xi'an, Shaanxi, P. R. China

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Abstract: Primitive neuroectodermal tumor (PNET) in the pelvic cavity is a relatively rare tumor of neural crest origin. We report a 35-year-old lady diagnosed as pelvic cavity PNET treated with 6 cycles of chemotherapy (VAC regimen then followed by IE regimen), and the final clinical effect was partial remission (PR). The patient did not show any characteristic symptoms except for left lower limb numbness and weakness. The diagnosis was confirmed by the imageology as well as immunohistochemistry of ultrasound-guided puncture and biopsy, which showed positive for Mic-2 (CD-99 antigen), vimentin, neuron-specific enolase (NSE), and Chromogranin A (CgA). She didn't have the chance to take the radical operation not only due to the size of the tumor, but invasion of the surrounding tissues. Therefore, she only received chemotherapy and was regularly followed for 18 months at our clinic without evidence of disease progression. The result of immunohistochemistry is a useful supplement in differential diagnosis, furthermore, CAV and IE alternating chemotherapy has high objective response rate for metastic PNET. The incidence of peripheral PNET (pPNET), their clinical and pathological features are discussed with a review of the literature.

Keywords: Ewing's tumor, primitive neuroectodermal tumor, therapeutics

Introduction

Primitive neuroectodermal tumors (PNETs) are small-blue-round-cell malignancies, predominantly arising in the soft tissues or bones in children and young adults [1]. PNETs belong to the Ewing's sarcoma family of tumors, based on shared chromosomal translocation at EWSR1 (Ewing sarcoma break point region 1). The peripheral primitive neuroectodermal tumor (pPNET), first recognized by Arthur Purdy Stout in 1918, is a member of the family of 'small round- cell tumors' [2]. Most of these tumors are diagnosed before the age of 35 years with a slight male preponderance, and primarily involve the central nervous system (CNS) [3]. Although PNET can occur in numerous solid organs such as the kidney, ovary, vagina, testis, uterus, cervix uteri, urinary bladder, parotid gland, heart, lung, rectum, pancreas and gall bladder, it is an extremely rare tumor entity [4]. Most of these are adolescent or young adult patients, with a male predominance [2, 5]. Primary PNETs in the pelvic cavity are relatively rare. To our knowledge, only a handful of cases have been reported in the literature. In this case report, we describe a PNET in the pelvic cavity in an adult woman, having a stable disease (SD) during the follow-up. Written informed consent was obtained from the patient's parent for publication of this case report and any accompanying images.

Case presentation

A 35-year-old female patient was admitted to our hospital with a chief complaint of left lower limb numbness and weakness and presented with a pelvic mass which she had palpated accidentally more than 5 months ago. Her past medical history was unremarkable. Prior to admission, the patient had no history of either hypertension or diabetes mellitus, and the family history was unremarkable. She also denied a family history of ovarian cancer or teratoma and the symptoms of neuroendocrine activity such as flushing, diarrhea, abdominal cramping, and palpitations. The physical examination showed

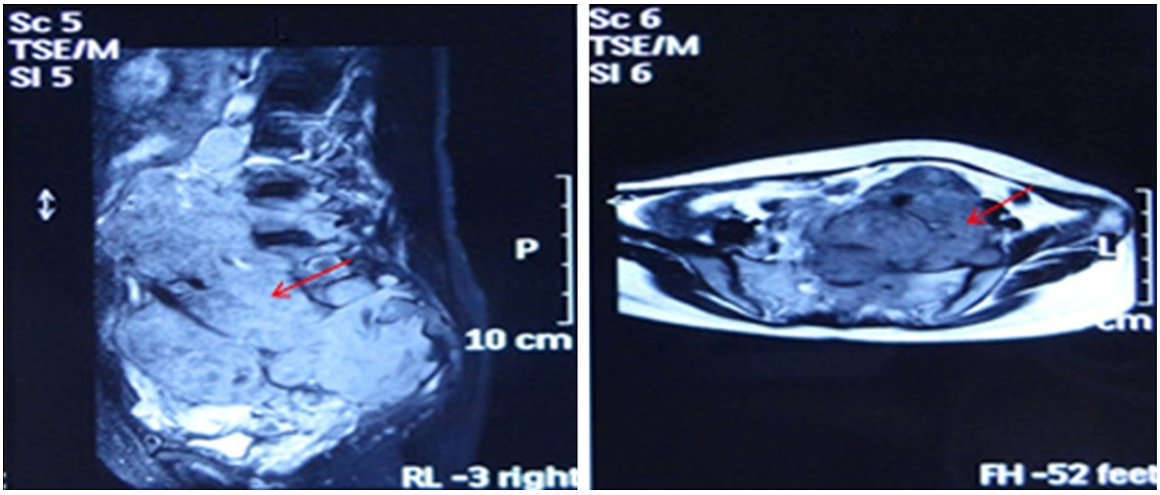

Figure 1. MRI scan. A large soft-tissue-mass (red arrows) in the pelvic cavity. The tumor invades the lumbosacral vertebrae and left-sided ureter.

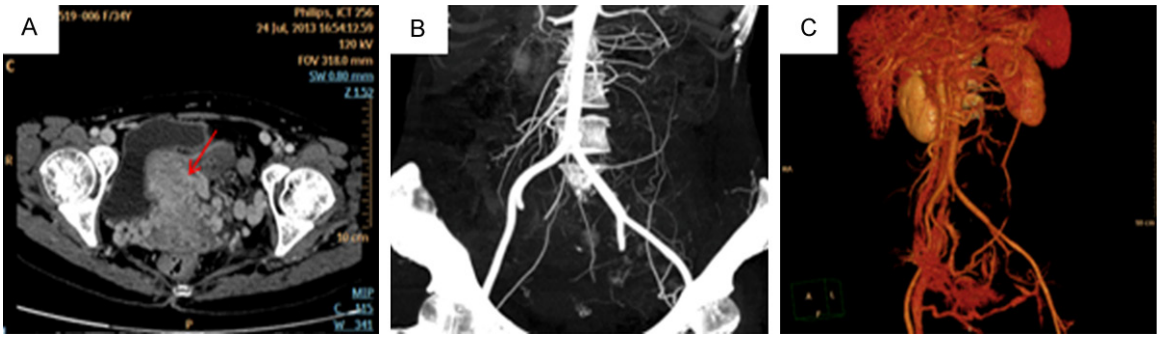

Figure 2. CTA scan. A: Demonstrated a large soft-tissue mass (red arrows) occupying the pelvic cavity. B and C: The left common iliac artery, internal iliac artery and external iliac artery were wrapped around the lesions, and many small vessels were sent into the mass. Local right common iliac artery was wrapped around the lesion. The lower part of the abdominal aorta was compressed and shifted to the right due to the mass.

that there was a solid irregular-shape-mass, measuring approximately 15 cm × 10 cm, on the left side of the pelvic, with close adhesion to adjacent tissue. Results of her routine blood tests were within the normal range. Because of the large lesion, the tumor cannot be resected completely, and the surgery and radiotherapy was not advocated. As a result of multidiscipline experts' group consultation, the patient was then referred to our department, where tumor markers for suspected malignancy revealed an elevated NSE level (86.71 ng/mL), with normal levels of lactate dehydrogenase (LDH), carcinoembryonic antigen (CEA), CA-199, alpha-fetoprotein (AFP) and beta-human chorionic gonadotropin (β-HCG).

She took the magnetic resonance imaging (MRI) in the local hospital on July 8th 2013 which showed a large soft-tissue-mass in the pelvic cavity with extensive seeding spreading on the surface. Furthermore, it also demonstrated lumbosacral vertebrae and left-sided ureter invasion by the tumor (**Figure 1**). Once admitted to our department, the presence of the tumor was subsequently confirmed by computed tomography angiography (CTA) scan (**Figure 2**). The intravenous pyelography (IVP) was performed, which revealed an absent sign of left kidney and left ureter. And the left wall of the bladder was compressed by the solid mass. The radioactive nephrogram showed the absent sign of left kidney and the right renal blood flow perfusion and function were normal.

On July 23th 2013, the patient took the ultrasound guided pelvic mass biopsy. The histopathology result of the ultrasound-guided punc-

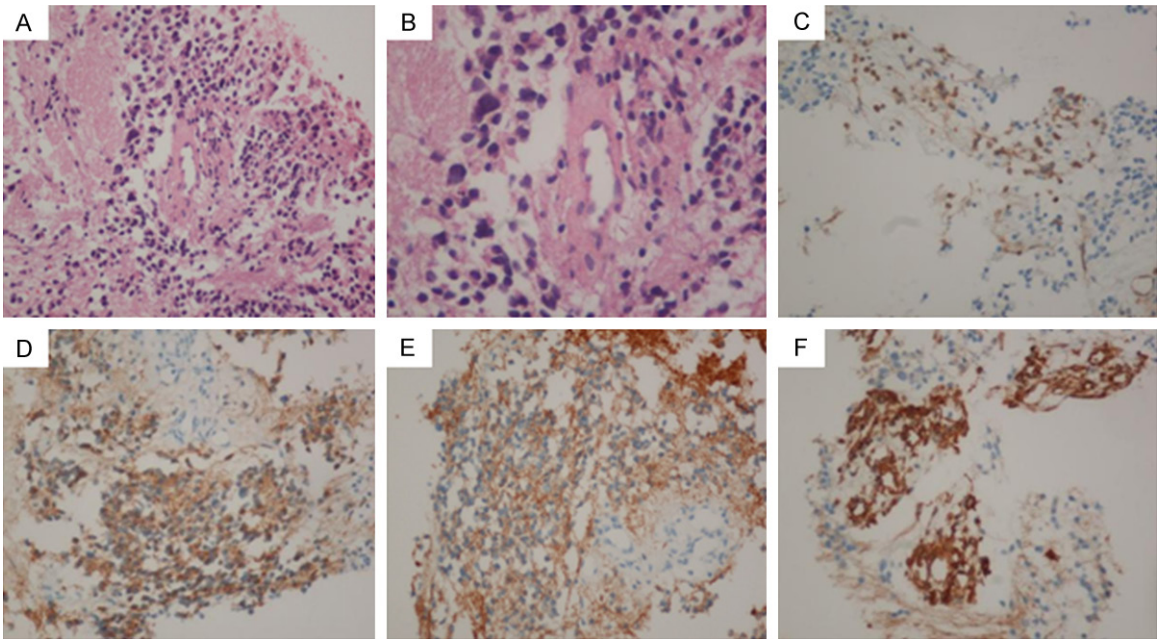

Figure 3. Histological findings. (A) Discohesive growth of small, blue, round to oval cells, the cytoplasm of the tumor cell was little and the nucleus was big and round in shape, characteristic of PNET (H&E, magnification × 20). (B) Characteristic scant, basophilic cytoplasm, ovoid or polygonal nuclei, fine granular chromatin texture and marked nucleoli. Some nuclei were hyperchromatic. (H&E, magnification × 40. (C) CD99 membranous expression, characteristic of PNET (magnification × 20). And expression of (D) CgA (magnification × 20) and (E) NSE (magnification × 20) and (F) vimentin (magnification × 20).

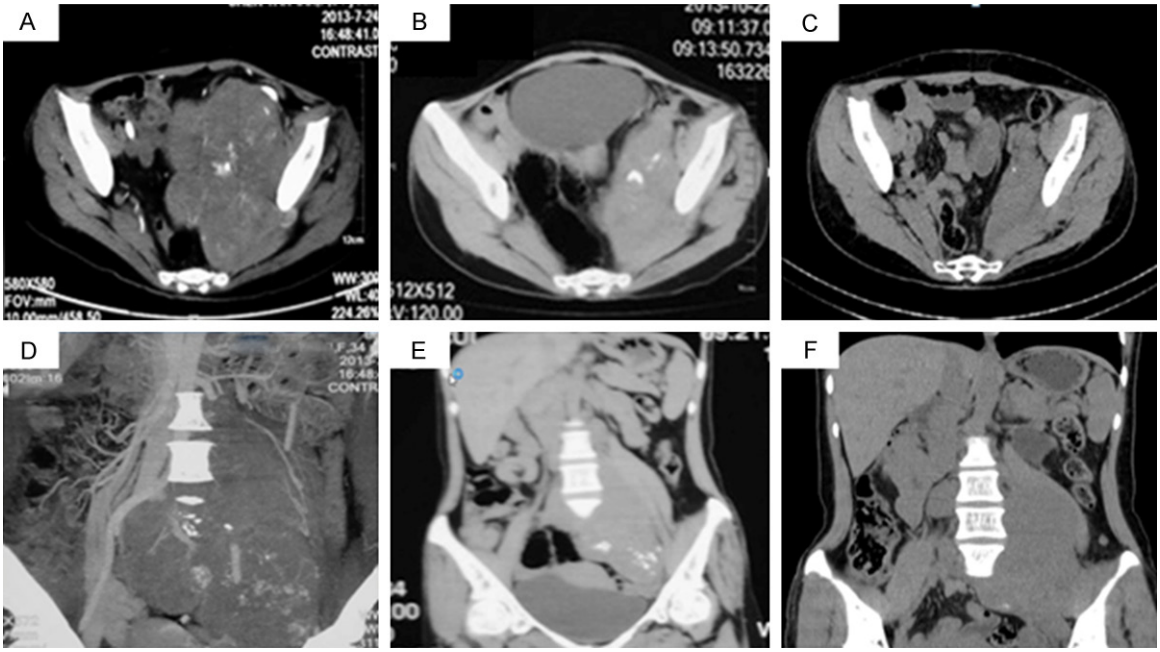

Figure 4. The follow-up CT scan (A) and (D) were taken on July 24th 2013; (B) and (E) were taken on October 22th 2013; (C) and (F) were taken on January 6th 2015.

ture and biopsy revealed a high cellularity, composed of small cells with hyperchromatic, round to oval nuclei and scanty to small amounts of cytoplasm arranged in lobules separated by fibrovascular septa, patternless sheets incompletely divided by fibrovascular

Table 1. Previous reports of primary primitive neuroectodermal tumor

Case	Year	Sex	Age	Tumor location	Immunohistochemical	Treatment	Follow up (months)	Reference
1 (2 cases)	2000	M	41	LUL	PAS(+), NSE(+), Leu(-) CgA(-), ProGRP(-) NSE(+), CgA(-), ProGRP(-)	VAI+SR	22	
		F	30	RLL		VAC+SR+EP	16	[26]
2	2007	NK	16	RLL	CD99(+)	VAIE	NK	[27]
3	2008	M	53	RLL	Vimentin(+), CD99(+), neurofilament(+), synaptophysin(+) CgA(-), S-100(-), cytokeratin 7(-)	VAC+IE	9	[28]
4	2013	F	20	LPH	Vimentin(+), CD99(+), CgA(-)	VAIE+SR+RT	15	[29]
5	2000	M	28	Kidney	NSE(+), S100(+), neurofilament(+), synaptophysin(+), Leu(-), Vimentin(-)	SR	NK	[30]
6	2012	M	50	Kidney	Vimentin(+), CD99(+), NSE(+)	SR+VAI	30	[31]
7 (2 cases)	2013	M	48	Kidney	Vimentin(+), CD99(+)	VAIE	12	[32]
		M	23	Kidney	Vimentin(+), CD99(+) synaptophysin(-), CgA(-)	VAIE	8	
8	2013	M	63	Adrenal Gland	CD99(+)	VAC/IE	13	[33]
9	2013	M	17	Adrenal Gland	CD99(+)	VAC/IE	NK	[34]
10	2012	F	28	Ovary	Vimentin(+), CD99(+) synaptophysin(-), CK(-)	SR+CT	18	[35]
11	2014	F	16	Ovary	synaptophysin(+), CD56(+) CgA (+), S-100(+) CD99(-)	SR+PT	13	[36]
12	2007	F	30	Uterus	Vimentin(+), CD99(+), CgA(+)	VAI+RT	16	[37]
13	2008	F	22	Uterus	CD99(+), NSE(+) Vimentin(-), S-100(-), CgA(-) synaptophysin(-)	SR+CT	10	[38]
14	2012	F	23	Cervix	Vimentin(+), CD99(+) S-100(-), CgA(-) synaptophysin(-)	SR+CT	NK	[39]
15	2013	F	27	Cervix	Vimentin(+), CD99(+), CD56(+) NSE(+) CD38(-), CD45(-), S-100(-)	CT+RT	6	[40]
16	2013	M	40	Thyroid	CD99(+)	EP+RT	27	[41]
17	2014	F	26	Cutis	Vimentin(+), CD99(+), S-100(+)	SR+CT+RT	6	[42]
18	2013	F	29	Prostate	CD99(+), CD45(-), S-100(-) synaptophysin(-)	SR+CT	12	[43]
19	2013	F	33	Breast	Vimentin(+), CD99(+) Synaptophysin(+), CgA(-)	VAC	NK	[44]
20	2012	F	13	Pancreas	CD99(+), NSE(+) CgA(-), PAS(-)	SR+CT+RT	41	[45]
21	2013	F	35	Pelvic cavity	Vimentin(+), CD99(+) NSE(+), CgA(+)	VAC/EP	18	This case

Abbreviations: M: male, F: female, LUL: left upper lung, RLL: right lower lung, PAS: periodic acid-Schiff, ProGRP: pro-gastrin-releasing peptide, CD: cluster of differentiation V: vincristine, A: doxorubicin, I: ifosfamide, SR: surgical resection, C: cyclophosphamide, E: etoposide, P: cisplatin, NK: not known, LPH: left pulmonary hilar, RT: radio therapy, CK: cytokeratin, CT: chemotherapy, PT: carboplatin+ paclitaxel.

septa or a trabecular/cord-like pattern; staining positive for CD-99 (Mic-2), vimentin, NSE, and CgA (Chromogranin A) (**Figure 3**).

Chemotherapy was performed. On August 2nd 2013, the patient was received the first cycle of combination chemotherapy (CAV: cyclophosphamide 1200 mg/m^2, adriamycin 75 mg/m^2, and vincristine 2 mg/m^2). All three drugs were given in a single day. After 2 cycles of the CAV regimen, the follow-up CT scan showed PR. Then the regimen was transferred to 2 cycles of IE (IE: ifosfamide 1.6 g/m^2 and etoposide 120 mg/m^2) and the clinical effect was stable disease (SD). During the treatment, NSE was decreased dramatically from 86.71 ng/ml to 29.87 ng/ml. Another 2 cycles of CAV regimen was performed after the 2 cycles of IE regimen were finished. And the follow-up CT scan was demonstrated in **Figure 4**.

Discussion

With regard to the low incidence of the disease, we searched on Pubmed to find 20 cases of pPNET in order to get a glimpse of the clinical and pathological features of this disease. An overview is given in **Table 1**. These 23 cases had a median age at diagnosis of 34 (range, 13-63). Time slots from the appearance of clinical manifestations to diagnosis are approximately 3.7 months. The performance of clinical symptoms are usually vague, diverse, and not obvious enough. They can be diagnosed based on histopathological morphology, immunohistochemical examination, and genetic testing. The use of immunohistochemical staining would make the diagnosis easier. CD99, a highly sensitive marker for PNET, was found in nearly all 23 (the last one is our patient, and case 1 and case 7 contain two patients respectively) of these patients. Multimodal treatment including surgery, chemotherapy, and/or radiotherapy is required. Eleven of these patients underwent surgical resection. Three patients underwent neoadjuvant chemotherapy followed by resection with or without adjuvant chemotherapy. The most commonly used chemotherapeutic drugs are: vincristine, doxorubicin, ifosfamide and cyclophosphamide, others include etoposide and cisplatin.

Epidemiological characteristics

PPNET was first reported occurring in the ulnar nerve by Stout in 1918. The tumor was composed of small round cells, which focally arranged as rosettes [6]. In 1921, an undifferentiated diffuse small round tumor occurring in the diaphysis of long bone was reported by Ewing. Later the condition was named Ewing's sarcoma (ES) [7]. This tumor represents 3%-6% of solid tumors and 1.4%-1.8% of malignant processes, with an incidence of 3 cases/million/year [8]. 90% of the cases appear between 5 and 30 years and is more common in men. PNET is usually seen along the central axis, particularly in the soft tissue or bone in children and young adults. This rare and aggressive tumor has also been described in the kidney, uterus, uterine cervix, pancreas, and adrenal Gland. Regardless of the point of origin, these tumors are highly aggressive, often quickly metastasizing to the lung and bone.

Recent studies showed that pPNET and ES have many similarities in histomorphology, immunohistochemistry, and molecular biology. Both tumors result from a reciprocal translocation of the long arms of chromosomes 11 and 22, t (11;22)(q24;q12), and are located at different neural differentiation stages [3, 9-11]. They are grouped as the ES/PNET family and are difficult to distinguish from each other on the basis of histomorphology, but can be differentiated by electron microscopy and immunohistochemical analysis.

Diagnosis

Imageological diagnosis

Making an accurate diagnosis is critical for optimal patients' management and prognosis. The most common clinical manifestation are manifested by pain (in the pelvis, femur or humerus) [12, 13], swelling and, sometimes, fever, weight loss, anemia and leukocytosis. Physical examination often reveals an abdominal or pelvic mass with recurrent abdominal pain. Imaging examination such as CT scan is able to provide important information regarding the size of the mass, the involvement of adjacent structures and the presence of metastasis.

Usually, the imaging tests such as CT scans of PNET exhibit heterogeneous masses, which often invade surrounding tissues, including bones. The most common origin sites are the long bones, such as the femur and humerus as well as the pelvic bones. CT findings reflect ero-

sions, periostitis and soft tissue masses, as in our case.

Histological and pathological diagnosis

Immunohistochemistry is important to establish the differential diagnosis [14]. Histologically, lymphoblastic lymphoma, neuroblastoma, rhabdomyosarcoma, poorly differentiated synovial sarcoma, and Wilms tumor are included in the differential diagnosis of ES/PNET because they are all small round cell tumors. Clinically, the most commonly used diagnostic criteria is as follows: CD99 positive, and at least two or more different neural markers (such as NSE, synaptophysin, S-100, VIM, NF) are positive, and lymphocyte common antigen (LCA) is negative, so does the markers of myogenic tumor immunohistochemistry (myoglobin, myosin) to exclude lymphoma and small round cell myogenic tumors. Immunohistochemistry combined with electron microscopic examination of cells containing neuroendocrine particles can confirm the diagnosis. In our case, immunohistochemical analysis showed positive for CD-99, vimentin, NSE, and CgA, according to which we finally make the diagnosis.

Treatment

The treatment of PNET should be various combinations of early surgical resection as well as adjuvant chemotherapy and radiation therapy [15]. As shown in a literature review concerning pulmonary PNET [16], 8 of 20 (40%) patients underwent resection and adjuvant chemotherapy with or without radiation, while 6/20 (30%) patients underwent neoadjuvant chemotherapy followed by resection with or without adjuvant chemotherapy, 5/20 (25%) patients underwent resection, and 1/20 (5%) patient only received chemotherapy. The 2-year survival rates for the first three groups were 33%, 66%, and 33%, respectively, and the patient who only underwent chemotherapy lived for 17 months following the initial diagnosis. To the best of our knowledge, our case is the first to report a patient with inoperable pelvic PNET who only underwent the chemotherapy, and lived for more than 17 months till the latest follow-up was made.

As far as we know, chemotherapy is considered to be one of the most efficient methods in tre ating metastasis PNETs. Chemotherapeutic agents such as cyclophosphamide, adriamycin, vincristine, ifosfamide, or etoposide have been used [17-19]. Recently, it has been noted that chemotherapy might be effective only for the first few cycles, and then the tumors develop resistance very quickly [17]. Adjuvant radiotherapy had been used for local recurrences as well as unresectable or incompletely resected PNETs [20]. A combination of surgery, radiotherapy and chemotherapy attain an increase in survival and disease-free survival [21, 22]. A high initial complete response of 94% was observed in patients of PNET treated with VAC chemotherapy plus local radiation therapy [23]. The best responses were reported with combinations based on anthracyclines (doxorubicin) and high doses of alkylating agents (cyclophosphamide or ifosfamide) [24]. Metastatic germ cell cancers are highly chemosensitive and have 80% cure rate with cisplatin-based chemotherapy. As mentioned in another article, the authors report the results of treatment of patients with malignant transformation to PNET with cyclophosphamide + doxorubicin + vincristine (CAV) alternating with ifosfamide + etoposide (IE). They noted that CAV and IE alternating chemotherapy has high objective response rate for PNET, as a result, they recommend adjuvant CAV alternating with IE chemotherapy for patients with PNET due to the high probability of recurrent disease and their high chemosensitivity to this regimen [25]. Regarding to the regimen mentioned above, the most common adverse reactions are myelotoxicity, nausea, vomiting and alopecia. In our case, the patient received 6 cycles of CAV/IE chemotherapy. And until nearest follow-up, the clinical effect was SD. This highlights the importance of achieving the correct diagnosis of these atypical tumors using all clinical, morphological, and ancillary methods available to allow for their correct and timely treatment.

Conclusion

Sharing some of the major features of pPNET tumor, herein we report a rare case of a female patient with pelvic cavity PNET. The clinical presentation is often vague and nonspecific and a definitive diagnosis depends on a combination of histology cytogenetic analysis, immunohistochemistry, and histology. Surgical resection remains the mainstay of treatment coupled with adjuvant chemoradiation. Reasonable

application of regimen CAV sequential IE may get a better prognosis in metastatic pPNET.

Disclosure of conflict of interest

None.

Address correspondence to: Jin Yang, Department of Medicine Oncology, The First Affiliated Hospital of Xi'an Jiaotong University, No. 277, Yanta West Road, Xi'an 710061, Shaanxi, P. R. China. Tel: +86-29-85323422; Fax: +86-29-85323422; E-mail: 1473-106133@qq.com

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