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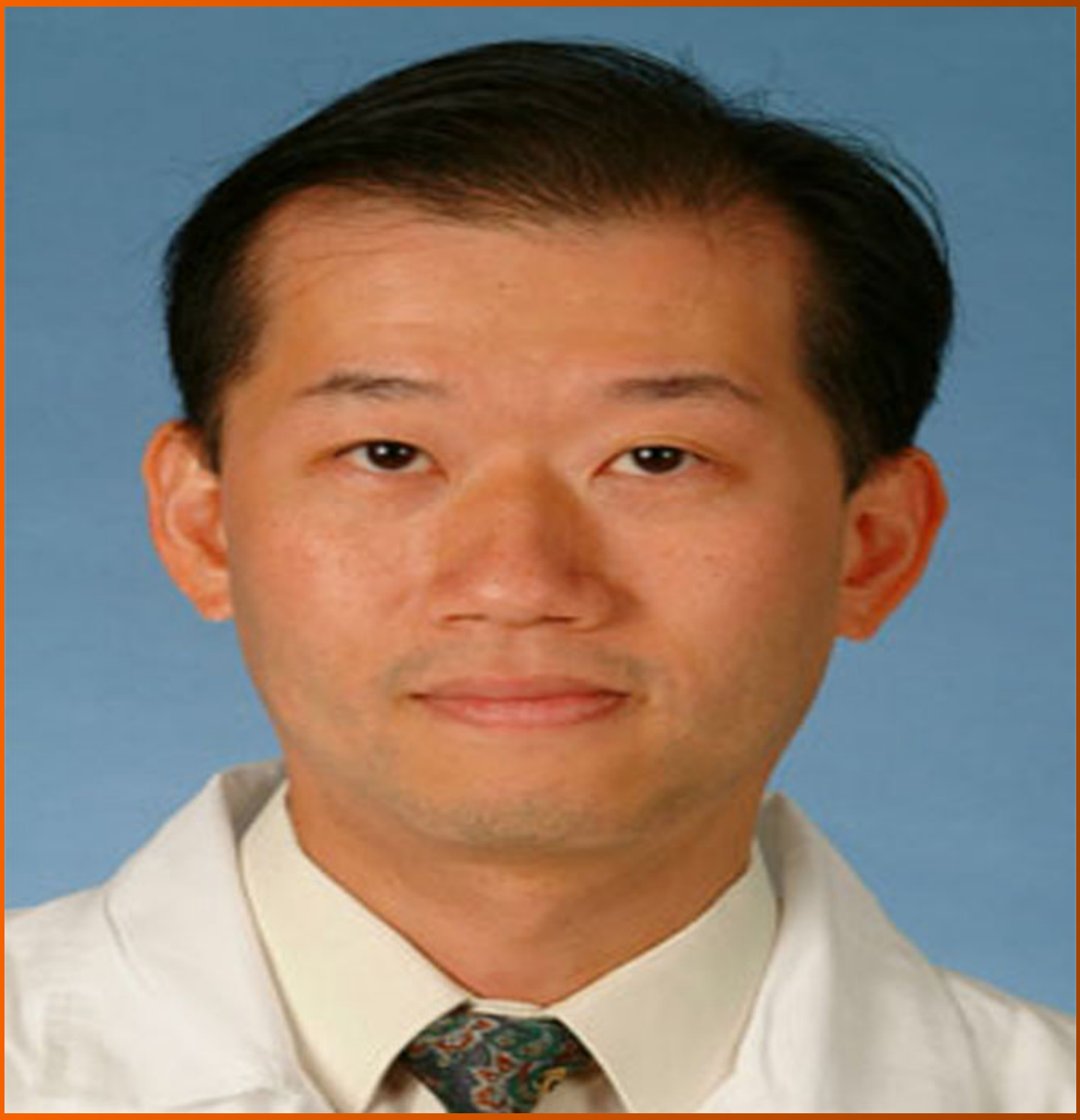

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CASE REPORT

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CASE REPORT

Primitive neuroectodermal tumors of the abdominal wall and vulva in children: Report of two cases and review of the literature

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Author contributions: Chen G, Dang YW, Luo YG, and Chen JB designed the research, supervised all experiments and corrected the manuscript; Xu QQ, Xing WW , Chen P, and Liang SW collected the clinical information, summarized literature, and performed histologic examination and immunohistochemistry; and Xu QQ wrote first version of the paper.

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Abstract

BACKGROUND

Primitive neuroectodermal tumors are rare, highly malignant small round cell tumors belonging to the Ewing sarcoma family. The purpose of this article is to present clinical manifestation, histology, treatment, and prognosis of two primitive neuroectodermal tumors (PNETs) in extremely rare anatomic locations, the abdominal wall and vulva.

CASE SUMMARY

Case 1 was a 66-month-old girl with lesions on the abdominal wall; tumor size was about 3.4 cm × 6.1 cm × 2 cm. The patient underwent radical resection of the tumor. After the operation, an alternating vincristine, doxorubicin, and cyclophosphamide/ifosfamide and etoposide (IE) regimen was given for eight cycles, and the patient survived for 66 mo without progression. Case 2 was a 40-month-old girl, with a vulvar lesion; tumor size was about 3.3 cm × 5 cm × 2.5 cm. The tumor was partially resected by surgery. The family left treatment after two cycles of vincristine, pirarubicin, and cyclophosphamide/IE chemotherapy, and the patient died at home six months after surgery.

CONCLUSION

PNET is a rare, fast-growing, highly malignant tumor that requires histologic and molecular analyses for exact diagnosis, and multimodal treatment is required to achieve a good prognosis.

Key words: Primitive neuroectodermal tumor; Therapy; Prognosis; Case report

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Core tip: Primitive neuroectodermal tumors (PNETs) are rare undifferentiated tumors with similar biological characteristics. They belong to the Ewing sarcoma family, accounting for 4% to 17% of all pediatric soft tissue tumors. PNETs usually occur in children and young adults under 25 years of age. We retrospectively analyzed two PNET cases at the First Affiliated Hospital of Guangxi Medical University from May 2012 to June 2014. Both patients were female with an age of onset at 66 and 40 mo. Both patients were provided inpatient visits, outpatient medical records, and telephone follow-ups for more than one year. In this report, we describe in detail the clinical manifestations, treatment protocols, pathological findings, and patient prognoses. This report provides an in-depth analysis of two cases of PNET at rare sites.

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INTRODUCTION

Primitive neuroectodermal tumors (PNET) are rare undifferentiated tumors with similar biological characteristics. They belong to the Ewing sarcoma family, accounting for 4% to 17% of all pediatric soft tissue tumors. Ewing sarcoma PNETs (ES/PNETs) usually occur in children and young adults under 25 years of age[1-3]. There are two main types according to cell location and origin: Central PNET (cPNET) and peripheral PNET (pPNET). The cPNETs are derived from the neural tube and mainly involve the brain and spinal cord, while the pPNETs are derived from the neural crest and occur outside the central nervous system, and often involve the sympathetic nervous system or soft tissue and bone[4]. We report on two patients presenting with PNETs located in the abdominal wall and vulva. As far as we know, only 13 cases of abdominal wall PNET and 37 cases of vulvar PNET have been reported.

We retrospectively analyzed these two PNET cases at the First Affiliated Hospital of Guangxi Medical University from May 2012 to June 2014. Both patients were female with an age of onset at 66 and 40 months. Both patients were provided inpatient visits, outpatient medical records, and telephone follow-ups for more than one year (Table 1). In this case report, we describe in detail the clinical manifestations, treatment protocols, pathological findings, and patient prognoses. This report provides an in-depth analysis of two cases of PNET at rare sites.

CASE PRESENTATION

Chief complaints

Case 1: A 66-month-old girl with no traumatic medical history presented with reported abdominal pain two months ago.

Case 2: A 40-month-old girl presented with a mass (the size of a broad bean) at the external vaginal orifice, accompanied by intermittent hemorrhage.

History of present illness

Case 1: Two months ago, the girl began to present with reported abdominal pain without any obvious causes.

Case 2: Eight months ago, the girl began to present with a mass (the size of a broad bean) at the external vaginal orifice, which was grown up and accompanied by intermittent hemorrhage.

History of past illness

Case 1: The patient has no special past history.

Case 2: The patient was diagnosed with pPNET *via* pathological biopsy at another hospital. But she did not receive surgical interventions.

Table 1 Follow-up data of two cases of primitive neuroectodermal tumor patients

Case	Age (mo)	Position	Size (cm)	Therapy	Survival (mo)	Outcome
1	66	Abdominal wall	3.4 × 6.1 × 2.0	S + Ch (VDC/IE)	66+	Survival
2	40	Vulva	3.3 × 5.0 × 2.5	S + Ch (VAC/IE)	14	Death

F: Female; S: Surgery; Ch: Chemotherapy.

Personal and family history
Both of the two patients had no significant personal or family history.

Physical examination upon admission
Case 1: Clinical examination revealed a large mass in her right abdomen approximately 5 cm × 5 cm.

Case 2: Physical examination revealed a 3 cm × 3 cm × 1 cm, red, irregular, soft mass located on the external vaginal orifice; no other symptoms were noted.

Laboratory examinations
Case 1: The patient had no significant laboratory test result.

Case 2: Laboratory examination revealed a hemoglobin level of 118 g/L (normal range: 120-160 g/L), blood platelet count of 422 × 10^9/L (normal range: 125 × 10^9/L to 355 × 10^9/L), neutrophil percentage of 20.9% (normal range: 40%-75%), neuronspecific enolase level of 61.96 ng/mL (normal range: 0.00-23.00), international normalized ratio of 0.75 (normal range: 0.80-1.40), and prothrombin time of 8.90 s (normal range: 9.00-15.00 s).

Imaging examinations
Case 1: Computed tomography (CT) showed a fusiform soft tissue density between the abdominis obliquus internus musculus and the abdominal transverse muscle in the right inferior abdominal, with a density measuring about 3.4 cm × 6.1 cm with irregular patchy calcification, evenly enhanced in the middle of the right lower abdominal wall (Figure 1). No evidence of metastatic disease was uncovered after a complete examination.

Case 2: Pelvic CT showed an irregular soft tissue density in the patient's vagina with some protruding lesions ranging over an area of about 3.3 cm × 5 cm × 2.5 cm. A boundary was not clear, nor was the inner wall of the normal vagina, and the enhancement scanning showed that the lesion was enhanced (Figure 2). No evidence of distant metastases was revealed upon head, chest, and abdominal CT.

TREATMENT

Case 1
During surgery to remove the mass, a gray-white mass measuring approximately 4 cm × 3 cm × 2 cm was excised from between the abdominis obliquus internus musculus and the abdominal transverse muscle. The mass was tough and unencapsulated, with a basal portion adhered to the abdominal transverse muscle. There was no invasion of the abdominal transverse muscle membrane. Incision of the tumor revealed calcification and a yellow, turbid liquid.

Case 2
During the surgery, a mass protruding into the vulva about 5 cm × 3 cm × 3 cm in size was visualized, presenting as red, irregular, soft tissue, leading inward to the vagina but clearly separated from the cervix with no cervical invasion. The tumor filled the vaginal orifice, invaded the hymen and urethral orifice, and covered the external urethral orifice. Partial resection was conducted due to the many invasion sites of tumor and the difficulty of complete resection. The operation was completed without complication.

FINAL DIAGNOSIS

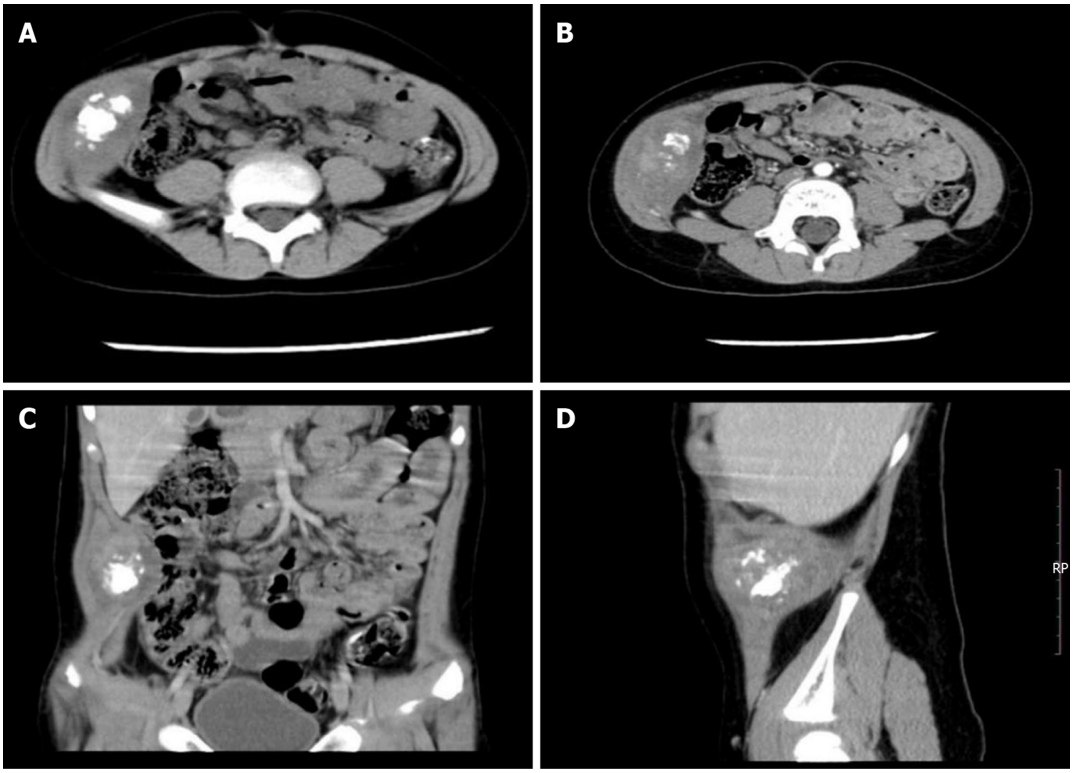

Figure 1 Computed tomography evaluation of Case 1. A: Axial computed tomography image showing a fusiform soft tissue density between the abdominis obliquus internus musculus and the abdominal transverse muscle in the right inferior abdomen; the middle of the mass has irregular patchy calcification; B: Enhanced image showing uniform enhancement of the mass; C and D: The mass involving the musculus transversus abdominis.

Case 1

Histologic examination revealed small round cells with a high nuclear cytoplasmic ratio. Immunohistochemistry showed that the tumor cells were positive for CD99 and Synaptophysin (Syn), and negative for CK, LCA, chromogranin A (CgA), vimentin, CD3, CD20, and CD56 (Figure 3). *EWSR1* gene rearrangement was positively confirmed by fluorescent *in situ* hybridization (FISH) analysis (Figure 4). The patient was thereby diagnosed with pPNET.

Case 2

Pathological examination demonstrated round and oval tumor cells in the shape of flakes or beams; cells were abnormal, and mitosis was apparent. On immunohistochemical analysis, the tumor cells were positive for CD99 and vimentin and negative for desmin, CgA, Syn, CD56, NSE, CK, LCA, myogenin, and actin. FISH analysis revealed heterotopic changes in the *EWSR1* gene (Figure 5). The diagnosis was pPNET. As the patient's sections were loaned out, we are unable to show the pathology images.

OUTCOME AND FOLLOW-UP

Case 1

One week after the operation, the patient began treatment with an alternating VDC/IE regimen for eight cycles of chemotherapy. VDC was provided as vincristine (1.5 mg/m^2), doxorubicin (37.5 mg/m^2), and cyclophosphamide (1.2 g/m^2); IE was ifosfamide (1.0 g days 1, 2, 3, 4, and 5) and etoposide (50 mg days 1, 2, 3, 4, and 5). The patient was followed for 66 mo with no relapse or metastatic disease, and currently attends school and functions normally. No treatments followed the eight cycles of chemotherapy.

Case 2

The patient received chemotherapy *via* an alternating VAC/IE regimen three weeks after surgery, as follows: VAC was vincristine (1 mg on day 1), pirarubicin (40 mg on day 1), and cyclophosphamide (700 mg on day 1); IE was ifosfamide (1.0 g on days 1, 2, 3, 4, and 5) and etoposide (50 mg on days 1, 2, 3, 4, and 5). The patient left treatment

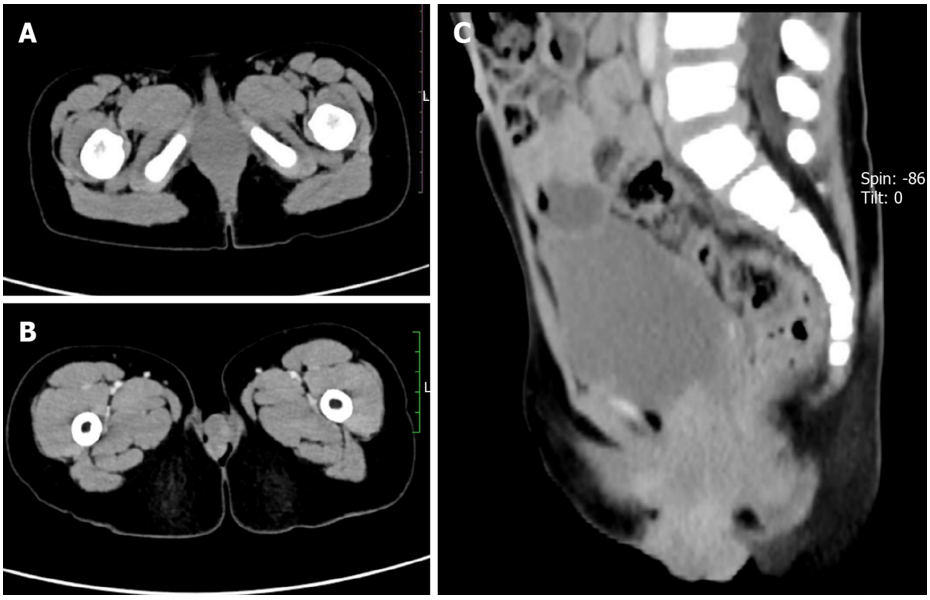

Figure 2 Computed tomography evaluation of Case 2. A: Axial computed tomography image showing an irregular soft tissue density in the vagina; B: Enhanced image showing non-uniform lesion enhancement; C: Sagittal image showing partial lesion protruding into the vagina.

after two cycles of chemotherapy and died at home 14 months after diagnosis, approximately six months after surgery. Cause of death was spread of the tumor throughout the body.

DISCUSSION

PNETs originate from the neuroectoderm and are highly malignant tumors with small round cells, first elucidated by Hart and Earle[5] in 1973. In 2013, the WHO pathological classification system updated and removed any differences between PNET and Ewing sarcoma because they demonstrate the same biological characteristics, reclassifying PNET as a member of Ewing sarcoma tumor family[6]. Campbell *et al*[7] compared the epidemiology, clinical features, and outcomes of patients with 3575 reported cases of Ewing sarcoma or PNET over a 40-year period, and proved that the classification by WHO was correct. PNET is known to be a rare disease that rarely occurs in the skin or subcutaneous tissue[8]. We reviewed the literature published over an extended period, and found only 13 cases of PNET in the abdominal wall (Table 2) with an average patient age of 27.5 years and maximum age of 65 years, with the youngest being 2 years. The abdominal wall PNET that we examined in this study is the second youngest of all such patients. Only 37 cases of PNET in the vulva were uncovered (Table 3) with an average age of 24.4 years and a maximum age of 65 years. The patient with vulvar PNET examined for this study was only 40 months of age and is the youngest case of vulvar PNET presently reported.

The mechanisms that underlie PNETs are unclear. Roncati *et al*[9] found a biological accumulation of copper, chromium, aluminum, and bismuth in an abdominal wall PNET patient who had been applying abdominal skin cream for a long period; interestingly, tests revealed that the cream contained aluminum and bismuth, suggesting that these metals were acting as intracellular carcinogens. The high magnetic resistance of bismuth might have interfered with cell electromagnetic equilibrium, and particularly impacted the electromagnetic balance found in neurons containing ES/PNET cells[10]. It is worth considering whether heavy metals such as aluminum and bismuth cause the formation of the PNET.

PNET differential diagnosis mainly involves the exclusion of other similar round blue cell tumors, as PNETs and those tumors have some of the same morphological characteristics, which can easily lead to misdiagnosis. Immunohistochemical, reverse transcription-polymerase chain reaction (RT-PCR), and fluorescence *in situ* hybridization (FISH) analyses are necessary for accurate diagnosis. CD99 and vimentin are the most commonly used immunomarkers in the diagnosis of PNET. Both of our cases were CD99-positive, and although Case 1 was negative for vimentin, we did show the translocation of the *EWSR1* gene in tumor cells *via* FISH analysis, confirming the PNET diagnosis. Because both patients' FISH analyses showed *EWSR1*

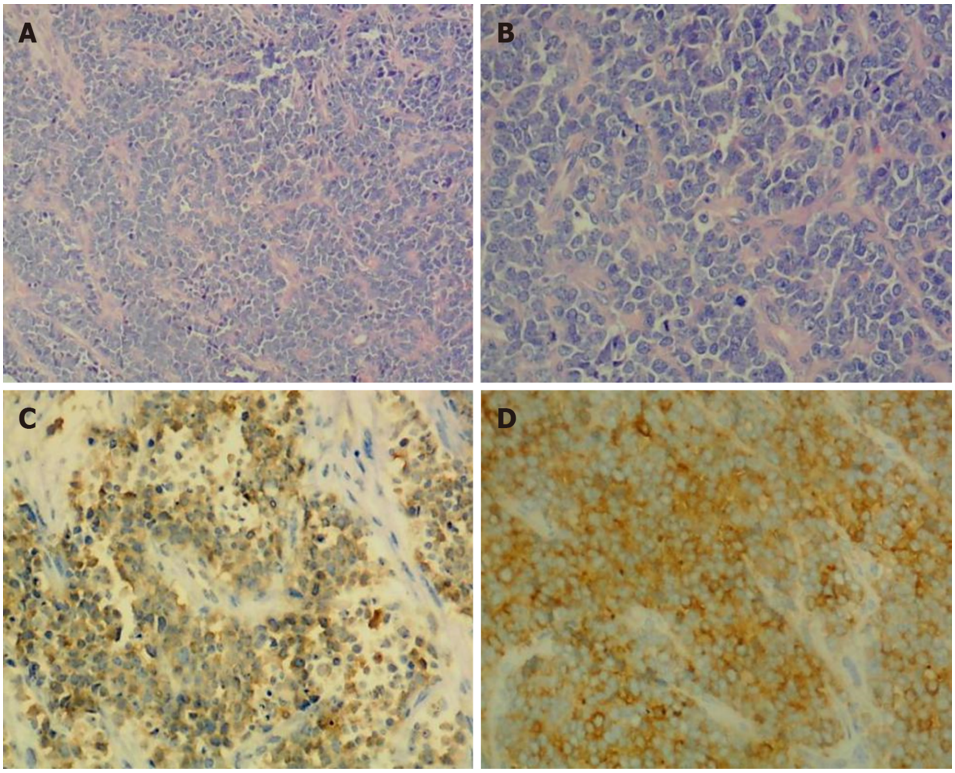

Figure 3 Case 1. A and B: Small round cells with large nuclei and little cytoplasm showing the characteristic perivascular rosette of primitive neuroectodermal tumors (H and E, 100× and 200×); C: Tumor cells positive for CD99 (DAB, 200×); D: Diffuse staining for Syn (DAB, 200×).

gene rearrangement, there was no need to test them by RT-PCR. In recent years, scientists have found high expression levels of cyclin D1 in PNET. A study comparing the expression patterns of cyclin D1 in ES/PNET and rhabdomyosarcoma demonstrated that all PNET patients showed a strong, diffuse, nuclear immune response to cyclin D1, while no expression of cyclin D1 was detected in any patients with rhabdomyosarcoma. Cyclin D1 is another highly sensitive immunological marker for the diagnosis of ES/PNET alongside CD99 and FLI-1 markers, although it is not recommended as an independent diagnostic marker thereof[10].

A main characteristic of PNET[11] is the detection of t (11; 22) (q24; q12) translocation, wherein an EWS-FLI-1 fusion gene is formed. In recent years, scientists have discovered other genetic changes in PNETs. Some researchers found histological and immunohistochemical tumor features consistent with those of PNET in a 15-year-old boy, but no EWSR1 rearrangement was found by FISH and RT-PCR molecular studies. Instead, a translocation of the long arms of chromosomes 18 and 19 was uncovered, resulting in a chromosome t (18; 19) (q23; q13.2) transposition. In addition, some researchers discovered that another translocation in chromosome t (4; 22) (q31n; q12) led to *EWSR1-SMARCA5* gene fusion in patients with PNET.

Because PNET is rare and little research has been performed on it, there is not enough epidemiological and evidence-based medical evidence to derive treatment standards. At present, the main chemotherapeutic drugs used in PNET are vincristine (V), doxorubicin (D), cyclophosphamide (C), actinomycin-D (A), ifosfamide (I), and etoposide (E); our methods of treatment are based on these drugs. Regimens of VAC/IE or VDC/IE are commonly used in chemotherapy, but their chemotherapeutic effects are not always satisfactory. Takigami *et al*[12] reported on a lung PNET case that had been treated with VDC/IE chemotherapy for 5 mo before surgery; after the tumor was resected, it recurred 1.5 mo later. In this case, adriamycin was replaced with actinomycin-D due to a cumulative dose of doxorubicin near 500 mg/m^2. Unfortunately, the tumor grew bigger, and the patient began taking pazopanib (800 mg/d); the tumor shrank four weeks later, and the patient survived for five months, eventually dying due to disease spread. When standard treatment fails, pazopanib can be another effective option. A randomized study of 120 cases of metastatic bone Ewing sarcoma and PNET in the Children's Cancer Group and the Pediatric Oncology Group in the United States showed that adding ifosfamide and etoposide to standard therapy does not improve outcomes for patients with bone Ewing sarcoma or PNET with metastases at diagnosis[13], although the addition of ifosfamide and etoposide to standard therapy can improve the prognosis of patients

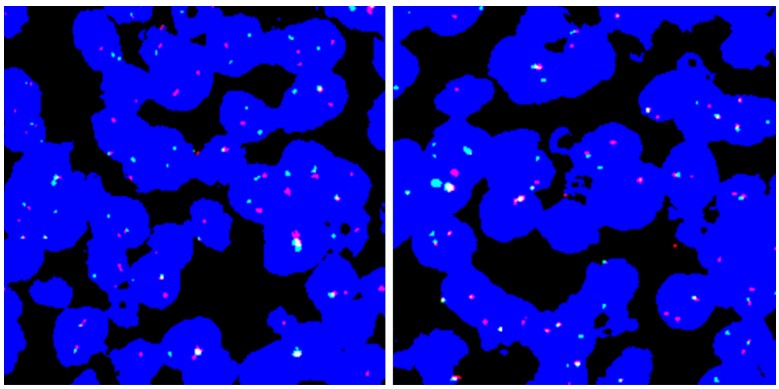

Figure 4 Fluorescent *in situ* hybridization in Case 1. The 5'-terminus of the *EWSR1* gene was labeled with red dye, and the 3'-terminus labeled with green dye. Results showed that the red and green signals were isolated from tumor cells, suggesting translocation of the *EWSR1* gene (DAPI, 1000×).

with no metastatic disease at the time of diagnosis[14]. The survival time of Ewing sarcoma patients was determined by whether the tumor had metastasized or not; a five-year survival rate was 33% in the case of tumor metastasis and 70% in those without metastasis. Extraosseous origin was an adverse prognostic factor for Ewing sarcoma[15-17]. A retrospective study of 975 patients with Ewing sarcoma in the European Intergroup Cooperative Ewing Sarcoma Study Group indicated that the presence of metastasis at diagnosis, exceptionally large tumors (volume ≥ 200 mL or largest diameter ≥ 8 cm), primary tumors located in the axial skeleton (especially the pelvis), and a histological response of less than 100% were strongly associated with poor survival in Ewing sarcoma[15]. The two cases we report on here were extraosseous in origin, but neither of them had metastasis at diagnosis, nor were they large tumors located in the axial skeleton. Case 1 completed standardized treatment and survived without progression for 66 mo; we consider that this is a successful treatment. Case 2, however, died 14 mo after diagnosis; extraosseous origin might have been an adverse prognostic factor, but the most likely cause of death was cessation of treatment.

In this paper, two extremely rare cases of PNET presenting a primary location in the abdominal wall and vulva are presented. A limitation of this report is the small number of cases reported on, which is due to the rarity of PNETs. We also only analyzed the specific diagnostic methods, pathological results, treatment plans, and follow-up. As survival analysis of the disease and prognostic indicators were lacking, specific tumor staging and treatment criteria could not be provided. When diagnosing PNET, immunohistochemistry is often not enough to provide us with satisfactory diagnostic information, and follow-up by FISH or RT-PCR can make the diagnosis more persuasive. For the treatment of PNET, we chose mass resection by surgery when conditions permit, followed by alternating VAC/IE or VDC/IE chemotherapy. We are increasing our efforts to collect more case data and improve diagnostic parameters for the treatment of PNET. Despite these limitations, we hope that our case report will help inform future clinical work.

Table 2 Thirteen reported cases of primitive neuroectodermal tumor in the abdominal wall

Ref.	Age	Sex	Size (cm)	Immuno-histo-chemistry	Molecular/ cyto-genetic analysis	MATOD	Therapy	Follow-up	Relapse	Outcome
Roncati *et al*[9], 2015	45 yr	M	1.5	NSE, CD99 (+)	FISH (+)	NA	NA	NA	NA	NA
Riccardi *et al*[18], 2010	15 yr	M	2.5-3.0	CD99, NB84a, vimentin (+)	FISH, RT-PCR (-)	No	S + Ch	NA	NA	NA
Betal *et al*[19], 2009	61 yr	F	NA	CD99, CD56, cytokeratins, S100 (+)	NA	No	S + Ch (6 cycles of VDC)	NA	No	NA
Taylor *et al*[8], 2000	33 mo	M	3.5 × 3.5 × 2.5	CD99 (+)	NA	No	S	10 yr	No	Survival
Soma *et al*[20], 2015	21 yr	F	6 × 4	CD 99, vimentin (+)	NA	No	S + Ch (VDC)	6 mo	No	Survival
Somers *et al*[21], 2004	16 yr	F	1.5	CD99, CD56, S100 (+)	FISH and RT-PCR (-)	No	Before metastasis: S + Ch (6 cycles of VAC); after metastasis: S + RT + CT (1 cycle of IE + CBP)	NA	Yes	Death
Savić *et al*[22], 2017	15 yr	M	3.8 × 2.6 × 3.7	CD99, vimentin, synaptophysin (+)	RT-PCR (+)	No	S + Ch (VAC) + RT	8m	No	Survival
Askri *et al*[23], 2008	35 yr	F	6.5 × 4	CD99 (+)	NA	NA	S + Ch (3 cycles of VDC)	NA	NA	NA
Gurria *et al*[24], 2011	23 yr	F	14 × 10 × 7	CD99, PAS (+)	NA	No	S + RT + Ch (VAC/IE)	8 mo	No	Survival
Aydinli *et al*[25], 2009	65 yr	M	5	CD99 (+)	NA	NA	S + Ch (6 cycles of VDCE)	1 yr	No	Survival
Wang *et al*[26], 2017	21 yr	F	5 × 4	CD99, vimentin, NSE (+)	NA	Yes	S + RT + Ch (VAC)	7 mo	Yes	Death
Zhan *et al*[27], 2012	2 yr	F	5.0 × 3.8 × 5.1	positive CD99, NSE, Ki67 (+)	NA	No	S + Ch (CTX + ADM + DDP)	1 yr	No	Survival
Present case, 2019	66 mo	F	3.4 × 6.1 × 2	CD99, Syn (+)	NA	No	S + Ch (VDC/IE)	66 mo	No	Survival

+: Positive; MATOD: Metastatic at the time of diagnosis; S: Surgery; Ch: Chemotherapy; RT: Radiotherapy; NA: Not available; FISH: Fluorescence *in situ* hybridization; RT-PCR: Reverse transcription-PCR; VAC: Vincristine and actinomycin D, cyclophosphamide; IE: Ifosfamide and etoposide; VDC: Vincristine, doxorubicin, and cyclophosphamide.

Table 3 Thirty-seven reported cases of primitive neuroectodermal tumor in the vulva

Ref.	Age	Size (cm)	Immunohistochemistry	Molecular/ cytogenetic analysis	MATOD	Therapy	Follow-up	Relapse	Outcome
Present case, 2019	40 mo	3.3 × 5 × 2.5	Vimentin, CD99 (+)	NA	No	S + Ch (VAC/IE)	14 mo	YES	Death
Pei *et al*[28], 2018	33 yr	0.5 × 0.5	PAS, CD99, vimentin (+)	EWSR1 gene (+)	Yes	S + RT + Ch	15 mo	NO	Survival
Chiang *et al*[29], 2017	65 yr	NA	CD99, NSE, SYN, CD56, S100, FLI-1 (+)	FISH (+)	NA	NA	NA	NA	NA

Kakoti *et al*[30], 2017	16 yr	15 × 10	CK, vimentin, CD99, FLI-1 (+)	NA	No	Ch (VDC/IE)	NA	NA	Death
Tunitsky-Bitton *et al*[31], 2015	15 yr	5	CD99 (+)	RT-PCR (+)	No	S + Ch (VDC/IE 14 cycles)	29	NO	Survival
Huang *et al*[32], 2015	20 yr	8 × 10 × 10	CD99, vimentin, NSE (+)	NA	Yes	S	2 wk	NA	Death
Rekhi *et al*[33], 2015	10 yr	12 × 8	MIC2/CD99, FLI-1(+)	FISH (+)	NA	S + Ch (VIME 5 cycles)	18 mo	YES	Survival
Narayanan *et al*[34], 2014	17 yr	3 × 2 × 2	MIC2 (+)	NA	NA	S + RT + Ch (VAC/IE)	22 mo	YES	Death
Matsuda *et al*[35], 2014	60 yr	NA	MIC-2, synaptophysin, NSE, neurofilament antibodies (+)	NA	No	S + RT + Ch (VACI)	48 mo	YES	Survival
Xiao *et al*[36], 2014	20 yr	NA	CD99, NSE (+)	NA	NA	Not done	NA	NA	Death
Xiao *et al*[36], 2014	36 yr	NA	CD99, Syn, NSE (+)	NA	NA	S + Ch (PEI, 4 cycles; PAC, 2 cycles)	13 mo	YES	Death
Che *et al*[37], 2013	37 yr	5 × 3.5 × 3; 3 × 2 × 1.2	CD99, vimentin, FLI-1 (+)	NA	NA	S + Ch (VAC)	12 mo	YES	Survival
Tang *et al*[38], 2012	17 yr	5.5 × 5 × 5	CD99 and FLI-1 (+)	NA	NA	S	NA	NA	LS
Tang *et al*[38], 2012	25 yr	2 × 2 × 2	CD99 and FLI-1 (+)	NA	NA	S	NA	NA	LS
Yang *et al*[39], 2012	20 yr	20 × 10 × 7	CD99 and NSE (+)	RT-PCR (+)	Yes	Ch	NA	NA	Death
Kelling *et al*[40], 2012	18 yr	1.7 × 9 × 1.5	CD99 and vimentin (+)	RT-PCR (+)	Yes	S + RT + Ch	3 mo	NA	Survival
Anastasiades *et al*[41], 2012	28 yr	3	CD99 (+)	NA	No	S + RT + Ch	12 mo	YES	Death
Dong *et al*[42], 2012	20 yr	11 × 7.7 × 6.5	CD99, NSE, CK (AE1/AE3) and Syn (+)	NA	Yes	S	3 mo	NA	Death
Dong *et al*[42], 2012	12 yr	3.1	CD99, NSE, CK (AE1/AE3) and Syn (+)	NA	Yes	NA	13 mo	NA	Survival
Dong *et al*[42], 2012	35 yr	NA	CD99 and NSE (+)	NA	Yes	S + Ch	20m	YES	Death
Halil *et al*[43], 2011	14 yr	NA	CD99 (+)	NA	NA	S + RT + Ch	9 mo	YES	Death
Boldorini *et al*[44], 2010	52 yr	NA	CD99, CK(AE1/AE3) and vimentin (+)	FISH (+)	No	S + RT + Ch(VAI/IE)	12 mo	NO	Survival
Dadhwal *et al*[45], 2010	20 yr	20 × 15 × 10	CD99 (+)	NA	Yes	S	20 d	YES	Death
Cetiner *et al*[46], 2009	23 yr	4 × 4	CD99 and vimentin (+)	RT-PCR (+)	Yes	S + R + Ch (VDC/IE)	7 yr	NO	Survival
Cetiner *et al*[46], 2009	29 yr	NA	CD99 and vimentin (+)	RT-PCR (-)	NA	S + Ch	51 mo	NO	Survival
Fong *et al*[47], 2008	17 yr	0.7 × 0.6 × 0.2; 2.1 × 1.7 × 1.5	CD99 and Fli-1 (+)	RT-PCR (+)	No	S + Ch (VDC)	48 mo	NO	Survival
McCluggage *et al*[48], 2007	19 yr	4	CD99 and FLI-1 (+)	RT-PCR and FISH (-)	NA	S + Ch	NA	NA	NA
McCluggage *et al*[48], 2007	20 yr	6.5	CD99 and FLI-1 (+)	FISH (+)	NA	S	NA	NA	Death
McCluggage *et al*[48], 2007	40 yr	3	CD99, FLI-1 (+)	FISH (+)	NA	S + Ch	12 mo	NA	Survival

Moodley *et al*[49], 2005	26 yr	4 × 5	NA	NA	No	Ch + RT	NA	YES	Death
Takeshima *et al*[50], 2001	45 yr	4 (at recurrence)	Neuron specific enolase, vimentin, HBA 71 (+)	NA	No	S	1 yr (at recurrence)	YES	Survival
Lazure *et al*[51], 2001	15 yr	20	CD99 (+)	RT-PCR (+)	NA	S + Ch	7 mo	NO	Survival
Vang *et al*[52], 2000	28 yr	0.9	CD99 (+)	RT-PCR (+)	NA	S + Ch	18 mo	NA	Survival
Paredes *et al*[53], 1995	29 yr	5	Vimentin (+)	NA	NA	S + RT + Ch (6 cycles of VAC)	8 mo	NO	Survival
Nirenberg *et al*[54], 1995	20 yr	12	PAS (+)	NA	NA	S + RT + Ch (VA)	10 mo	YES	Death
Scherr *et al*[55], 1994	10 yr	6.5 × 5.5 × 2.0	HBA-71 (+)	NA	No	S	NA	NA	NA
Habib *et al*[56], 1992	23 yr	1.5	CK, EMA (+)	NA	NA	NA	NA	NA	NA

+: Positive; MATOD: Metastatic at the time of diagnosis; S: Surgery; Ch: Chemotherapy; RT: Radiotherapy; NA: Not available; FISH: Fluorescence in situ hybridization; RT-PCR: Reverse transcription-PCR; PEI: Cisplatin, ifosfamide, and etoposide; PAC: Cisplatin, cyclophosphamide, and actinomycin D; VAC: Vincristine, actinomycin D, and cyclophosphamide; IE: Ifosfamide and etoposide; VDC: Vincristine, doxorubicin, and cyclophosphamide; VIDE: Vincristine, ifosfamide, doxorubicin, and etoposide; VIME: Vincristine, ifosfamide, mesna, and etoposide; LS: Loss to follow-up.

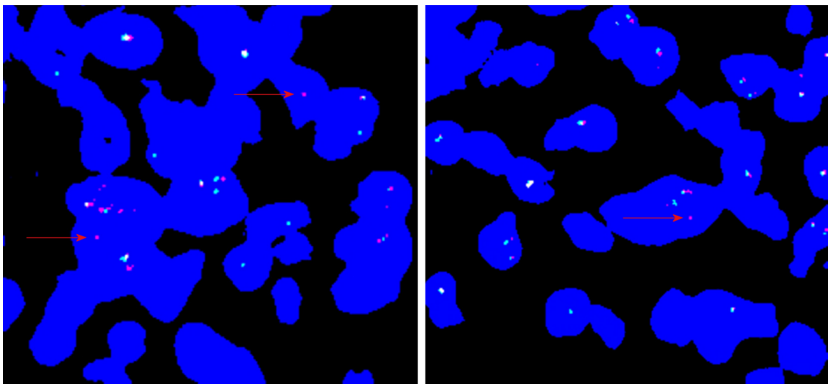

Figure 5 Fluorescent *in situ* hybridization in Case 2. The separation of the red and green signals indicates that the *EWSR1* gene is translocated. Red arrows indicate the translocation change of the *EWSR1* gene (DAPI, 1000×).

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