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Research Letter

Clinical analysis of primitive neuroectodermal tumors in the female genital tract: A report of three cases

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Dear Editor,

Primitive neuroectodermal tumors (PNETs) in the genital tract are very rarely encountered by gynecologists. They are a member of the Ewing sarcoma family. Primitive neuroectodermal tumors encompass a group of embryonic tumors with varying degrees of neuronal differentiation. Peripheral PNETs are small round blue cell tumors with a neuroectodermal origin [1]. Most PNETs of the genital tract occur in the ovary, and more rarely in the vulva, vagina, cervix, and uterus. The literature has only a few case reports and small case series on PNET of the genital tract. In this correspondence, we report three rare cases of PNET with genital tract origin that arose from the vulva, uterus, and ovary, and we assess the clinical characteristics of these neoplasms.

We have diagnosed three cases of PNET of the genital tract, the clinical features of which are described in Table 1. The PNETs had a wide range of age distribution and clinical presentation. The tumors ranged in size from 4 cm to 35 cm.

Medical histories and imaging studies alone are inadequate for a definitive diagnosis of PNET. Primitive neuroectodermal tumors can be diagnosed via histological investigation of a sample. It is characterized as small round cells with the formation of pseudo-rosettes and Homer–Wright rosettes [2]. Endometrial stromal sarcoma, granulose cell tumors, carcinoid tumor, rabdomyosarcoma, neuroblastoma, osteosarcoma, Wilms tumor, and malignant melanoma should be considered in the differential diagnosis Distinction between these tumors can be very difficult without additional studies such as histochemistry and electron microscopy. An exact differential diagnosis can be determined with the assistance of immunohistochemical studies. The transmembrane protein CD99 is a very useful and sensitive immune histochemical marker; however, other small round blue cell tumors such as small cell carcinoma, rhabdomyosarcoma, and lymphoma can also show immunoreactivity to CD99. For most patients, routine histologic and immunohistochemical examination is sufficient to render a correct diagnosis of PNET; however, genetic analysis is very helpful when morphology is inconclusive or when these tumors appear in an unusual clinical setting.

There is no standard treatment regimen for peripheral PNET because of the rare number of cases, which appear in different locations. Treatment for peripheral PNET has included complete surgical excision of the lesion, chemotherapy, and, occasionally, radiation therapy. Local surgery or radiotherapy alone has a high risk of mortality and recurrence. Some authors have advocated 6 weeks of chemotherapy before and after surgery and radiotherapy to increase disease-free survival rates [3]. Chemotherapy regimen consisting of iphosphomide, doxorubicin, cyclophosphomide, etoposide and cysplatin can be used in the treatment. However, certain protocols cannot be established for these rare cases because of their sporadic nature.

After 18 months of follow up without any recurrences, we lost contact with one of the three patients. The other two patients have shown no signs of recurrences during their follow up. All three patients did not indicate any serious complications with the surgery or the chemotherapy regimen.

In summary, peripheral PNET in the pelvic region is a rare and aggressive tumor with a high rate of recurrence. It clinically resembles a benign condition. For patients with heterogeneous large volume masses, PNET should be considered in the differential diagnosis. Advanced pathological investigation techniques

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Table 1
Clinical presentation, treatment, and follow up of primitive neuroectodermal tumors in the female genital tract.

Case No.	Age (y)	Disease site	Clinical presentation	Surgery	Cd99	S100	Vimentin	Nse	Cd117	Additional therapy	Follow up
1	42	Vulva	Vulvar mass	Bilateral radical vulvectomy & femoral lymph node dissection + chemotherapy + radiotherapy	(+)	(+)	(+)	(−)	(−)	Carboplatin + adriamycin + external radiotherapy	18 mo disease-free (last information in Feb 2014)
2	51	Uterus	Pelvic pain	Mass excisions, left hemicolectomy, appendectomy, & omentectomy	(+)	(−)	(+)	(+)	(−)	Etaposide & cycplatine	14 mo disease-free (no signs of recurrence at her last visit in Jul 2014)
3	33	Ovary	Abdominal tenderness	Mass excision	(+)	(−)	(+)	(−)	(+)	Adriamycin + cisplatin + external radiotherapy	10 mo disease-free (no signs of recurrence at her last visit in Jun 2014)

should be performed for a definitive diagnosis. A definitive treatment regimen can be developed by the presentation of cases and their prognosis in the literature.

Conflicts of interest

The authors have no conflicts of interest relevant to this article.

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