



Primary Ewing Sarcoma Presenting as a Vulvar Mass: A Case Report

Xu Guocai* and Zhang Bingzhong

Department of Gynecologic Oncology, Sun Yat-sen Memorial Hospital, Sun Yat-sen University, China

Abstract

A 33-year-old woman presented with a 5-month history of left vulva mass. Pathological biopsy diagnosis was Ewing's sarcoma accidentally. PET/CT revealed there were many solid masses distributed in left adnexa area and the pelvic wall with the left groin multiple lymph node metastasis. The patient was treated with three cycles of neoadjuvant therapy before the surgery. And the combined chemotherapy and radiotherapy were given postoperatively. Adjuvant chemotherapy consisting of epirubicin and ifosfamide. She also received radiation therapy for local control. The patient is currently alive without any evidence of recurrence or metastasis.

Keywords: Ewing's sarcoma; Chemotherapy; Radiation therapy

Introduction

Ewing Sarcoma (ES) and Primitive Neuro Ectodermal Tumor (PNET) are a group of tumors that generally affect the skeletal system. Extra osseous ES and PNET presenting in the genitourinary tract are highly uncommon. Few cases of primary vulvar and vaginal cases of ES have been published to date. We present a case of a primary ES in the labium minorum with pelvic metastasis.

Case Presentation

A 33-year-old woman presented with a 5-month history of left vulva mass without fever and pain. The local hospital treated the mass as a Bartholin's cyst, and performed a fistulation surgery, but the suspected diagnosis was Ewing's sarcoma accidentally. The later Positron emission tomography-computed tomography (PET/CT) revealed there were many solid masses distributed in left adnexa area and the pelvic wall, with the left groin multiple lymph node metastasis. A plan was made for three cycles of neoadjuvant therapy (EI, Epirubicin 100 mg+ Ifosfamide 7.5 g) and then local control with surgery in our hospital, considering its multiple metastatic lesions. The left internal iliac artery embolization was performed to control severe hemorrhage before the surgery. After the surgery, another three cycles of chemotherapy (EI, Epirubicin 100 mg+ Ifosfamide 7.5 g) were given. Post treatment pelvic MRI showed solid masses distributed in left adnexa area and the pelvic wall. So another surgery of left pelvic mass resection and hysterectomy with left adnexectomy plus right salpingectomy and right ovarian transposition was given. Brachytherapy was planned followed by one additional cycles of chemotherapy (EI, Epirubicin 100 mg+ Ifosfamide 7.5 g). The patient is currently alive without any evidence of recurrence or metastasis.

Discussion

EWS is a highly malignant, small, round cell tumor that originates from the primitive neuro ectodermal cells. It was first described by James Ewing in 1921 [1]. There is a slightly male predominance with a male-to-female ratio of 1.5:1 [2]. In general, approximately 20% to 30% of EWS patients present with metastases at the time of their diagnosis [3-4]. EWS has extremely low incidence in the vulva of the female genital tract. Here we report a case of EWS of the vulva in a 33-year-old woman and give a review of this rare disease.

The main morphology of EWS is small round cells, ill-defined borders, scanty cytoplasm, and high mitotic index. CD99, a cell surface glycoprotein, and FLI-1, a DNA-binding transcription factor, markers are positive in the majority of ES/PNET neoplasms [5]. CD99 is expressed with a membranous distribution in virtually all neoplasms in the ES/pPNET and has been demonstrated to be extremely useful in diagnosis of cases [6]. Detecting FLI-1 protein expression by immunohistochemical stain is another useful method to diagnose ES/pPNET. Because the genetic characteristic of ES/pPNET is the presence of the translocation t (11;22) (q24; q12), which creates

OPEN ACCESS

*Correspondence:

Xu Guocai, Department of Gynecologic Oncology, Sun Yat-sen Memorial Hospital, Sun Yat-sen University, China, E-mail: xuguocai1018@126.com

Received Date: 19 Aug 2018

Accepted Date: 15 Sep 2018

Published Date: 29 Sep 2018

Citation:

Guocai X, Bingzhong Z. Primary Ewing Sarcoma Presenting as a Vulvar Mass: A Case Report. Clin Oncol. 2018; 3: 1529.

Copyright © 2018 Xu Guocai. This is an open access article distributed under the Creative Commons Attribution License, which permits unrestricted use, distribution, and reproduction in any medium, provided the original work is properly cited.

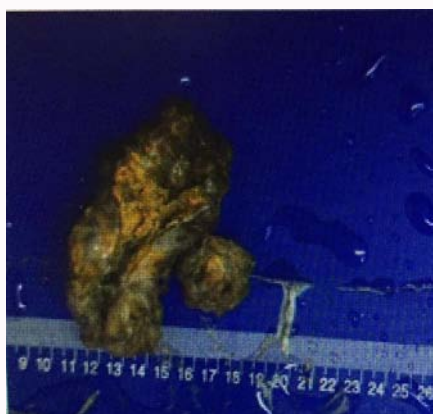


Figure 1: A mass of labium minorum after complete resection.

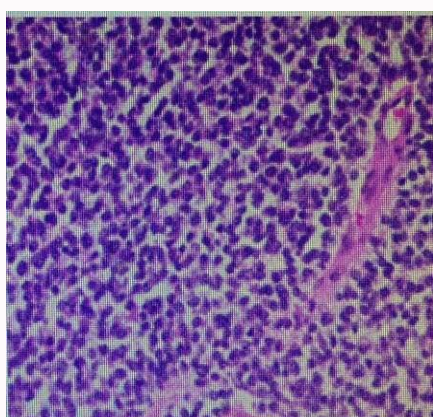


Figure 2: Histopathology slide demonstrating the typical appearance of ES.

the EWS/FLI1 fusion gene and results in the expression of a chimeric protein. The sensitivity of FLI-1 was greater and specificity was more than 90% [7]. Our reported case presented both CD99 and FLI-1 staining positive, strongly suggesting that it is a case of EES.

We summarized the clinical details of previously described cases involving the vulva together with our cases [8-11]. EES usually occur in relatively young women, a similar age of predilection for these neoplasms when they involve more usual sites. In all the reported cases, the mean age at the time of the initial diagnosis of previous cases is 27.6 years (median 26, range 10-52), but our patient was older than this [12].

The treatment of ES requires a multidisciplinary approach. In addition to surgical resection, the patients are typically treated with multiagent chemotherapy and, occasionally, with radiation therapy to improve local control. In the cases of vaginal ES, it is unclear if a hysterectomy is indicated. In two reported cases, hysterectomy and partial vaginal excision were performed as part of the treatment [13,14]. These women were 30 and 39 years old and had completed childbearing. Given the lack of clear recommendations, this approach may not be warranted in a younger woman interested in future childbearing.

Because extra osseous ES is rare, the treatment guidelines for adjuvant therapy mirror the recommendations for osseous ES. The multiagent standard chemotherapy regimen typically includes doxorubicin, vincristine, and cyclophosphamide, alternating with ifosfamide and etoposide [15]. Neoplasms in the EES were thought to

be aggressive with poor prognosis. Pelvic ES/PNET has an unfavorable prognosis and survival rate, particularly in the case of metastatic disease [16,17]. In the absence of metastatic disease, primary vulvar and vaginal ES neoplasms likely have a more favorable prognosis. Our patient underwent surgery, and then received six cycles chemotherapy. About 23 months after diagnosis, she is alive and well. This relatively favorable outcome may be because they usually arise in cutaneous or superficial sites, with smaller size, superficial location, early detection, and complete surgical removal of the lesion. Given the infrequent nature of this disease and a relatively short follow-up, it is difficult to comment on long-term survival and recurrence rates.

In conclusion, ES/PNET tumors are exceedingly rare in the female tumor. But it should be important to consider them as part of the differential diagnosis of a vulvar or vaginal mass. Complete surgical resection should be the first step in managing a firm mass in the vulva or vagina. Appropriate molecular and histopathological studies are crucial in determining the correct diagnosis. Specific treatment recommendations are lacking for extraosseous ES, and the treatment management should follow the recommended therapy for osseous ES, which includes complete resection, multiagent chemotherapy, and, possibly, the addition of local radiation therapy.

References

1. Ewing J. Diffuse endothelioma of bone. *Pathol Soc.* 1921;21:17-24.
2. Yeshvanth SK, Ninan K, Bhandary SK, Lakshinarayana KP, Shetty JK, Makannavar JH. Rare case of extra skeletal Ewings sarcoma of the sinonasal tract. *J Cancer Res Ther.* 2012;8(1):142-4.
3. Cotterill SJ, Ahrens S, Paulussen M, Jürgens HF, Voûte PA, Gadner H, et al. Prognostic factors in Ewing's tumor of bone: analysis of 975 patients from the European Intergroup Cooperative Ewing's Sarcoma Study Group. *J Clin Oncol.* 2000;18(17):3108-14.
4. Bacci G, Longhi A, Ferrari S, Mercuri M, Versari M, Bertoni F. Prognostic factors in non-metastatic Ewing's sarcoma tumor of bone: an analysis of 579 patients treated at a single institution with adjuvant or neoadjuvant chemotherapy between 1972 and 1998. *Acta Oncol.* 2006;45(4):469-75.
5. McCluggage WG, Sumathi VP, Nucci MR, Hirsch M, Dal Cin P, Wells M, et al. Ewing family of tumors involving the vulva and vagina: report of a series of four cases. *J Clin Pathol.* 2007;60(6):674-80.
6. Stevenson A, Chatten J, Bertoni F. CD99/p30/32/MIC2/neuroectodermal/Ewing's sarcoma antigen as an immunohistochemical marker: Review of more than 600 tumors and the literature experience. *Appl Immunohistochem.* 1994;2:231-40.
7. Folpe AL, Hill CE, Parham DM, O'Shea PA, Weiss SW. Immunohistochemical detection of FLI-1 protein expression: A study of 132 round cell tumors with emphasis on CD99- positive mimics of Ewing's sarcoma/primitive neuroectodermal tumor. *Am J Surg Pathol.* 2000;24(12):1657-62.
8. Vang R, Taubenberger JK, Mannion CM, Bijwaard K, Malpica A, Ordonez NG, et al. Primary vulvar and vaginal extra osseous Ewing's sarcoma/peripheral neuroectodermal tumor: Diagnostic confirmation with CD99 immunostaining and reverse transcriptase-polymerase chain reaction. *Int J Gynecol Pathol.* 2000;19(2):103-9.
9. Habib K, Finet JF, Plantier F. A rare lesion of the vulva. *Arch Anat Cytol Pathol.* 1992;40:158-9.
10. Lazure T, Alsamad IA, Meuric S, Orbach D, Fabre M. Primary uterine and vulvar Ewing's sarcoma/peripheral neuroectodermal tumors in children: Two unusual locations. *Ann Pathol.* 2001;21(3):263-6.
11. McCluggage WG, Sumathi VP, Nucci MR, Hirsch M, Dal Cin P, Wells M, et al. Ewing family of tumours involving the vulva and vagina: Report of a

- series of four cases. *J Clin Pathol*. 2007;60:674-80.
12. Shao-Min Che, Pei-Long Cao, Hong-Wei Chen, Liu Z, Meng D. Primary Ewing's sarcoma of vulva: A case report and a review of the literature. *J Obstet Gynaecol Res*. 2013;39(3):746-9.
13. Liao X, Xin X, Lu X. Primary Ewing's sarcoma-primitive neuroectodermal tumor of the vagina. *Gynecol Oncol*. 2004;92(2):684-8.
14. Bancalari E, de Alava E, Tardio JC. Primary vaginal Ewing sarcoma: case report and review of the literature. *Int J Surg Pathol*. 2012;20(3):305-10.
15. Grier HE, Krailo MD, Tarbell NJ. Addition of ifosfamide and etoposide to standard chemotherapy for Ewing's sarcoma and primitive neuroectodermal tumor of bone. *N Engl J Med*. 2003;348:694.
16. Al-Tamimi H, Al-Hadi AA, Al-Khater AH, Al-Bozom I, Al-Sayed N. Extra skeletal neuroectodermal tumour of the vagina: a single case report and review. *Arch Gynecol Obstet*. 2009;280(3):465-8.
17. Tunitsky-Bitton E, Uy-Kroh MJ, Michener C, Tarr ME. Primary Ewing Sarcoma Presenting as a Vulvar Mass in an Adolescent: Case Report and Review of Literature. *J Pediatr Adolesc Gyn*. 2015;28(6):e179-83.