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Article begins on next page

Lymphoma Presenting as a Mass of the Vulva-report of a case of a rare vulvar neoplasm not treated by surgery

Qing Wang, MD*, Bernadette Cracchiolo, MD, MPH**, Debra Heller, MD*,**

From the Departments of Pathology & Laboratory Medicine*, and Obstetrics, Gynecology, and Women's Health**, Rutgers-New Jersey Medical School, Newark, NJ

Address Correspondence to:

Debra S. Heller, MD

Dept of Pathology-UH/E158

Rutgers-New Jersey Medical School

185 South Orange Ave

Newark, NJ, 07103

Tel 973-972-0751

Fax 973-972-5724

hellerds@njms.rutgers.edu

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Tables 0, Figures 2

Institutional Review Board approval is not required at our institution for case reports.

Precis:

Vulvar lymphoma is exceedingly rare, and must be considered in the differential diagnosis of a blue cell tumor.

Keywords: vulvar neoplasms, lymphoma, vulva

Introduction:

Vulvar lymphomas are exceedingly rare, and may be primary or part of systemic disease. They fall under the category of “small round blue cell tumor”, which includes a large number of neoplasms that need to be distinguished, but appear similar on hematoxylin and eosin stain. We present a case and discuss this entity. Only consent for treatment was obtained from this patient, and therefore both the description and the gross image have been de-identified to protect privacy.

Case Report

A woman in her 70s was seen for new onset vulvar mass. Her history was significant for pelvic exenteration 5 years prior to the current presentation. At that time, she had been diagnosed with poorly differentiated adenocarcinoma on biopsy of a cervical mass, where endometrial primary was favored over cervical primary based on immunohistochemistry and morphology, and she was diagnosed as Stage II endometrial adenocarcinoma. The tumor was determined at that time to be unresectable and she underwent radiation therapy, for a total of 4500 cGy. There was no significant clinical response, and she subsequently underwent the exenteration a few months after radiation. Evaluation of the exenteration specimen revealed that the tumor showed some treatment effect, and invaded about 75% of the endometrium (with foreign body giant cell reaction, which may have been prior tumor, going deeper into myometrium), and half the cervix, however a profuse foreign body giant cell reaction on the rectosigmoid was suggestive of prior tumor. Pelvic lymph nodes were negative.

During the current evaluation, the patient was seen to have a 3 cm polypoid right labia majora mass with intact overlying skin. The clinical impression was metastatic endometrial adenocarcinoma to the vulva, and a biopsy was taken. After the results of the vulvar biopsy disclosed lymphoma, she underwent subsequent PET-CT which showed widely disseminated systemic disease, including in lymphoid regions in the pelvis, mesentery, and upper abdomen, and bone involvement including clavicle, rib, vertebrae in thorax and lumbar regions, femur and humerus, interpreted as consistent with disseminated lymphoma. The findings were compatible with stage IV lymphoma. She is currently undergoing chemotherapy.

Pathology:

The H&E stained histologic sections of the right labial mass biopsy showed a diffuse subcutaneous infiltration of medium-to-large sized atypical lymphoid cells with round nuclear contours, dispersed chromatin, small nucleoli and scant cytoplasm. There are numerous mitotic figures and apoptotic bodies. There were scattered tinged body macrophages with a starry-sky appearance (figures 1,2). Immunohistochemistry was performed, and the neoplastic lymphoid cells were positive for CD20, PAX-5, CD10, Bcl-6, CD45, vimentin (subset), but negative for Bcl-2, cyclin D1, CD3, CD56, CD99, pan cytokeratin, CK20, synaptophysin, chromogranin, FLi-1, HMB45, HHF35 and EMA. Fluorescence In Situ Hybridization (FISH) was negative for rearrangement of bcl-2, c-myc, and bcl-6 genes. The diagnosis rendered was diffuse large B-cell lymphoma, germinal center B-cell type, with a high proliferation index (95-100%) based on Ki-67 stain.

Discussion:

Non-Hodgkin lymphoma (NHL) is a group of lymphoid cancers that includes all types of lymphoma except Hodgkin lymphoma. Diffuse large B-cell lymphoma is a subtype of NHL. Non-Hodgkin's lymphoma (NHL) of systemic origin can rarely involve the vulva (1), although systemic lymphoma is much more likely to spread to the ovary (2). Primary vulvar lymphomas are exceptionally rare, with only a few cases reported, and even fewer of them diffuse large B-cell lymphoma (2,3). In a literature review of 29 primary vulvar NHLs (2), only 8 were diffuse large B cell lymphomas. In a series of 6 cases of NHL involving the vulva, two diffuse large B cell lymphomas were thought to have arisen primarily in the vulva (3). Women in Vang's series (3) were 40 to 70 years old, slightly younger than our case.

The classification of lymphomas is complex, and not all of the reported cases were fully classified. There are not enough cases reported to assess prognosis or optimal therapy. What is important is that surgery is not the primary mode of therapy for lymphomas, where chemotherapy is generally first line therapy. Hence it is imperative to establish the correct diagnosis. Making the correct diagnosis is complicated by the fact that lymphomas belong to a group of neoplasms described in pathology as "small round blue cell tumors" for their similar appearance on routine histology. Immunohistochemistry and molecular genetic studies are utilized in distinguishing the neoplasms, which can include lymphoma, rhabdomyosarcoma, Ewing's sarcoma/Peripheral neuroectodermal tumor (PNET), and Merkel cell tumor, an aggressive neoplasm of skin, all of which can rarely occur on the vulva. Poorly differentiated squamous cell carcinomas, adenocarcinoma, melanoma and skin adnexal malignancies must also be considered (3). Underdiagnosis with interpretation of the lesion as inflammatory is also possible (2). There has

been some evidence that prior radiation therapy may slightly increase the risk of developing subsequent NHL(4).

We present a case of NHL of the vulva, initially thought clinically to be metastatic uterine adenocarcinoma. As the tumor was found to be disseminated shortly after, the vulva may or may not be the primary site of tumor. It is possible that systemic lymphoma was first detected as a vulvar mass. It is important for clinicians and pathologists to be aware of the possibility of vulvar NHL, which has a wide differential diagnosis. As it is an unusual neoplasm in this area, involvement of clinicians experienced in the treatment of lymphoma would be prudent in these cases.

Abbreviations and Acronyms

CD-Cluster of differentiation

FISH- fluorescence In Situ Hybridization

PAX-Paired box gene

Fli-1- Friend leukemia integration 1 transcription factor

Tdt- Terminal deoxynucleotidyl transferase

Ki-67- antigen identified by monoclonal antibody Ki-67

Hhf35- muscle-actin-specific monoclonal antibody

C-MYC-cellular MYC(a protooncogene)

EMA-epithelial membrane antigen

Bcl-2- B-cell lymphoma 2

PNET- Peripheral neuroectodermal tumor

PET-CT-Positron Emission Tomography - Computed Tomography

NHL-non-Hodgkin lymphoma

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