

Metastatic genital tract adenocarcinoma to the vulva: a rare occurrence presenting as painful vulvar enlargement

Rutgers University has made this article freely available. Please share how this access benefits you.
Your story matters. <https://rucore.libraries.rutgers.edu/rutgers-lib/48966/story/>

This work is an **ACCEPTED MANUSCRIPT (AM)**

This is the author's manuscript for a work that has been accepted for publication. Changes resulting from the publishing process, such as copyediting, final layout, and pagination, may not be reflected in this document. The publisher takes permanent responsibility for the work. Content and layout follow publisher's submission requirements.

Citation for this version and the definitive version are shown below.

Citation to Publisher Heller, Debra & Cracchiolo, Bernadette. (2016). Metastatic genital tract adenocarcinoma to the vulva: a rare occurrence presenting as painful vulvar enlargement. *Journal of Lower Genital Tract Disease* 20(2), e16-e17. <http://dx.doi.org/10.1097/LGT.000000000000187>.

Citation to this Version: Heller, Debra & Cracchiolo, Bernadette. (2016). Metastatic genital tract adenocarcinoma to the vulva: a rare occurrence presenting as painful vulvar enlargement. *Journal of Lower Genital Tract Disease* 20(2), e16-e17. Retrieved from [doi:10.7282/T30P1223](https://doi.org/10.7282/T30P1223).

Terms of Use: Copyright for scholarly resources published in RUcore is retained by the copyright holder. By virtue of its appearance in this open access medium, you are free to use this resource, with proper attribution, in educational and other non-commercial settings. Other uses, such as reproduction or republication, may require the permission of the copyright holder.

Article begins on next page

Metastatic Genital Tract Adenocarcinoma to the vulva-a rare occurrence presenting as painful vulvar enlargement.

Debra Heller, MD*,** Bernadette Cracchiolo, MD, MPH.**

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

Word count: Précis:22, Body of Text: 765

Running title: Adenocarcinoma vulvar metastasis

Disclosures: none

Conflicts of interest: none

Tables 0, Figures 3

Precis:

Vulvar metastasis of genital tract adenocarcinoma is extremely rare, and highlights the need for liberal vulvar biopsy and knowledge of prior history.

Keywords: Endometrial neoplasms, neoplasm metastasis, vulva

Abstract:

Objectives: Pathologist-clinician communication has been an ongoing topic in the literature. Pathology reports are geared to assisting clinicians with patient therapy, but sometimes there are barriers to communication. This survey aims to explore clinicians' understanding of their pathology reports in the membership of the International Society for Vulvovaginal Disease(ISSVD).

Methods: An email survey was sent to all members of the ISSVD.

Results: Surveys were emailed to 397 members, with 91 responding(23 %). Most(76%) of the respondents were gynecologists, with 13% dermatologists and 6% advanced practice nurses. 40% of respondents did not always understand their pathology reports, 62% did not know the difference between levels and recuts, 71% were unclear as to why levels rather than recuts would be ordered, and 26% was not familiar with the term "spongiosis". Over 94% read the gross description on a pathology report. Over 90% reported speaking with their pathologist, which they considered important. They considered having a pathologist with specialty expertise important.

Conclusions: Clinician members of the ISSVD are particularly attuned to the importance of pathology consultation in the care of women with vulvovaginal conditions. There are still areas for potential improvement in educational efforts, particularly providing information on how pathology laboratory processes may impact the report, as well as in further education in dermatopathology terminology for those unfamiliar.

Introduction

Adenocarcinoma of the vulva is uncommon, and usually arises from Paget's disease, sweat glands, or anogenital mammary-like tissue. Adenocarcinoma of Bartholin's gland is in the differential but is often diagnosed late, after the tumor has overgrown the characteristic criteria. Metastatic adenocarcinoma to the vulva from an endometrial or cervical primary is rare. We describe a case, presenting with painful vulvar enlargement. High index of suspicion, knowledge of prior history by both clinician and pathologist, and liberal biopsy are needed to establish the diagnosis.

Case Report

A woman in her mid-60s presented to the Emergency Department(ED) with vaginal bleeding and severe vulvar pain. She had been treated one year prior to admission outside the United States with a supracervical hysterectomy for postmenopausal bleeding, and diagnosed with grade 2 Stage IB endometrial adenocarcinoma. She was referred for adjuvant radiation, but did not comply. Shortly prior to the current admission, she developed irregular vaginal spotting. This progressed to profuse bleeding, maceration of the vulvar epithelium, vulvovaginal swelling and bilateral lower limb edema. The patient was evaluated by a gynecologist outside our institution, and a pap smear reportedly showed glandular cells worrisome for endocervical adenocarcinoma. Human papillomavirus testing was negative, and the ultrasound at that time revealed a prominent large cervical stump. The patient presented to our ED, with an enlarged inguinal lymph node and pronounced diffuse swelling of both labia majora with firm induration, extending to her upper thighs. She could not tolerate examination. Examination under anesthesia was performed and the impression was of tumor recurrence encompassing the

entire labia majora, the distal urethra, and the vagina. The cervix could be palpated as also having tumor. On rectal examination, there was a tumor nodule extending through the muscularis, and a 2 cm right inguinal lymph node. She underwent biopsies of the right and left labia majora. Computerized axial tomography showed multiple pulmonary metastases. She underwent palliative chemoradiation but expired shortly thereafter.

Pathology: Overlying native vulvar squamous epithelium was uninvolved (figure 1), consistent with metastatic disease. Histology showed poorly differentiated adenocarcinoma, with focal squamoid features (figure 2). The tumor showed 5% staining with estrogen receptor, and was negative for progesterone receptor. The lesions stained for epithelial-related antigen (MOC-31, monoclonal antibody 31), consistent with adenocarcinoma, and with vimentin, consistent with endometrial origin. p16 staining was noted, and p53 was strongly positive, which can sometimes be seen in poorly differentiated endometrial carcinoma. Carcinoembryonic antigen was focally positive. Hence the immunoprofile overlapped, but given the prior history, endometrial was favored, while cervical carcinoma could not be ruled out.

Discussion

Vulvar adenocarcinoma is uncommon. When it does occur, it is usually of primary origin, arising as invasive Paget's disease, sweat gland adenocarcinomas, or breast-like adenocarcinomas arising in anogenital mammary-like tissue(1). Bartholin's gland adenocarcinomas are also in the differential.

Metastatic carcinoma to the vulva is rare, representing only about 5-8% of vulvar carcinomas(2). In a series of 66 patients, the most common presenting symptoms were mass, pain, and ulceration(3). Gynecologic and non-gynecologic primaries were about evenly distributed(46.9% vs 43.9%), with a few tumors of unknown origin(3). Breast cancer has recurred in the vulva as much as 20 years after initial diagnosis(4), demonstrating the importance of knowing prior history, and supplying it to the evaluating pathologist. The lack of an in-situ component of the overlying epithelium, as seen in our case, is also helpful as well in suspecting metastatic disease(3). Other sites of origin of carcinomas metastatic to vulva include cervix(3), vagina(3),ovary(3,5), colorectal and anal(3)(figure 3), appendiceal(6), cutaneous melanoma, lung, lymphoma, bladder, urethra, and pancreas(3),

Metastatic endometrial and endocervical adenocarcinoma to the vulva are extremely rare. In Neto's series(3), 6(9%) of cases arose in the endometrium, and 23% from cervix. Giordano et al(2) reviewed the literature, and found 13 reported endometrial cases, presenting with mass, pain, ulceration and bleeding. The majority succumbed to their disease. In one highly unusual case, the lesion metastasized to a squamous cell carcinoma of the vulva(7).

While the current patient's diagnosis was unsurprising, based on the suboptimal therapy she had undergone prior to arrival at our institution, it is important to consider the full prior history of patients presenting with a vulvar lesion suspicious for neoplasia. It is also important to provide that history to the pathologist. Adenocarcinoma of unknown primary is a common diagnostic problem in medicine, and pathologists are able to narrow down the likely primary sites with a

focused battery of immunohistochemical stains based on history. Vimentin, estrogen receptor, CEA and p16 are helpful in distinguishing endometrial adenocarcinoma from endocervical adenocarcinoma, but there is overlap, as seen here. For clinicians, biopsy of all suspicious lesions is important.

Abbreviations and Acronyms

ED- Emergency Department

CM-centimeter

MOC-31- Monoclonal antibody 31,

References:

1-Kajal B, Talati H, Daya D, Alowami S. Apocrine adenocarcinoma of the vulva. *Rare Tumors* 2013;5:e40

2-GiordanoG, Gnetti L, Melpignano M. Endometrial carcinoma metastatic to the vulva: a case report and review of the literature.*Pathol Res Pract* 2005;201:751-6.

3-Neto AG, Deavers MT, Silva EG, Malpica A. Metastatic tumors of the vulva: a clinicopathologic study of 66 cases. *Am J Surg Pathol.* 2003;27:799-804.

4-Alligood-Perccoco NR, Kessler MS, Willis G. Breast cancer metastasis to the vulva 20 years remote from initial diagnosis: a case report and literature review. *Gynecol Oncol Reports* 2015;13:33-5.

5-Achimas-Cadariu P, Vlad C, Fetica B, Zgaia A, Cainap C. Unusual skin metastasis in a patient with recurrent micropapillary serous carcinoma-a case report and review of the literature. *Clujul Med* 2015;88:237-40.

6-Cheung KW, Cheung VY. Recurrence of carcinoma of the appendix presenting as vulvar swelling: a case report. *J Obstet Gynaecol Can* 2014;36:904-6.

7-Wimmer JL, Coffey DM, Kaplan AL, Ayala AG, Ro JY. Tumor-to-tumor metastasis with endometrial carcinoma metastatic to squamous cell carcinoma of vulva. *Arch Pathol Lab Med* 2013;137:1825-1828.

Legends:

Figure 1-Overlying vulvar squamous epithelium showed no evidence of neoplasia

Figure 2-The tumor was poorly differentiated, and focally squamoid, with areas of glandular differentiation(inset)

Figure 3-A case of metastatic colon cancer to the right labium majus. Courtesy Micheline Moyal-Barracco, MD



