Angiosarcoma of the vulva

Ashley Lafever, MD,[1] Eric Tyler, MD,[1] Colleen K Stockdale, MD, MS[1]

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Abstract

Objective: Angiosarcoma is a rare neoplasm typically presenting in the skin and superficial soft tissues, with an equal female-to-male ratio. We present a woman with recurrent vulvar lesions and a history of melanoma found to have angiosarcoma.

Case: A 67-year-old woman presented with a seven month history of recurrent vulvar lesions with symptoms of discomfort and bleeding. The histopathological diagnosis revealed high-grade angiosarcoma. Computed tomographic imaging with contrast and positron emission tomography revealed disseminated disease. She underwent chemotherapy and radiation but died within one year of diagnosis.

Conclusions: The prognosis for patients with angiosarcoma is generally poor, especially with disseminated disease. Early detection remains the key element in maximizing patient outcomes.

[1]University of Iowa Hospitals and Clinics, Department of Obstetrics and Gynecology, Iowa City, Iowa

Introduction

Angiosarcomas comprise less than 2% of all sarcomas, are commonly aggressive, and disseminate widely.[1-3] They often present with a bruise-like appearance which can progress to nodular and ulcerated lesions. While angiosarcomas can arise anywhere in the body, they rarely occur in the female genital tract.[4] The lung and liver are the most common sites of metastases.[1] Primary angiosarcomas arise without recognized associated risk factors.[2] Certain familial syndromes such as Li Fraumeni and neurofibromatosis, as well as environmental factors such as radiation exposure, chronic inflammation, and foreign body material are associated with development of secondary angiosarcomas.[1,2,4,5] We present a case of angiosarcoma occurring in a woman with a history of melanoma.

Case Report

A 67-year-old gravida 5 para 4 woman presented to our tertiary referral center with a seven month history of recurrent vulvar lesions. She reported onset and recurrence of a single vulvar lesion following incision and drainage on two occasions; once by her dermatologist

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Corresponding author: Colleen K. Stockdale, MD, MS, University of Iowa Hospitals and Clinics, Department of Obstetrics and Gynecology, 200 Hawkins Drive, Iowa City, IA 52242. Telephone: 319-384-6849. Fax: 319-384-8620. colleen-stockdale@uiowa.edu.

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and once by her gynecologist. Subsequently, she noted the presence of additional similar lesions which prompted referral for further evaluation given a history of melanoma excised from her back four years previously.

At her initial office visit with our clinic, the patient reported intermittent, mild to moderate vulvar discomfort as well as bleeding from the lesions. Her history was notable for: total abdominal hysterectomy and bilateral salpingo-oophorectomy for symptomatic fibroid uterus in 2010, normal Pap smear three years prior with no history of abnormal pap smears, and melanoma treated with excisional biopsy four years prior. Her general physical examination was remarkable only for multiple black, protuberant vulvar lesions, concerning for melanoma. A lesion on the right mons was approximately 1.8 cm in diameter (Figure 1).

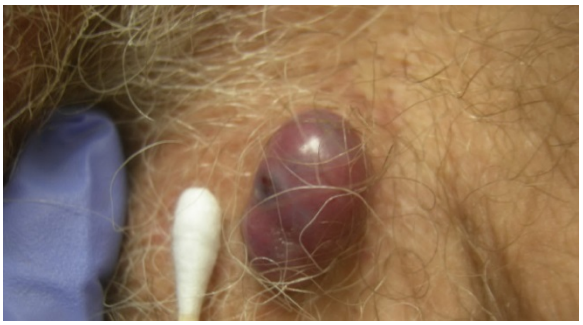

Figure 1. Right mons lesion

A second lesion on the left inferior lateral labia majora was approximately 2.5 cm in diameter. A 0.5 cm diameter nodular lesion of the right inferior labia majora was also present (Figure 2). The mons also had a hyperpigmented erythematous and brown patch. Speculum exam revealed normal vaginal mucosa with no lesions or excoriations identified. There was no inguinal lymph node adenopathy palpable.

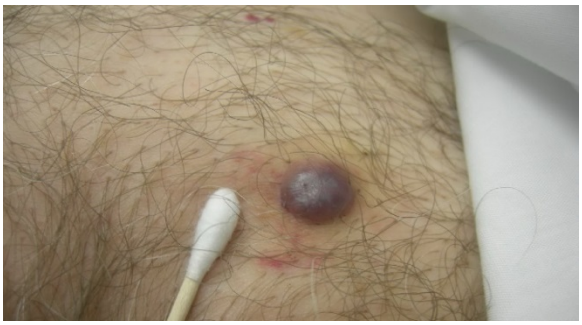

Figure 2. Right labia majora lesion

Due to the number of lesions present, excisional biopsy was limited to the two largest lesions. Microscopic evaluation showed large, hyperchromatic, pleomorphic tumor cells. Immunohistochemistry showed the tumor cells were positive for CD34 and CD31. Immunohistochemical analysis of the lesions showed scattered dermal staining for IgG and IgM. C3 and fibrinogen were identified in the walls of superficial vessels. The final histopathological diagnosis was high-grade angiosarcoma.

Patient was referred to gynecology-oncology and hematology-oncology. Concurrent evaluation locally for abdominal pain and by the referral services following histopathological diagnosis confirmed disseminated angiosarcoma with hepatic, pancreatic, and renal involvement on computed tomographic imaging with contrast and positron emission tomography. Given the disseminated disease and poor

response to treatment, she underwent several interventions including chemotherapy with paclitaxel; a trial study using paclitaxel, valproic acid, and bevacizumab; adrimycin; and radiation. Significant disease course complications included pathologic left hip fracture and aortic stenting for metastatic lesion involving the descending thoracic aorta, in addition to multiple admissions for fever and infections. Within one year of diagnosis, she died of complications from her metastatic disease.

Discussion

A recent case study and review of the literature identified 51 patients with primary angiosarcoma of the female genital tract.[6] Two of the 51 cases presented with vulvar lesions; both had localized disease unlike our case in which disseminated disease was present at the time of diagnosis.

While a broad differential may be formulated for vulvar lesions, multiple findings should increase the suspicion of malignancy as in this case presentation. These include: asymmetry, border irregularity, color variation, rapid change, lack of healing, and bleeding. Biopsy is necessary to differentiate between benign and malignant lesions. Although melanoma has not been identified as a specific risk factor, a recent analysis from the Surveillance, Epidemiology, and End Results (SEER) program identified 26% (of 434 patients with angiosarcoma) had a prior primary cancer.[3] An increased index of suspicion for malignancy may have prompted early biopsy diagnosis and improved outcome.

In addition to surgery to obtain negative margins, treatment may include radiation and/or chemotherapy. Extensive local or radical excision is the preferred treatment for angiosarcoma when localized.[7] Chemotherapy utilizing adrimycin plus ifosfamide has been the initial treatment for soft tissue sarcoma.[8] Recently, multiple different chemotherapy combinations have been tested for the treatment of angiosarcoma.[4,8,9]

The prognosis for patients with vulvar sarcomas has been generally poor due to lack of clinical experience, the aggressive nature of the disease, and the poor response to chemotherapy and radiotherapy employed. The overall course appears to be greatly influenced by the size and extent of infiltration of the tumor. Most cases have an aggressive behavior with a median survival of seven months, and 5 year survival of about 35%.[5] Palliative chemotherapy for disseminated disease is hampered by toxicities and poor response rates.

Conclusion

Given the overall poor prognosis of angiosarcoma, early detection remains the key element in maximizing patient outcome.

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