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Vulvar Leiomyosarcoma Masquerading as a Bartholin Gland Cyst

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Abstract

INTRODUCTION: Primary sarcomas of the vulva are rare and make up 1-3% of all vulvar malignancies, with the most common among these being leiomyosarcomas. They usually present in women 40 to 50 years old, but cases have been reported in those as young as 14 years old. The most frequent presentation is that of an enlarging mass over the labia majora, followed by Bartholin gland area, and are often mistaken for Bartholin gland cysts. This report is to present a 29-year-old female who presented with a vulvar mass, initially suspected to be Bartholin gland cyst, found to be vulvar leiomyosarcoma.

DESCRIPTION: Patient is a 29 year old G2P1011 who presented with a right vulvar mass, initially measuring 9.7 cm on ultrasound, with subsequent incision and drainage and word catheter placement in the Emergency department. She had a history of Bartholin gland cyst, with no other pertinent medical history. She underwent resection of right Bartholin gland cyst and hematoma with subsequent pathology revealing cellular spindle cell neoplasm, favor leiomyosarcoma.

Pathologic features include 10 x 5.5 x 1.6 cm mass excised, with multiple areas of coagulative tumor necrosis and variable mitotic activity, mostly infrequent but up to 8-9/10 hpf. A mild proportion of the tumor showed moderate atypia. Immunostains performed showed positivity for desmin, SMA, ER and PR (80-90%.) She was found to have a 1 cm residual mass at 2 week follow up. PET scan was completed with nonspecific perineal uptake without metastasis and likely physiologic uterine and ovarian uptake.

She underwent right modified radical vulvectomy and right sentinel groin lymph node dissection and dilation and curettage. Pathology confirmed leiomyosarcoma with extension to medial margin and within 1 mm from deep margin, with negative endometrial and endocervical curettage. MRI was completed showing no evidence of developing metastatic disease. She underwent another modified right radical vulvo-vaginectomy with pathology revealing squamous epithelium and underlying connective tissue with fibrosis, chronic inflammation, with no diagnostic evidence of residual leiomyosarcoma. She then received radiation to vulva, for a total of 60 Gray in 33 fractions. She is undergoing surveillance every 4 months currently.

DISCUSSION: Vulvar leiomyosarcomas remain underrepresented in the literature due to their rarity. The largest case series to date, published in 2021, included 44 cases of vulvar leiomyosarcoma3. Because of this paucity of data, they are often overlooked and mistaken for other vulvar pathology, including bartholin gland cysts/abscess, leading to delay in diagnosis and treatment. Additionally, as vulvar leiomyomas primarily affect women who are premenopausal, they escape pathologic evaluation upon initial presentation.

A pathologic criteria to differentiate between leiomyomas and leiomyosarcoma was developed by Nielsen in 1999, using a clinicopathologic study of 25 patients4. This was further expanded to include factors associated with higher rates of local

recurrence, including lesions over 5 cm with infiltrating margins, extensive necrosis and over 5 mitoses per 10 high power fields.

In subsequent case studies, these criteria have been used to guide management with recommended management as primarily excisional with limited postoperative radiation therapy considered for local control. Recommended margins for excision are 2 cm. Chemotherapy appears to have a limited or experimental role.

Data from Akrivi shows that of 44 cases, 3 patients underwent radiotherapy in addition to excision with it used in another 5 patients for recurrence (7/44.) An additional 3 studies were seen, showing use of radiotherapy following excision with no local recurrence for 1, 1.5 and 5 years respectively. Due to this, decision was made for postoperative radiation due to positive margins seen on initial surgery, as well as size on presentation based on the role of radiation therapy in uterine leiomyosarcomas. Because recurrence rates remain as high as 65-77%, including other tumor sites, further studies are needed to understand best methods to prevent recurrence.