

Leiomyoma of the Vulva

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Leiomyoma of the Vulva

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

Precis:

Vulvar leiomyomas are uncommon, and criteria for malignancy are more stringent than for the uterine counterpart.

Keywords: vulvar neoplasms, leiomyoma, vulva

Introduction:

Vulvar leiomyomas are uncommon, and should be distinguished from vulvar fibromas and other solid lesions of the vulva. 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 thirties presented with dyspareunia and a vulvar mass in the region of the Bartholin's gland. There was no reported fever or drainage from the mass. She reported that the mass appeared to be growing over six months. The clinical impression was initially a Bartholin's cyst, as the mass was spongy and compressible. No inguinal adenopathy was noted. Incision in the office to place a Word catheter was performed, but the solid nature of the lesion became apparent, with no drainage of fluid. The patient subsequently underwent excision of a 5.1 cm firm solid mass.

Pathology: Cut surface of the lesion showed a well-circumscribed, bulging lesion which was white, whorled, and rubbery, without hemorrhage or necrosis (figure 1). The lesion was histologically composed of whorled smooth muscle fibers (figure 2). Smooth muscle actin immunostain showed diffuse cytoplasmic staining, confirming the smooth muscle nature of the lesion (figure 2 inset). There was no significant atypia, necrosis, or discernible mitotic activity.

Discussion:

Vulvar leiomyomata are much less frequent than their uterine counterparts.

Histogenesis is unclear, but the lesions may arise from smooth muscle cells in vascular walls or around hair follicles(1). Labia majora lesions may arise from the dartos muscle fibers in the vulva. Vulvar leiomyomata may be asymptomatic, or present with mass, erythema, pain or itching(2). An unusual association that appears to have a genetic basis is vulvar leiomyoma with esophageal leiomyoma(2,3). Estrogen and progesterone receptors have been demonstrated in vulvar leiomyomas, and some have grown with pregnancy(2).

It is important to distinguish benign from malignant smooth muscle lesions of the vulva, and the criteria are more stringent than for the uterine counterpart, with lower mitotic counts associated with aggressive behavior. Aside from obvious metastasis, it has been postulated that infiltrative margins signify malignancy(4). Other features besides infiltrative margins associated with recurrence risk include a size of 5 cm or more, mitoses of 5 or more per ten high power fields(hpf), and significant(moderate to severe) nuclear atypia. Nielsen et al diagnosed leiomyosarcoma if three or more of these features were met, atypical leiomyoma for two features, and leiomyoma for one or less(2). Our case, which measured 5.1 cm, had none of the other features of concern, and so qualifies as a leiomyoma.

The differential diagnosis includes other vulvar masses, including Bartholin's cyst or abscess when located in the location of the gland, and other solid lesions that can affect the vulva such as aggressive angiomyxoma, angiomyofibroblastoma, and fibroma.

Bartholin's cysts and abscesses will be located in the characteristic location, and will be

cystic, rather than solid. Aggressive angiomyxoma and angiomyofibroblastoma have a completely different histologic appearance and immunoprofile, and can thus be distinguished. Although they may stain for desmin, both aggressive angiomyxoma and angiomyofibroblastoma are usually negative for smooth muscle actin, which is positive in leiomyoma(2). Degeneration with hydropic and myxoid change in a vulvar leiomyoma may make distinction from aggressive angiomyxoma a challenge. The stellate cells of aggressive angiomyxoma differ from the spindle cells of leiomyoma(5). Fibromas of the vulva tend to be large, pedunculated, and not hormonally responsive. They would not be expected to stain for smooth muscle actin(6), nor have the cigar shaped nuclei of smooth muscle cells. Other neoplasms of the vulva which can have a spindled appearance that can be excluded with immunohistochemical panels include the spindle cell variant of squamous cell carcinoma, melanoma, monophasic synovial sarcoma, malignant peripheral nerve sheath tumor, epithelioid sarcoma, rhabdomyosarcoma, and granular cell tumor(5).

Transvaginal ultrasound and MRI may be helpful in establishing the nature of vulvar leiomyoma, and MRI may be helpful in distinguishing it from leiomyosarcoma (3).

Treatment is surgical excision.

In conclusion, leiomyoma of the vulva is an uncommon lesion that must be distinguished from Bartholin's cysts and abscesses, as well as other solid vulvar lesions. The criteria for malignancy have been established and are more stringent than the uterine counterpart. As recurrence has been reported as much as 10 years after excision of vulvar leiomyoma(2), long term follow-up is indicated in these patients.

Abbreviations and Acronyms

MRI-magnetic resonance imaging

HPF-high power fields

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