Massive Cellular Angiofibroma of the Vulva

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Word count: Précis: 22, Body of Text: 1033

Running title: Vulvar Massive Cellular Angiofibroma

Disclosures: none

Conflicts of interest: none

Tables 0, Figures 5

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

Precis:

Cellular angiofibromas of the vulva are uncommon, and usually small and circumscribed. A massive cellular angiofibroma extending into the pelvis is described.

Key words: vulvar neoplasms,cellular angiofibroma, mesenchymal neoplasms

Introduction:

Cellular angiofibroma is a rare benign mesenchymal tumor, occurring almost exclusively in the superficial soft tissues of the genital region. It is usually small and well circumscribed. This is one of the features to consider in distinguishing it from the usually more infiltrative aggressive angiomyxoma of the vulva. We describe a challenging case of a massive and infiltrative cellular angiofibroma, which presented a major surgical challenge.

Case:

A woman in her sixth decade was referred to a surgical oncologist for evaluation of a left vulvar mass that had been progressively increasing in size over ten years. She had undergone a partial excision of the mass ten years prior, at an outside institution. At that time, the mass extended to the ischiorectal fossa , and the excision was aborted secondary to hemorrhage. The pathology showed a questionable hemangioma and the patient was told to follow up accordingly. She presented to our institution desiring surgical excision.

Computerized tomography (CT) scan images without IV contrast and magnetic resonance imaging (MRI) of the abdomen and pelvis from 2012 were reviewed and compared to CT images of the abdomen and pelvis performed with IV contrast one month prior to surgery. Imaging findings were significant for an 8.4 x 2.6 x 7.6 cm lobulated soft tissue mass in the left perineum with posterior extension into the buttock and anterior extension into the vulva. This mass contained mixed soft tissue and fatty components. The medial margin of the mass abutted the left lateral margin of the anus and vagina without frank invasion. There was no direct infiltration of the levator ani muscles. There was a second lobulated component to this mass within the medial subcutaneous fat of the left buttock which measured 4.0 x 2.9 x 5.0 cm. These masses were unchanged in extent when compared with the prior MRI performed in 2012 (Figures 1,2).

Examination under anesthesia (EUA) showed a left sided superficial rubbery mobile mass lateral and posterior to the anus with no palpable mass within the rectal vault. The mass was palpable deep and lateral to the vagina on the left side, but there was no intravaginal extension. The patient underwent complete excision of the mass, which was well circumscribed and was dissected free without complication. The resected mass measured 12 x 8 cm, was lipomatous and multi lobulated, with a cystic component attached to the superior portion of the lipomatous mass. The excision was followed by complex closure of the vulvar wound by plastic surgery. A Jackson Pratt drain was left in place and was removed one week postoperatively. The patient had an uncomplicated hospital course and was discharged home in stable condition on the second postoperative day.

Pathologic diagnosis of cellular angiofibroma with fatty overgrowth was made. The tumor was comprised of bland spindle cells, and thick vessels, with a lipomatous component. (figures 3,4,5). Immunohistochemical stains were performed; the tissue was found to be smooth muscle actin positive, HMB-45 negative.

Discussion:

A variety of soft tissue tumors occur in the vulvovaginal region of reproductive-aged women. Most are small, hormonally responsive, and clinically indolent; often mistaken clinically for a Bartholin gland cyst or a labial cyst [1,4] . Some of these tumors are predominantly site-specific while others have a more generalized distribution and are not confined to the vulvar region. The most common specialized genital stromal neoplasms are superficial angiomyxoma, angiomyofibroblastoma, and cellular angiofibroma[1] . Aggressive angiomyxoma is also in the differential, and is more worrisome because of its more infiltrative nature and tendency to recur, particularly after incomplete excision. Precise diagnosis can be difficult due to overlapping histological and immunohistochemical features; however, accurate classification is important as these tumors differ in prognosis and treatment, and some may recur locally if incompletely excised. Other mesenchymal neoplasms that have a more generalized distribution and behave in a clinically benign fashion include leiomyoma, lipoma, neurofibroma, schwannoma, granular cell tumor, glomus tumor, and hemangioma.[1]

Cellular angiofibroma of the vulva is a benign mesenchymal tumor of the vulva with histologic features suggestive of spindle cell lipoma, first described by Nucci et al in 1997[1]. The term "cellular angiofibroma" was chosen to emphasize the cellular spindle cell component and the prominent blood vessels characteristic of this tumor [1]. When first described, this type of tumor was characteristically reported in middle-aged women (median age at diagnosis of 47.5) [1]; however, according to recent literature both women and men seem to be equally affected, most often in the fifth and seventh decade, respectively [2,5]. In women, this tumor usually arises in the subcutaneous tissues of the vulva and presents as a small (<3cm) mass often described as a painless swelling. In men, it usually arises in the inguinal and scrotal regions [2]. Histologically, the tumor is composed of a uniform, monotonous spindle cell proliferation, often admixed with prominent vessels with thick and hyalinized walls, and clusters of

mature adipocytes. It is usually well circumscribed but can be focally infiltrative; it is not associated with aggressive clinical behavior [1]. Local excision with clear margins appears to be adequate in the treatment of these lesions; however, data on long term follow up is not currently available [5].

Diagnosis of soft tissue tumors of the vulva, including cellular angiofibroma, can be challenging and frequently necessitates the use of CT or magnetic resonance imaging for further characterization. These imaging modalities not only help narrow the differential diagnosis but can also aid surgical planning. Cellular angiofibroma is almost always encapsulated, well circumscribed, and hypervascular [2], thus, surgical removal of these tumors requires thorough knowledge of pelvic anatomy, as well as precise and careful dissection to minimize blood loss. One of the most important early surgical steps in the removal of the mass is identification of its capsule [2]. Surgical removal of the mass with its capsule not only helps guarantee complete excision and prevent its recurrence, but also minimizes blood loss [3]. A multidisciplinary approach may be preferred, depending on the extent and size of the lesion as well as the surgical challenges of each individual case.

In summary, we present a massive cellular angiofibroma of the vulva that extended into the pelvis. Evaluation of imaging and a multidisciplinary approach permitted complete excision of this surgically challenging lesion.

Disclosures: none

Abreviaciones and Acronyms

CT-Computerized axial tomography

MRI-Magnetic resonance imaging

HMB-45-Human melanoma black 45

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