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Case Report

Vulvar Aggressive Angiomyxoma in a Young Woman: A Case Report

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Abstract

Aggressive angiomyxoma is a locally infiltrative tumor with a variable chance of recurrence after surgical excision and rarely causes distant metastasis. Due to its variable presentation, clinical diagnosis is complicated and can be misdiagnosed. These tumors are more common in women of reproductive age and are usually present in the pelvis and perineum and cases in males are less common. This report describes a 19-year-old woman with a painless vulvar mass that rapidly developed over a few weeks. Imaging demonstrated no pelvic invasion. The surgical excision was without complications. Histopathologic, and immunohistochemical studies confirmed the final diagnosis.

Keywords: *Soft Tissue Tumor; Mesenchymal Tumor; Angiomyxoma; Vulvar Mass*

Introduction

Aggressive Angiomyxoma (AAM) is a rare mesenchymal tumor that occurs predominantly in women of reproductive age, with a peak incidence in the fourth to fifth decades of life [1]. First described in 1983 by Steeper., *et al*. these tumors involve the deep soft tissues of the lower genital tract, perineum and pelvis [2]. The term aggressive is used because the tumor is locally infiltrative with high chances of recurrence but rarely cause distant metastasis [3,4]. Following surgical excision, the possibility of a local recurrence exists from a few months to several years later (2 months to 15 years) [5]. Common sites are the vulva, perineum, profound pelvic (paravaginal, perirectal and ischiorectal), gluteal region, and retroperitoneum. In men, this condition has been reported to involve the scrotum, paratesticular, penile shaft, pelvic region, perineum, and inguinal area [6,7]. This disorder is more common in women with a female-to-male ratio of 6.6 to 1 [8,9]. The tumor often develops in either the perineal or the pelvic area and it typically manifests as a lump that is painless and usually does not grow rapidly [4] but large-sized multi-lobular (often larger than 10 cm in size at the time of diagnosis) or polypoid mass with finger-like projections have been reported [1,9]. Due to its variability in the presentation, AAM can clinically be misdiagnosed as other pathologies, including vulvar carcinoma, Bartholin gland cyst, vulvar abscess, hernia, vaginal polyp, leiomyoma and lipoma. Misdiagnosis can lead to incomplete surgical excision [1].

Case Presentation

A 19-year-old female college athlete with no significant medical history presented with a rapidly growing painless mass on the vulva over one month. The patient reported regular menses with menarche at age 13. She had no history of abdominal or pelvic trauma. She denied any prior or current sexual intercourse. Physical exam was notable for a large, pedunculated, nontender 18 cm mass of the left labia majora with a base of 6 cm and about 1.5 cm in width (Figure 1A and 1B). The skin was intact but scalloped at the base of the mass, which extended to a smooth pedunculated distal end. There was no inflammation at the base or the distal end. The tumor extended to the middle of the labia majora and was non-tender to tension. The remainder of the pelvic examination was unremarkable. The external genitalia, including the Bartholin's, urethral and skene's glands, were normal, limited pediatric speculum examination was normal and digital vaginal and rectal exams were normal with palpable normal uterus and adnexal structures.

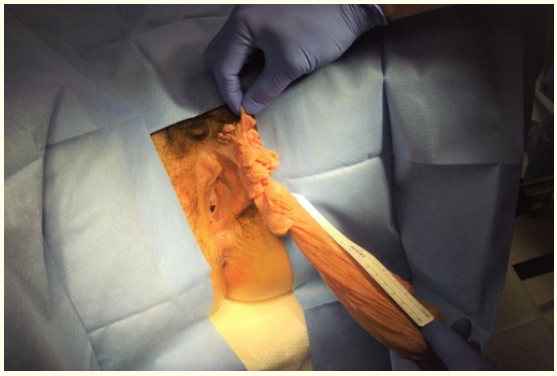

Figure 1A: *Intraoperative view of mass prior to resection.*

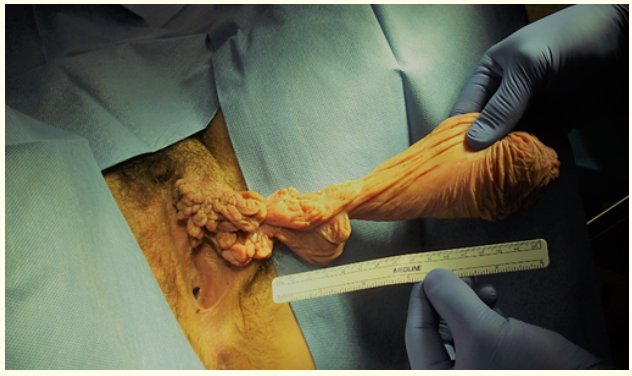

Figure 1B: *Intraoperative view of mass prior to resection.*

Imaging/Investigation

The patient had already been assessed at a separate hospital system's emergency department before she arrived at the outpatient appointment. She received a computed tomography (CT) of the abdomen and pelvis with contrast, followed by an ultrasound of the pelvis and ultrasound of the external genital mass.

CT of the abdomen and pelvis, the lower lung fields, heart, liver, spleen, gallbladder, kidneys, adrenal glands, and pancreas were all within normal limits. The bowel was unremarkable. There was no evidence of lymphadenopathy or free fluid in the abdomen or pelvis. An external mass lesion was identified extending inferiorly from the left labia majora and measuring 11 cm in length (Figure 2).

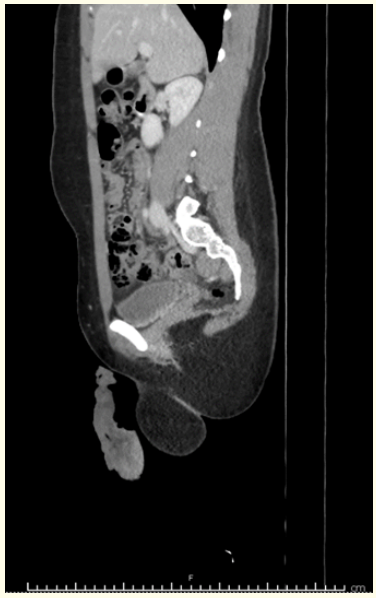

Figure 2: *CT Abdomen/Pelvis with contrast.*

Pelvic ultrasound demonstrated a uterus measuring 7.5 x 5 cm, an endometrial stripe of 10 mm, unremarkable ovaries, and no appreciable ovaries fibroids. Ultrasound imaging of the left labia majora mass was notable for increased vascular flow on color doppler and identified an intra-tumor cystic structure measuring 0.1 x 0.3 x 0.8 cm (Figure 3).

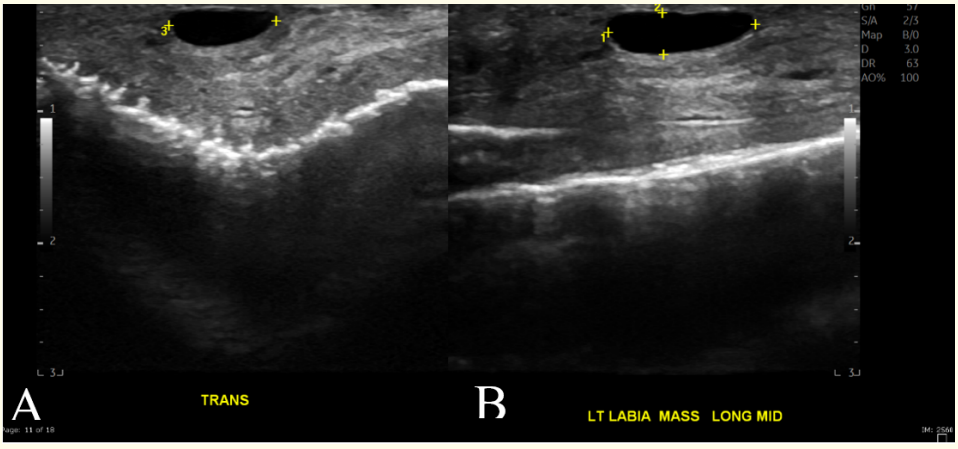

Figure 3: *Transabdominal pelvic ultrasound of the mass.*

Treatment and follow-up

Because the CT scan did not demonstrate pelvic infiltration of the mass and no obvious signs of malignancy, complete excision as an outpatient planned. An elliptical incision around the mass base, approximately 7 cm in length was created. Sharp dissection was used to separate the skin from the underlying subcutaneous fat. The underlying fatty tissue was pushed away from the skin appendage using delicate, painstaking dissection until it could be removed. The mass was sent *in toto* with a margin of approximately half a centimeter of skin and subcutaneous tissue for pathologic analysis. The wound was explored, and there was no evidence of a hernia. The wound was closed in layers. The post-operation aspect is seen in figure 4.

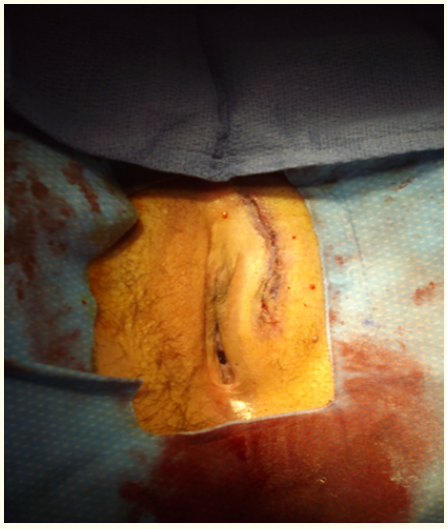

Figure 4: *Posterior operation aspect.*

The patient was discharged the day of surgery and rest of her postoperative recovery was without complication. At her 2-week post-operative follow-up visit, the patient was doing well. Further follow-up was planned for a repeat physical examination at six and twelve months. Given the lack of intraperitoneal invasion, it was decided that the use of any imaging such as magnetic resonance imaging (MRI) to assess for recurrence would depend on physical exam findings at follow-up appointments.

Discussion

This rare benign tumor can infiltrate neighboring structures, displace them, and recur locally [10]. Distant metastasis is extremely rare, with just three reported cases involving metastasis to the lung and mediastinum [3]. In most cases histopathology and immunohisto-chemistry (IHC) is essential for final diagnosis, but imaging exams can help establish the diagnosis. Chemotherapy and hormone therapy guidelines have not been defined, and decisions are made case by case.

The most common imaging modalities used in the workup of AAM are ultrasound, contrast-enhanced CT, and MRI. On ultrasound, AAM has a hypoechoic or cystic appearance. A hypo attenuating or isoattenuating mass will be seen on CT. Of these, only 50% of CTs will also show a classic 'swirling' appearance suggestive of internal laminated architecture. The finding of a laminated or swirling pattern on T2-weighted MRI is considered pathognomonic for AAM. This makes MRI the imaging modality of choice for diagnosis and early detection of

recurrence. In this case, the patient received CT and US at an outside hospital. However, a diagnosis was not made at that time. The inclusion of MRI may have helped to decrease the time to diagnosis. In the post-surgical setting, MRI is the modality of choice for detecting early recurrence - especially in tumors with intraperitoneal invasion. While most reported cases of AAM have an intraperitoneal invasion, this was absent in the case of our patient. For this reason, her post-surgical follow-up consisted of serial exams with MRI only if needed [8].

Histopathology, IHC, and hormone receptor status are helpful in both diagnosis and management decisions for AAM. Pathology gross examination showed un-encapsulated locally infiltrative mass. Microscopic examination by H&E stain (Figure 5) demonstrates hypocellular neoplasm with spindle to satellite tumor cells and delicate cytoplasmic processes. Tumor cell nuclei have bland chromatin and occasional small nucleoli. No mitosis is identified. Stroma shows areas of myxoid changes with delicate collagen fibers and dilated capillaries, irregular, infiltrative borders without capsule. No necrosis or hemorrhage is identified. Immunohistochemical stain HMGA2 (Figure 6) demonstrates strong nuclear staining in tumor cells which confirms the diagnosis. Infiltrative, ill-defined tumor borders and positive HMGA2 exclude main differential diagnosis, angiomyofibroblastoma. Lastly, patient's tumor was estrogen receptor positive (Figure 7).

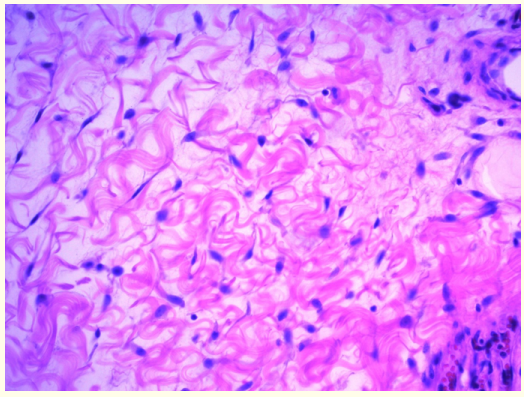

Figure 5: *H&E stain shows spindle cells with bland chromatin in delicate stroma, magnification 200x.*

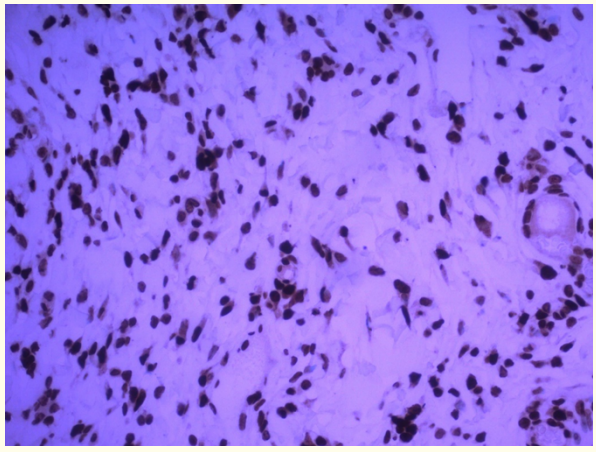

Figure 6: *Immunohistochemical stain HMGA2 shows strong nuclear positivity, magnification 200x.*

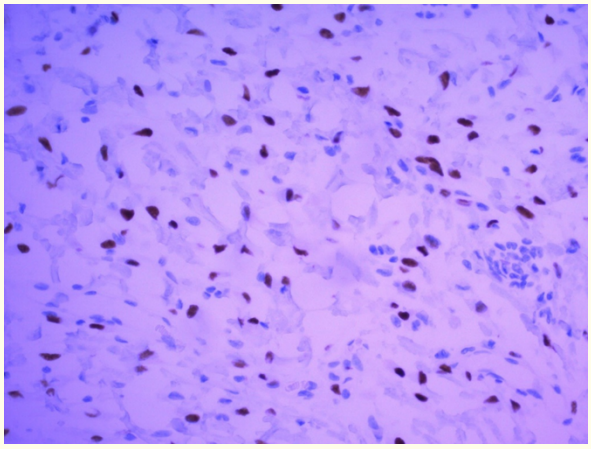

Figure 7: *Immunohistochemical stain estrogen receptor (ER) shows nuclear positivity in neoplastic cells, magnification 200x.*

Due to the challenge of diagnosing AAM, histopathology with IHC is needed for the diagnosis, wherefor surgery needs to be done. Tumor resection with wide margins remains the first-line treatment [1,4]. There is no consensus regarding the pre- or post-surgical use of chemotherapy and hormone therapy. This is probably due to the rarity of this neoplasm. Treatment with chemotherapy and radiation is limited because of low mitotic activity of the tumor. However, the radiotherapy has been reported in recurrent diseases after poor results with surgical excision [1,7]. As an adjuvant treatment, angiographic embolization can be used preoperative to decrease the tumor size, especially in hormone-negative tumors [1,3].

Hormone-positive tumors tend to respond to therapy with gonadotropin-releasing hormone (GnRH) agonists, raloxifene, and tamoxifen in both preoperative and postoperative settings, not as a stand-alone therapy. With the lack of consensus, treatment must be individualized on a case-to-case basis after discussion with a multidisciplinary team [1,3,7,11].

Conclusion

Aggressive angiomyxoma is a rare mesenchymal tumor with a predilection for pelvic region of women of reproductive age. Frequently presenting as a painless mass, this tumor can be locally infiltrative with a variable risk of recurrence, and in rare cases, distant metastasis can occur. Imaging is recommended to assess the extension. Currently, the MRI swirling pattern on T2-weighted is considered pathognomonic for this tumor and is the best modality to detect recurrences. Wide excision is the standard treatment, and hormonal therapy can be an adjuvant in hormone-positive tumors. Histopathology examination and immunohistochemistry studies demonstrate strong positivity for HMGA2 stain.

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