Case Report

Vaginal Angiomyofibroblastoma: A Case Report and Review of Diagnostic Imaging

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1. Introduction

Angiomyofibroblastoma (AMFB) is a rare, indolent mesenchymal tumor that most commonly occurs in the female genital tract in premenopausal women, most frequently in the vulva and vagina. AMFB typically measures less than 5 cm; however, case reports describe tumors of up to 23 cm in size [1]. Few cases have been reported affecting the fallopian tube, ischiorectal fossa, cervix, and bladder, as well as similar tumors in the male spermatic cord, scrotum, and perineum [2–6]. AMFB was first described in the literature by Fletcher et al. in 1992 as an important distinction from aggressive angiomyxoma (AA), which is an infiltrative myxematous mesenchymal tumor with the potential for local recurrence [7].

Preoperative diagnosis of AMFB and distinction from other soft tissue tumors is often difficult, as there is limited information on characteristic imaging findings. The differential diagnosis for a vaginal or vulvar mass includes Bartholin’s gland cyst, epidermal inclusion cyst, Gartner’s duct cyst, fibroma, lipoma, hemangioma, leiomyoma, and alternative rare mesenchymal tumors. Here, we describe a case report of a vaginal AMFB and relevant radiologic features to aid in diagnosis and distinction from aggressive angiomyxoma.

2. Case

A 30-year-old gravida 1 para 1 female presented to our Emergency Department complaining of a vaginal mass present since the birth of her child 4 years earlier. At that time, she underwent an uncomplicated vacuum-assisted vaginal delivery and was unaware of significant lacerations or repairs. She felt that the mass had not changed significantly in size since the postpartum period, but she had never been evaluated by a physician. She complained of acutely worsening discharge over the previous month, described as watery yellow to pink and occasionally blood tinged. She denied changes in bowel movements, dysuria, hematuria, fevers, chills, night sweats, changes in appetite, or weight loss. She complained of both entry and deep dyspareunia.

Physical exam was notable for copious serosanguinous fluid within the vaginal vault. A well-circumscribed, smooth cystic structure approximately 4 cm in diameter was noted
along the posterior vaginal wall. There was also a 0.5x1.0 cm exophytic lesion overlying the mass with serosanguinous drainage (Figure 1). On rectal exam, the mass was noted to be separate from the cervix and within the rectovaginal septum. Rectal involvement was not appreciated.

Tissue biopsies were taken; however, they were of limited diagnostic value, showing fibrous tissue with acute and chronic inflammation and squamous debris, consistent with cyst wall and contents.

A pelvic ultrasound (US) was performed and showed a complex vaginal mass, inseparable from the cervix, measuring 5.1 x 3.8 x 5.4 cm (Figure 2(a)). Color Doppler demonstrated minimal peripheral vascularity (Figure 2(b)). Magnetic resonance imaging (MRI) was subsequently performed for further characterization (Figure 3). The mass measured 4.7 x 4.8 x 4.9 cm and appeared to arise from the posterior wall of the vagina, separate from the cervix. The mass was heterogeneously hyperintense on T2-weighted (T2W) images and hypointense on T1-weighted (T1W) images. Postcontrast sequences demonstrated enhancement of the wall, absent internal enhancement superiorly, and bulky, nodular, hyper-enhancement inferiorly, consistent with a complex cystic mass.

The patient was taken to the operating room for an uncomplicated surgical excision. Histology was notable for hypocellular edematous myxoid stromal tissue alternating with hypercellular areas of stromal cells clustered around thin walled small to medium-sized vessels, consistent with angiomyofibroblastoma. Stromal cells were noted to be spindled with eosinophilic cytoplasm. Nuclei were round or oval with fine chromatin (Figure 4). Rare mitotic figures were noted. Immunohistochemical stains were negative for desmin, alpha-smooth muscle actin (α-SMA), and progesterone receptor but demonstrated focal estrogen receptor positivity.

3. Comment

Although AMFB is a rare diagnosis, it is an important consideration in the premenopausal and perimenopausal patient presenting with a vulvovaginal mass given its predilection for this region of the female genital tract. In a review of the literature in 2015 by Wolf et al., 125 cases of female AMFB had been previously reported, of which 92% were either vulvar (N = 98) or vaginal in origin (N = 17). Median age was 45, and the majority of cases were observed in women less than 60 years of age [8]. Since 2015, there have been 10 additional cases including ours describing AMFB in females, six of which were located in the vulva, one cervical, one in the broad ligament, and one on the patient’s foot [2, 9–17]. All women were under the age of 50.

Differential diagnosis for AMFB includes Gartner’s duct cyst, epidermal inclusion cyst, leiomyoma, and fibroepithelial polyps among the more common etiologies. It is most important to distinguish AMFB from aggressive angiomyxoma (AA), which can commonly be misdiagnosed based on anatomic and pathologic similarities (Table 1). Although there have been no reports of distant metastasis, AA is known to recur in 33-72% of cases and is locally invasive, often entrapping nerves and mucosal glands [18, 19]. Surgical approach to AA requires wide local excision given the infiltrative nature of the lesion versus simple excision of AMFB; thus, it is important to attempt differentiation between the two prior to surgery.

Diagnostic utility of preoperative imaging for AMFB remains controversial. However, as the number of reported cases increases in the literature, more data exists on common radiographic features (Table 2).

3.1. Ultrasonography. US is typically the first imaging modality used to evaluate vaginal masses due to its intrinsic high resolution, availability, and cost effectiveness. US can be utilized to distinguish vulvar and vaginal AMFB from AA and other mesenchymal tumors [20, 21]. AMFB shows hyperechoic areas with irregular and small hypoechoic cystic areas interspersed within homogenous echogenic stroma [8, 20–23]. Minimal vascularity in AMFB is occasionally noted [8, 22]. Absence of prominent vascularity on color Doppler is consistent histologically with the predominant capillary-like vascular component of AMFB. Echogenic areas have been shown to represent hypocellular stroma on histology, while hypoechoogenicity represents hypercellular areas [8, 21]. In our case, US characteristics were similar to those previously described.

In contrast, US appearance of AA is typically a hypoechoic mass with homogenous echogenicity and occasional echogenic septa correlating to fibrous bands on pathology [24, 25]. These fibrous bands can create a layered or swirling appearance similar to that seen on MRI and CT imaging of AA [26]. Vascularity can also be much more prominent on color Doppler of AA than AMFB [26, 27].
Figure 2: Transvaginal ultrasound. (a) Gray scale image shows a mixed echogenicity mass (arrows) with small hypoechoic cystic areas (3.0 MHz) 10 cm. (b) Color Doppler image shows minimal vascularity. Magnitude: 3 MHz.

Figure 3: (a) Sagittal T2 weighted image shows a predominantly hyperintense mass (arrows) with small central areas of hypointensity. (b) On T1 weighted axial image the mass is homogeneously hypointense (arrows). (c) Sagittal contrast enhanced image demonstrates heterogeneous hyperenhancement inferiorly (dashed arrow) and absent enhancement superiority (•). Cervix: cx.
Figure 4: (a) Mesenchymal lesion with alternating hypercellular and hypocellular areas (4x magnification; hematoxylin and eosin stain). (b) Higher magnification demonstrating ovoid and spindle-shaped cells aggregated around small blood vessel (40x magnification).

Table 1: Clinical and pathologic features of AMFB and AA.

|                      | AMFB                                                   | AA                                                    |
|----------------------|--------------------------------------------------------|-------------------------------------------------------|
| **Clinical features**|                                                        |                                                       |
| Age (years)          | 16 – 86 (Median 45)                                    | 16 – 70 (Median 37)                                   |
| Size (cm)            | < 5 (0.5 – 23)                                         | > 5 (1-60)                                            |
| Location             | Female pelvis and perineum: predominantly vulvar/vaginal | Female pelvis and perineum                           |
| Onset                | 1 month to 8 years                                     | 1 month – 5 years                                     |
| Recurrence           | None reported                                          | 33-72%                                                |
| **Pathology**        |                                                        |                                                       |
| Low-power features    |                                                        |                                                       |
|                      | Well-defined                                          | Infiltrative                                          |
|                      | No entrapment of mucosal glands or nerve bundles       | Entrapped mucosal glands and nerve bundles            |
|                      | Alternating hypocellular and hypercellular areas       | Stromal cells distributed throughout                 |
| Vasculature           |                                                        |                                                       |
|                      | Abundant thin-walled vessels, mostly capillary-like    | Small to medium-sized vessels, mostly thick-walled or hyalinized |
|                      | Abundant                                              | Low cellularity                                       |
| Stromal cells         |                                                        |                                                       |
|                      | Spindle, plump spindle, or oval                       | Thin delicate spindle or stellate                     |
|                      | Perivascular aggregation                              | Mitotic figures typically absent                     |
| Stroma                |                                                        | Myxoid, hyaluronic-acid rich                          |
|                      | Mucin-poor, containing delicate, wavy collagen fiber   |                                                       |
| **Immunostaining**   |                                                        |                                                       |
| Desmin               | Positive (50-60%)                                      | Positive (Up to 73%)                                  |
| α–SMA                | Negative (positive in up to 15%)                       | Positive                                              |
| S-100                | Negative                                              | Negative                                              |
| Vimentin             | Positive                                              | Positive                                              |
| Estrogen receptor     | Positive                                              | Positive                                              |
| Progesterone receptor | Positive                                              | Positive                                              |

To complete the differential diagnosis, vaginal wall cysts are anechoic masses without internal vascularity; vaginal epidermoid cysts are homogeneous hyperechoic masses without internal vascularity; and heterogeneous solid masses are often malignancies or leiomyoma [22].

3.2. Magnetic Resonance Imaging. On MRI, AMFB is typically well circumscribed [9]. It is rare to see central necrosis or degeneration but it does not rule out AMFB [9]. AMFB is most often described as hypointense on T1 weighted (T1W) images, similar to that of skeletal muscle, hyperintense on
| Study             | Sex | Age | Size (cm) | Location          | MRI                                                                 | TVUS                                                                 |
|-------------------|-----|-----|-----------|-------------------|----------------------------------------------------------------------|-----------------------------------------------------------------------|
| Wolf et al.       | F   | 38  | 9x6x1.5   | Vagina            | T1 weighted: Homogenous hypointense                                   | Well-demarcated, homogenous mass, medium echogenicity, few septations |
|                   |     |     |           |                   | T2-weighted: Homogenous hypointense mass                               | Doppler: several small intralesional vessels                           |
| Souza et al.      | M   | 19  | 2.8       | Scrotum           | T1-weighted: Central hypointensity Intermediate hyperintensity, capsule | Well-defined, heterogeneous echo texture, small hypoechoic cysts        |
|                   |     |     |           |                   | T2-weighted: Intermediate homogenous hyperintensity, Hypointense capsule | Doppler: minimal flow                                                  |
|                   |     |     |           |                   | Central heterogeneous mixed hypo and hyperintensity                   |                                                                       |
| Shoji et al.      | F   | 50  | 8x7x5     | Vulva             | T1-weighted: Hypointense mass                                         |                                                                       |
|                   |     |     |           |                   | GA-T1: Homogenous enhancement, poorly enhanced area at center         |                                                                       |
|                   |     |     |           |                   | T2-weighted: Mildly hyperintense                                      |                                                                       |
| Geng et al.       | F   | 46  | 8.5x6x6   | Paravaginal       | T1-weighted: signal intensity similar to skeletal muscle              |                                                                       |
|                   |     |     |           |                   | GA-T1: Homogenous hyperintensity                                      |                                                                       |
|                   |     |     |           |                   | T2-weighted: Hyperintensity, mild hyperintensity, mild hypointensity   |                                                                       |
| Kim et al.        | F   | 40  | 9x5.5x2.5 | Vulva             |                                                                       |                                                                       |
|                   |     |     |           |                   | N/A                                                                  |                                                                       |
| Qiu et al.        | F   | 32  | 13.2x5.8  | Posterior cul-de-sac | T1-weighted: N/A                                                       |                                                                       |
|                   |     |     | x7.8      |                   |                                                                      | Moderately echoic mass, small hypoechoic area                         |
| Kitamura et al.   | F   | 24  | 3 x 2.8 x 2.8 | Urethra         | T1-weighted: N/A                                                       |                                                                       |
|                   |     |     |           |                   |                                                                      | N/A                                                                  |
| Lim et al.        | F   | 48  | 3.8x3.5x2.8 | Posterior perivesical space | T1-weighted: Signal intensity similar to skeletal muscle               |                                                                       |
|                   |     |     |           |                   |                                                                      |                                                                       |
| Maruyama et al.   | M   | 72  | 7.2x5.5x2.2 | Scrotum          | T1-weighted: Similar to or lower signal intensity than skeletal muscle |                                                                       |
|                   |     |     |           |                   |                                                                      | Mixed echogenicity, with several small hypoechoic cystic areas         |

Table 2: Characteristic imaging features of AMFB on MRI and US.
| Study          | Sex | Age | Size (cm) | Location     | MRI                                | TVUS                                                   |
|---------------|-----|-----|-----------|--------------|-----------------------------------|--------------------------------------------------------|
| Wang et al.   | F   | N/A | N/A       | Anterior vaginal | N/A                               | Poorly defined, irregularly shaped, hypoechoic, band-like echoes of various width Doppler: positive blood flow |
| Wang et al.   | F   | N/A | N/A       | Urethral     | N/A                               | Well-defined, hypoechoic cystic solid mass Doppler: positive blood flow |
| Mortelet al.  | F   | 46  | 2.5x3.5   | Perineal     | T1-weighted: hypointense, inhomogeneous mass with small focus of hyperintensity representing fat GA-T1: Strong homogenous uptake T2-weighted: Hyperintense homogenous signal similar to surrounding fat | N/A                                                   |
T2 weighted (T2W) images, and with homogenous hyperenhancement on gadolinium chelate (Gd-C) enhanced images [1, 9, 23, 28]. Differences in T1W and T2W signal intensity may be attributable to variations in lipid and collagenous content in AMFB [8, 23, 28–31].

Contrast enhanced imaging was only reported in 5 studies. Four showed homogenous hyperenhancement and one demonstrated heterogeneous enhancement [23]. Hyperenhancement is thought to be related to prominent vascularity associated with myofibroblast tumors and AMFB in particular [1, 9, 23, 28, 30–32].

Our case demonstrated heterogeneously high T2 and low T1 signaling intensity on MRI which is consistent with the literature. Postcontrast sequences demonstrated enhancement of the wall, absent internal enhancement superiorly, and bulky nodular enhancement inferiorly, consistent with a complex cystic mass.

When correlated histologically, hyperintensity on T2W and absent enhancement correspond to hypocellular areas with abundant collagenous stroma. Areas of hyperenhancement correspond to areas of hypercellularity and vascularity, with little collagenous stroma and water content. Intermediate intensity on T2W and less avid enhancement are thought to represent areas of intermediate cellularity and collagenous stroma [28].

Data on MRI characteristics of AA is limited. AA has similar findings to AMFB on T1W and T2W images but is thought to be differentiated best by its enhancement characteristics. AA is described as having an intense, swirled or layered pattern. Similar swirling and layered enhancement is also described on contrast enhanced CT [26–28, 33–36]. AMFB does not typically show infiltrative pattern but it can grow around structures. If infiltration and swirled or layered pattern are observed, AA is much more likely than AMFB [28, 34–36].

4. Conclusion

In conclusion, in the patient with a vaginal or vulvar mass, it is important to consider AMFB among the differential diagnoses, particularly because of the importance of distinguishing AMFB from AA for treatment purposes. Postoperative histologic examination is needed for definitive diagnosis; however, we propose that ultrasound and MRI are increasingly useful diagnostic tools, particularly as the number of reported cases increases. Given the cost effectiveness of ultrasound, initiating imaging workup of a solid vaginal or vulvar mass with ultrasonography is suggested.

Additional Points

Precis. Ultrasound and magnetic resonance imaging are useful modalities in the diagnosis of angiomyofibroblastoma, a rare mesenchymal tumor. Teaching Points. (1) AMFB occurs in the vulva and vagina in over 90% of reported cases. AMFB should be considered on the differential diagnosis of benign appearing vulvar and vaginal masses. (2) Transvaginal ultrasound is the recommended first line imaging for AMFB. MRI is recommended as second-line imaging and is preferable to CT. (3) It is important to attempt preoperative distinction from aggressive angiomyxoma in order to optimize surgical approach.

Conflicts of Interest

All the authors have no conflicts of interest for this study.

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