CASE REPORT

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Case Report of Vaginal Angiomyofibroblastoma and Differential Diagnosis of Other Benign Mesenchymal Tumors in the Female Genitalia

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ABSTRACT A wide variety of mesenchymal lesions, benign and malignant tumors can involve the vulvovaginal region. However, it is not easy to make a list for differential diagnoses by imaging modalities or gross findings. Angiomyofibroblastoma (AMFB) is a rare, benign mesenchymal tumor. Most cases of AMFB occur in the female genital tract. We reported a case of a 20-year-old female patient with AMFB on the anterior vaginal wall. The patient visited emergency clinic because of sudden vaginal bleeding that progressively increased. Ultrasound and magnetic resonance imaging revealed an approximately 7.7×5.0 cm mass between the cervix and vaginal wall. Incision was made at the anterior vaginal wall and local excision was performed under general anesthesia. She was discharged without complications and vaginal structure was well recovered. In addition, we reviewed other mesenchymal tumors of the female genitalia for their differential diagnosis.

Keywords: Genitalia, female; mesoderm; neoplasms; mesenchymoma; diagnosis, differential

A wide variety of mesenchymal lesions, benign and malignant tumors can involve the vulvovaginal region. However, it is not easy to make a list for differential diagnosis by imaging modalities or gross findings.

Angiomyofibroblastoma (AMFB) is a rare, benign soft tissue tumor of the mesenchymal tumor group, which is comprised of blood vessel and stromal cells.[1] Most patients with AMFB are young to middle-aged women. AMFB generally originates from the female genital tract, especially the vulva and vagina. Infrequently, it can be found in the peri-vesicular space or the Douglas pouch.[2] A few cases of AMFB have been reported in the male inguinoscrotal region, and one case each in the nasal cavity and mediastinum.[3-5] Clinically, AFMB presents as a slow growing mass, with no other characteristic symptom that distinguishes it from other female genital tract tumors. In total, 143 cases of AMFB have been reported, including this case. According to the published reports, AMFB rarely recurs after simple excision and one case alone showed sarcomatous transformation.[6]

CASE REPORT

A 20-year-old virgin female patient visited our emergency room because of vaginal bleeding. The patient had a history of irregular menstrual intervals and her last menstrual period occurred 2 weeks prior to the visit. Five days ago, she experienced a sudden vaginal bleeding that progressively increased. Trans-abdominal and trans-rectal ultrasound (USG) revealed

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an approximately 7.7×5.0 cm mass between the cervix and vaginal wall (Figure 1). The mass was a well circumscribed, heterogeneous echoic lesion, with focal hypervascularity in its distal part. In addition, the endometrial thickness was 15 mm.

Magnetic resonance imaging (MRI) indicated an 8-cm submucosal, anterior paravaginal wall mass with multiple tumor vessels derived from the anterior capsule; the lesion showed heterogeneous T2 weighted signal intensity with multifocal small cystic changes (Figure 2).

By pelvic exam, the mass was palpable in the anterior vaginal wall, but not in the intravaginal space. The following day, an informed consent was obtained and trans-vaginal mass excision was performed under general anesthesia with the patient in the lithotomy position. First, we made a 4 cm incision on the anterior vaginal wall; and subsequently, excised part of the mass for frozen biopsy. The frozen biopsy result indicated a benign tumor with spindle cells. After obtaining the frozen biopsy result, the mass was excised to a few fragments using a scalpel. A Jackson-Pratt (JP) drain was inserted in the space and the anterior vaginal wall was sutured by interrupted suture with Polysorb™ (Covidien™, Dublin, Ireland). The patient was discharged without any complication at 7-days post-surgery. The JP drain was removed on the day before discharge. The final pathology report confirmed the diagnosis of AMFB (Figure 3). Informed consent was obtained from the patient to use her medical records for academic purposes.

DISCUSSION

Usually, patients with AMFB do not experience marked symptoms, and the tumor is detected on physical examination. Clinically, majority of AMFB

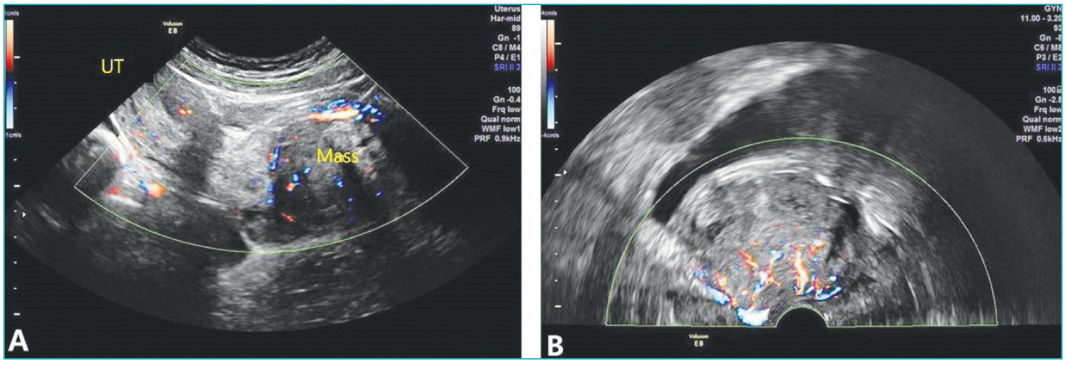

FIGURE 1: A) Trans-abdominal ultrasound image shows a well circumscribed of between cervix and anterior vaginal wall; **B)** Trans-rectal ultrasound image shows lower genital tract mass with hetero-echogenicity and focal vascularity on color Doppler.
UT: Uterus.

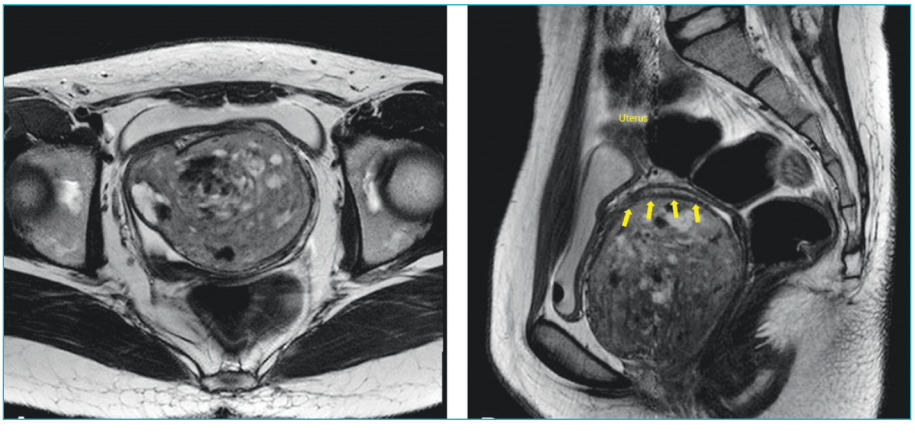

FIGURE 2: T2-weighted magnetic resonance images (**A:** coronal view, **B:** sagittal view) show well demarcated mass with heterogeneous signal intensity with multifocal small cystic changes. Arrows are indicating that the mass was compressed the vagina and uterus.

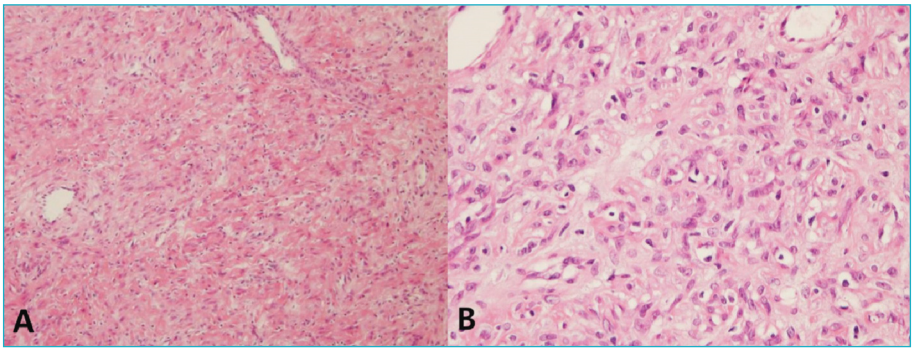

FIGURE 3: **A)** Photomicrograph shows small to medium-sized vessel with variable cellularity, hypocellular areas are noted at the left upper part and hypercellular areas are located at the right lower part (Hematoxylin-Eosin stain, original magnification, x100); **B)** Spindle-shaped to ovoid cells with uniform nuclei and there was no abnormal mitosis neither atypical cell (Hematoxylin-Eosin stain, x400).

cases are misdiagnosed as Bartholin cyst or labial cyst. Most cases of reported AMFB were found as a non-tender, movable mass. The differential diagnosis of palpable vulvovaginal mass include leiomyoma, lipoma, Bartholin cyst, labial cyst, abscess, aggressive angiomyxoma (AAM), granular cell tumor, superficial angiomyxoma, extramammary myofibroblastoma, prepubertal vulval fibroma, sarcoma including leiomyosarcoma, and tumor like lesions including inguinal hernia, urethral diverticulum, rectocele, nodular fasciitis, postoperative spindle cell nodule, and fibroepithelial polyps.[3,6] Other mesenchymal tumors could be confused pathologically with AMFB (Table 1).

Differential diagnosis is important to determine the extent of excision according to the possibility of recurrence and metastasis. AAM is the first to be distinguished from AMFB, because AAM requires extensive resection while AMFB can be treated by simple local excision. Gross examination can be useful for a differential diagnosis of AMFB with AAM, since AMFB is well-circumscribed, but AAM is frequently adhered to adjoining fat, muscle or regional structures.[7] AMFB and AAM is a slowly progressive tumor that commonly grows in the female genital tract. AAM is a benign tumor, but has marked tendency for local recurrence with occasional reports of metastasis to the lungs.[8] AAM is named for its frequent recurrence characteristic (up to 70%), often years after excision; in addition, some cases show distant metastasis that results in fatality.[9,10] Therefore, the treatment of choice for AAM includes optimal surgery, wide local excision with 1 cm margins, and long term follow-up is recommended whereas AMFB requires only simple local resection.

However, pathologic findings may overlap with or mimic other mesenchymal tumors, which causes difficulty in distinguishing between the tumors. AMFB and AAM have similar immunohistochemistry findings that include CD34, desmin, estrogen receptor, and progesterone receptor positivity. AMFB is characterized pathologically as a well demarcated, but unencapsulated tumor that is comprised of an admixture of numerous small and delicate thin-walled vessels and plump round to spindle-shaped cells, and the absence of necrosis and mitosis as compared to AAM (Figure 3).

There are other mesenchymal tumors that need to be differentiated, and most of them can be treated with local excision except AAM (Table 1).

Fibroepithelial stromal polyp is a benign, polypoid, or pedunculated lesion that commonly occurs in the vulvovaginal region in premenopausal females. It is hormone-sensitive and commonly occurs in pregnancy. Its pathologic features typically include a central fibrovascular core and s stellate and multinucleated stromal cells.[11]

Cellular angiofibroma is also a benign soft-tissue tumor, which mainly arises in the genitourinary region of both genders. It is well circumscribed with wispy collagen bundles and prominent vessels that are often hyalinized.[3]

TABLE 1: Differential diagnosis with angiomyofibroblastoma.

	Invasive potential	Pathologic finding	Treatment of choice
Angiomyofibroblastoma	Benign, Rare recurrence	Well demarcated, consist of numerous small delicate thin-walled vessels and plump round to spindle-shaped cell Reactive for vimentin and desmin[2,4]	Simple excision
Aggressive angiomyxoma	Benign High recurrence Rare metastasis	Grossly ill-defined margin, consist of spindle and stellate-shaped cell, variable internal blood vessel size; medium to large. Reactive for vimentin, desmin, CD34[7]	Wide local excision including with 1 cm margin
Fibroepithelial stromal polyp	Benign Rare recurrence	Central fibrovascular core and contains stellate and multinucleated stromal cells Reactive for vimentin, desmin, actin, estrogen and progesterone receptor[10]	Simple excision
Cellular angiofibroma	Benign Rare recurrence	Bland spindle shaped cell, wispy collagen bundle, prominent small to medium sized, often hyalinized vessels Reactive for CD34, negative for S-100 protein[3]	Simple excision
Prepubertal vulvar fibroma	Benign Frequent recurrence after incomplete resection	Poorly marginated, hypocelluar spindle cell, reactive for CD34[11]	Simple excision Especially with clear margin
Extramammary myofibroblastoma	Benign Rare recurrence	Well circumscribed, myofibroblastic differentiated spindle shaped cell Reactive for desmin, CD34[12]	Simple excision

Prepubertal vulvar fibroma usually presents as a painless swelling of the vulva. The pathologic finding is a poorly circumscribed lesion with proliferation of hypocellular, bland spindle-shaped, and "patternless" cells.[12]

Extramammary myofibroblastoma is a benign spindle cell lesion which involves the inguino-groin region in elderly male or postmenopausal women. It is a slowly growing tumor that causes no pain symptoms. The tumor is mainly composed of spindle-shaped cells with myofibroblastic differentiation.[13]

Imaging including MRI and USG can be used as tools for the differential diagnosis of vulvovaginal mass; but there is no current consensus on the typical findings. On USG, AMFB is described as a well demarcated with inhomogeneous echogenicity and multiple hypoechoic areas within an echogenic stroma, similar to the findings in our case. However, in some reports, AMFB is described as homogenous, with medium echogenicity and without solid cystic features.[3] Also, MRI findings of AMFB differ by case.[14,15] For an accurate diagnosis, pathologic confirmation is necessary.

A few cases of recurrence have been reported that were initially diagnosed as AMFB. In each case of recurrence, there was sarcomatous change or misdiagnosis. A vulvar AMFB that showed sarcomatous change at the same site after the surgery was reported.[6] In another case, a mass in the anterior vaginal wall showed recurrence at the same site after 2 years; however, the authors concluded that the first diagnosis was incorrect.[7]

AMFB rarely shows recurrence or malignant transformation. Simple excision with clear margin is the best surgical approach. Especially, for a vulva lesion, radical or extended excision is not recommended for women of reproductive age because of cosmetic reasons. The extent of surgery is determined according to whether the tumor is malignant or not. Thus, the accuracy of results from the frozen biopsy in the operating room or fine needle aspiration biopsy before surgery are critical to determine the extent of the surgery.

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Conflict of Interest

No conflicts of interest between the authors and / or family members of the scientific and medical committee members or members of the potential conflicts of interest, counseling, expertise, working conditions, share holding and similar situations in any firm.

Authorship Contributions

Idea/Concept: *Yong-Soon Kwon, Jae Young Kwack;* ***Design:*** *Yong-Soon Kwon;* ***Control/Supervision:*** *Yong-Soon Kwon, Jae Young Kwack;* ***Data Collection and/or Processing:*** *Soojin Kim, Jeong Soo Lee;* ***Analysis and/or Interpretation:*** *Soojin Kim, Jeong Soo Lee;* ***Literature Review:*** *Jae Young Kwack;* ***Writing the Article:*** *Jae Young Kwack;* ***Critical Review:*** *Soojin Kim, Jeong Soo Lee;* ***References and Fundings:*** *Yong-Soon Kwon;* ***Materials:*** *Yong-Soon Kwon.*

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