

Original Article

Aggressive Angiomyxoma of the Vulva in a Woman

Xiaojing Li, Siyu Cao, Xuechai Bai and Liang Wang*

Department of Gynecology, The 2nd Affiliated Hospital, Zhejiang University School of Medicine, Hangzhou, Zhejiang, China

Summary

Background: Aggressive Angiomyxoma (AAM) is a rare invasive mesenchymal stromal tumor predominantly in women at reproductive age. The diagnosis is based on clinicopathologic and immunohistochemical features. The disease is known as a mesenchymal tumor of premenopausal women and it is extremely rare in men. It has a predilection at the vulvovaginal region, and which may be misdiagnosed as Angiomyofibroblastoma (AMF). Before the surgical resection, we can't confirm the diagnosis. AAM tends to relapse locally and be differentially diagnosed from the other mesenchymal tumors.

Case presentation: This is a case report of massive vulvar AAM in a 42-year-old Chinese woman with right labia majora mass, which has been developing within the previous 12 months, exhibiting the AAM clinical impression. The mass measured 16 × 10 × 3 cm, without diffuse ulcer and purulent discharge.

Interventions: We performed a wide surgical treatment with the mass. Other treatment options, such as hormonal therapy and radiotherapy, can be the potential alternatives.

Outcomes: The patient discharged 12 days after the surgery therapy. There was no AAM recurrence or metastasis in a period of 12-month follow-up. It is necessary with a long-term follow-up.

Conclusion: The vulvar AAM is an aggressive and benign mesenchymal tumor. In this case, we present the diagnosis, treatment, and prognosis for vulvar AAM. The tumor was removed completely by the surgery, but a long-term follow-up is requisite for surveilling on recurrence.

Keywords: Aggressive angiomyxoma; Mesenchymal tumor; Vulva

*Corresponding author: Liang Wang, Department of Gynecology, The 2nd Affiliated Hospital, Zhejiang University School of Medicine, No.88 Jiefang Road, Hangzhou, Zhejiang 310009, China, Tel: +0086 57187783128; E-mail: 2196042@zju.edu.cn

Citation: Li X, Cao S, Bai X, Wang L (2020) Aggressive Angiomyxoma of the Vulva in a Woman? J Reprod Med Gynecol Obstet 5: 063.

Received: November 27, 2020; **Accepted:** December 14, 2020; **Published:** December 21, 2020

Copyright: © 2020 Li X, et al. This is an open-access article distributed under the terms of the Creative Commons Attribution License, which permits unrestricted use, distribution, and reproduction in any medium, provided the original author and source are credited.

Abbreviations

AAM: Aggressive Angiomyxoma

ER: Estrogen Receptor

HE: Hematoxylin and Eosin

SMA: Smooth Muscle Actin

CA: Cancer Antigen

CEA: Carcino-Embryonic Antigen

AFP: α -Fetoprotein

SCC: Squamouscell Carcinomas

Introduction

First reported in 1983, aggressive angiomyxoma is a rare and slow growing mesenchymal tumour [1]. AAM usually has no clinical symptom and grows in a insidious manner as well as possesses a moderate-to-high risk of local relapse. Its diagnosis is still difficult because of its non-specific clinical and radiological aspects. It has poor long-term prognosis. AAM could appear in vulva, perineal region, buttocks, or pelvis in women at reproductive age [2,3]. So far, the underlying causes for AAM remain unclear. A few of recent studies suggest that AAM may be associated with chromosome alteration in 12q13-15 region [4,5]. Here, we describe a case of a giant vulvar AAM and take the treatment procedure for the patient, together with a literature review on AAM.

Case Report

We present the case of a 42-year-old female patient with a giant solid mass on the right vulva for 1 year, with no specific past medical history noted and has never been pregnant. No treatment was given because she did not pay any attention to it at the beginning. The mass grows and increases quickly, without fever, redness, secreta, dysmenorrhea, burning sensation, bleeding, abdominal pain, perineum pain, dysuria, difficulty in defecation, or other symptoms. Her complaints were merely of discomfort when seated and a discharge with a fishy odor. She denied having had oral, anal or vaginal intercourse when questioned privately. She had no history of surgery, inflammatory disease, medications, or trauma and her family history was unremarkable. The mass was round, well-circumscribed, pedunculated, soft, spongy in consistency and nontender. The size of the mass was about 16cm-10cm with a pedicle of 3cm (Figure 1).

No enlarged inguinal lymph nodes were palpated bilaterally. Bi-manual examination revealed that no nodule appeared on vaginal, cervix, uterus, or bilateral accessory. Surface body ultrasound showed that several vessels were observed in the perineum of the mass with a sign of slight blood flow signal. In her pelvic and abdominal cavity, no abnormality was found in enhanced magnetic resonance imaging (Figure 2).

The laboratory white blood cell, alanine amino transferase and C-reactive protein levels data revealed no significant abnormalities. The tumor markers, including cancer antigens Cancer Antigen

(CA)-125, CA-199, Carcino-Embryonic Antigen (CEA), CA-153, α -Fetoprotein (AFP) and Squamous Cell Carcinomas (SCC), were all in Fig normal ranges. We present a total excision of a tumor mass with clear margins at the second Affiliated Hospital, Zhejiang university school of medicine, with no evidence of any relapse to date during the follow-up (Figure 3).

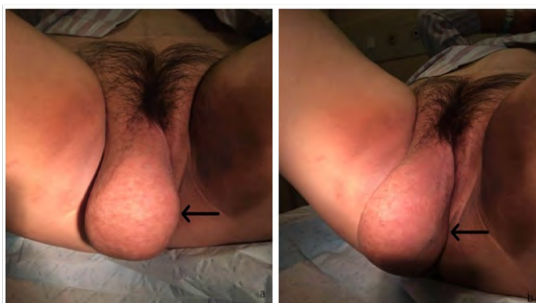


Figure 1: A mass in the perineum (arrows). The size of the mass was about 16cm-10cm with a pedicle of 3cm.

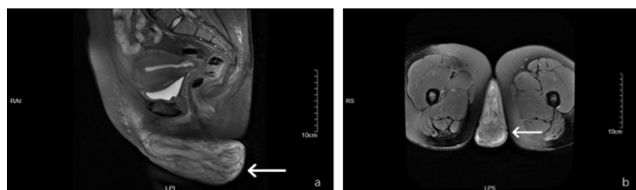


Figure 2: MRI of the pelvis showing the perineal mass (white arrow).



Figure 3: Reposition of mucosal flap and suture an incision site after tumor removal.

The histopathologic examination of the resected mass confirmed the diagnosis of AAM (Figure 4). The patient was discharged 12 days after a surgery with satisfactory outcomes. No evidence of recurrence or distant metastasis was observed during the 24-month follow-up period.

Discussion

The AAM mainly occurs on the vulva, vagina, pelvic cavity, hips, perineum, and crissum in reproductive female aging from 30 to 40 years old. Occasionally, AAM occurs in men. The morbidity rate of men versus women is about 1:6. It is aggressive because of its nature

with local infiltration and recurrence. Its relapse rate varies from 35% to 72%. The AAM primely occurs in the perineal and pelvic regions, which may lead to a possible misdiagnosis as Bartholin gland cyst or hernia. In addition, AAM is also difficult to differentiate from angio-myofibroblastoma due to similar morphology. Therefore, to diagnose AAM needs the evidence of clinical features and histologic pathologies.

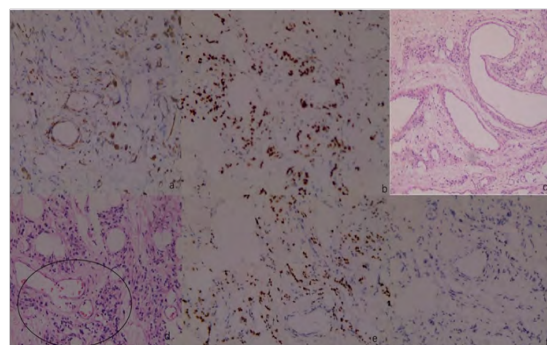


Figure 4: The pathologic and immunohistochemical characteristics of the mass. (A) Desmin was positive for immunohistochemistry. Magnification $\times 200$. (B) Strong and diffuse positivity for Estrogen Receptors (ER) (IHC for ER, $\times 200$). (C) HE staining; magnification, $\times 100$. (D) HE staining; magnification, $\times 200$. Small to medium-sized parenchyma vessels and more thick-walled small vessels. (E) Strong nuclear positivity in PR ($\times 200$). (F) S-100 stain was positive.

AAM is described as having both smooth and adherent margins that infiltrate the host's tissues. The histopathological characteristics consist of a population of hypocellular spindle cells sparsely spread in a loose myxoid matrix with collagen bundles. In conclusion, AAM is a locally benign and aggressive mesenchymal entity, and the surgical removal of the mass can cure it.

Findings

None.

Availability of Data and Materials

The data used and/or analysed during the current study are available from the corresponding author on reasonable request.

Ethical Approval

This study was approved by the ethics committee of the 2nd Affiliated Hospital, Zhejiang University School of Medicine (Hangzhou, People's Republic of China) and was permitted to be published. Written informed consent to have the case details and accompanying images published was obtained from the patient and her husband. All clinical investigations were conducted in accordance with the principles expressed in the Declaration of Helsinki.

Consent for Publication

Written informed consent was obtained from the patients for the publication of this case report and any accompanying images. A copy of the consent form is available for review by the Editor of this journal.

Competing Interests

The authors declare that they have no competing interests.

References

1. Steeper TA, Rosai J (1983) Aggressive angiomyxoma of the female pelvis and perineum. Report of nine cases of a distinctive type of gynecologic soft-tissue neoplasm. *Am J Surg Pathol* 7: 463-475.
2. Mccluggage WG, Connolly L, McBride HA (2010) HMGA2 is a sensitive but not specific immunohistochemical marker of vulvovaginal aggressive angiomyxoma. *Am J Surg Pathol* 34: 1037-1042.
3. Giraudmaillet T, Mokrane FZ, Delchier-Bellec MC, Motton S, Cron C, et al. (2015) Aggressive angiomyxoma of the pelvis with inferior vena cava involvement: MR imaging features. *Diagn Interv Imaging* 96: 111-114.
4. Elkattah R, Sarkodie O, Otteno H, Fletcher A (2013) Aggressive angiomyxoma of the vulva: A précis for primary care providers. *Case Rep Obstet Gynecol* 2013: 183725.
5. Srinivasan R, Mohapatra N, Malhotra S, Rao SK (2007) Aggressive angiomyxoma presenting as a vulval polyp. *Indian J Cancer* 44: 87-89.



- Advances In Industrial Biotechnology | ISSN: 2639-5665
- Advances In Microbiology Research | ISSN: 2689-694X
- Archives Of Surgery And Surgical Education | ISSN: 2689-3126
- Archives Of Urology
- Archives Of Zoological Studies | ISSN: 2640-7779
- Current Trends Medical And Biological Engineering
- International Journal Of Case Reports And Therapeutic Studies | ISSN: 2689-310X
- Journal Of Addiction & Addictive Disorders | ISSN: 2578-7276
- Journal Of Agronomy & Agricultural Science | ISSN: 2689-8292
- Journal Of AIDS Clinical Research & STDs | ISSN: 2572-7370
- Journal Of Alcoholism Drug Abuse & Substance Dependence | ISSN: 2572-9594
- Journal Of Allergy Disorders & Therapy | ISSN: 2470-749X
- Journal Of Alternative Complementary & Integrative Medicine | ISSN: 2470-7562
- Journal Of Alzheimers & Neurodegenerative Diseases | ISSN: 2572-9608
- Journal Of Anesthesia & Clinical Care | ISSN: 2378-8879
- Journal Of Angiology & Vascular Surgery | ISSN: 2572-7397
- Journal Of Animal Research & Veterinary Science | ISSN: 2639-3751
- Journal Of Aquaculture & Fisheries | ISSN: 2576-5523
- Journal Of Atmospheric & Earth Sciences | ISSN: 2689-8780
- Journal Of Biotech Research & Biochemistry
- Journal Of Brain & Neuroscience Research
- Journal Of Cancer Biology & Treatment | ISSN: 2470-7546
- Journal Of Cardiology Study & Research | ISSN: 2640-768X
- Journal Of Cell Biology & Cell Metabolism | ISSN: 2381-1943
- Journal Of Clinical Dermatology & Therapy | ISSN: 2378-8771
- Journal Of Clinical Immunology & Immunotherapy | ISSN: 2378-8844
- Journal Of Clinical Studies & Medical Case Reports | ISSN: 2378-8801
- Journal Of Community Medicine & Public Health Care | ISSN: 2381-1978
- Journal Of Cytology & Tissue Biology | ISSN: 2378-9107
- Journal Of Dairy Research & Technology | ISSN: 2688-9315
- Journal Of Dentistry Oral Health & Cosmesis | ISSN: 2473-6783
- Journal Of Diabetes & Metabolic Disorders | ISSN: 2381-201X
- Journal Of Emergency Medicine Trauma & Surgical Care | ISSN: 2378-8798
- Journal Of Environmental Science Current Research | ISSN: 2643-5020
- Journal Of Food Science & Nutrition | ISSN: 2470-1076
- Journal Of Forensic Legal & Investigative Sciences | ISSN: 2473-733X
- Journal Of Gastroenterology & Hepatology Research | ISSN: 2574-2566
- Journal Of Genetics & Genomic Sciences | ISSN: 2574-2485
- Journal Of Gerontology & Geriatric Medicine | ISSN: 2381-8662
- Journal Of Hematology Blood Transfusion & Disorders | ISSN: 2572-2999
- Journal Of Hospice & Palliative Medical Care
- Journal Of Human Endocrinology | ISSN: 2572-9640
- Journal Of Infectious & Non Infectious Diseases | ISSN: 2381-8654
- Journal Of Internal Medicine & Primary Healthcare | ISSN: 2574-2493
- Journal Of Light & Laser Current Trends
- Journal Of Medicine Study & Research | ISSN: 2639-5657
- Journal Of Modern Chemical Sciences
- Journal Of Nanotechnology Nanomedicine & Nanobiotechnology | ISSN: 2381-2044
- Journal Of Neonatology & Clinical Pediatrics | ISSN: 2378-878X
- Journal Of Nephrology & Renal Therapy | ISSN: 2473-7313
- Journal Of Non Invasive Vascular Investigation | ISSN: 2572-7400
- Journal Of Nuclear Medicine Radiology & Radiation Therapy | ISSN: 2572-7419
- Journal Of Obesity & Weight Loss | ISSN: 2473-7372
- Journal Of Ophthalmology & Clinical Research | ISSN: 2378-8887
- Journal Of Orthopedic Research & Physiotherapy | ISSN: 2381-2052
- Journal Of Otolaryngology Head & Neck Surgery | ISSN: 2573-010X
- Journal Of Pathology Clinical & Medical Research
- Journal Of Pharmacology Pharmaceutics & Pharmacovigilance | ISSN: 2639-5649
- Journal Of Physical Medicine Rehabilitation & Disabilities | ISSN: 2381-8670
- Journal Of Plant Science Current Research | ISSN: 2639-3743
- Journal Of Practical & Professional Nursing | ISSN: 2639-5681
- Journal Of Protein Research & Bioinformatics
- Journal Of Psychiatry Depression & Anxiety | ISSN: 2573-0150
- Journal Of Pulmonary Medicine & Respiratory Research | ISSN: 2573-0177
- Journal Of Reproductive Medicine Gynaecology & Obstetrics | ISSN: 2574-2574
- Journal Of Stem Cells Research Development & Therapy | ISSN: 2381-2060
- Journal Of Surgery Current Trends & Innovations | ISSN: 2578-7284
- Journal Of Toxicology Current Research | ISSN: 2639-3735
- Journal Of Translational Science And Research
- Journal Of Vaccines Research & Vaccination | ISSN: 2573-0193
- Journal Of Virology & Antivirals
- Sports Medicine And Injury Care Journal | ISSN: 2689-8829
- Trends In Anatomy & Physiology | ISSN: 2640-7752

Submit Your Manuscript: <https://www.heraldopenaccess.us/submit-manuscript>