Case Report

iMedPub Journals
www.imedpub.com

Gynaecology & Obstetrics Case report
ISSN 2471-8165

2020
Vol.6 No.1:7

DOI: 10.36648/2471-8165.6.1.88

A 19 cm Angiomyxoma of the Vulva in a 45-Year-Old Woman: A Case Report

Ietto C[1]*, Giannini A[2], Perutelli A[2], Pistolesi S[2], Pancetti F[2], Mannella P[2] and Simoncini T[2]

1 Department of New Technologies and Translational Research, Division of Pathology, University of Pisa, Italy
2 Department of Experimental and Clinical Medicine, Division of Obstetrics and Gynecology, University of Pisa, Italy

***Corresponding author:** Ietto C

chiaraietto90@gmail.com

Department of New Technologies and Translational Research, Division of Pathology, University of Pisa, Italy.

Tel: 393333819817

Abstract

Angiomyxoma is a rare, benign, locally infiltrative mesenchymal tumor that occurs in women at reproductive age. We present a 45-year-old female's case with a four years history of vaginal growth which has been progressively increasing in size. The patient was misdiagnosed of Bartholin's duct cysts and was asymptomatic. Aggressive angiomyxoma is a frequently misdiagnosed tumor that tends to recur locally. The prefix "aggressive" was added to emphasize this tumor characteristic. The diagnosis was based on histopathological and immuno-histochemical results. Treatment involves surgery that should aim wide local excision. However, over the years alternative treatment was proposed, in particular, hormonal therapy based on the female hormone receptors of this mesenchymal tumor. Application of imaging method, like ultrasounds, could be useful both for differential diagnosis and to evaluate the tumor before surgery. The frequent local recurrence of this disease requires a long-term follow-up.

Keywords: Angiomyxoma; Mesenchymal tumor; Vulvar tumor

Received: February 10, 2020; **Accepted:** March 03, 2020; **Published:** March 09, 2020

Citation: Ietto C, Giannini A, Perutelli A, Pistolesi S, Pancetti F, et al. (2020) A 19 cm Angiomyxoma of the Vulva in a 45-Year-Old Woman: A Case Report. Gynecol Obstet Case Rep Vol.6 No.1:7

Highlights

- Angiomyxoma is a rare, locally aggressive, mesenchymal tumor that arises in the perineal region and usually occurs in females at reproductive age.
- Patients often present no symptoms, except for a palpable mass of the vulva.
- Surgery is the main treatment option.
- Long-term follow up is necessary for the frequent local recurrence of the disease.

Introduction

Aggressive angiomyxoma is a rare, benign, locally infiltrative mesenchymal tumor found usually in women during reproductive age, which was first described by Steeper and Rosai in 1983. They reported a case series of 9 female patients, who presented a benign-appearing myxoid and vascular tumor that was infiltrative and had a propensity for local recurrence, hence the term aggressive [1]. It is a slow-growing, low-grade neoplasm involving the pelvis and perineum with a high risk for local recurrence, which often occurs after many years [2]. The typical characteristics include gelatinous appearance and locally infiltrative nature without evidence of nuclear atypia or mitosis. Microscopically, sections showed many walled vessels of various sizes, a loose myxoid and collagenous stroma and stellate and spindle-shaped neoplastic cells. Immunohistochemically, the neoplastic cells showed strong positivity for vimentin and desmin and moderate positivity for CD34 and hormone receptors for estrogen and progesterone [3]. Ultrasound can be a valuable imaging method for the preoperative diagnosis, evaluation of scope, and follow up of angiomyxoma [4]. Surgery is the main method for the treatment of symptomatic angiomyxoma; it's a benign disease originating from mesenchymal tissues with a tendency for local aggression [5].

Case Report

A 45-year-old female with a four years history of left-side vaginal and vulvar growth which has been progressively increasing in size. The swelling was not painful and there was no vaginal discharge or bleeding. The mucosa was intact and no pus discharge or

© Under License of Creative Commons Attribution 3.0 License

bleeding was found during preoperative examination. There was no history of perineal surgery, however, she had surgery for endometriosis ovarian cyst and the intrauterine device placed 5 years ago. Vaginal examination revealed a soft, fluctuant and non-tender mass on the left lateral vaginal wall measuring 7 × 4 cm, with no associated lymphadenopathy. The cervix, uterus, and adnexa were normal. Ultrasonography was performed and showed a unilocular mass that was oval, well-circumscribed, heterogeneous, located under the skin in the middle to lower part of the labia major. She was diagnosed as a left Bartholin's gland cyst and inserted into the pre-hospitalization path to plan the surgery since its size. Given the suspected non-malignancy, a preoperative biopsy was not performed. The patient underwent surgery, with the spinal anaesthesia, to remove the neoformation that extended from the medial surface of the left labia major, deeply, to the ischio-rectal fossa. An incision was made and subsequently following the cleavage plane of the neoformation wall it was possible to remove the entire para-vaginal mass. The neoformation showed a parenchymatous aspect, length of 19 cm, weight 175 g. Macroscopically, the tumor was a bulky, poorly circumscribed mass with surrounding tissues and had a tan-pink colour and rubbery consistency (**Figure 1**). At the end of the surgery blood losses were minimally. During the surgery, drainage was placed as prevention of bleeding and removed after 24 hours. The postoperative observation period was regular, without major complications. The patient was discharged after 48 hours with oral antibiotics for 5 days and subcutaneous heparin for 6 days. Five days after discharge, the patient was hospitalized for hyperpyrexia and vulvar hematoma with pus and blood leakage. The wound and blood culture at the febrile peak was performed. Endovenous ciprofloxacin was initially administered. After the swabs result the antibiotics have been replaced with Piperacillin/ Tazobactam endovenous 3 times a day for 10 days and Tigecycline endovenous twice a day for 6 days. Swabs were positive for *Enterococcus faecalis* and *Bacterioides fragilis.* Instead, blood culture resulted in negative. During hospitalization, the patient was monitored with blood tests and daily medications. The patient was discharged after 12 days with oral antibiotics and heparin at home. At the discharge, the patient was in good condition, apyretic, and she showed a significant reduction of the vulvar and perineal hematoma. The result of the histological examination of the vulvar formation was: angiomyxoma of the vulva. From a pathologic standpoint; microscopically the tumor was formed by fibro-myxoid stroma and fusiform spindle-shaped tumor cells with no atypia and mitosis, no necrosis or hemorrhagic areas (**Figure 2**). Immunohistochemistry revealed reactivity for desmin, vimentin, CD34, CD3 and Ki-67 activity of less than 1%. According to the histological report a close follow-up was recommended at least for 2 years for the risk of local recurrence.

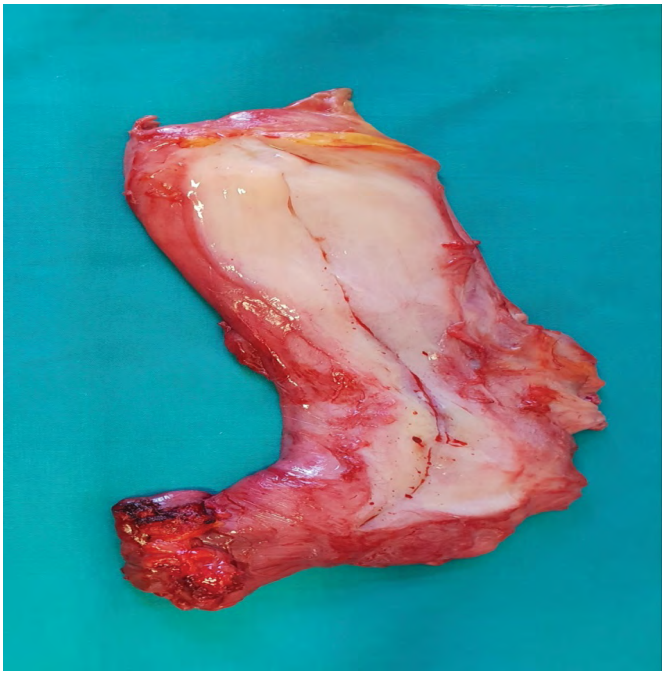

Figure 1 Macroscopic image of excised lesion.

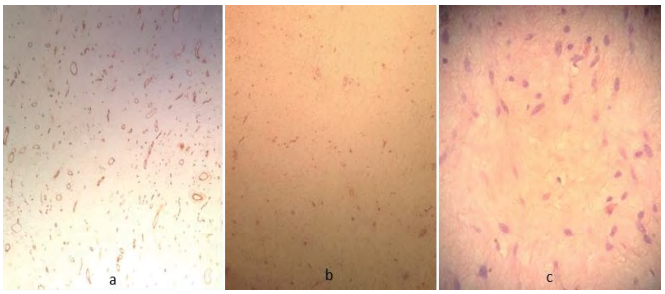

Figure 2 Microscopic images: (a) Rich vasculature evidenced by CD34 (IHC, Ventana Medica System, 4x), (b) Hypocellular mixoid area. H-E (4x), (c) Spindled mesenchimal cells with no atypia in a loose stroma (25x).

Discussion

Angiomyxoma is a benign neoplastic disease that originates from mesenchymal tissues with extensive local invasiveness and a high recurrence rate [5]. The rate of relapse varies from 35% to 72% [6]. It mainly occurs on the vagina, vulva pelvic cavity, perineum, hips and crissum in reproductive females, ageing from 30 to 40 years old. However, there were also cases occurring in the scrotum or spermatic cord of males [7,8]. Owing to its localization is often misdiagnosis as Bartholin's gland cyst, vaginal cyst, abscess, leiomyoma, lipoma or hernia [9]. In any case, the diagnosis of certainty can only be obtained with the histology. Tumor size is highly variable and ranges from 1 cm to 60 cm. The tumor is often tan-pink to tan-grey, bulky, and has a rubbery consistency with a glistening, gelatinous cut surface. Areas of congested blood vessels, haemorrhage, or fibrosis may be present [10]. Microscopically, sections showed many walled vessels of various sizes, a loose myxoid and collagenous stroma and stellate and spindle-shaped neoplastic cells. The positive immunohistochemistry results of strong positivity for vimentin and desmin and moderate positivity for CD34 and hormone receptors for estrogen and progesterone [11]. Cytogenetic studies of aggressive angiomyxoma are scares, therefore, recently, it has been shown that the architectural transcription factor HMGA2,

located on chromosome 12q13-15, is sometimes rearranged in this kind of tumor and this may result in aberrant HMGA2 protein expression [12]. HMGA2 is a member of the family of high mobility group proteins, which are architectural transcription factors expressed primarily during embryogenesis. However, HMGA2 is not a specific marker of aggressive angiomyxoma as it can be found in other vulvovaginal mesenchymal lesions (for example leiomyomatous neoplasm), but it could be useful in assessment of margins and in the detection of small foci of residual or recurrent tumor in reexcision specimens [13]. Besides, a novel translocation HMGA2-YAP fusion was described in a woman diagnosed of aggressive angiomyxoma, who was responsive to estrogen antagonism. This will allow the development of new target therapies [14]. As for the treatment, there is no global consensus on the preferred management for angiomyxoma, but surgical resection represents the mainstay of care. A complete and wide local excision is crucial, due to its recurrent nature. To achieve complete surgical excision and minimal invasiveness, it's essential to apply an imaging method. Multiple imaging modalities have been used to evaluate angiomyxoma preoperatively, including computed tomography (CT), magnetic resonance imaging (MRI), and ultrasonography. Appearance on imaging may be variable, but has been noted to have a distinct “swirling” pattern on MRI, related to its myxoid composition. On CT, it may be hypodense to muscle, or have both cystic and solid components. A combination of transabdominal, transperineal and transvaginal ultrasound exam could be helpful for the proper assessment that describes the lesion as a hypoechoic and heterogeneous mass [15]. Pre-operative imaging may thus provide clues to increase suspicion for angiomyxoma, but unfortunately for timely diagnosis, it is important to have a high index of suspicion and misdiagnosis rates have been reported as high as 82% [16]. Therefore, since estrogen and progesterone receptors are commonly positive, recently, hormonal therapy, such as GnRH agonists, has been used as a potential therapeutic target [17]. The use of hormonal modulating therapies has shown success in both neoadjuvant and adjuvant modality, with a resolution in residual or recurrent tumor growth [16]. Long-Term surveillance is further required for these patients since the recurrence and local invasion are the principal causes of morbidity [18].

Conclusion

In conclusion, angiomyxoma is a rare, locally aggressive tumor, which originates from mesenchymal tissues and can easily be misdiagnosed with other diseases just like Bartholin’s gland cyst. Surely MRI and ultrasound can be useful but not decisive to resolve doubts, because the diagnosis is histological. So, it is mandatory to keep in mind angiomyxoma when an asymptomatic and slow-growing vulvar mass is detected in young females.

Author Contribution

All authors made a substantive contribution to the information or material submitted for publication.

Conflict of Interest

The authors declare that they have no conflict of interest regarding the publication of this case report.

Funding

This research did not receive any specific grant from funding agencies in the public, commercial, or not-for-profit sectors.

Patient Consent

Written informed consent was obtained from the patient for the publication of this case report and the accompanying images.

Provenance and Peer Review

This case report was peer-reviewed.

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 Rashmi B, Anish K, Vanita S, Raje N (2007) Aggressive Angiomyxoma of the Vulva in Pregnancy: A Case Report and Review of Management Options. Med Gen Med 9(1):16.

3 Varras M, Akrivis C, Lekkou P, Kitsiou E, Demou A, et al. (2006) Aggressive angiomyxoma of the vulva: our experience of a rare case with review of the literature. Eur J Gynaecol Oncol 7(2):188-192.

4 Chen-Yang Z, Na S, Yu-Xin J, Yang M (2018) Application of ultrasound in aggressive angiomyxoma: eight case reports and review of literature. World J Clin Cases 6(14):811-819.

5 Salman MC, Kuzey GM, Dogan NU, Yuce K (2009) Aggressive angiomyxoma of vulva recurring 8 years after initial diagnosis. Arch Gynecol Obstet 280:485-487.

6 Methieson A, Chandrakanth S, Yousef G (2007) Aggressive angiomyxoma of the pelvis: A case report. Can J Surg 50:228-9.

7 Chihara Y, Fujimoto K, Takada S, Hirayama A, Cho M, et al. (2003) Aggressive angiomyxoma in the scrotum expressing androgen and progesterone receptors. Int J Urol 10:672-675.

8 Morag R, Fridman E, Mor Y (2009) Aggressive angiomyxoma of the scrotum mimicking huge hydrocele: case report and literature review. Case rep Med 2009:157-624.

9 Sutton BJ, Laudadio J (2012) Aggressive angiomyxoma. Arch Pathol Lab Med 136:217-221

10 Chan YM, Hon E, Ngai SW, Ng TY, Wong LC (2000) Aggressive angiomyxoma in females: is radical resection the only option? Acta Obstet Gynecol Scand 79(3):216-220

11 Sun Y, Zhu L, Chang X, Chen J, Lang J (2017) Clinopathological Features and Treatment analysis of rare aggressive angiomyxoma of the female pelvis and perineum - A retrospective study. Pathol Oncol Res 23:131-137.

12 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(7):1037-1042.

13 Rabban JT, Dal Cin P, Oliva E (2006) HMGA2 rearrangement in a

© Under License of Creative Commons Attribution 3.0 License

case of vulvar aggressive angiomyxoma. Int J Gynecol Pathol 25(4):403-407.

14 Lee MY, Da Silva B, Ramirez DC, Maki RG (2019) Novel HMGA2-YAP1 fusion gene in aggressive angiomyxoma. BMJ Case Rep 28;12(5).

15 Wiser A, Korach J, Gotlieb WH, Fridman E, Apter S, et al. (2006) Importance of accurate preoperative diagnosis in the management of aggressive angiomyxoma: report of three cases and review of the literature. Abdom Imaging 31:383-386.

16 Bai HM, Yang JX, Huang HF, Cao DY, Chen J, et al. (2013) Individual managing strategies of aggressive angiomyxoma of female genital tract and pelvis. Eur Oncol 39(10):1101-1108.

17 Poirier M, Fraser R, Meterissian S (2003) Case 1 Aggressive angiomyxoma of the pelvis: response to luteinizing hormone-releasing hormone agonist. J Clin Oncol 21:3535-3536.

18 Ma j, Wan J, Jiang B (2018) Huge abdominal and perineal aggressive angiomyxoma: A misdiagnosed case report and literature review. Cell Mol Biol 64:110-113.