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Case Report

Aggressive Angiomyxoma of the Labia Majora

João Brito Jaenisch Neto* and Caroline Rodrigues Cardoso

Vaginal Aesthetic Surgeon of Hospital Mãe de Deus, Menino Deus, Porto Alegre, Brazil

***Corresponding Author:** João Brito Jaenisch Neto, Vaginal Aesthetic Surgeon of Hospital Mãe de Deus, Menino Deus, Porto Alegre, Brazil.

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Abstract

Aggressive Angiomyxoma has been reported as less than 200 cases worldwide. It is a rare, infiltrative mesenchymal tumor of the vulvovaginal, pelvis and perineal areas. The purpose of this article is to present common history, presentation and findings, differential diagnosis, the management of this unusual tumor, and the prognosis after treatment.

Keywords: *Aggressive Angiomyxoma; Labia Majora*

Introduction

First described by Steeper and Rosai in 1983 [1], this aggressive, slow growing tumor arises in vulvovaginal region pelvis and perineum, with nonspecific symptoms [3,6].

Aggressive Angiomyxoma presents in the reproductive age woman with symptoms of a vulvovaginal mass, pelvic fullness, vulvar and vaginal pressure and the pain, dyspareunia, vulvar asymmetry, and unappealing vulvar appearance [2,5,8]. The differential diagnosis includes angiomyofibroblastoma, myxoid lipomatous tumors and lipomas, angiomyxoma, abscess, Bartholin Cysts, inclusion cyst, fibroepithelial stromal polyps, hernias, myxoids leiomyomas. The diagnosis is easily made by microscopic histopathologic examination [4,9]. Surgical excision is the treatment of choice both for definitive diagnosis, appearance, and personal comfort. Long term follow up is necessary due to the high incidence of recurrence [7].

Case Report

A healthy sexually active 36 year old female, G2P2, came into the office as a referral for a vulvar mass seen on ultrasound located on the right labia majora. This patient had two cesarean sections and no vaginal deliveries or vaginal/vulvar trauma in her history. She had a history of a neurogenic bladder due to a spinal cord injury 15 years ago. Her most pressing concern was the bulky aesthetic appearance of her right labia majora and the asymmetry it caused. It gave her great embarrassment that affected her interpersonal relationships in a negative manner. She felt abnormal and unwanted due to the strange appearance she internalized. The right labia majora had been growing for four years without pain except for the discomfort during intercourse.

On initial physical exam the patient presented with irritative and protuberant swelling in the right labia majora measuring 12 cm x 5 cm. The asymmetry was very pronounced on the front standing view. The skin was smooth and normal in appearance but there was a palpable smooth and spongy mass with minimal mobility entering into the vaginal canal and pressing inward onto the vaginal wall. The

internal vaginal canal was normal in texture and moisture with no abnormal lesions found. The adnexal and rectal exams were normal. An ultrasound was performed which showed a large non-specific mass taking up the entire space of the labia majora.

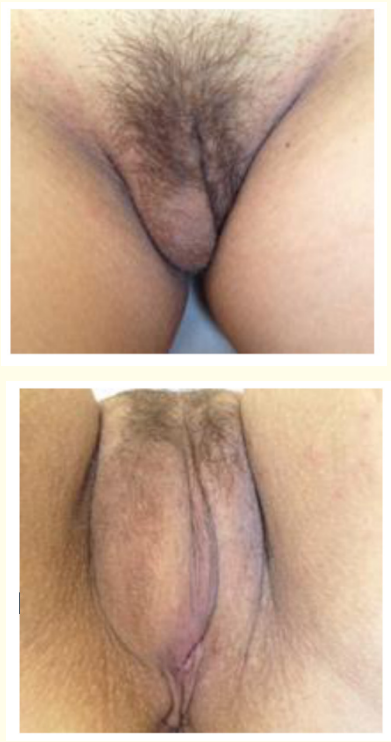

Figure 1

Surgery technique

Surgical excision and biopsy of the unknown mass was decided upon. A prior liposuction attempt had been unsuccessful. The patient was taken to the operating room and given general anesthesia and prophylactic Cephalosporin antibiotics. Measurements were carefully taken and marking were placed on a vertical plane to match the contours of the mass. The labia majora has multiple small branches of internal pudendal vessels with multiple small branches of nerves originating from the internal pudendal nerve. Local anesthetic with epinephrine given with a 30 gauge needle helped reduce the bleeding and post op pain without distorting the anatomic landmarks.

The vertical skin incision was made with a size 10 blade and blunt dissection with tonsil clamps and Metzenbaum scissors was carried out. Great care was taken to attempt and remove them mass intact and have the lesion "shell out" as much as possible while at the same time ensure complete excision with no abnormal tissues left that were grossly visible. A larger dissection was needed as the mass infiltrated the surrounding structures like an iceberg. Deeper dissection encountered brisk bleeding as larger vessels encountered needed both cautery and suturing. The dissection took 90 minutes. Closure of the wound was performed in several layers to eliminate the dead space and control any bleeding. Inverted mattress sutures and subcuticular suturing preceded interrupted skin closure using delayed absorbable sutures. Great care was taken to achieve an aesthetically pleasing closure with minimal tension on the edges. Post-operative

local anesthesia was given and the patient was discharged in stable condition 18 hours later. There was mild bruising but no hematoma formation. Ice packs and gentle pressure also helped with comfort.

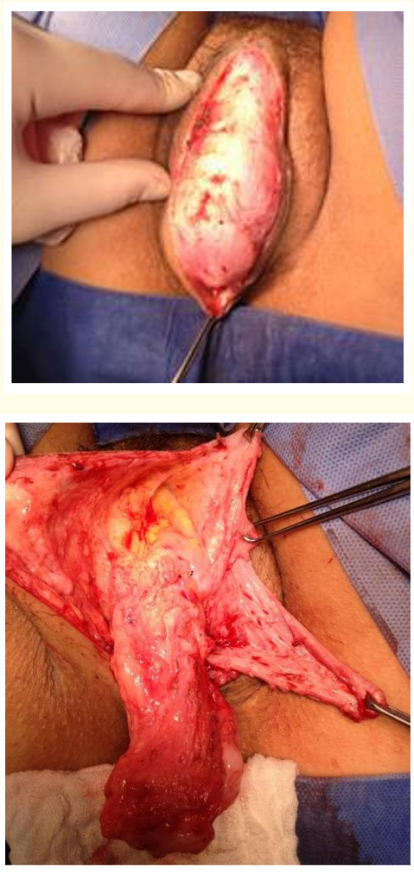

Figure 2

Histopathology: Mesenchymal neoplasm, fibroblast standard with aedematosous stroma richly vascularized.

Immunohistochemical

Material: Skin and fat of right labia majora.

Panel of antibody(s)

- Deomina: (D33): Positive
- RE (SP1): Positive
- Actina ML (1 A 4): Negative
- S100 (P): Negative
- Ki-67 (30-9): Proliferative index less than 1%

Conclusion: Compatible with Aggressive Angiomyxoma.

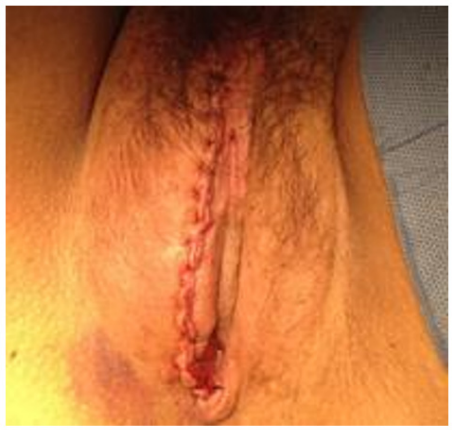

Figure 3

Discussion

Aggressive Angiomyxoma is a rare mesenchymal neoplasms that affect women in their reproductive age and can be mistaken clinically and microscopically for several other masses. The tumor is locally infiltrative, slow growing, with nonspecific symptoms, and arises in the vulvovaginal region, pelvis, and perineum. Surgery is the gold standard to treat this aggressive tumor. Surgery is often radical, at times mutilating, that results in heavy bleeding due to the rich vascularity of the involved areas of the pelvis.

Pre-operative diagnosis is difficult due to rarity and absence of diagnostic features. The pathogenesis of AA is poorly understood and histogenesis has been controversial.

Recurrences are not rare in the same location and can be seen months to years after the excision. Other alternatives treatments, such as Radiotherapy, Chemotherapy, or Angiography embolization also help after excision. Liposuction is of little benefit.

The long-term follow up essential as you can expect an average of one recurrence during the patient's lifetime.

Conclusion

Aggressive Angiomyxoma is a very rare benign neoplasm of the reproductive age years that occurs in the vulvovaginal regions such as the labia majora. The treatment Gold Standard is local excision and debulking. Local recurrences (80% in the first 5 years) are common but the long term prognosis is good to excellent.

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