Case Report

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Angiographic Embolization followed by Excision of Large Cellular Angiofibroma of the Vulva: A Case Report

Torres SB^{1*}, Diez Brito M¹, Peremateu MS¹, Wernicke A², Saez Perrotta C² and Marchitelli C¹

¹Departamento de Ginecología, Hospital Italiano de Buenos Aires, Juan D. Perón 4190 (C1181ACH) Buenos Aires, Argentina

²Departamento de Anatomía Patológica, Hospital Italiano de Buenos Aires, Juan D. Perón 4190 (C1181ACH) Buenos Aires, Argentina

***Correspondence:** Sofia Belen Torres, Departamento de Ginecología, Hospital Italiano de Buenos Aires, Juan D. Perón 4190 (C1181ACH) Buenos Aires, Argentina, E-mail: sofia.torres@hospitalitaliano.org.ar

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Abstract

Introduction: Cellular angiofibroma is a rare benign mesenchymal tumor, occurring almost exclusively in the superficial soft tissues of the genital region. Some of these lesions presents as large, hypervascularized masses. There are no reports in the literature to date on the challenges of resection of hypervascularized vulvar masses, or of hypervascularized cellular angiofibroma.

Materials and methods: We describe a case of a cellular angiofibroma of the vulva and the surgical approach with tumoral vessel embolization and posterior resection.

Case: A 47-year-old women was referred to vulvar pathology section of Italian Hospital of Buenos Aires for evaluation of left vulvar mass. A pelvic angiography was performed 5 days prior to surgery. It showed a hypervascularized formation at the level of the left vulva at the expense mainly of branches of the left internal pudendal artery. A complete resection was performed afterwards at our institution. The histopathological diagnosis was compatible with cellular angiofibroma.

Conclusion: We consider that surgical planning is essential and has to be performed with a multidisciplinary approach. Angiographic embolization should be taken into account as a valid resource when planning a surgery with a high risk of blood loss in order to decrease morbidity.

Keywords: Cellular angiofibroma, Mesenchymal tumor, Vulvar mass, Vulvar tumor, Angiographic embolization

Introduction

Cellular angiofibroma is a rare benign mesenchymal tumor, occurring almost exclusively in the superficial soft tissues of the genital region [1].

It was first described by Nucci and colleagues in 1997. These authors described four cases involving the vulva and added two in addendum [2,3]. The lesions were cellular and well circumscribed but unencapsulated and

were composed of spindle-shaped cells with characteristic prominent blood vessels with thick hyalinized walls. Entrapped fat was usually present around the periphery. Some of these lesions present as large, hypervascularized masses and they generate symptoms such as localized pressure and pain.

There are no reports in the literature to date on the challenges of resection of hypervascularized vulvar masses, or of hypervascularized cellular angiofibroma.

Materials and Methods

We describe a case of a cellular angiofibroma of the vulva and the surgical planning with tumoral vessel embolization and posterior resection. Patient has signed informed consent for publication.

Case Presentation

A 47-year-old woman was referred to the vulvar pathology section of Hospital Italiano de Buenos Aires for evaluation of a left vulvar mass (Figure 1). She had no relevant pathological personal records. The patient reported a history of pain in the last months, which led her to consultation.

Figure 1: Tumoral mass located in left labia majora of the vulva.

She had previously undergone a partial excision of the mass 2 months prior to the visit to our Institution. In the former surgery, surgeons were forced to interrupt the procedure as a consequence of excessive bleeding, performing an incomplete resection. The specimen was analyzed by our institution pathologists and the result was a cellular angiofibroma.

Images were requested for better characterization of the mass. Imaging findings were the following: Magnetic Resonance Imaging (MRI) scan showed increase in size of the left labia majora, solid formation

with abundant vascular component and signs of edema (hyperintensity in T2, without restriction in diffusion), measures approximately 77 × 48 × 29 mm. Intense enhancement in arterial and venous phases in perfusion sequences. It is interpreted as a tumor of the vulva with high vascularization. No regional or pelvic lymph nodes are observed (Figure 2a, 2b). A Doppler ultrasound evaluation was performed which confirmed the presence of a hypoechogenic formation of defined borders with abundant vascularization, suggestive of tumoral origin.

Figure 2a: MRI Coronal view, note vulvar mass encompassing left labia majora.

The case was presented at multidisciplinary tumor board and a decision was made to perform catheter embolization of the tumor feeding vessels prior to surgery to reduce the possibility of intra-surgical bleeding.

A pelvic angiography was performed 5 days prior to surgery. It showed a hypervascularized formation at the level of the left vulva at the expense mainly of branches of the left internal pudendal artery (Figure 3). Selective catheterization was deemed the most appropriate option; it was performed with a microcatheter that embolized the vessels with 700-900 micron spheres. The angiographic control showed the exclusion of the lesion and permeability of the proximal branches of the internal pudendal artery (Figure 4). The procedure was well tolerated without complications.

Figure 2b: MRI Sagittal View.

A complete resection was performed afterwards at our institution. A tumor formation measuring $6 \times 4 \times 2$ cm surrounded by fat tissue was recognized. The prior embolization undoubtedly facilitated the surgery, however towards the end of the surgical procedure it was difficult to identify the plane of dissection between the tumor and the vaginal wall, leading us to remove part of the vaginal mucosa to remove the whole tumor and to control hemostasis. After the excision a simple closure was performed and a Jackson Pratt drain was left in place. The patient was discharged after 1 day of hospitalization without complications. The drain was removed 1 week after the surgery (Figure 5).

Figure 4: Internal pudendal angiographic embolization.

Figure 3a, 3b: Pelvic angiography. Coronal and sagittal view.

The macroscopic appearance of the tumor revealed a whitish coloration with hemorrhage foci, defined edges and soft-elastic consistency. Microscopically, the tumor presented as a circumscribed formation constituted

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Figure 5: After the tumoral mass excision: primary closure and drainage placement.

Figure 6: Cellular angiofibroma H&E.

Figure 7: Immunohistochemistry.

Immunohistochemistry was performed, Cd 34: Positive RE: Positive Desmin: Negative (Figure 7). The histopathological diagnosis was compatible with cellular angiofibroma and margins of the surgical resection were negative.

In the first post-operative visit the patient presented fever and vulvar redness and swelling, which was interpreted as an incipient surgical site infection so antibiotic treatment was indicated (amoxicillin-clavulanic acid). She presented a good response with complete resolution.

Discussion

Cellular angiofibroma that was first described by Nucci et al. in 1997 is a benign mesenchymal neoplasm characterized by 2 main components: bland spindle cells and prominent small to medium-sized vessels with mural hyalinization.

It is found most frequently in middle-aged women around the fifth decade [2], however Amadhnia et al. reported a case of a 20-year-old woman [4]. It occurs equally in both sexes and most often arises in the inguinoscrotal or vulvovaginal regions, although cases occurring in extragenital locations such as perineum, chest wall and retroperitoneum have also been described [5,6].

In some cases it's difficult to make differential

diagnosis with other entities that manifest clinically as vulvar masses such as: Bartholin gland, labial, and sub mucosal cysts.

Vulvar angiofibroma usually occurs as a small lesion (usually less than 4 cm) with well-circumscribed margins [4]. In our case, the tumor was larger and did not have a well-defined capsule, also, at the distal pole of the tumor, a clear plane of cleavage with the vaginal wall was not found, making the resection difficult at this point and having to remove part of the vaginal mucosa.

With regard to histopathological differential diagnosis, there are many mesenchymal tumors of the vulva (also known as soft tissue or connective tissue tumors) that show similarity to cellular angiofibroma; they comprise the following: leiomyoma, hemangioma, and lipoma. Furthermore, spindle cell tumors include neurofibroma, schwannoma, and smooth muscle sarcoma. Less frequent masses include angiomyofibroblastoma, angiomyxoma, and cellular angiofibroma. Morphologic diagnosis can be done in most cases, but immunohistochemical studies are sometimes essential to confirm it [7].

Most of these tumors are specific to the genital region while others may be widely distributed. Aggressive angiomyxoma (AA), angiomyofibroblastoma (AMF), smooth muscle neoplasm's, fibromatosis and solitary

fibrous tumour (SFT), benign nerve sheath tumors, perineurioma, among others less frequent [8,9].

Cellular angiofibroma shares morphologic and immunophenotypic overlap with mammary-type myofibroblastoma and spindle cell lipoma. These 3 entities have also been shown to share genetic changes with involvement of 13q14,7,11, presenting themselves as variations of a single genetic entity [9,10].

The best treatment choice seems to be local excision with negative margins. Usually, this tumor does not recur, although it was once reported in a 49-year-old woman undergoing a simple excision of a 4 cm tumor. She subsequently developed a recurrent swelling at the site of the previous excision [8]. Nevertheless there is not currently available data on long-term follow up. We have also found no evidence about experience in the preoperative management of large and hypervascularized tumoral masses of the vulva. To the best of our knowledge, prophylactic angiographic embolization has not been described in the surgical approach for these kind of tumors.

Regarding follow-up, the patient has not had major complications related to the surgical procedure. A long-term control is suggested in order to monitor recurrences.

Conclusion

Cellular angiofibroma is a rare benign vulval mesenchymal lesion with limited potential for local recurrence. We consider that surgical planning is essential and has to be performed with a multidisciplinary approach, using images such as CT or RMI to further characterization of the tumor. Also, angiographic embolization should be taken into account as a valid resource when planning a surgery with a high risk of blood loss in order to decrease morbidity.

Declarations

The patient has given written consent for publication. Approval was obtained from the institutional ethical committee.

The author declares no financial support or conflict of interest.

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