Painful cellular angiofibroma of the vulva: case report

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Abstract

Introduction

Cellular angiofibroma is a rare benign mesenchymal tumour that occurs in the subepithelial myxoid stromal zone of the vulvovaginal region. We report a case of painful angiofibroma in a 52-year-old morbidly obese female, in the left labia.

Case report

A 52-year-old morbidly obese (body mass index of 67) female was referred to our gynaecologic oncology clinic with a 3-year history of a large, painful, pendulous left vulvar mass. Magnetic resonance imaging of the lesion helped confirm that the mass did not represent a hernia and was fibrous in nature. Resection revealed a 12 × 12 × 9 cm, a 187-gram encapsulated mass. Histologically, it was a well circumscribed tumour composed of hyalinised blood vessels and collagen bundles, exhibiting characteristic immunohistochemical features of a cellular angiofibroma.

Conclusion

Angiofibroma of the vulva is a rare benign encapsulated mass, yet hypervascular: Its diagnosis in a morbidly obese patient is challenging and necessitates magnetic resonance imaging or CT scan. Its surgical removal needs precision and meticulousness especially in a patient with multiple comorbidities.

Introduction

Cellular angiofibroma of the vulva is a benign mesenchymal lesion originally described by Nucci et al.1. It is found equally in men and women, with women affected earlier in life than men2. When found in the vulva, its average diameter is 3 cm3. Complete local excision is the adequate treatment of cellular angiofibroma, although data on long-term follow-up is not currently available4,5. There are no reports in the literature to date on the challenges of diagnosis and resection of cellular angiofibroma in morbidly obese patients. This case report describes the challenges of the diagnosis and surgical resection of a large angiofibroma in a morbidly obese patient.

Case report

A 52-year-old morbidly obese (body mass index of 67) female was referred to our gynaecologic oncology clinic with a 3-year history of a large, painful, pendulous left vulvar mass. Her past medical history was significant for diabetes mellitus, hypertension, chronic kidney insufficiency, sleep apnoea and bilateral lower extremity lymphedema that limited her mobility completely for years and leaving her wheelchair bound. Because of complex comorbidities she was denied surgical resection by other institutions.

On examination, her abdominal pannus measured 40 cm below her waist down to her knees and a 20 cm size mons pubis that covered her vulva and upper thigh. After exposing the perineum, her left soft and irreducible vulvar mass measured 10 cm, covering her left vulva, urethral meatus, entroitus and anus. The skin over the mass had demonstrated pressure changes, as this was the most dependent part of her body. The diagnosis necessitated MRI, which helped confirm that the mass did not represent a hemia and was fibrous in nature.

Understanding the complexity of her surgery and possible postoperative morbidity and mortality, special operative and recovery room teams were assembled including a high-risk anaesthesiology and nursing team.

After the induction of anaesthesia, the patient was positioned in a high lithotomy position (Figure 1A) and the surgical excision was performed carefully removing the mass with its capsule, with meticulous haemostasis. Resection revealed a 12 × 12 × 9 cm, a 187-gram encapsulated mass. The surgical bed was closed in three layers and the skin was closed using delayed absorbable stitch (Figure 1B).

Microscopic examination showed hyalinised blood vessels and collagen bundles. Fibrous areas were interspersed with mature adipose tissue (Figure 2A). Immunostains showed that the lesion was...
diffusely oestrogen receptor positive (Figure 2B) and focally progesterone receptor positive (Figure 2C) leading to the pathologic diagnosis of cellular angiofibroma.

The patient was discharged to her residence at a local skilled nursing facility after 3 days. Despite our best surgical effort in closing up the layers the surgical incision separated, we believe it is due, secondary, to her obesity, poor nutrition and being wheelchair bound. One month follow-up revealed her incision site healing by secondary intention, without signs of infection. After a minor adjustment in her oral pain control regimen, she was pain-free at six-week follow-up.

Discussion

Various benign mesenchymal lesions may occur in the vulva area such as cellular angiofibroma, angiomyofibroblastoma, aggressive angiomyxoma, lipoblastoma-like tumour of the vulva, fibroepithelial stromal polyp and superficial cervicovaginal myofibroblastoma. Cellular angiofibroma usually arises in subcutaneous tissue of the vulva in women and in inginal and scrotal regions of men. It has, however, been reported in the subcutaneous tissue of the extragenital locations such as chest wall, retroperitoneum and the oral cavity. Angiofibromas typically present as a painless vulvar mass and are often misdiagnosed as a Bartholin gland or submucosal labial cyst.

Pathologically, cellular angiofibroma is a well circumscribed cellular spindle cell neoplasm composed of uniform stromal cells admixed with prominent hyalinised vessels, giving rise to its name based on the two most prominent components of this tumour: the blood vessels and the uniform stromal cells. Cellular angiofibroma may show cellular atypia or morphologic features of sarcomatous transformation. The distinction of cellular angiofibroma from other common vulvar region neoplasms is primarily by morphology, although immunohistochemistry studies may contribute. Cellular angiofibromas are characterised by vimentin positivity with negative staining for desmin, α smooth muscle actin and S-100 protein. Negative staining for desmin and α smooth muscle actin helps to exclude myofibroblastic differentiation, while the lack of staining for S-100 protein excludes a nerve sheath tumour.

When first described, cellular angiofibroma was characteristically found in middle-aged women; however recent literature shows that this tumour occurs equally in women and men, with women affected most often in the fifth decade, and males in the seventh decade. The patient’s age at diagnosis ranges from 22 to 78 years old. Angiofibroma of the vulva is characteristically small, with an average diameter of 3 cm. The size of this tumour in general varies from 0.6 to 25 cm (with a median of 2.7 cm); however, our case had an angiofibroma that was 9 cm above the median.

Clinically, cellular angiofibroma most commonly follows a benign course. Local excision with clear margins is usually adequate in the treatment of these lesions, although data on long-term follow-up is not currently available. Recently, one report of local vulvar recurrence after six months was postulated in a recent review to be reflective of continued growth of residual disease, rather than any intrinsic local aggression. Complete surgical resection is of paramount importance in angiofibroma, as this tumour is almost always encapsulated and hypervascular. Surgical removal of the mass with its capsule does not only guarantee complete surgical removal and hence prevent its recurrence, but minimised blood loss. Identifying the capsule of the tumour is the most important early step in the surgical removal. This step becomes a very important one in a patient with such complex comorbidities like the one in this case report. Given the sensitivity of the common anatomic sites, there is no justification for using a wide surgical margin.

Conclusion

Angiofibroma of the vulva is a rare benign encapsulated mass, yet hypervascular. Its diagnosis in a morbidly obese patient is challenging and necessitates a MRI or CT scan. Its surgical removal needs precision and meticulousness especially in a patient with multiple comorbidities.
Case report

Consent
Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

References

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