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Angiomyofibroblastoma of the vulva: A case report and review of the literature

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ABSTRACT

Angiomyofibroblastoma (AMFB) represents a rare, benign mesenchymal tumor with a predilection for the vulvovaginal region. It is usually diagnosed in middle-aged women. Histopathology and immunohistochemical study remain the key to diagnosis. Like other benign mesenchymal vulval tumors, AMFB shows indolent behavior and rarely recurs after complete surgical excision. Herein, we present a case of vulvar AMFB in a 51-year-old woman to highlight the diagnostic difficulties when considering this rare entity.

1. Introduction

Angiomyofibroblastoma (AMFB) is a rare benign mesenchymal tumor that typically occurs in the female genital tract, particularly in the vulva and vaginal wall. It has occasionally been reported in the male scrotum and groin [1]. First described by Fletcher et al. in 1992 [2], AMFB is often diagnosed in middle-aged women [2]. The tumor usually presents as a slow-growing asymptomatic mass. The main clinical differential diagnosis is Bartholin's cyst and perineal hernia. Definitive diagnosis depends on histology, which shows a proliferation of bland-looking spindle cells, associated with thin-walled abundant vasculature. Herein, we present a case of vulvar AMFB in a 51-year-old woman to highlight the diagnostic difficulties when considering this rare entity.

2. Case Presentation

A 51-year-old Moroccan woman, gravida 5 para 5, presented to the department of gynecology with a one-year history of a slow-growing painless mass at the right vulva. She had no previous history and a regular cycle. Upon physical examination, she had a well-circumscribed pedunculated polypoidal mass of the right labia majora measuring 4 cm × 3 cm × 3 cm with normal overlying skin (Fig. 1). There was no palpable node in the inguinal area. The rest of the examination did not show any abnormality. Pelvic ultrasound revealed a 5 cm isthmic fibroma type 4. Vaginal smear was normal. Magnetic resonance imaging (MRI) revealed a well-circumscribed polypoid mass with heterogeneous intensity and enhancement in the subcutaneous tissue of the vulva. Complete blood count and biochemical parameters were all within normal range.

A complete surgical removal was performed and the specimen was sent for pathological examination. Macroscopic examination showed a polypoid, greyish-white, well-circumscribed, non-encapsulated lesion measuring 4 cm × 3.5 cm × 3 cm. There was no necrosis (Fig. 2).

Histologically, the tumor had two components: the first consisted of thin-walled capillaries arranged in lobules separated by fibrous septa with a focal myxoid changes; the second consisted of a proliferation of bland-looking spindle cells arranged haphazardly (Fig. 3(A and B)).

Immunohistochemically, almost all neoplastic cells were positive for desmin and smooth muscle actin (SMA), and negative for cytokeratin (AE1/AE3), CD34, and S100 protein. Thus, the definitive diagnosis was angiomyofibroblastoma (Fig. 3 (C and D)).

At one-year follow-up there was no evidence of recurrence.

3. Discussion

Angiomyofibroblastoma (AMFB) is benign mesenchymal tumor first described by Fletcher et al. in 1992 [2]. It commonly involves the female external genital tract, with few reports among men [3,4]. The prognosis is excellent; however, sarcomatous transformation has been reported in one case [5]. AMFB typically occurs in middle-aged women (age range

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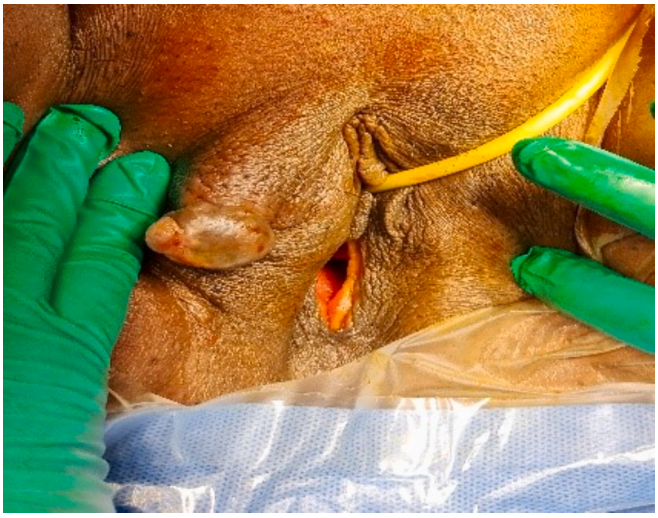

Fig. 1. Physical examination: pedunculated angiomyofibroblastoma of the vulva.

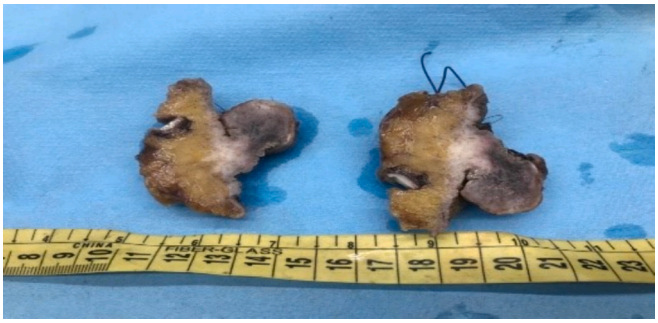

Fig. 2. Gross findings: polypoid, greyish-white, well-circumscribed, non-encapsulated mass.

from 30 to 50 years) [6]. In this case, the patient was 51 years old.

Clinically, it presents as a slow-growing, painless, subcutaneous mass; thus, it can be mistaken for Bartholin gland or labial cyst, lipoma or hydrocele [7].

On MRI, AMFB appears as solid, well-circumscribed variably heterogeneous soft-tissue masse [8]. Radiological findings are essential to draw surgical resection margins and for differential diagnosis before surgical removal.

On gross examination, AMFB present as pinkish to yellow lesions with mucoid or myxedematous cut surface, ranging from 0.5 to 12 cm in largest diameter [2]. Microscopically, the tumor classically shows alternating hypocellular and hypercellular areas, consisting of spindle cells associated with scattered capillary-sized blood vessels with focal hyalinization. The stroma might be abundant and myxedematous to collagenized. The nuclei show minimal atypia with scattered mitotic figures. In some cases, the neoplastic cells might be multinucleated, plasmacytoid and epithelioid [9]. The presence of an adipocytic component had been reported.

On immunohistochemistry, tumor cells express vimentin, desmin, and variably smooth muscle actin. They are negative for estrogen and/or progesterone receptors and negative S100 and cytokeratin [10]. CD34's expression is uncommon, and has been reported essentially in the lipomatous variant.

The differential diagnosis is broad, including malignant mesenchymal tumors, as well as aggressive angiomyxoma, cellular angiofibroma, fibroepithelial stromal polyp and superficial cervicovaginal myofibroblastoma.

Aggressive angiomyxoma (AAM) is a locally infiltrative tumor usually exceeding 10 cm, consisting of small spindle- to stellate-shaped cells embedded in abundant myxoid matrix with a "pushing" infiltrative borders. The cells are usually monomorphic and more uniformly distributed than in angiomyofibroblastoma [4]. Immunohistochemically, AAM and AMFB are positive for vimentin, estrogen receptor and progesterone receptor and negative for S-100, but their positivity for desmin and α-SMA is variable. Conversely, a recent molecular study supports that AAM and AMFB are distinct neoplasms. The HMGA2 gene arrangement has been described in a third of AAM cases but not in AMFB [11]. Differential diagnosis also includes cellular angiofibroma, which shows more thick-walled blood vessels and does not express hormone receptors. Other diagnoses which have to be ruled out are inflammatory

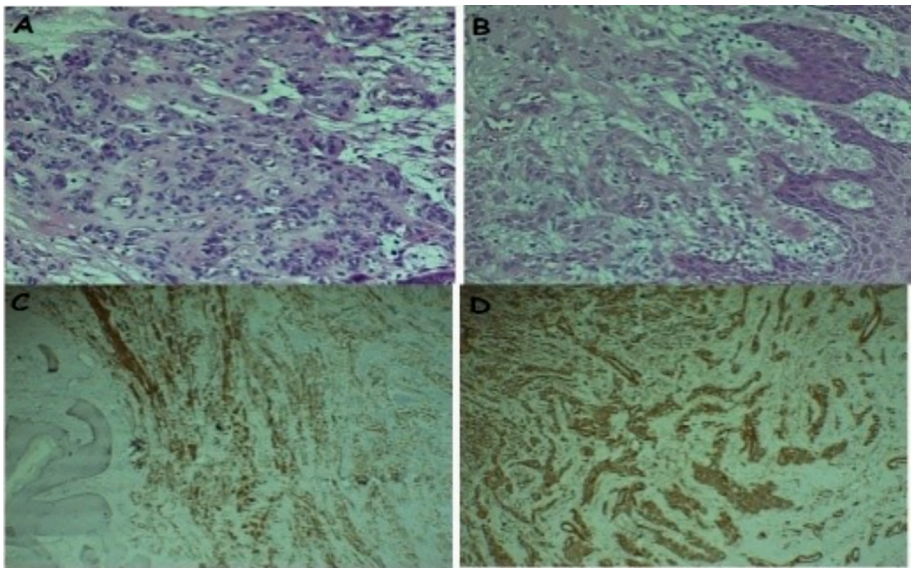

Fig. 3. A and B: Histopathological findings: The tumor had two components: the first consisted of thin-walled capillaries arranged in lobules separated by fibrous septa with a focal myxoid changes; the second consisted of a proliferation of bland-looking spindle cells arranged haphazardly (hematoxylin and eosin ×400). C and D: Immunohistochemistry staining: positivity for desmin and smooth muscle actin (SMA) (immunohistochemical stain ×400).

myofibroblastic tumor and inflammatory fibroid polyp [12].

Surgical removal is the gold standard treatment. In the present case, the patient underwent simple excision with clear margins.

The prognosis is generally favorable as AMFB has a very low risk of sarcomatous transformation [13]. Recurrence may occur if the lesion is incompletely excised.

4. Conclusion

In summary, we have reported the clinical and histopathologic features of the vulval angiomyofibroblastoma, which is frequently misdiagnosed before surgery. The complete excision followed by histopathology and immunohistochemistry findings confirm the diagnosis.

Contributors

Samia Sassi contributed to patient care, conception of the case report, acquiring and interpreting the data, undertaking the literature review and drafting the manuscript.

Chaimaa Nadim contributed to the conception of the case report, acquiring and interpreting the data and revising the article critically for important intellectual content.

Rihane El Mohtarim contributed to drafting the manuscript, acquiring and interpreting the data and undertaking the literature review.

Lamiae Rouas contributed to drafting the manuscript, acquiring and interpreting the data and undertaking the literature review.

Yousfi Mounia contributed to drafting the manuscript, acquiring and interpreting the data and undertaking the literature review.

Najat Lamalmi contributed to undertaking the literature review and revising the article critically for important intellectual content.

Fatima El Hassouni contributed to undertaking the literature review and revising the article critically for important intellectual content.

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Patient consent

Obtained.

Provenance and peer review

This article was not commissioned and was peer reviewed.

Conflict of interest statement

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

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