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Superficial Angiomyxoma of the Vulva: A Case Report

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Abstract

Superficial angiomyxoma is a rare benign mesenchymal tumor that mainly occurs in the genital region. We report the case of a 51-year-old woman with a painless vulvar mass, well circumscribed on ultrasound. On gross finding, it was a polypoid and bilobed mass, partially encapsulated. On histological examination, it was a proliferation of non-atypical spindle cells with an abundant, myxoid stroma and numerous medium-sized blood vessels. The diagnosis was superficial angiomyxoma. The clinical features do not often lead to the diagnosis of superficial vulvar angiomyxoma. It is based on histological examination and immunohistochemistry is helpful to differentiate it from other myxoid tumors.

Keywords

Angiomyxoma, Mesenchymal Tumor, Myxoïd Stromal, Tumor of Vulva

1. Introduction

Superficial angiomyxoma is a rare benign mesenchymal tumor of cutaneous location. This lesion is most often observed on the trunk, limbs, head or neck. Genital localization, mainly in the vulvar region, is less frequent [1] [2]. This pathology was first described as a soft tissue tumor associated with Carney's complex [2]. Currently, this entity has been established as an independent soft tissue tumor regardless of whether any genetic disease, including Carney's complex, exists or not [3].

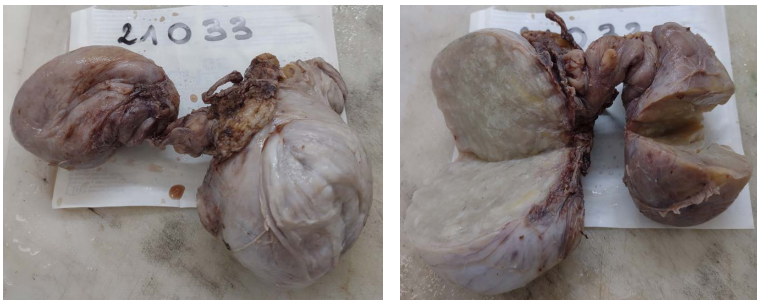
We report a case of superficial angiomyxoma of the vulva in a 51-year-old woman.

2. Observation

This was a 51-year-old postmenopausal woman, G2 P2, who came to the gynecological department of the Soavinandriana Hospital (CENHOSOA) Antananarivo for a vulvar swelling that had been evolving for 1 year with progressive increase in volume. The clinical examination showed a mass in the area of the labia majora, not painful to palpation. The skin surface opposite the lesion was macroscopically normal, without ulceration. Ultrasound revealed a subcutaneous mass, well circumscribed.

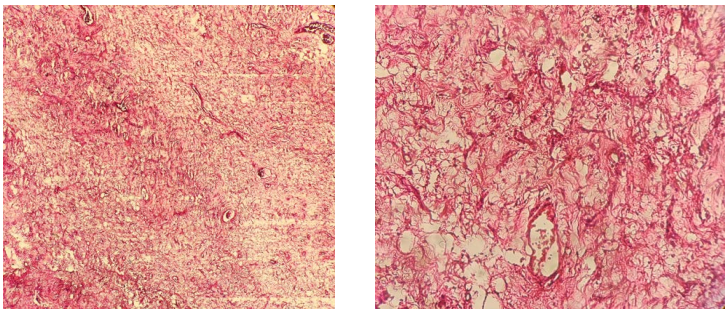
The patient benefited from an excision of the tumor.

On the macroscopic apparence, the mass was polypoid, bilobed with a smooth external surface, partially encapsulated and of soft consistency. Each lobe measured 8 × 5 × 4 cm and 7 × 4 × 3 cm, respectively. The two lobes were connected by a fibrous-like cord measuring 4 cm. On section, it was a homogeneous, yellowish-white, gelatinous fleshy nodule (Figure 1). The histological features of the nodule corresponded to a tumor proliferation of monomorphic spindle or star-shaped cells, lacking cyto-nuclear atypia with an abundant, myxoid stroma and numerous medium-sized blood vessels (Figure 2). In periphery of the lesion, striated muscle cells and adipose lobules of normal structure were observed, not invaded by the lesion. No mitosis figures were observed.



(a) (b)

Figure 1. Mass of the vulva, bilobed, partially encapsulated with a fibrous-like cord connecting the lobes (a); Homogeneous, yellowish-white, gelatinous fleshy nodule (b). Source: CENHOSOA.



(a) (b)

Figure 2. Proliferation of monomorphic spindle cells, without cyto-nuclear atypia with abundant, myxoid stroma and numerous medium-sized blood vessels HE ×40 (a); HE ×200 (b). Source: CENHOSOA.

Immunohistochemical examination was requested but was not honored by the patient (search for estrogen and progesterone receptor antibodies, desmin, actin and CD34).

The diagnosis was a superficial angiomyxoma.

The postoperative follow-up, in the short term, was without complication. After 6 months of follow-up, no tumor recurrence was observed.

3. Discussion

Superficial angiomyxoma is a relatively rare soft tissue tumor [2]. This entity was first described in 1986 by Carney *et al.* in their series on multifocal superficial myxoid tumors occurring in the Carney complex [3]. Benign tumors of the vulva account for 22% of vulvar diseases. Superficial angiomyxoma is a benign tumor rarely reported in the genital region and the vulvar localization is very rare [4].

This pathology can occur at any age with a peak of frequency in the 4th decade [5]. Our patient was 51 years old at the time of diagnosis.

Clinically, superficial vulvar angiomyxoma presents as a polypoid or papulo-nodular skin mass of superficial location (involvement of the dermis, hypodermis or both). It is a slow-growing, painless lesion that may evolve between 2 months and 10 years [1]. In our patient, the clinical presentation was consistent with that described in the literature and the lesion had been evolving for 1 year.

Because of its small size and rarity, this pathology may be clinically misdiagnosed as a labial cyst, polyp, Bartholin's cyst, Gartner's duct cyst or perineal hernia [5].

According to the literature, the lesion generally measures less than 5 cm (between 0.9 and 4 cm). However, some authors such as Mucize Ozdemir *et al.* [5], Hyun-Soo Kim *et al.* [1] have reported tumors larger than 5 cm in diameter. Their cases were respectively 6 cm and 12.5 cm in long axis. Macroscopically, the lesion is well limited and multilobulated, white or grayish in color, and myxoid or gelatinous in appearance on section. The section slices sometimes show hemorrhagic remodeling [6]. Our case was close to what has been described in the literature but its particularities were to be bilobed.

On histological examination, superficial angiomyxoma corresponds to a proliferation of tumor with a clear boundary, hypocellular, organized in lobules. It is made of spindle or star-shaped cells with monomorphic nuclei, lacking cyto-nuclear atypia. The stroma is abundant and myxoid. The lesion presente numerous small to medium-sized thin-walled blood vessels with an inflammatory infiltrate, especially neutrophils [6]. According to Hyun-Soo Kim *et al.* [1], in 25% - 30% of cases, an epithelial component within the tumor proliferation can be seen, usually in the form of cysts, lined by a squamous-like lining or clusters of basaloid cells. These foci probably result from tumor invasion of adnexal structures. Mitoses are rare [1]. Concerning our case, there is no inflammatory infiltrate and epithelial component.

There is a differential diagnosis according the morphological appearance of superficial angiomyxoma with other mesenchymal tumors of the vulva, such as

aggressive angiomyxoma, angiofibromyxoma.

For aggressive angiomyxoma, the tumor size, most often the neoplasm has a diameter greater than 10 cm. Macroscopically, the lesion is non-encapsulated, soft, polypoid and gelatinous in appearance on section. Histologically, it is a hypocellular tumor, poorly limited. It is composed of fibroblasts and myofibroblasts [6] and is characterized by the infiltrative aspect of the lesion invading deeply into the perilesional soft tissues [1] [4] [7] [8]. The cells are spindle-shaped, small and uniform, with pale and poorly bounded cytoplasm, within an abundant and myxoid stroma with numerous large to medium-sized vessels with thick, hyalinized walls, which is not identified in a superficial angiomyxoma [8], these morphological aspects were not observed in our case.

Angiofibromyxoma is a well-limited cystic lesion, usually less than 5 cm in diameter, which clinically makes one suspect a Bartholin's cyst [5]. It is characterized on histological examination by the presence of alternating hypo- and hypercellular areas. The latter has a much higher stromal cellularity than that of a superficial angiomyxoma. The tumor cells have an eosinophilic cytoplasm and a plasmacytoid-looking nucleus. Numerous thin-walled capillary-like vessels are present and tumor cells tend to aggregate around the blood vessels [1].

Apart from morphological features, immunohistochemical examination is an aid in deciding between these diagnostic entities.

It has been reported that spindle cells of superficial angiomyxomas do not show immunoreaction with actin and desmin [5] [9], estrogen and progesterone receptors [1]. They express vimentin, CD34 and CD44, S-100 [1] [10]. CD44 immunostaining is characteristic of aggressive angiomyxoma. In contrast, the absence of ER and PR labeling points to superficial angiomyxoma, as aggressive angiomyxoma and angiofibromyxoma are usually positive for one or both of these receptors [1] [11]. The tumor cells of these two diagnostic entities are usually positive for desmin [1].

In our case, the diagnosis was based on the morphological appearance of the lesion, in the absence of immunohistochemical results.

Superficial angiomyxoma is a benign lesion, however, local recurrence is possible, requiring a healthy resection margin. The local recurrence rate is approximately 30% - 40% and is associated with incomplete resection [1] [12]. In our patient, after 6 months of follow-up, there was no tumor recurrence, which supports our diagnosis.

4. Conclusion

Vulvar superficial angiomyxoma is a rare benign tumor. The diagnosis is often not suspected on clinical examination. It is based on histological examination. The morphological features can sometimes be confused with other myxoid tumors of the vulva. Superficial angiomyxoma is characterized by the lobulated appearance of the tumor, hypocellularity of the proliferation with a clear boundary, and the absence of large, thickened, hyalinized vessels. Immunohisto-

chemical examination is helpful to diagnosis, the absence of ER and PR staining points to a superficial angiomyxoma.

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

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