

## Unknown Tumor of the Vaginal Wall: Case Report and Literature Review

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### Abstract

**Background:** Superficial myofibroblastoma of the lower female genital tract is a rare, recently described tumor, which has a distinctive clinico- pathological profile. It is a diagnosis that needs to be thinking for appropriate treatment.

**Case:** We report a 21-year old patient who presented with a pelvic mass, with no significant medical history. The radiological assessment showed a pelvic cystic tumor suppressing up the uterus. A surgical excision was performed with a diagnosis of superficial myofibroblastoma of the lower female genital tract. The patient received no adjuvant therapy after surgery and remains well with no signs of local recurrence at 16-month follow-up

**Conclusion:** This tumor should be differentiated from other mesenchymal lesions, which may arise in this area. Treatment and prognosis are different according to histological type. The authors discuss this finding through the diagnostic difficulties and the evolution of this entity.

**Keywords:** Myofibroblastoma; Vagina; Pathology; Immunohistochemistry

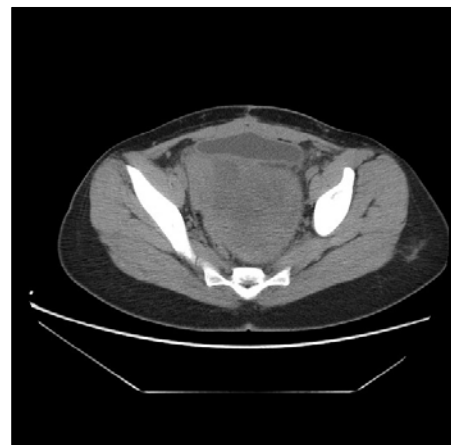
### Introduction

Superficial myofibroblastoma of the lower female genital tract (SMFGT) is a rare neoplasm which is preferentially situated in the vagina and cervix [1-3] and less frequently in the vulva. In 2001, Laskin et al. [1] firstly described a series of 14 seemingly distinctive mesenchymal tumors that occurred exclusively in the superficial lamina propria of the vagina and cervix of middle to old-aged women, They proposed the term “superficial cervicovaginal myofibroblastoma (SCVM) to highlight the unique features of this tumor: the superficial subepithelial location and myofibroblastic differentiation of the tumor cells. In this paper we describe a new case -to the best of our knowledge, the first-one in Moroccan literature-, discuss the main histological features of this rare entity and the problem of differential diagnosis it raises.

### Case Presentation

The patient, a 21-year old female, presented with a two year-history of a pelvic mass that enlarged gradually. The patient had consulted a private structure where she was treated surgically without histological evidence. 6 months later the patient consulted again in our hospital for the same problem. Clinical examination showed a fluid hypogastric mass, fixed deep plane. Ultrasonography showed cystic well-defined mass with mild heterogeneity, measuring 10 cm in maximum dimension. The scan confirms the size and cystic nature of mass (Figure 1), with regular contours, driving forward bladder, uterus and rectum laterally, without enhancement after injection of contrast. Radiologists have suggested a cystic lymphangioma or leiomyoma with cystic alterations. After multidisciplinary meeting, surgical resection was decided. Surgical excision was performed. The cystic mass was received fragmented. Microscopic examination showed cystic walls lined by mesenchymal cells. The mass was made up of several cystic cavities. The lesions were located in the lamina propria of the vagina (Figure 2) and separated from the overlying epithelium by a vague rim of uninvolved stroma. There are alternatively hypo and hypercellular areas (Figure 3) with heterogeneous cellularity and diverse growth patterns. In the superficial areas, the lesions were less cellular, composed of uniform spindle to stellate shaped cells that arranged mostly in a haphazard fashion. A lacelike or sievelike pattern was observed focally in these areas. The central or deep areas were more cellular and appeared collagenized, composed of slender spindle cells frequently arranged in waving fascicles with paralleling collagen fibers.

A gradual transition between the superficial areas and the central cellular areas could be observed. On high power examination, the spindle or stellate cells had a bland appearance with oval to fusiform nuclei and pale eosinophilic cytoplasm. Mitotic figures or necrosis were absent. There are scattered inflammatory cells mainly represented by lymphocytes and plasmocytes, the lesion contained a few small to medium-sized vessels. Immunohistochemical studies were performed on paraffin-embedded sections with a panel of antibodies using the standard EnVision technique. Appropriate positive controls were run simultaneously. The results showed that the lesion was diffusely positive with desmin (Figure 4), calponin and estrogen receptor (ER)



**Figure 1:** CT: multilocular pelvic cystic mass 10 cm, with regular contours, driving forward bladder, uterus and rectum laterally.

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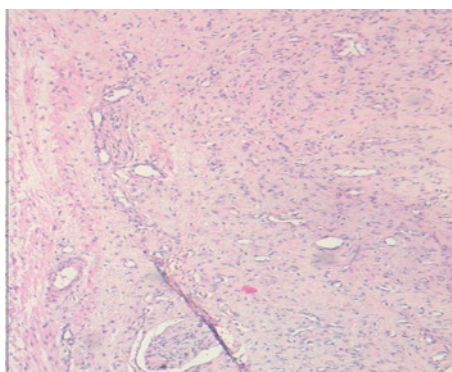


Figure 2: HES X 10. Lesions were located in the lamina propria of the vagina.

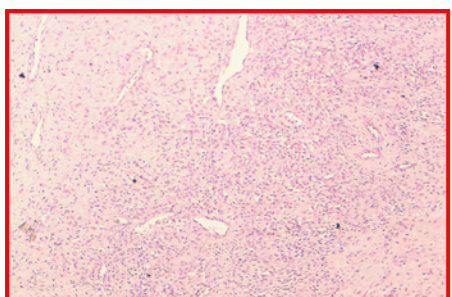


Figure 3: HES x 10. Hypo and hypercellular areas.

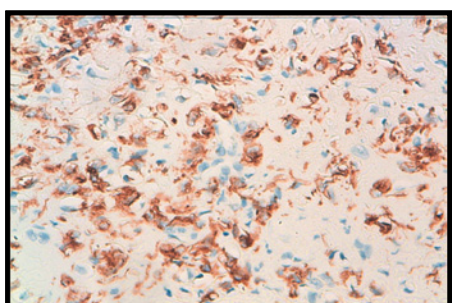


Figure 4: Immunohistochemical reactivity with desmin.

(Figure 5). CD34, HMGA2, h caldesmon, S100 protein, CD99, inhibin, calretinin and smooth muscle actin are negative. The features are those of a superficial myofibroblastoma of the lower female genital tract with central cystic degeneration. The patient received no adjuvant therapy after surgery and remains well with no signs of local recurrence at 16-month follow-up.

## Discussion

A wide variety of mesenchymal lesions occur in the lower female genital tract [4,5]. Broadly, these mesenchymal lesions can be separated into two groups. The first group includes several well-characterized tumors that show a marked tendency to occur in the lower female genital tract, such as aggressive angiomyxoma, angiofibroblastoma (AMF) and cellular angiofibroma (CA) [6-8]. These tumors can also be called as relatively site-specific. The second group embraces a wide range of heterogeneous lesions that frequently occur in this region, but arise in other anatomic sites as well, with examples

as fibroepithelial stromal polyp (FSP), superficial angiomyxoma. SCVM represents a new member of the first group. SCVM appears under recognized. To our knowledge, current case raises the number to 36. This rare entity affects women ranging in age from 23 to 80 years [9], which is preferentially situated in the vagina and cervix and less frequently in the vulva [1-3]. The pathogenesis of SCVM is not well understood. However, nearly one third of SCVM patients had a history of Tamoxifen and pregnancy or hormone-replacement therapy, suggesting that hormones may possibly have a role in the development of the tumor. It has been postulated that the subepithelial mesenchymal cells located in the perivascular or in connective tissue (lamina propria) of the lower female genital tract, which show a "native" positivity for oestrogen and progesterone receptor [4], tend to be the origin of most site-specific mesenchymal tumors that arising in this site [1]. Clinically, the most common presentation was an asymptomatic polypoid nodule or mass, with cysts or polyps being the most preoperative diagnosis [10]. Most SCVM were described as polyps or had a polypoid outline, measuring 1.0 cm to 8.0 cm (mean, 2.5 cm) in diameter, not well limited, encapsulated and lobulated with cystic features. Histological examination shows a well-circumscribed but unencapsulated lesion covered by unremarkable or hyperplastic squamous epithelium. Deep to the surface epithelium there is usually an uninvolved Grenz zone [9], although rarely the lesion extends up to the epithelial-subepithelial junction. There is typically moderate cellularity with cells with bland ovoid, spindle or stellate nuclei, often with a wavy appearance, embedded in a finely collagenous stroma, sometimes with thicker collagen bundles. Multiple patterns, including lacelike, sieve-like and fascicular, are a characteristic feature, as are myxoid or oedematous foci. There are no or few mitotic figures. Tumor cells in SCVM typically expressed vimentin, desmin, CD34, ER and PR (progesterone receptor); with desmin and CD34 accentuating the dendritic processes of the cytoplasm, whereas actin was consistently negative. Due to some histological and immunohistochemical overlapping features, SCVM can be confused with other mesenchymal lesions of the lower female genital tract. The principal differential diagnosis is FSP, especially the cellular variant [8]. The younger population of FSP, absence of distinct Grenz zone in FSP, lack of heterogeneous cellularity and diverse growth patterns, are crucial features to distinguish SCVM from FSP [8]. The other tumor enters the differential diagnosis is AMF, it is also characterized by alternating hypercellular and hypocellular areas. However, unlike SCVM, AMF contains abundant small to medium sized blood vessels. The tumor cells in AMF also have a propensity to grow around the vessels. Furthermore, SCVM is frequently negative for  $\alpha$ -smooth muscle actin. In contrast, AMF is usually positive for actins [8]. Cellular angiofibroma (CA) is another site-specific mesenchymal tumor that should be considered in the differential diagnosis. CA usually involves the vulva and the inguinoscrotal region. CA differs

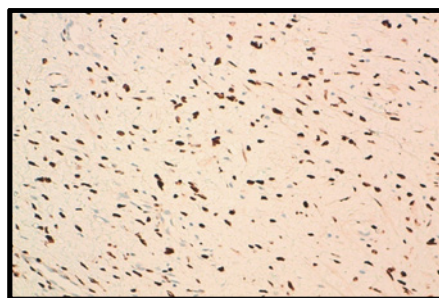


Figure 5: Immunohistochemical reactivity with estrogen receptor<sup>1</sup>.

from SCVM by its numerous small- to medium-sized thick-walled vessels and negative staining for desmin. Other lesions that may be confused with SCVM include solitary fibrous tumor (SFT) and mammary-type myofibroblastoma. SFT which may display alternating cellularity. In contrast to SCVM, SFT is typically positive for CD34 and negative for desmin. Mammary-type myofibroblastoma [11] is a spindle cell tumor with morphological and immunophenotypic features identical to myofibroblastoma of breast. Mammary-type myofibroblastoma shows a predilection for the inguinal area of older men. Based on the reported cases, superficial myofibroblastoma of the lower female genital tract is a benign lesion, although local recurrence 9 years after excision has been reported in one case. Metastasis has not been reported [9]. Superficial myofibroblastoma of the lower female genital tract diagnosis based on clinical, radiological and morphological patterns. Adjuvant therapy with radiotherapy in invasive forms is recommended [8]. Summarizing, we report a new case of vaginal superficial myofibroblastoma and describe the family of mesenchymal lesions, which may arise in this area.

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