

## CASE REPORT

# Vaginal superficial myofibroblastoma. Case report and review of the literature

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### Abstract

Superficial myofibroblastoma of the lower female genital tract is a rare, recently described tumor, which has a distinctive clinicopathological profile. We report a 63-year old patient who presented with a polypoid vaginal mass. Histopathological examination revealed a moderately cellular tumor composed of bland spindled and stellate cells. Immunohistochemical investigations showed reactivity for vimentin, desmin and CD34. This tumor should be differentiated from other mesenchymal lesions, which may arise in this area.

**Keywords:** myofibroblastoma, vagina, pathology, immunohistochemistry.

### Introduction

In 2001, Laskin WB *et al.* [1], based on the observation of 14 patients, described a distinctive tumor of the lower female genital tract, which they named "superficial cervicovaginal myofibroblastoma".

Recent publications added 18 new cases [2–4]. As in the series of Ganesan R *et al.* [2] the tumors occurred in the vulva and vagina rather than in the cervix and vagina, the authors proposed the term "superficial myofibroblastoma of the lower female genital tract".

In this paper we describe a new case – to the best of our knowledge, the first-one in Romanian literature –, discuss the main histological features of this rare entity, and the problems of differential diagnosis it raises.

### Material and methods

The patient, a 63-year old female, presented with a two year-history of a vaginal mass that enlarged gradually.

No hormonal therapy was mentioned in her medical record.

Gynecologic examination revealed a polypoid tumor, 4 × 3.5 cm, attached to the right lateral wall of the vagina by a short stalk. The lesion was excised and submitted to the Department of Pathology.

The specimen was fixed in 10% formalin and embedded in paraffin. The slides were stained with hematoxylin-eosin.

Immunohistochemistry was performed with Dako reagents for vimentin, desmin, alpha-smooth muscle actin ( $\alpha$ -SMA), S-100 protein, estrogen and progesterone receptors and CD34.

### Results

*Gross examination* showed a well-circumscribed, unencapsulated, lobulated, firm tumor, with a pinkish-gray cut surface.

*Microscopic examination* revealed a superficially located tumor, separated from the epithelium by a band of uninvolved tissue. The tumor, moderately cellular, was composed of spindled (Figure 1) and stellate cells. The nuclei were ovoid or wavy, with a finely dispersed chromatin and inconspicuous nucleoli (Figure 2). No mitoses were seen. The cells were embedded in a finely cellular stroma. Some areas were more cellular and showed a fascicular pattern of growth (Figure 3), while in other areas the neoplastic cells were dispersed in a more dense collagenous stroma (Figure 4). There were no necrotic foci. The tumor was penetrated by capillary-like or somewhat larger vessels with thin walls (Figure 5).

Immunohistochemical investigations showed a diffuse and strong reactivity for vimentin (Figure 6) and desmin (Figure 7), and a less intensely staining for CD34 (Figure 8). Desmin stain decorated the cytoplasmic processes of the cells, giving the impression of a syncytium. Stains for  $\alpha$ -SMA, S-100 protein and estrogen and progesterone receptors were negative.

### Discussions

Superficial myofibroblastomas of the lower female tract are rare tumors, which are preferentially situated in the vagina and cervix [1–4] and less frequently in the vulva [2].

A history of Tamoxifen treatment was documented both in myofibroblastoma and in other mesenchymal lesions of the lower female genital tract, suggesting a possible role of hormones in the pathogenesis of these lesions.

It has been suggested that superficial myofibroblastoma, aggressive angiomyxoma, superficial angiomyxoma, cellular angiofibroma, angiomyofibroblastoma and fibroepithelial polyp are closely related entities, which have a common histogenesis, originating in the lower female genital tract, possibly from a pluripotential primitive cell located around the vessels of connective tissue [5–10].

These cells show a native positivity for estrogen receptor and progesterone receptor, which is shared by the neoplastic elements derived from these cells. Two cases occurring in the vulva [2] and the vaginal tumor that we have studied were estrogen and progesterone negative, possibly arising from a population of hormone receptor negative cells.

The microscopic features of the cases reported so far

typify a quite distinctive neoplasm. It is well circumscribed, but unencapsulated, superficially located and separated from the epithelium by a rim of collagenous or myxoid stroma (“grenz zone”). The cells, spindle or stellate-shaped, are placed in a collagenous stroma, which may contain dense areas. In some cases there were hypocellular, myxoid areas, resulting in a lace-like pattern. Mitotic activity is minimal or absent. Thin-walled vessels tend to concentrate towards the center of the lesion. The clinical course is usually uneventful [1–3].

One patient developed, however, a local recurrence nine years after the initial treatment, so that a prolonged follow-up was recommended [3].

The results of immunohistochemical investigations are summarized in Table 1. Most tumors were reactive for vimentin, desmin, CD34, estrogen receptor and progesterone receptor. The percentage of reactivity for  $\alpha$ -SMA and muscle specific actin is lower. The experience with calponin, CD99, and bcl-2 is still limited.

**Table 1 – Results of immunohistochemical investigations**

Reference	CD34	Vimentin	Desmin	$\alpha$ -SMA	Muscle specific actin	Calponin	CD99	bcl-2	ER	PrR
Laskin WB <i>et al.</i> [1]	11/13	5/5	13/13	5/11	2/8				10/10	10/10
Ganesan R <i>et al.</i> [2]	6/12	11/11	9/12	0/12					9/11	
Stewart CJR <i>et al.</i> [3]	5/5	5/5	5/5	2/5		5/5	5/5	5/5	5/5	5/5
González Martínez MB <i>et al.</i> [4]	1/1	1/1	1/1						1/1	1/1
<b>Total</b>	<b>22/31</b>	<b>22/22</b>	<b>27/30</b>	<b>7/28</b>	<b>2/8</b>	<b>5/5</b>	<b>5/5</b>	<b>5/5</b>	<b>25/27</b>	<b>16/16</b>

$\alpha$ -SMA – alpha smooth muscle actin; ER – estrogen receptor; PrR – progesterone receptor

Superficial myofibroblastoma of the lower female genital tract raises difficult problems of differential diagnosis with other mesenchymal lesions, which may arise in this area.

Superficial angiomyxoma are small, lobulated lesions, composed of stellate or bipolar fibroblasts set in a myxoid stroma, which contains an inflammatory infiltrate with collection of neutrophils. Almost one quarter of cases, contain epithelial structures. Immunohistochemically, tumor cells were negative for desmin and showed a variable expression of S-100 [11, 12].

Aggressive angiomyxoma are large and gelatinous and infiltrate the surrounding tissues. The lesions are composed of spindle or stellate cells scattered in a myxoid stroma. There are numerous, medium-size, thick-walled vessels. Delicate, elongated bundles of smooth muscle cells, often present in the stroma, tend to concentrate around blood vessels. Immunohistochemically, cells stain for vimentin, actin, and desmin, but are negative for S-100 [13–15].

Angiomyofibroblastoma occurs mainly in the vulvar region, but it may also arise in the vagina. It usually presents as a small, well circumscribed, but unencapsulated lesion. Histologically, there is a considerable variation in cell density, with alternating hyper- and hypocellular areas. The cells are cytologically bland and show a spindle, epithelioid or plasmocitoid appearance. Occasional multinucleated cells may be seen. There are numerous blood vessels, usually thin-walled and capillary-like, which may assume an arborizing configuration; other vessels may have thick, muscular walls. Neoplastic cells tend

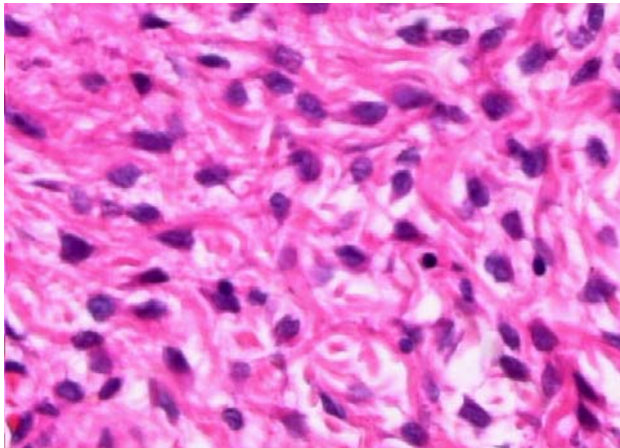
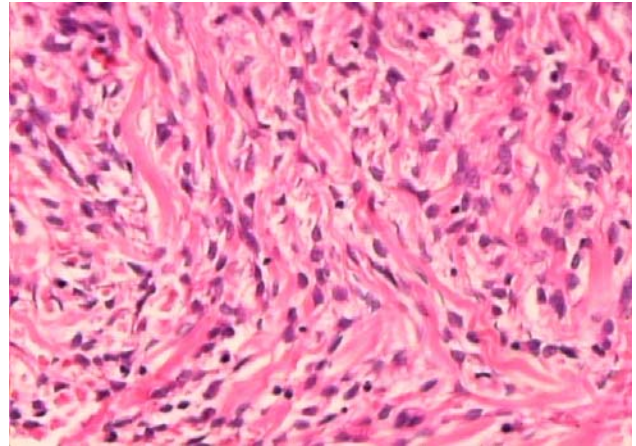
to concentrate around blood vessels. Mitotic activity and necrosis are absent. Immunohistochemical investigations showed reactivity for vimentin and, in most cases, for desmin; some cases showed positivity for HHF35 and/or  $\alpha$ -SMA. Both estrogen and progesterone receptors are diffusely expressed [16–26].

The fibroepithelial stromal polyps are subepithelial, poorly circumscribed lesions, lacking an identifiable lesional margin. The stromal cells may be spindle, stellate or multinucleated, the later being more frequently seen close to the epithelium. Mitotic activity is usually low. Immunohistochemically, the cells are vimentin-positive, and often desmin-positive. The results concerning the reactivity for SMA are conflicting. Some cases show positivity for estrogen and progesterone receptors [27–29]. Polyps that show marked hypercellularity, marked cytologic pleomorphism and high mitotic counts may be misdiagnosed as sarcoma [30]. A worrisome hypercellularity and a pseudosarcomatous pattern may be seen during pregnancy or hormone therapy [31].

Cellular angiomyofibroma are rare lesions, which show a predilection for the vulva. The lesions, which are well circumscribed, consist of bland spindle cells, numerous thin or thick, often hyalinized vessels and mature lipocytes. Phenotypically, the cells express vimentin and are positive for estrogen and progesterone receptors. Desmin and S-100 are negative [32–37].

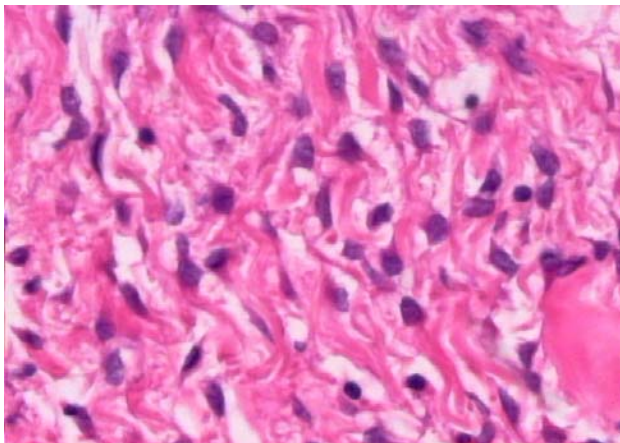
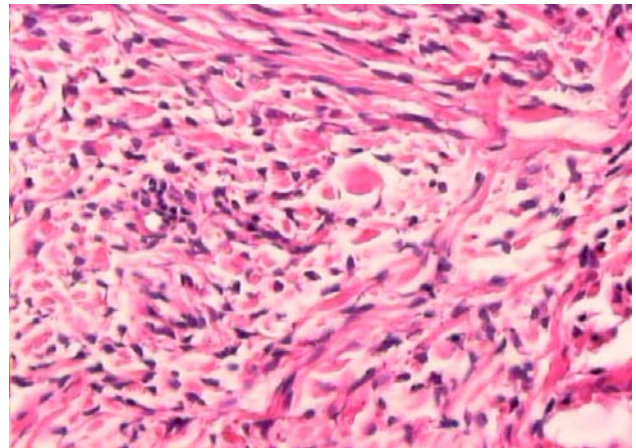
Solitary fibrous tumor is a cellular lesion, which shows a fascicular arrangement of spindle cells, a pronounced stromal collagenisation and a hemangiopericytoma pattern.

**Figure 1 – Vaginal myofibroblastoma.** A dense tumoral area composed mainly of spindle-shaped cells separated by undulated collagen bundles (HE stain,  $\times 50$ )



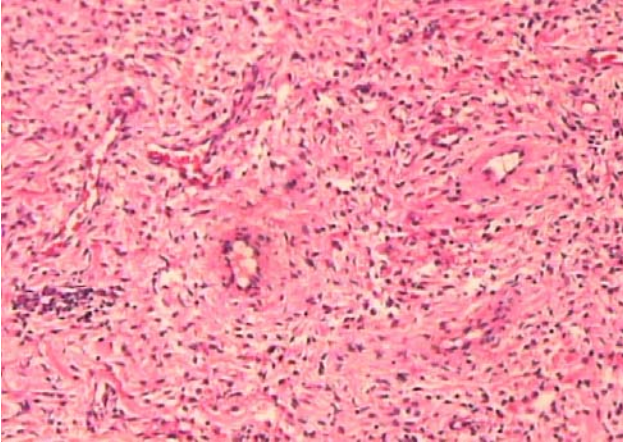
**Figure 2 – Vaginal myofibroblastoma.** The tumor cells had bland nuclei, with delicate chromatin and inconspicuous nucleoli (HE stain,  $\times 100$ )

**Figure 3 – Vaginal myofibroblastoma.** Fascicles of spindled cells with stromal hyaline deposits (HE stain,  $\times 50$ )



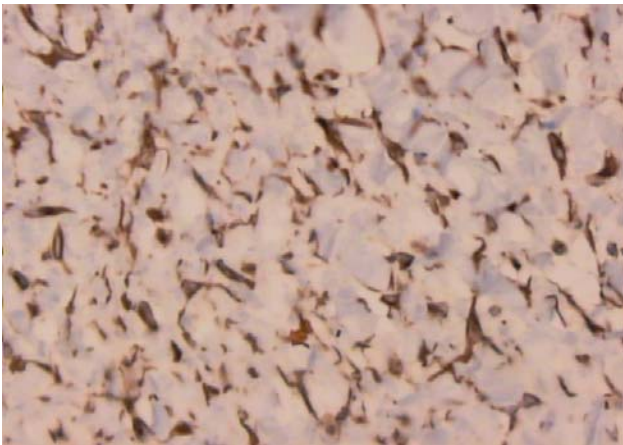
**Figure 4 – Vaginal myofibroblastoma.** Higher magnification of the previous area. One can see the spindle shaped tumor cells, with fibrillary cytoplasm, and a large hyaline mass in the bottom-right corner of the picture (HE stain,  $\times 100$ )





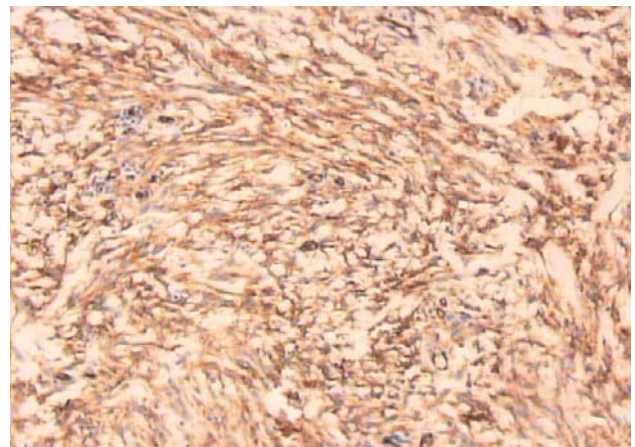
**Figure 5 – Vaginal myofibroblastoma. Numerous blood vessels were observed on the tumoral surface (HE stain, ×25)**

**Figure 6 – Vaginal myofibroblastoma. Tumor cells showed intense immunopositivity for vimentin, ×25**



**Figure 8 – Vaginal myofibroblastoma. Moderate positivity for the CD34 immunostaining, ×25**

**Figure 7 – Vaginal myofibroblastoma. The staining for desmin highlighted the stellate-shape of some tumor cells and the syncytium-like pattern of growth, ×50**



Multinucleated giant cells and areas of necrosis have been found in some cases. Tumor cells show a diffuse vimentin and CD34 staining, but are negative for desmin [38, 39].

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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