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Case report

Low-grade fibromyxoid sarcoma of the vulva presenting as a cystic mass: A case report and review of literature

Ghada Sahraoui [a], Farah Sassi [a,*], Lamia Charfi [a], Montassar Ghallab [b], Karima Mrad [a], Raoudha Doghri [a]

[a] Pathology Department, Salah Azaiez Institute, Tunis, Tunisia
[b] Surgical Oncology Department, Salah Azaiez Institute, Tunis, Tunisia

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ABSTRACT

Introduction: Low-grade fibromyxoid sarcoma (LGFMS) is a tumor with a propensity for late recurrence which is rarely described in the vulva.
Case presentation: A 22-year-old woman presented with a growing right vulvar cystic mass that had been present for 2 months. She underwent surgical wide excision. The final pathologic diagnosis revealed LGFMS of the vulva and a right radical hemivulvectomy with negative margins was performed.
Results: The patient has not experienced a local or metastatic recurrence after 2-years follow- up.
Conclusions: Despite being rare, LGFMS of the vulva should be taken into account when making a diagnosis of vulvar lesions. Definite diagnosis is based on pathological examination. MUC4 positivity is characteristically expressed. To prevent future recurrences, radical excision is necessary.

1. Introduction

Low-grade fibromyxoid sarcoma (LGFMS) is a rare malignant fibroblastic neoplasm [1]. Due to its propensity to mimic other soft tissue neoplasms histologically and the lack of ancillary diagnostic biomarkers, the true incidence of LGFMS is underrated, according to the 5th edition of World Health Organization of Soft Tissue and Bone Tumors [1]. The trunk and proximal extremities are the most often affected areas; they typically present as an isolated mass subfascial in depth [2]. LGFMS arising in the vulva is rare and limited data have been published yet. To date, there have been 9 cases reported in the English literature of LGFMS of the vulva: three individuals case reports [2–4] and a series of seven cases of which six were in the vulva [5]. This report aims to present the tenth case, to the best of our knowledge, of a LGFMS developing from the vulva region and discuss throughout a review of literature its clinical presentation, emphasize the importance of differential diagnosis when confronted with subsequent vulvar neoplasm and prognosis over the long period.

2. Case presentation

A 22-year-old woman, with no past medical history, presented to another hospital with a painless, growing cyst of the vulva that had been present for two months. Physical examination revealed a well-defined cyst of the right labia majora measuring 3 cm. The overlying skin had a crusty surface, hard consistency without pigmentation. No inguinal lymphadenopathy was palpable whereas a physical pelvic examination likewise revealed no concerns. The diagnosis of Bartholin cyst was suggested. Complete blood count was normal. Local excision was performed under local anesthesia. The diagnosis of an undifferentiated carcinoma with positive margins was first retained in the outside hospital. The patient was referred to our institute by her attending gynecologist to seek a second opinion and possibly a broader immunohistochemical study. The histological examination revealed a mesenchymal proliferation of spindle cells occupying the dermis with positive margins. The tumor consisted of cellular myxoid nodules and collagenous hypocellular areas with abrupt transition (Fig. 1). Tumor cells were organized in short fascicles. They had plump spindled to ovoid nuclei with mild hyperchromasia (Fig. 2). Atypical mitotic figures were scarce, <2 per 10 high power fields (Fig. 3A). Arcades of small vessels with perivascular sclerosis were observed focally (Fig. 3B). Tumor necrosis and lymphovascular invasion were absent. Immunohistochemical study showed no staining for AE1/AE3, EMA, hormone receptors, SMA, h-Caldesmone, Desmin, CD-34, S-100 protein and CD-10. Ki-67 was

* Corresponding author.
E-mail address: sassi.farah@outlook.fr (F. Sassi).

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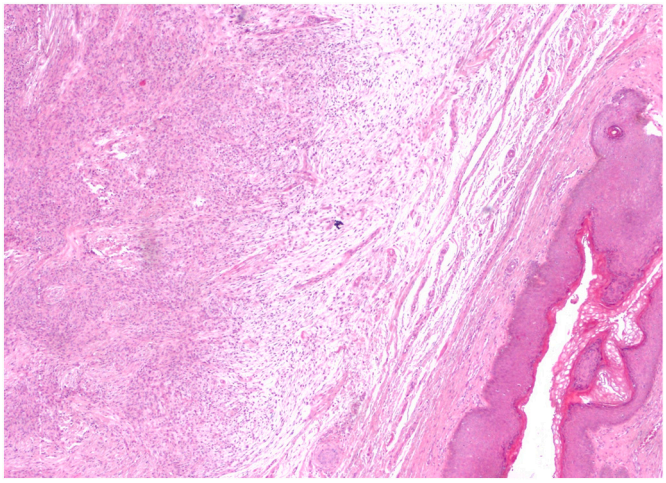

Fig. 1. Low-power appearance of LGFMS (HE×40). The tumor consisted of lobulated cellular myxoid and collagenous hypocellular areas with abrupt transition.

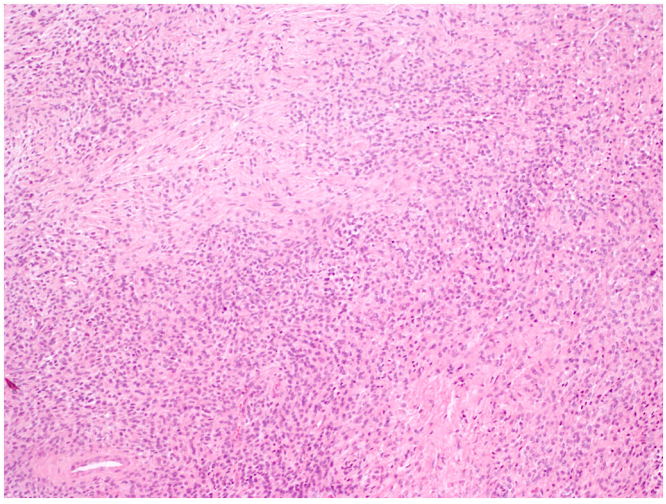

Fig. 2. HE×200. Tumor cells had plump spindled to ovoid nuclei with mild hyperchromasia.

evaluated at 10 %. These patterns suggested a LGFMS. MUC 4 was strongly and diffusely positive in tumor cells, confirming the diagnosis of LGFMS (Fig. 4). Computed Tomography (CT) scan showed no distant metastasis. A right radical hemivulvectomy was carried out. There was no residual tumor, and the margins were negative, according to the pathologic analysis. Additional adjuvant therapy was not deemed necessary to finish the course of treatment after consideration in a meeting with the oncology multidisciplinary team due to the indolent clinical behavior and the complete resection of the tumor. The post-operative course was uneventful, and the patient was discharged from the hospital after 15 days. After 2-years follow-up, the patient remains free of local or metastatic recurrence by physical examination and CT scan of the chest, abdomen, and pelvis.

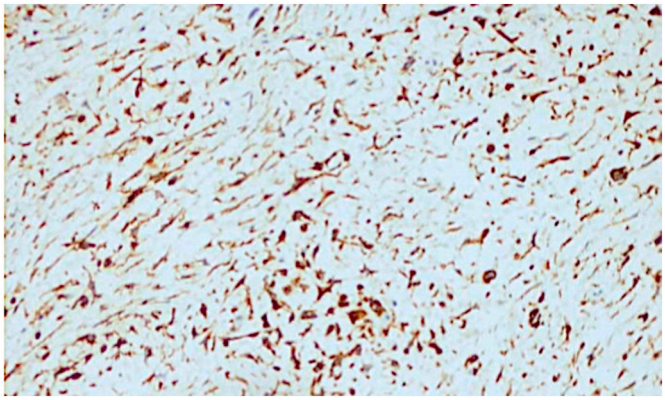

Fig. 4. HE 200. Strong and diffuse cytoplasmic expression of MUC4 in tumor cells.

The SCARE 2020 guideline was used to report clinical progress for this case report [6].

3. Discussion

This case illustrates that vulvar LGFMS mimics the morphologic, immunophenotypic, and molecular characteristics reported in more common regions.

Median age at presentation was 40 years ranging between 22 and 59 years [2–5]. Our patient was the youngest among the reported cases (Table 1).

All histologic forms of vulvar neoplasia share comparable symptoms. Most patients have a solitary, fleshy, nodular, or warty vulvar plaque, ulcer, or mass on the labia majora; the labia minora, perineum, clitoris, and mons are less frequently affected [3]. VanSandt et al. [2] described the case of a patient diagnosed with a LGFMS of the vulva who initially presented with a vulvar mass presumed to be a Bartholin gland cyst. In our case, the patient presented with a 3-cm cystic mass in the right labia majora.

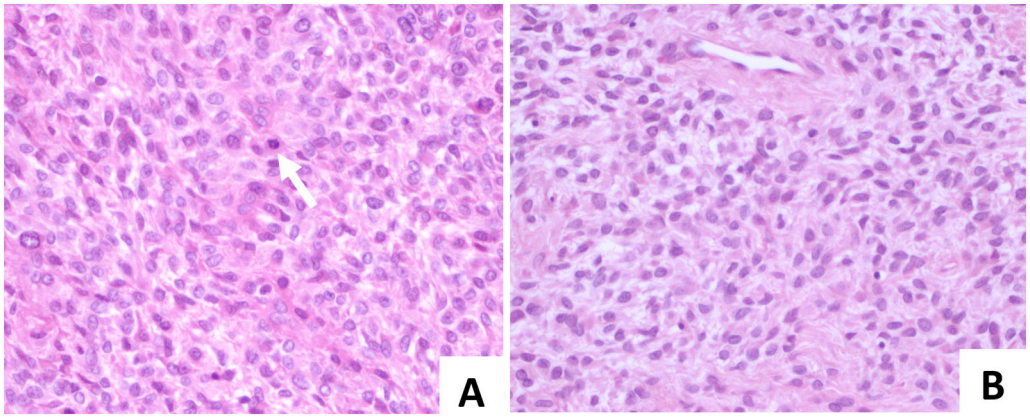

Fig. 3. A: HE×400. Atypical mitotic figures (Arrowhead) B: HE×400. Arcades of small vessels with perivascular sclerosis were observed focally.

Table 1
Reported cases of low-grade fibromyxoid sarcoma of the vulva.

Year	Authors	Number of cases	Age (years)	Site	Tumor size (cm)	Treatment options	Adjuvant therapy	Follow-up (months)	Outcome
2012	Barnhill et al. [4]	1	45	Left VM	5	Surgical excision + Left radical hemivulvectomy + ipsilateral inguinal lymph-node resections	No	12	Disease-free
2013	Vansandt et al. [2]	1	36	Right VM	6	Surgical excision completed by right hemivulvectomy	No	24	Disease-free
2018	Cenjik et al. [3]	1	45	Left VM	2	Left radical hemivulvectomy	No	24	Disease-free
2022	Costigan et al. [5]	6	59, 37, 40, 46, 39,34	3 Right 2 Left 1 NM VM	2,5, 8.7, 1 0.2 (3 NM)	5 local excision of which 3 were reexcised (1 not known)	No	141- 16- 57 and 10 (2 not known)	Disease-free
2022	Current case	1	22	Right VM	3	Radical hemivulvectomy	No	24	Disease-free

V.M: vulvar mass; NM: not mentioned.

Vulvar tumors that are slow-growing and painless are diagnosed with both benign and malignant diseases. Common benign etiologies include lipomas, Canal of Nuck cysts (peritoneal cysts), Bartholin gland cysts and abscesses [2]. However, malignancies comprise squamous cell carcinoma, melanoma, and Bartholin gland adenocarcinoma [2]. Its occurrence in the lower female genital tract is rare, and thus a wide range of differential diagnoses must be considered, such as deep angiomyxoma, superficial angiomyxoma, perineurioma, myxoid smooth muscle neoplasia, cellular angiofibroma, and myxoid dermatofibrosarcoma protuberans [5,7]. It is crucial to keep a clinical awareness for the less common etiology given this wide range of differential diagnoses, each with a unique clinical therapy and prognosis.

On macroscopic examination, LGFMS's tumor size varies between 1.2 and 8.7 cm [5]. They were well-circumscribed, white to yellow [5], yellow to pink [2], gray pasty [3], predominantly firm to focally gelatinous cut surfaces [5]. One tumor showed central hemorrhage, but none showed necrosis [5]. In our case, the surface was crusty with hard consistency.

Microscopically, Costigan et al. [5] noticed often a mild perivascular lymphocytic infiltrate with no appreciable inflammation in the remaining. In fibrous areas, tumor vasculature predominantly comprised small arterioles, whereas myxoid foci contained curvilinear capillaries, which formed characteristic “arcades”. Perivascular hyalinization was prominent in one tumor. Other rare findings included myoid bundles noted away from the tumor-stromal interface and foci of extravasated red blood cells each noted in one case. Mitoses were rare in the data already published and no tumor necrosis was noticed (Table 2).

It is important to note that MUC4 immunohistochemistry is a useful auxiliary tool because it is only positive in LGFMS and a small subset of monophasic synovial sarcomas [8]. However, to date, there is currently no information on MUC4 staining in superficial angiomyxomas, cellular angiomyxoma, and angiomyofibroblastomas [5]. EMA was expressed in tumor cells in 4 cases out of 6 in a case series [5]. In difficult cases, molecular testing highlighting FUS or rarely EWSR115 fusion is the diagnostic key of LGFMS [9]. Costigan et al. [5] found a FUS rearrangement in 5 out of 7 cases.

In the current case, the patient was first diagnosed with undifferentiated carcinoma but then with the uncertainty of the proposed diagnosis, a second opinion was sought and the diagnosis of LGFMS was retained with MUC4 expressed in tumor cells. Therefore, when the clinical presentation and predicted sample results are discordant, the differential diagnosis should be broadened.

Similar to the cases described in the studies by VanSandt et al. [2] and Barnhill et al. [4], the current patient underwent a radical hemivulvectomy to maximize the extent of the massive tumor's excision. While Barnhill et al. [4] did an inguinal lymphadenectomy to check for any indications of metastatic dissemination, inguinal lymph node dissection was not done in the current patient nor in the other reports [2,3,5]. It is unclear whether adjuvant therapy may benefit patients with recently discovered LGFMS. No adjuvant treatment was given in the current case nor the other researches [2–5].

Patients with local recurrences typically have an excellent prognosis after reexcision of the new tumors [10]. The prognosis of the current patient after complete surgical resection of the lesion and a radical hemivulvectomy with negative margins seems good without developing local or distant recurrence after 2-year follow-up.

4. Conclusion

As one of the less frequent histologic subtypes of sarcomas developing in the lower genital female tract, LGFMS can be misdiagnosed due to its benign appearance. Radical excision after pathological confirmation of the diagnosis is mandatory, as well as long-term follow-up of the patient, since the disease tends to develop late metastases years after the initial diagnosis.

Ethical approval

No.

Research registration number

No.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the

Table 2
Summary of pathology features for low-grade fibromyxoid sarcoma's diagnosis in the vulva.

Authors	Mitoses	Necrosis	Capillary “arcades”	EMA	MUC4	S100	SMA	FUS rearrangement
Vansandt et al. [2]	Present	Absent	Present	NM	NM	NM	NM	Present
Cenjik et al. [3]	4/10 HPF	Absent	NM	NM	NM	–	+	Not done
Costigan et al. [5]	<1/10 HPF	Absent	3/7	+4/6 cases	+5/6 cases	NM	NM	+ 5/7 cases
Current case	Atypical mitoses focally	Absent	Present	–	+	–	–	Not done

NM: not mentioned.

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Credit authorship contribution statement

All the authors read and approved the final version of the manuscript.

Ghada Sahraoui (MD): conception, acquisition of clinical data, and revising the manuscript.

Farah Sassi (MD): conception, acquisition of data, literature research and preparing the manuscript.

Lamia Charfi (MD): revising the manuscript.

Montassar Ghallab (MD): Supervision and acquisition of clinical data.

Karima Mrad (MD): revising the manuscript critically.

Raoudha Doghri (MD): manuscript editing and revising the manuscript critically.

Declaration of competing interest

The authors report no declarations of interest.

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