An apparently benign vulvar mass: possibly a rare malignancy

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Summary

Background: Vulvar dermatofibrosarcoma is a rare fibrous tumor of intermediate grade malignancy, with a tendency for local recurrence, and rarely metastasizes. Management should be multidisciplinary. This is a report of an apparently benign vulvar mass with delayed diagnosis of vulvar dermatofibrosarcoma. *Case report:* A 42-year-old woman was referred to our hospital because of a vulvar tumor lasting 16 years, although several gynecological procedures and a total laparoscopic hysterectomy had been performed two years before. During this long period the lesion did not change morphological features and remained asymptomatic. Only a benign vulvar mass was diagnosed. Then, the swelling became evident showing erythematous skin with an aspect of "peau d'orange", leading the patient to consult a specialist. A firm vulvar swelling was observed in the anterior third of right labia majora continuing with about 3 cm of cord on top, quite movable above the underlying tissue but not on the overlying tissue. A wide excision was performed. The pathological examination showed positive margins. One month later an extensive deeper excision was performed. Histology confirmed a diagnosis of dermatofibrosarcoma. Immunohistochemistry was strongly positive for CD34. *Conclusion:* Vulvar lesions always require complete pathologic examination even in case of features of benign tumor to exclude a dermatofibrosarcoma. The role of the pathologist is essential to ensure negative microscopic margins and to avoid local recurrence.

Key words: Vulvar neoplasms; Dermatofibrosarcoma; Vulvar mass.

Introduction

Dermatofibrosarcoma protuberans (DFSP) is a rare cutaneous soft tissue sarcoma, accounting for approximately < 1% of all these tumours with an incidence of 0.8-5 cases per million per year, occurring most commonly during the fourth and fifth decades of life [1]. DFSP is considered locally aggressive with a high rate of local recurrence but its metastatic potential is low, with rare local and distant metastasis, usually via hematogenous spread to the lungs. Of all DFSP 85-90% are low-grade lesions; the others contain a high-grade fibrosarcomatous component and are considered to be intermediate-grade lesions [2].

DFSP exhibits an indolent growth pattern and in many cases symptoms are long-lasting [2]. The lesion mainly affects the trunk and lower extremity but it has also been found in the upper extremity, head and neck, and groin area. Some lesions develop in sites of previous trauma, particularly scars. The vulva is an uncommon site of involvement, and only approximately 28 cases are reported in the literature [3]. We report a case of a vulvar DFSP to discuss clinical and histopathological features, and the appropriate management.

Case Report

A 42-year-old woman, gravida 2, caesarean section 2, was referred to our hospital because of a vulvar tumor lasting 16 years. Total laparoscopic hysterectomy was performed two years before because of myomatosis.

The anamnesis revealed a firm vulvar swelling in the anterior third of the right labia majora continuing with about 3 cm cord on top (Figure 1). The mass was quite movable above the underlying tissue but not on the overlying tissue. It had been quiescent until six months before, when the swelling became evident with skin erythema. Ultrasound scan showed a complex capsulated mass of 21 x 9 mm continuing with hyperechogenic cord-like echos. A wide excision was performed, and the pathological examination showed positive margins. One month later an extensive deeper excision was performed.

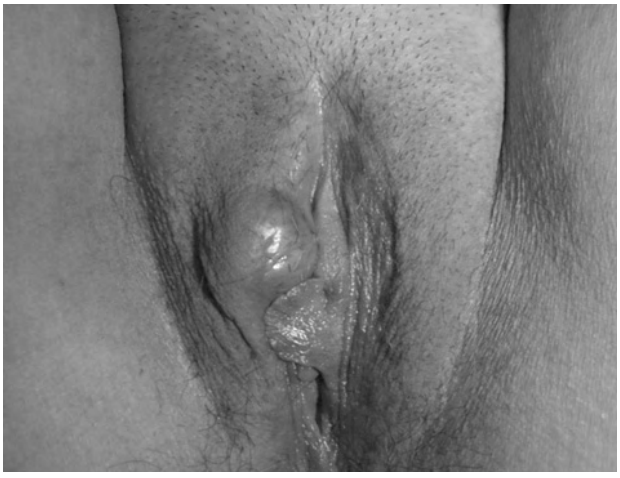

Figure 1. — A firm, erithematous, subcutaneous nodule over the anterior third of the right labia majora.

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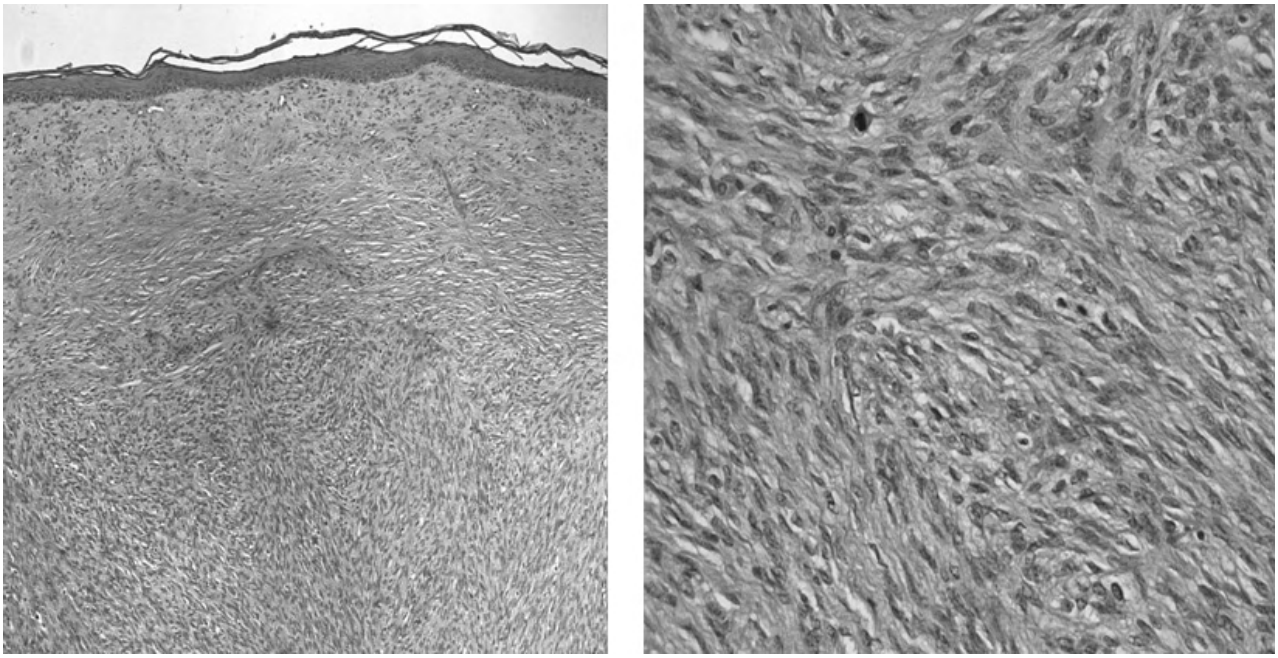

Figure 2. — A dense uniform proliferation of monomorphous spindle-cells separated from the overlying epidermis (H&E 10x).

Grossly, the tumour appeared as a gray-white nodule with focal erosion of the overlying skin. Lesion size was approximately 4 cm and it was not pigmented. The tissue sample was fixed in formalin, then routinely processed and embedded in paraffin. The sections were stained with haematoxylin-eosin. Additional sections were cut and subjected to immunohistochemical studies using antibodies to the following antigens: CD34; CD31, S100; desmin, actin, CD68, vimentin, HMB45, Mart-1, Pan-CK, Ki67. Histological examination showed dense, uniform and monomorphous spindle-shaped tumour cells (Figure 2) arranged in a storiform pattern with little pleomorphism (Figure 3). It was poorly circumscripted with diffuse and intricate infiltration of both the dermis and the subcutaneous adipose tissue in a typical honeycomb pattern. Honeycomb was characterised by tumour cells extending between and surrounding groups of preexisting fat cells, which remained viable, a pathognomonic feature of DFSP. The overlying epidermis was thinned, atrophic with focal erosion. Nerve and muscle invasion was not observed, nor was necrosis or angiolymphatic invasion present. Focal infiltration of lymphocytes and plasma cells involved the periphery of the tumour. No areas of fibrosarcomatous differentiation were found. Mitotic activity was moderate (< 5/10 high-power fields). Immunohistochemically DFSP cells were strongly positive for CD34 (Figure 3) and negative for S100, desmin. In view of these morphological and immunohistochemical findings, the diagnosis of DFSP of the vulva was made.

Discussion and Conclusion

Vulvar DFSP is rare: a total of 28 cases have been reported in the literature as reviewed by Ghorbani and colleagues [3]. This is the first report of vulvar DFSP with delayed diagnosis, although the lesion was discovered 16 years before and the patient had undergone several gynaecological procedures. During this long period the lesion did not change morphological features and remained asymptomatic. A benign vulvar mass was diagnosed, therefore allowing the progression of the lesion. Then, the swelling became evident showing erithematous skin with an aspect as "peau d'orange", leading the patient to consult the specialist.

Clinical presentation of vulvar DFSP is characterised by ulceration, pain, bleeding and the appearance of a nodule, but most patients are asymptomatic for a long period as in our report. Indeed, due to its indolent nature, the tumour is often undetected in the early stages and usually misdiagnosed at presentation [4]. Therefore, in cases of vulvar lesions it is mandatory for a complete examination even if the feature is like a benign tumour because DFSP can be excluded only with a pathological examination.

The treatment of choice is surgical excision with wide margins of 3-5 mm up to 5 cm of normal skin [5]. High recurrence rates despite "negative" margins can occur because large portions of the true margins are not evaluated on standard histological specimens and because it is difficult to identify "finger-like" projections, responsible for tumor recurrence. Mohs' micrographic surgery has been advocated to ensure precise margin control with microscopic examinations of deep and lateral margins [6]. This procedure is associated with reduced recurrences [7] and offers preservation of normal tissue in the vulva, which is crucial for cosmetic and functional reasons. Mohs' micrographic surgery was not performed in our patient because the first histological specimen showed "positive" margins only in one specific direction, at the supero-lateral right margin deep in the pubic adipose tissue.

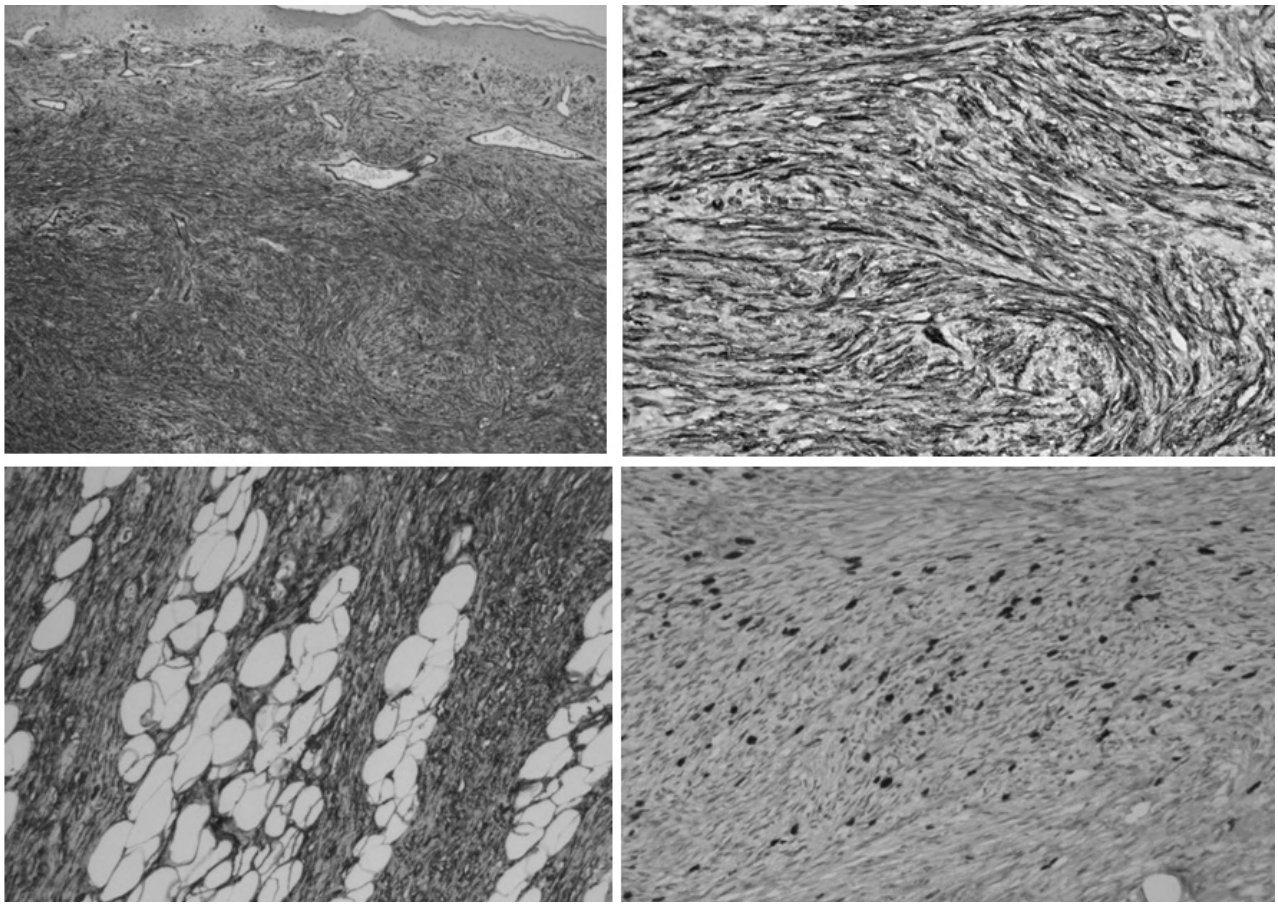

Figure 3. — Top-left. Strong, diffuse CD 34 immunoreactivity in vulvar DFSP.
Top-right. CD 34. Tumour cells arranged in a storiform pattern.
Bottom left. Infiltration of subcutaneous tissue in a characteristic honeycomb pattern.
Bottom right. Ki 67. Moderate mitotic activity.
Bottom. High cellularity and little pleomorphism of spindle-cells (H&E 40x).

The presence of a spindle cell proliferation within the skin allows a differential diagnosis with invasive spindle cell squamous cell carcinoma, spindle cell melanoma, neural tumours, smooth muscle tumours (leiomyosarcoma) and angiosarcoma [8]. Strong positivity for CD34 staining in this histologic context is diagnostic of DFSP, while the lack of staining with cytokeratins argues against a carcinoma [9, 10]. Likewise, negativity for S100 and HMB45 argues against neural and melanocytic origin, respectively. The lack of staining with smooth muscle actin and desmin excludes leiomyosarcoma and the lack of staining with CD31 in this context excludes angiosarcoma [8]. The overlap of histological features makes the differential diagnosis difficult between DFSP and dermal fibrous histiocytoma [10]: foam cells, giant cells and inflammatory cells were better represented in fibrous histiocytomas, as well as more ovoid cells and increased mitotic activity. The typical infiltration of fat is usually observed in DFSP. Most fibrous histiocytomas are only focally positive for CD34 and histiocytes express a stronger immunoreactivity for CD163 than DFSP [11]. Areas of fibrosarcomatous differentiation have been associated with unfavourable cause, higher tendency to recurrence and increased risk of metastasis [12].

Because DFSP tends to spread in microscopic projections away from the visible lesion, very wide local excision is required for tumour control. Adjuvant radiotherapy administered either before or after surgery, significantly reduces the risk of local recurrence in patients who have close or positive surgical margins [13].

In conclusion, vulvar DFSP is a subtle pathology with asymptomatic clinical presentation. Any vulvar lesion always requires a complete pathologic examination even in case of features of benign tumour to exclude a DFSP. The role of the pathologist is essential to ensure negative microscopic margins and to avoid local recurrence.

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