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Vulvar Dermatofibrosarcoma Protuberans in a 42-Year-Old Female: A Case Report and Review of Recent Literature

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1. Abstract

Dermatofibrosarcoma Protuberans (DFSP) is a rare, slow-growing skin sarcoma with a high rate of local recurrences. It most commonly occurs on the trunk and limbs, whereas cases of vulvar DFSP are extremely rare. We present the case of a 42-year-old woman with grade I obesity, in whom an asymptomatic 7 mm nodule was found on the left mons pubis. The initial excision revealed a spindle cell mesenchymal tumour with neoplastic tissue at the deep margin. As a result, the patient was referred to a specialised oncology centre for radicalisation of the lesion with a 5-centimetre margin. The final histopathological examination confirmed the absence of residual neoplastic tissue. This case highlights the importance of early diagnosis and the implementation of appropriate treatment, which in this case was surgical removal of the lesion with a wide margin, to improve treatment outcomes and reduce the risk of local recurrence in vulvar DFSP.

2. Introduction

Dermatofibrosarcoma protuberans (DFSP) is a rare mesenchymal skin sarcoma of low malignant potential, characterised by slow growth, a low propensity for metastasis, and a high rate of local recurrence (20% to 50%) [1]. The tumour typically arises in the dermis and infiltrates the subcutaneous tissue, most commonly presenting as an asymptomatic, indurated plaque; in more advanced stages, it may lead to the development of nodules and ulcerations [2].

The estimated incidence is 3 to 5 cases per million people, with a similar distribution between males and females [3]. DFSP most commonly occurs on the trunk and extremities, whereas vulvar involvement is exceptionally rare, with fewer than 100 cases reported in the English-language literature to date [2]. In a review of vulvar DFSP cases, the most common site was the labia majora (approximately 52% of cases), followed by the mons pubis (approximately 11%) [4].

Due to the disease's slow progression and its similarity to benign lesions or other malignancies, definitive diagnosis requires a core needle or excisional biopsy, followed by histopathological and Immunohistochemical (IHC) evaluation [4,5]. The primary treatment for DFSP is surgical resection with negative margins, most commonly achieved by Mohs Micrographic Surgery (MMS) or Wide Local Excision (WLE) [6,7]. In cases with positive margins or a high risk of recurrence, postoperative radiotherapy is recommended [8]. For patients with unresectable disease or multiple recurrences, targeted therapy with tyrosine kinase inhibitors, such as imatinib, may be considered [9]. Despite the high rate of local recurrences, the overall prognosis for patients with DFSP is

excellent [8].

In this case report, we present a rare case of vulvar DFSP.

3. Case presentation

A 42-year-old woman with class **I** obesity (BMI 30.42 kg/m^2) presented to the gynaecological clinic on April 22, 2024, due to a nodular lesion on the left side of the pubic mound that had persisted for about two years. The lesion was round, firm, and mobile, measuring approximately 7 mm, and had the characteristics of a fibroma/sebaceous cyst. The patient reported no accompanying symptoms, including pain. In the collected history, the patient reported two natural births. The last menstrual period was April 13, 2022. In March 2024, cervical cytology was performed, yielding a normal result. The patient denied drug

allergies, chronic diseases, previous hospitalizations, and surgical procedures. She was chronically taking venlafaxine. There was no family history of neoplastic diseases. The patient denied smoking tobacco, consuming alcohol, or using other stimulants. The patient's general condition at the time of admission was good. Lymph nodes were non-palpable. Vital signs upon admission were also normal. The physical examination revealed no significant abnormalities.

In laboratory tests performed before hospitalisation, no significant deviations were found: RBC $4.9 \times 10^6/\mu L$, WBC $4.36 \times 10^3/\mu L$, APTT 31.2 s, prothrombin time 11 s, sodium 139 mmol/L, potassium 4.35 mmol/L, creatinine 0.83 mg/dL, glucose 99 mg/dL, and TSH 1.88 µIU/mL.

The patient was referred to the gynaecological ward for removal of the lesion. Under aseptic conditions and under local anaesthesia, an elliptical excision of the vulvar lesion was performed. Postoperatively, hyaluronic acid cream was applied three times daily.

The histopathology result was obtained on June 3, 2024. The examination revealed a spindle-cell mesenchymal tumour, most likely Dermatofibrosarcoma Protuberans (DFSP). In the immunohistochemical study there was, negative expression of SOX, HMB 45, and desmin, with positive expression of vimentin. The Ki-67 proliferation index was 20%. The lateral margin was 4 mm, while at the base of the specimen, the lesion was along the line of cut, suggesting a failure of the procedure.

After obtaining the histopathology result, the patient resigned from further treatment at the original centre and reported to the National Institute of Oncology, where a radicalisation of the post-operative scar was performed with a wide tissue margin (5 cm). After resection, no residual neoplastic tissue was found.

Currently, the patient remains under oncological supervision with follow-up examinations every 6 months. No features of local recurrence were detected. The patient was considered cured. A schematic overview of the patient's clinical course and treatment is summarized in Figure 1.

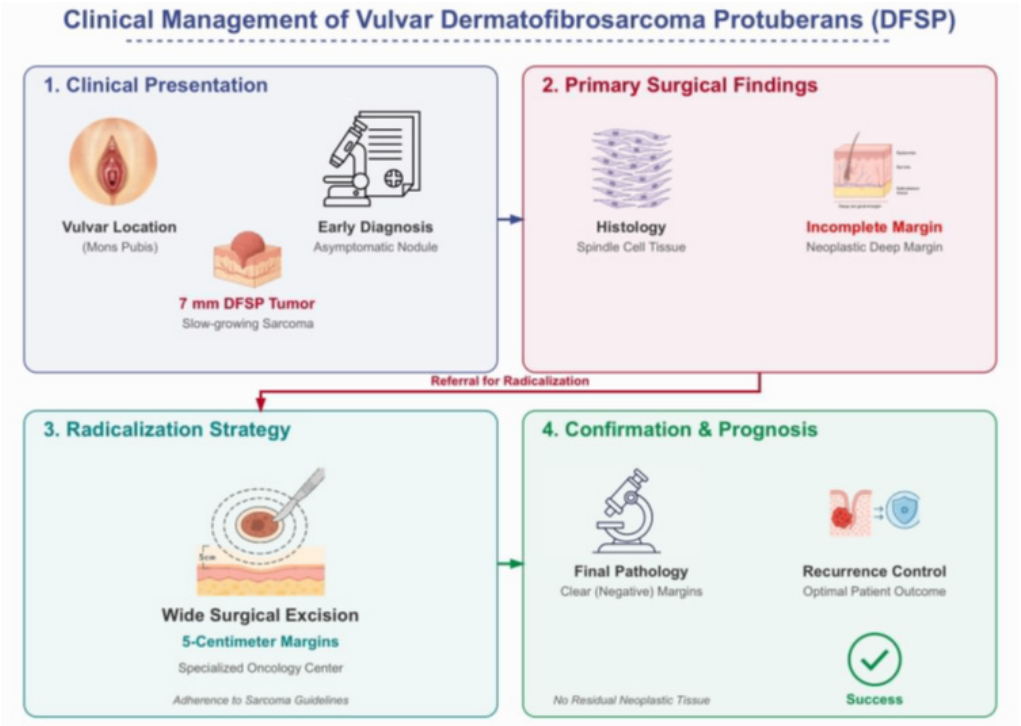

Figure 1: Clinical management diagram of the presented vulvar DFSP case, detailing the diagnostic path, the primary incomplete surgical margins, the following wide local excision strategy, and the definitive disease-free outcome.

4. Discussion

DFSP of the vulva is extremely rare. The profile of our 42-year-old patient is consistent with demographic trends, in which the median age of onset is approximately 46 years. The most frequent site of vulvar DFSP manifestation is the labia majora (52.2%), followed by the mons pubis (11.6%), which was the location of the lesion in our patient. The mean tumour size reported in the literature is 5.32 cm; in the described patient, the result is significantly lower, with the lesion's diameter being only 7 mm. Such early diagnosis at a small size is rarely encountered and represents a significant favourable prognostic factor.

Vulvar DFSP is exceptionally rare. In this study, we compared our patient's case with literature reports from 2000 to 2026, summarised in Table 1. The clinical profile of this case is highly consistent with the characteristics documented in the literature.

Our patient is a 42-years-old, which is close to the typical age range for this malignancy. Although her age is slightly lower than the median age of onset (47 years) observed in our analysis, it remains consistent with overall demographic trends. Our data shows that the most frequent site for vulvar DFSP is the labia majora (78,6%), followed by the mons pubis (28,6%). The last

one was the location of the tumour in our patient. A unique feature of this case is the tumour size, which was only 7 mm in its largest dimension, which is much smaller than the mean size of 5.32 cm reported in the literature. For this comparison, we used the largest reported diameter of each tumour. Diagnosing DFSP at such an early stage and small size is rarely encountered and serves as a significant favourable prognostic factor.

Literature review Table 1 indicates that CD34 (Cluster of Differentiation 34) expression is the defining feature of vulvar DFSP and is present in the vast majority of cases reported in the literature. Although CD34 assessment was not performed in our patient, positive vimentin and negative desmin expression supported the diagnosis of DFSP. This immunohistochemical profile is consistent with several reports presented in our literature review [2,3,10–12]. This suggests that these antigens may also serve as supportive markers for DFSP, and, combined with the anatomical location, clinical presentation, and histopathological findings, allows for a definitive diagnosis. Furthermore, it is worth noting that a complete Immunohistochemical (IHC) panel is not always strictly mandatory for an accurate diagnosis, as evidenced by other reported cases in which IHC data were either limited or not specified [4,13].

The clinical presentation in our patient was typical of most reported cases, with the lesion being asymptomatic. However, in some cases, symptoms such as bleeding, pain, or itching may occur [14–17]. Complete surgical excision with clear margins remains the mainstay of treatment for vulvar DFSP. In our patient's case, the primary procedure revealed neoplastic tissue at the base of the specimen (positive deep margin), necessitating further intervention. In the literature, the local recurrence rate for positive margins is 18,5%. These data fully justify the decision to refer the patient to a referral centre for radicalisation of the surgical scar, which allowed for complete local control and a favourable oncological prognosis.

Table 1: Summary of clinicopathological characteristics of vulvar DFSP cases reported in the literature (2000–2026). Continued.

Author, Year	Age	Location	Tumor size (cm)	IHC panel	Margin status	Symptoms	Treatment	Outcome
Alsaleh, 2025 [1]	47	Left labia majora to gluteal fold	6.8	N/R	Negative	None	Wide local excision, radical vulvectomy, left inguinofemoral lymphadenectomy	Complete resolution
Arab, 2023 [2]	53	Right labia majora, clitoris	10 x 7 x 5	CD34(+), Ki67(+)	Negative (after re-excision with 2-3 cm margins)	None; later: pain, rapid growth (recurrence)	Radical vulvectomy, bilateral inguinofemoral lymphadenectomy	Disease-free (18 months)
Bernárdez, 2015 [3]	39	Left labia majora, perianal area	12 x 5	CD34(+), Factor XIIIa(-), SMA(+), Desmin(-)	Positive (affected)	Pain, malaise	Radical excision, reconstruction, adjuvant radiotherapy	Disease-free (12 months)
Bogani, 2015 [4]	50	Left labia majora	2 x 1	CD34(+), Ki67(4%)	Negative (after re-excision)	Itching	Excisional biopsy, re-excision (4 cm margins)	Disease-free (24 months)
Detrés, 2026 [5]	55	Left labia majora	5.1 x 5	CD34(+)	Negative (0.3 cm deep margin)	None, later burning	Radical hemivulvectomy, V-Y advancement flap	Disease-free
Doufekas, 2009 [6]	39	Left labia majora	1.0-1.5	N/R	Negative	None	Debulking, MMS	Disease-free (3 years)
Goyal, 2021 [7]	35	Right labia majora, mons pubis	6 x 6 (main), 1.5 x 1	CD34(+)	Negative (3 cm)	Discomfort	Radical hemivulvectomy, resection of deep nodule	Disease-free (24 months)
Hammonds, 2010 [8]	59	Right labia majora	4 x 2	N/R	Negative	None	MMS, Burow's triangle excision	Disease-free (2.5 years)
Jeremic, 2019 [9]	55	Mons pubis, clitoris, labia majora	18 x 10 x 8	CD34(+), Desmin/Actin/SMA /CD117/S100(-), Ki-67 (45%), p53 (60%)	Negative	Ulceration	Radical excision (3 cm margins), flap reconstruction	Lung/chest wall metastasis; death
Jozwik, 2024 [10]	92	Mons pubis, left labium	10 x 9	Low Ki67/MIB1	Negative	None	WLE (3 cm margins), abdominal rhomboid flap reconstruction	Disease-free (14 mo)

Kholová, 2001 [11]	31	Left labia majora	2	CD34(+), Factor VIII(-), CD31(-), Ki-67 (±)/ MIB-1(+)	Negative	N/R	Radical excision, local radiotherapy	Disease-free after treatment of the seventh recurrence
Mahajan, 2025 [12]	47	Mons pubis, right labia majora	12 x 7	CD34(+), Desmin(+), S100(+), Bcl-2(+), SMA(+), ER(+), PR(+)	Negative	Difficulty in walking	WLE (2cm margins)	Disease-free after WLE with normal genital function
Mancari, 2024 [13]	64	Labia majora	4	CD34(+)	Positive	None	WLE	Repeat WLE with no subsequent local recurrence
Moodley, 2000 [14]	39	Left labia majora	8 x 12	CD34(+)	Negative	Underlying swelling	Reexcision with a 3-cm margin	No recurrence at 3-month follow-up
Neff, 2019 [15]	57	Mons pubis, left labia majora	20 x 15	CD34(+), S100 (-), SOX10 (-), GFAP(-)	Negative	Bleeding	Radical vulvectomy with complex wound closure using VRAM, one-year adjuvant treatment with imatinib 400mg	No evidence of recurrence for over twelve months
Vanni, 2000 [16]	39	Vulva, with extension to the perineal raphe	6	CD34(+), SMA(+), HHF-35 (+), Desmin (-), S100 (-)	Negative	None	Local excision with wide margins	No evidence of recurrence
Vathiotis, 2018 [17]	72	Right labia majora	N/R	CD34(+), vimentin(+), CD10(+), Ki67(30%)	Primary margins: positive; post-WLE margins: negative	Chest pain on right hemithorax, productive cough (2 months), diffuse expiratory rhonchi	Primary vulvar tumor excision; recurrence at 18 months treated with WLE; radical vulvectomy performed after second local recurrence	Lung metastases of FS-DFSP
Vasudevan, 2013 [18]	62	Vulva, extending medially to labia minora	6 x 9	CD34(+), S100(-), SMA(-)	Positive	Swelling	Local excision	N/R
Wiszniewska, 2016 [19]	44	Right labia	5 x 3.5 x 3	CD34(+), S100/SMA/Desmin /WT1/ER/PR (-)	Negative	Burning pain	Excision, re-excision	Disease-free (18 months)
Xie, 2025 [20]	27-73 (±44,3)	Mons pubis, labia majora	2–6,5	CD34(+), S100(-), Desmin(-), SMA(-)	Negative: 5; Positive: 1; N/A:1	None	WLE in every case	No evidence of disease at 12-54 months; 1 local recurrence at 28 months
Zemni, 2019 [21]	47	Left labia majora	6	CD34(+), Ki67(+)	Negative	Pain	Radical excision (5-cm margins)	No evidence of recurrence
Sermpetzoglou, 2012 [22]	66	Mons pubis	Three nodules: 1; 2; 2,5	CD34(+), Vimentin (+), CD117(-), S100P(-), ER(-), PR(-)	N/R	Erythematous induration (8 x 5 cm)	Vertical excision with the reconstrction	No evidence of recurrence

Table legend: ER: Estrogen receptor; FS-DFSP: Dermatofibrosarcoma protuberans with fibrosarcomatous transformation; IHC: Immunohistochemical; MMS: Mohs Micrographic Surgery; N/A: Not Available; N/R: Not Reported; PR: Progesterone receptor; WLE: Wide Local Excision.

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