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Case Report

Vulvar dermatofibrosarcoma protuberans: a case series

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Abstract: Dermatofibrosarcoma protuberans (DFSP) is a rare, superficial low - to intermediate-grade sarcoma that typically presents as an asymptomatic slow-growing indurated dermal plaque. Although DFSP has a relatively high local recurrence rate, distant metastasis is rare. DFSP primarily occurs on the trunk and extremities but is seldom reported in the vulva. Due to its rarity, the clinical characteristics, pathological diagnosis, prognosis, and optimal management of vulvar DFSP remain poorly characterized. We retrospectively analyzed vulvar DFSP cases from our institution (January 2010 - January 2023). Clinical data and follow-up were obtained from hospital records, and imaging studies were reviewed via the picture archiving and communication system. Seven patients were included, with a median symptom-onset age of 44.3 years (range, 27-73). Patients typically presented with firm, asymptomatic masses. The labia majora was most commonly affected (n = 6, 85.7%). Tumor size averaged 4.3 cm (range, 2.0-6.5). All patients underwent excisional biopsy followed by wide local excision; none received lymphadenectomy. Surgical margins were documented in six patients, with negative margins achieved in five after initial wide excision. Over a mean follow-up of 27.3 months (range, 12-54), one patient (14.3%) experienced local recurrence. No recurrence occurred in patients with negative margins. Vulvar DFSP predominantly affects young and middle-aged women, manifesting as vulvar masses of variable size. Wide local excision is the primary treatment. While vulvar DFSP has a propensity for local recurrence, widely negative margins appear protective. Long-term follow-up is recommended to monitor for recurrence.

Keywords: Dermatofibrosarcoma protuberans, vulva, wide local excision, Mohs micrographic surgery, case series

Introduction

Dermatofibrosarcoma protuberans (DFSP) is a rare, superficial low - to intermediate-grade sarcoma that typically originates in the dermis and infiltrates subcutaneous tissue [1, 2]. Histologically, it is characterized by spindle cells arranged in a storiform pattern with diffuse, strong CD34 immunopositivity. DFSP primarily occurs on the trunk and extremities, affecting young and middle-aged adults; it typically presents as an asymptomatic, slow-growing indurated dermal plaque [3-5]. Ulceration and nodule formation may develop in advanced stages. Although DFSP exhibits a relatively high local recurrence rate, distant metastasis is rare. Vulvar involvement by DFSP is exceptionally uncommon, with fewer than 100 cases reported in the English literature. Due to the rarity of this presentation, the clinical characteristics, pathological diagnosis, prognostic factors, and optimal management strategy for vulvar DFSP remain poorly characterized.

To contribute to the existing literature, we conducted a retrospective review of 7 vulvar DFSP cases diagnosed and treated at our institution between January 2010 and January 2023. This study aims to characterize the clinical and pathological features, treatment approaches, and long-term outcomes of vulvar DFSP.

Materials and methods

All vulvar DFSP cases were identified through the Anatomical Pathology Department archives at West China Second University Hospital, Sichuan University. We retrospectively analyzed

cases diagnosed at our institution between January 2010 and January 2023. Clinical characteristics and follow-up data were extracted from hospital records, and imaging studies were reviewed using the picture archiving and communication system. Patient age at diagnosis, presenting symptoms, tumor location and size, surgical approach, margin status, and outcomes were analyzed. All surgical specimens underwent independent histopathological review by two specialized gynecologic pathologists to confirm DFSP diagnosis according to World Health Organization criteria. The following parameters were recorded: overlying epidermal appearance, involved tissue type (dermis, subcutaneous tissue, skeletal muscle), mitotic index (per 10 high-power fields [HPFs]), and presence of multinucleated cells or fibrosarcomatous transformation. Patients with the following conditions were excluded: one or two specialized gynecologic pathologists deny the diagnosis, patients without complete follow-up data.

Results

A total of ten patients were identified. After histopathological review, one case was excluded due to diagnostic disagreement by both specialized pathologists, and two were excluded for incomplete follow-up data. Seven patients meeting the inclusion criteria were ultimately analyzed.

Clinical features of vulvar dermatofibrosarcoma protuberans

Clinical characteristics are summarized in **Table 1**. Patients presented with vulvar DFSP at a mean age of 44.3 years (range, 27-73) and typically had firm, asymptomatic masses. All seven tumors were primary vulvar DFSP. Lesions were located on the labia majora and mons pubis, with the labia majora most commonly affected (n = 6, 85.7%). Presentations included a pigmented lesion (n = 1) and tumor nodules (n = 6). The mean tumor size was 4.3 cm (range, 2.0-6.5).

Treatment of vulvar dermatofibrosarcoma protuberans

All seven patients underwent excisional biopsy followed by wide local excision; none received lymphadenectomy (**Table 1**). Surgical margin data were unavailable for one patient. Among the six patients with documented margins, five achieved negative margins after initial wide local excision. The patient with a positive margin did not undergo re-excision, radiotherapy, or chemotherapy due to advanced age and serious comorbidities, and died shortly after of unrelated diseases.

Outcome of vulvar dermatofibrosarcoma protuberans

Follow-up data were available for all seven cases (**Table 1**). The mean follow-up duration was 27.3 months (range, 12-54). Among the seven patients, one (14.3%) experienced recurrence. The relapsed patient was the individual with a positive surgical margin described earlier. This elderly woman developed local recurrence at 28 months after surgery and died shortly after of unrelated diseases. None of the five patients with negative surgical margins have experienced recurrence to date. The patient with undocumented surgical margins remains recurrence-free at last follow-up.

Histopathologic characteristics of vulvar dermatofibrosarcoma protuberans

Histopathologic features are summarized in **Table 2**. All tumor lesions exhibited typical histological features of DFSP (**Figures 1A-C** and **2A-D**). In six patients, tumors comprised spindle cells arranged in a storiform pattern (**Figure 1B**), with DFSP components involving subcutaneous tissue and dermis (**Figure 1A**). Typical DFSP cases showed mitotic indices ranging from 1 to 20 per 10 HPFs (median, 5 per 10 HPFs). Fibrosarcomatous components were absent in six lesions, excepting Case 7 where this data was unavailable.

Immunohistochemical characteristics and fluorescence in situ hybridization results of vulvar dermatofibrosarcoma protuberans

The immunohistochemical characteristics and fluorescence in situ hybridization (FISH) results of vulvar DFSP are summarized in **Table 3**. All seven cases were CD34-positive (**Figure 2A**), with strong positivity in five. Estrogen receptor staining was positive in one of four tested cases. Progesterone receptor staining was negative in all four tested cases. Additional immunohistochemical studies for S-100,

Table 1. Clinical features of vulvar dermatofibrosarcoma protuberans

Case Number	Age	Site	Presenting symptom	Tumor type	Tumor size (cm)	Treatment	Margin status	Outcome upon follow-up
1	49	Rt. Labium majus	Rt. Labium majus pigmented lesion	Primary	3.0	Wide local excision	NEG	No evidence of disease at 36 months
2	73	Rt. Labium majus	Rt. Labium majus mass	Primary	6.5	Wide local excision	POS	Local recurrence at 28 moths. Dead of other causes
3	27	Lt. Labium majus	Lt. Labium majus mass	Primary	4.0	Wide local excision	NEG	No evidence of disease at 18 months
4	29	Mons pubis	Mons pubis mass	Primary	3.5	Wide local excision	NEG	No evidence of disease at 22 months
5	49	Lt. Labium majus/mons pubis	Lt. Labium majus/mons pubis mass	Primary	6.0	Wide local excision	NEG	No evidence of disease at 54 months
6	29	Lt. Labium majus	Lt. Labium majus mass	Primary	5.0	Wide local excision	NEG	No evidence of disease at 12 months
7	54	Rt. Labium majus	Rt. Labium majus mass	Primary	2.0	Wide local excision	N/A	No evidence of disease at 21 months

Rt., right; Lt., left; NEG, negative; POS, positive; N/A, not available.

Table 2. Histopathologic characteristics of vulvar dermatofibrosarcoma protuberans

Case Number	Cellular appearance	Tissue affected	Mitosis (×10 HPF)	Fibrosarcomatous Component
1	Spindle cells, storiform pattern	Subcutaneous tissue	3	No
2	Spindle cells, storiform pattern	Dermis	3-5	No
3	Spindle cells, storiform pattern	Dermis	>10	No
4	Spindle cells, storiform pattern	Dermis	18-20	No
5	Spindle cells, storiform pattern	Subcutaneous tissue, dermis	>10	No
6	Spindle cells, storiform pattern	Subcutaneous tissue, dermis	<3	No
7	N/A	Dermis	1-2	N/A

HPF, high power fields; N/A, not available.

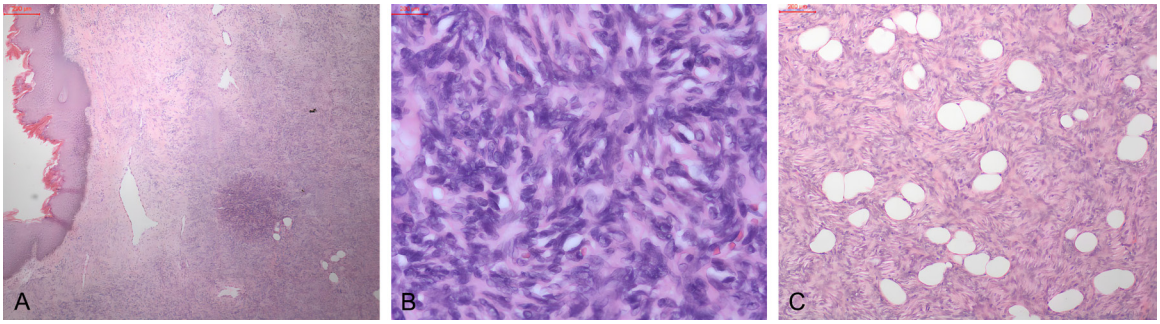

Figure 1. Histopathologic characteristics of vulvar dermatofibrosarcoma protuberans. Histopathological examination revealed a dermal tumour composed of spindle cells extending to subcutaneous tissue (A), spindle-shaped cells arranged in a storiform pattern (B), and spindle cells infiltrating surrounding fatty tissues forming a honeycomb-like structure (C). Original magnification: (A) ×20; (B) ×400; (C) ×100.

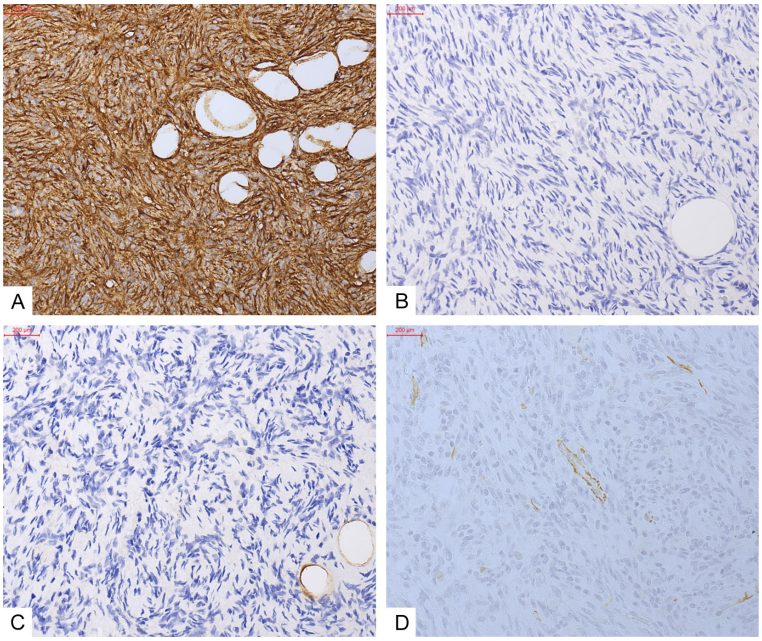

Figure 2. Immunohistochemical characteristics of vulvar dermatofibrosarcoma protuberans. (A) Strong and diffuse CD34 immunoreactivity. (B) Negative desmin expression. (C) Negative S-100 expression. (D) Negative smooth muscle actin (SMA) expression. Original magnification: ×200 (A-D).

Desmin, and smooth muscle actin (SMA) were negative in all seven cases (**Figure 2B-D**). Among three STAT6-stained cases, one was positive. Anaplastic lymphoma kinase (ALK) staining was negative in four cases. Six cases underwent CD10 staining: two were negative, while among four positive cases, one showed strong positivity, two focal positivity, and one moderate positivity. Fluorescence in situ hybridization-platelet derived growth factor receptor (FISH-PDGFR) staining was positive in five of six tested lesions (**Figure 3**).

Discussion

DFSP is a rare, well-differentiated fibrocutaneous tumor of low - to - intermediate grade malignancy. Metastases are rare (reported rates: 1%-5%) [6-8]. DFSP can affect adults of all ages but occurs most frequently in young and middle-aged adults, particularly during the fourth decade of life [3-5]. Although DFSP may involve any body site, vulvar involvement is exceptionally rare, accounting for 1.5% to 5% of vulvar malignancies [9]. Our results indicate vulvar DFSP predominantly affects adult women, with a mean presentation age of 44.3 years (range, 27-73), consistent with prior studies [10, 11].

DFSP may be asymptomatic, though vulvar DFSP can present with bleeding, ulceration, or pain. Similar to our cases, most vulvar DFSP lesions manifest as solitary subcutaneous masses; however, multiple nodules or plaque-like lesions with peripheral red-to-blue discoloration may occur [12]. While vulvar DFSP can involve any perineal site, the labia majora is the most common location (left > right), with reported sizes ranging from 1.5 to 15 cm [13]. In our series, 85.7% (6/7) of tumors originated in the labia majora (three left-sided, three right-sided). The mean tumor size was 4.3 cm (range,

Table 3. Immunohistochemical characteristics and fluorescence in situ hybridization results of vulvar dermatofibrosarcoma protuberans

Case Number	CD34	PR	ER	S100	Desmin	SMA	STAT6	CD10	ALK	FISH-PDGFR
1	SP	-	+	-	-	-	N/A	SP	-	N/A
2	SP	-	-	-	-	-	-	-	-	-
3	SP	N/A	N/A	-	-	-	-	N/A	-	+
4	SP	N/A	N/A	-	-	-	N/A	FP	N/A	+
5	FP	-	-	-	-	-	N/A	MP	-	+
6	SP	N/A	N/A	-	-	-	+	-	N/A	+
7	MP	-	-	-	-	-	N/A	FP	N/A	+

SMA, smooth muscle actin; ALK, anaplastic lymphoma kinase; PR, progesterone receptor; ER, estrogen receptor; FISH-PDGFR, fluorescence in situ hybridization-platelet derived growth factor receptor; SP, strong positive; MP, moderate positive; WP, weak positive; FP, focal positive; N/A, not available.

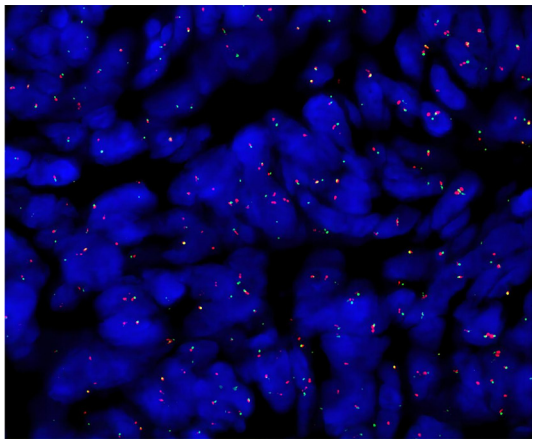

Figure 3. Representative result of fluorescence in situ hybridization-platelet derived growth factor receptor (FISH-PDGFR) staining. Original magnification: ×1000.

2.0-6.5), consistent with prior reports [10, 14, 15].

Due to its painless, slow growth, vulvar DFSP is often overlooked by patients. Clinically, it may be mistaken for a sebaceous or Bartholin cyst. Like other vulvar tumors, definitive diagnosis requires confirmation through excisional or core needle biopsy. Simple palpation cannot assess invasion depth; therefore, preoperative MRI/CT is necessary [6, 14]. Histopathogically, DFSP features spindle-shaped cells arranged in a storiform ("cartwheel") pattern. Immunohistochemistry shows diffuse strong positivity for CD34 and vimentin, but negativity for desmin, smooth muscle actin, S-100, factor XIII and keratin [16-18]. DFSP tumor cells lack nuclear pleomorphism and typically exhibit fewer than 5 mitoses per 10 high-power fields (HPFs) [10, 19]. In this study, our cases demonstrated typical DFSP features: immunohistochemical positivity for CD34 and negativity for desmin, SMA, and S-100. Most cases had mitotic rates below 5/10 HPFs. Differential diagnoses include smooth muscle-derived, neurogenic, and solitary fibrous tumors. Negative staining for desmin/SMA excluded smooth muscle tumors, S-100 negativity ruled out neurogenic tumors, and STAT6 negativity excluded solitary fibrous tumors.

Wide and deep local excision is recommended for both primary and recurrent DFSP, as microscopic projections of the tumor often extend beyond macroscopically recognizable margins [9, 10, 20]. When incompletely or marginally excised, DFSP exhibits locally aggressive behavior with recurrence rates of 20-49% [21, 22]. Multiple studies confirm that large free margins reduce recurrence risk [10, 23, 24]. Mohs micrographic surgery provides microscopic margin evaluation to ensure complete excision in high-risk DFSP. Growing evidence supports frozen-section margin control (e.g., Mohs surgery) as safe and effective for vulvar DFSP [22, 25, 26]. In our series, all patients underwent wide local excision. No recurrences occurred in patients with negative margins during follow-up. The single recurrence occurred in the patient with a positive surgical margin. Mean follow-up was 27.3 months (range, 12-54).

Although vulvar DFSP prognosis is relatively favorable, risks of local recurrence and distant metastasis persist. Therefore, post-treatment follow-up is essential. Optimal surveillance intervals remain undefined, but current recom-

mendations include clinical examination every 6 months during the first 3-5 years and annually thereafter.

The limitations of this study include its retrospective design, small sample size, and lack of randomization. Future studies with larger patient cohorts and long-term follow-up are necessary.

Conclusions

In summary, vulvar DFSP is rare and frequently unsuspected clinically. It predominantly affects young and middle-aged women, typically manifesting as external genital masses of variable size. Histologically, vulvar DFSP is characterized by spindle cells arranged in a storiform pattern with diffuse strong CD34 immunopositivity. Wide local excision represents the primary treatment. Although vulvar DFSP exhibits a tendency for local recurrence, patients with widely negative margins are unlikely to experience recurrence. Mohs micrographic surgery may ensure negative margins and reduce recurrence rates. Careful long-term follow-up for local and distant recurrence is recommended after initial surgery.

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Written consent to publish this information was obtained from the patients.

Disclosure of conflict of interest

None.

Abbreviations

DFSP, Dermatofibrosarcoma Protuberans; SMA, Smooth Muscle Actin; ALK, Anaplastic Lymphoma Kinase; HPFs, High-Power Fields; MRI, Magnetic Resonance Imaging; CT, Computed Tomography.

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