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***PDGFB* Rearrangements in Dermatofibrosarcoma Protuberans of the Vulva: A Study of 11 Cases Including Myxoid and Fibrosarcomatous Variants**

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Abstract

Dermatofibrosarcoma protuberans (DFSP) is a low grade fibroblastic sarcoma that tends to arise in young to middle age adults and involve the trunk and proximal extremities. Rare examples of vulvar DFSP have been reported, including myxoid, myoid and fibrosarcomatous variants, but detection of the characteristic t(17;22)(q22;q13) that produces *COL1A1-PDGFB* gene fusion has not been evaluated in a large series of primary vulvar tumors. The clinical, morphologic, immunohistochemical and molecular cytogenetic features of 11 cases were examined. Patient age ranged from 29 to 75 years (mean 46, median 43). Seven tumors were purely classic DFSP, one was purely myxoid DFSP and the remaining three had varying quantities of fibrosarcomatous DFSP. All cases of classic DFSP had diffuse expression of CD34 and low level p53 immunoreactivity. Myxoid variants had strong, but reduced expression of CD34. Fibrosarcomatous DFSP showed focal CD34 expression and increased p53 reactivity. Nine of 11 tumors (82%) had rearrangement of *PDGFB* by fluorescence *in situ* hybridization. The two non-rearranged tumors were a classic DFSP and a myxoid DFSP with fibrosarcomatous transformation. Follow up was available for 9 patients (82%) and ranged from 1 to 108 months (mean 30, median 21). Eight patients had tumors with positive margins, one of which developed local recurrence after no further therapy. No patient developed metastasis. The high frequency of *PDGFB* rearrangement in vulvar DFSP provides a useful exploit in diagnostically challenging cases and genetic evidence of probable clinical response to targeted therapeutics in cases of locally advanced or metastatic vulvar DFSP.

Introduction

Dermatofibrosarcoma protuberans (DFSP), previously considered in the World Health Organization Classification of Tumors of Soft Tissue(1) as a neoplasm of fibrohistiocytic

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differentiation, is now listed as a tumor of fibroblastic/myofibroblastic differentiation in the 2013 edition of the Classification(2). It is a superficial low grade sarcoma of intermediate malignancy that typically arises on the trunk and proximal extremities of young adults and has a high frequency for local recurrence due to diffuse infiltration of the dermis and subcutis(2). Distant metastasis is rare, but can occur, particularly when tumors have transformed to fibrosarcoma or pleomorphic sarcoma.

In addition to tumors that have progressed to high grade sarcoma, several low grade morphologic variants of DFSP have been described including myxoid, myoid and pigmented forms(2). Myxoid DFSP(3, 4) shows a varying degree of myxoid change to its stroma, often resulting in loss of the characteristic storiform growth pattern of classic DFSP, and can morphologically mimic myxoid genital stromal neoplasms when arising in the vulva. Myoid DFSP(5, 6) is so named due to the presence of occasional small, localized myoid nodules in otherwise classic DFSP that are thought to represent non-neoplastic myointimal proliferations of entrapped blood vessels. Uncommon cases of classic DFSP can contain scattered pigmented dendritic melanocytic cells and are given the designation of pigmented DFSP(7).

Most examples of classic DFSP and its variants have a unique underlying unbalanced chromosomal translocation, t(17;22)(q22;q13), that generates a supernumerary ring chromosome containing a *COL1A1-PDGFB* gene fusion(8). This genetic alteration can be detected by karyotype, fluorescence *in situ* hybridization (FISH) or reverse transcription polymerase chain reaction. The *COL1A1-PDGFB* fusion provides not only a useful exploit for diagnostics, but also for treatment purposes as well since the mechanisms of tumorigenesis by which the neoplastic cells proliferate render them sensitive to tyrosine kinase inhibition. The resulting *COL1A1-PDGFB* chimeric protein is indistinguishable from normal platelet derived growth factor (PDGF) subunit β , and stimulates tumor cell proliferation by an autocrine mechanism since PDGF β receptors are present on the tumor cells of DFSP(9). Imatinib mesylate, a tyrosine kinase inhibitor, has been reported as an effective pre-operative, adjuvant or palliative therapy in cases of advanced DFSP(10, 11).

Studies of extra-vulvar DFSP indicate that their characteristic t(17;22)(q22;q13) or resulting fusion transcript can be detected in a majority of tumors. However, the detection rate of this genetic aberration has not been evaluated in a large series of DFSP of the vulva, particularly in myxoid and fibrosarcomatous tumors which can pose diagnostic difficulty. To better understand the frequency of *PDGFB* rearrangement in vulvar DFSP and its variants, we evaluated a series of tumors by FISH.

Material and Methods

The pathology archives of Mayo Clinic, Brigham and Women's Hospital and The Johns Hopkins Hospital were searched for cases of DFSP arising in the vulva. Clinical data, pathologic data and patient status at last follow up were obtained from institutional medical records, referring pathologists or clinicians. All available hematoxylin and eosin slides were retrieved and evaluated by two of the authors (KJ and JKS). Formalin-fixed paraffin-

embedded tissue blocks were obtained for immunohistochemistry (IHC) and FISH at 5 µm sections.

Unstained slides underwent antibody-specific epitope retrieval using the Dako EnVision automated detection system (Dako, Carpinteria, CA) for antigens: CD34 (QBEnd/10; 1:50; Leica Biosystems, Buffalo Grove, IL), smooth muscle actin (1A4; 1:50; Dako), desmin (DERII; 1:100; Leica Biosystems) and p53 (DO-7; prediluted; Ventana Medical Systems, Tucson, AZ). Immunoreactivity was scored by percentage of tumor cell expression: 0 (negative); 1+ (1% to 33%); 2+ (34% to 66%); 3+ (67% to 100%) and intensity: weak (W), heterogeneous (H) or strong (S).

PDGFB rearrangement status was evaluated in all cases by a dual color probe set according to a previously published protocol(12). In brief, following denaturation of DNA from unstained sections, flanking BAC probes were hybridized to the *PDGFB* locus at 22q13 with the 5' telomeric region in SpectrumGreen (Abbott, Abbott Park, IL) and the 3' centromeric region in SpectrumOrange (Abbott) fluorochromes. Two technologists scored 200 interphase nuclei. The typical unbalanced rearrangement pattern (2R2F) was required in in at least 10% of tumor cells to be interpreted as positive.

In two cases, tissue was processed for karyotyping according to a previously published method(13).

Results

Clinical and pathologic findings are summarized in Table 1. A total of 11 tumors from 11 patients were identified. Patient age ranged from 29 to 75 years (mean 46, median 43). Clinical impression was available for 10 patients and varied from vulvar mass (n=6) to Bartholin gland mass (n=1), sebaceous cyst (n=1), vulvar cyst (n=1) or abscess (n=1). Gross size was known for 10 tumors, ranging from 2 to 6.3 cm (mean 4, median 4). Submitted diagnoses were provided in seven consultation cases: spindle cell neoplasm (n=2), neurofibroma (n=2), DFSP (n=2) and low grade dedifferentiated liposarcoma (n=1). Simple excision was the initial surgical procedure for 9 patients (82%) which resulted in positive excision margins in 8 patients (89%). The two remaining patients (18%) underwent wide local excision, and both of these specimens had positive margins.

Seven cases (64%) were purely classic DFSP. These tumors showed a dermal-based proliferation of uniformly spindled cells with eosinophilic cytoplasm and mildly atypical, wavy nuclei arranged in a storiform or pinwheel-like pattern (Figs. 1A,1B). Tumor cells extensively infiltrated fibrous septae and interdigitated fat cells of subcutaneous tissue to form lobules, resulting in a honeycomb-like pattern. Delicate, thin-walled vessels were the more prevalent type of vasculature, with scattered medium-sized vessels also present. Adnexal structures in the cutis and subcutis were frequently encased by tumor cells. The quantity of interstitial collagen ranged from minimal to moderate. Necrosis was not identified. Mitotic activity was low, between 0 to 3 figures per 10 high power fields.

One case (9%) was purely myxoid DFSP. This infiltrative tumor exhibited extensively myxoid stroma that resulted in loss of storiform growth and greater prominence of small to

medium sized blood vessels (Fig. 1C). The tumor cells retained uniformly low grade cytology, but took on a more fibrillary or neural quality to the cytoplasm (Fig. 1D). Mitotic figures were not seen and necrosis was not present.

Three cases (27%) showed progression to fibrosarcomatous DFSP, one of which was classic DFSP intermixed with fibrosarcoma, one was myxoid DFSP intermixed with fibrosarcoma and the remaining was purely fibrosarcoma. In the two tumors with components of classic or myxoid DFSP, the transition to fibrosarcoma was gradual rather than abrupt. In the areas of fibrosarcoma, the tumors had a fascicular or herringbone architecture with greater cytologic atypia (uniformly high grade) and mitotic activity (between 8 to 15 figures per 10 high power fields) (Figs. 1E and 1F). Mostly small to occasionally medium-sized vessels were interspersed. Necrosis was not identified in any tumor.

Immunohistochemical findings are summarized in Table 2. All cases of classic DFSP exhibited diffuse (strong, 3+) expression of CD34 (Fig. 2A) and uniformly low level (heterogeneous, 1+) expression of p53 (Fig. 2B). Myxoid DFSP had strong, but reduced or patchy (2+) CD34 expression and low level p53 (heterogeneous, 1+). Fibrosarcomatous DFSP demonstrated no more than focal CD34 (strong, 1+) and increased p53 expression (heterogeneous, 3+) (Figs. 2C,2D). Focal SMA positivity was identified in two tumors with fibrosarcomatous transformation, varying from weak to strong intensity, but only in the areas of fibrosarcoma. The remaining tumors were negative for SMA. Desmin was negative in all tumors.

Nine of 11 tumors (82%) demonstrated rearrangement of *PDGFB* (Fig. 2E). The two non-rearranged tumors were a classic DFSP and a myxoid DFSP with fibrosarcomatous transformation. Conventional G-banded chromosome analyses were available for two *PDGFB* rearranged tumors, both of which demonstrated a supernumerary ring chromosome (derivative chromosome 22) containing the centromere of chromosome 22 and interspersed with fusion sequences from chromosomes 17 and 22 (Fig. 2F).

Follow up was available for 9 patients (82%) and ranged from 1 to 108 months (mean 30, median 21). Of the 8 patients with involved margins, four patients (nos. 3, 4, 7 and 9) underwent additional therapeutic procedures (50%). One of these patients (no. 3) had classic DFSP with clinical evidence of local recurrence within 6 months and underwent two subsequent wide local excisions to achieve negative margins. Although two patients (nos. 4 and 7), one with classic DFSP (no. 4) and one with myxoid DFSP with fibrosarcomatous transformation (no. 7), had no clinical evidence of recurrence, they underwent wide local excision for clearance of margins (no. 4 by Mohs micrographic surgery). A separate patient (no. 9) received adjuvant radiotherapy after removal of a classic DFSP by wide local excision with positive margins. The remaining four patients with positive margins (50%) and one patient with negative margins had no further clinical intervention and no evidence of recurrence at last follow up.

Discussion

While our clinical and pathologic findings are in keeping with other studies of DFSP(14–16) such as the tendency to arise in young to middle age women, usual tumor size of approximately 4 to 5 centimeters and conventional microscopic features including a spectrum of variants, one unusual result of our series is the low incidence of recurrence. Despite margin involvement in 10 of 11 cases, local recurrence was clinically documented in only one patient. This could be attributed to awareness of the importance of complete excision of DFSP since two patients without clinical evidence of disease opted for re-excision of the tumor bed. Another explanation for the low rate of recurrence could be limitation of comprehensive follow up given that several cases were consultations. An additional possibility is that our follow up time period is insufficient to capture recurrence events. A large survey of recurrent DFSP found that a majority of patients experienced recurrence within approximately three years of diagnosis, sooner if any portion of the tumor had fibrosarcomatous transformation(17, 18).

In the three cases of fibrosarcomatous DFSP in our series, whether pure or as a component within a tumor containing areas of classic DFSP, the fibrosarcomatous component was unrecognized by both general and gynecologic pathologists. The prognostic significance of fibrosarcomatous or other high grade sarcomatous transformation of DFSP is debated. Studies of tumors exhibiting morphologic evidence of progression to fibrosarcoma or pleomorphic sarcoma found that transformed DFSP was associated with a higher frequency of metastasis and risk of patient death than purely classic DFSP(19, 20). Goldblum and colleagues(21) contested this concept, suggesting that such conclusions about transformed DFSP were based on variably and inadequately excised tumors. In their own study of 18 cases of transformed DFSP uniformly treated by wide local excision with negative margins, no patient developed metastasis during the follow up period. The authors offered that these improved patient outcomes were likely a result of entirely eliminating metastatic potential, but they could not exclude the possibility that their cases may reflect a less aggressive subset of transformed DFSP simply because these tumors could be fully excised with negative margins (and akin to the generally improved prognosis of high grade sarcomas that occur in the extremities). A recent review of the literature by Liang and investigators(22) looked at outcome data in 1422 patients with DFSP and 255 patients with fibrosarcomatous DFSP. Patients with transformed tumors had an increased risk of local recurrence (29.8% versus 13.7%), distant spread (14.4% versus 1.1%) and death of disease (14.7% versus 0.8%). They also found that any quantity of fibrosarcomatous change led to poorer patient outcome. The clinical significance of fibrosarcomatous transformation may be controversial, but recognition and documentation of high grade sarcomatous transformation in DFSP is prudent until additional data about its impact on patient outcome are available. If morphologic findings such as herringbone architecture, higher grade cytologic features and increased mitotic activity are equivocal, increased expression of p53(23) is helpful in differentiating fibrosarcomatous transformation from classic DFSP (compare Figs. 2B and 2D).

Whether the tumor is classic DFSP, a low grade variant or transformed high grade sarcoma, the initial management is surgical with the goal being complete resection to achieve negative

margins(18). In comparison to wide local excision, some studies have reported greater success in margin clearance by use of Mohs micrographic surgery, especially in anatomic locations where tissue conservation is critical or where risk of incomplete excision is potentially higher due to need to preserve physiologic or cosmetic structures. A systematic review by Foroozan and investigators(24) of 23 non-randomized clinical trials contrasting micrographic surgery to wide local excision found a lower recurrence rate of patients treated by micrographic surgery (1.11%) versus wide local excision (6.32%), leading some centers to consider micrographic surgery as preferred surgical therapy for DFSP. However, financial expense of mapping surgery is a factor, and in anatomic regions where cosmesis is less critical, wide local excision with histologically negative margins remains an acceptable method(25).

Due to the protuberant, infiltrative growth and low grade cytologic features of classic DFSP and the clinical significance of negative margins, accurate microscopic assessment of margins can be challenging. CD34 has been shown to be a useful stain for differentiating histologic mimics of DFSP such as scar tissue. Prieto et al(26) examined CD34 immunoreactivity in DFSP re-excision specimens containing foci of residual tumor adjacent to scar. The tumor cells were consistently and strongly positive for CD34 while the surrounding scar tissue was negative. They noted endothelium, periadnexal cells and scattered dendritic cells were also positive for CD34, but these findings did not interfere with appropriate classification of cells as neoplastic or non-neoplastic. In our experience, CD34 is an excellent and reliable stain to capture the full extent of tumor infiltration and to evaluate subtle margin involvement (Figs. 3A,3B).

In surgically unresectable tumors or advanced stage patients, imatinib, a tyrosine kinase inhibitor, is a potentially efficacious therapeutic targeted modality due to its interruption of constitutive activity of PDGFB receptor found on tumor cells(27). A study by McArthur and colleagues(28) examined the effect of imatinib in ten patients: eight with locally advanced DFSP and two with metastatic disease (both metastatic tumors had undergone fibrosarcomatous transformation). Four patients experienced complete clinical response, all of whom had cytogenetic confirmation of t(17;22)(q22;q13). However, in one patient with metastasis and a fibrosarcomatous tumor with t(17;22)(q22;q13), in addition to other complex karyotypic changes, had only a partial response with disease progression 7 months later. Further, in the other patient with metastatic disease whose fibrosarcomatous tumor lacked cytogenetic evidence of t(17;22)(q22;q13) had no response to imatinib. The authors concluded that targeted therapy can be beneficial in advanced stage DFSP, but that the presence of t(17;22)(q22;q13) may be influential in predicting response to targeted therapy.

Protocols of paraffin-based techniques using multiplex reverse transcription PCR or FISH to identify *COL1A1-PDGFB* fusion have been published(12, 29–32). Of these, reported detection rates or evidence of *COL1A1-PDGFB* fusion range from 83 to 100%. By FISH, our rate of *PDGFB* rearrangement was 82% or 9 of 11 cases, inclusive of variants and high grade tumors. With this high frequency of underlying genetic aberration, analysis of t(17;22)(q22;q13) or *COL1A1-PDGFB* fusion transcript provides a useful exploit in diagnostically difficult cases arising in the vulva due to DFSP's morphologic overlap with a variety of

tumors such as fibrous histiocytoma, peripheral nerve sheath tumors, smooth muscle tumors, melanoma and deep angiofibroma since none of these entities share this genetic aberration.

In its classic form, DFSP can be confused with fibrous histiocytoma, a benign tumor of fibrohistiocytic differentiation, due to both showing a proliferation of cytologically bland spindle cells with a fibroblastic or histiocytoid appearance. However, in contrast to DFSP, fibrous histiocytoma typically maintains an overlying Grenz zone associated with epidermal hyperplasia and basilar hyperpigmentation, entraps bundles of collagen fibers at the periphery of the lesion and does not exhibit the same pattern of extensive infiltration into septae of subcutaneous fat lobules(33). DFSP is immunohistochemically different from fibrous histiocytoma by its consistently diffuse expression of CD34 and lack of CD163 expression, a pattern that is reversed in fibrous histiocytoma(34).

Peripheral nerve sheath tumors such as diffuse neurofibroma and malignant peripheral nerve sheath tumor are important diagnostic considerations. Neurofibroma is usually formed by a loose arrangement of small, elongated and wavy spindled cells set in a collagenous extracellular matrix and lacks the storiform architecture of DFSP. Although the diffuse variant of neurofibroma is an expansive lesion that can entrap adnexal structures and extend into septae of subcutaneous fat, it does not widely infiltrate the adipocytes to create a honeycomb pattern(35, 36). Also, Wagner-Meissner bodies which are distinctive spherical clusters of spindled cells with ample cytoplasm are commonly found in diffuse neurofibroma and not in DFSP. In contrast to DFSP, neurofibroma exhibits only patchy to focal CD34 positivity and strong S100 protein positivity. Malignant peripheral nerve sheath tumor is a neurogenic neoplasm that typically has an alternating hyper- and hypocellular fascicular architecture, that can result in a herringbone configuration in some examples, and is formed by high grade cells with retained elongated and somewhat wavy nuclei, numerous mitotic figures and regions of coagulative necrosis(37). It can be distinguished from fibrosarcomatous DFSP by identification of a respective precursor component such as neurofibroma or classic DFSP. Malignant peripheral nerve sheath tumor exhibits reduced, but at least focal, S100 protein expression whereas fibrosarcomatous DFSP is negative for this marker(38).

Smooth muscle tumors can be differentiated from DFSP by morphology or immunohistochemistry. Leiomyoma is distinguished from classic DFSP by its absence of storiform growth and tendency to be well circumscribed. If leiomyoma is infiltrative, it tends to have a more broad extension of fascicles into surrounding non-neoplastic tissue rather than develop a tendril-like pattern of invasion seen in DFSP(39). Although the myoid variant of DFSP can exhibit expression of smooth muscle markers, the extent of positivity is focal and limited to myoid nodules(6). Leiomyosarcoma can morphologically overlap with fibrosarcomatous DFSP, but is distinguished by the presence of fascicular rather than herringbone architecture, usually greater degree of nuclear pleomorphism and expression of desmin(20).

Melanoma and deep angiofibroma can also resemble DFSP and its variants such as pigmented DFSP and myxoid DFSP, respectively. Although desmoplastic melanoma is an infiltrative tumor of cytologically low grade spindle cells, in contrast to pigmented DFSP, it

is an overall ill defined neoplasm of relatively low cellular density with stromal desmoplasia that often has an in situ component (lentigo maligna) and is S100 protein positive and CD34 negative(40). Deep angiomyxoma is a genital stromal tumor with a propensity for local recurrence. It is uniformly hypocellular with an extensively myxoid matrix that is occupied by a population of low grade spindle or stellate cells(41). Occasional smooth muscle bundles or myoid bundles located adjacent to blood vessels that immunoreact to smooth muscle markers can also be seen. Although deep angiomyxoma can also be CD34 positive, in contrast to myxoid DFSP, it is typically desmin positive and expresses some degree of hormone receptors(42). Further, if present, identifying a component of classic DFSP is useful in separating these entities. In diagnostically challenging cases, FISH or RT-PCR to identify *COL1A1-PDGFB* in DFSP can be helpful.

In summary, we present findings in a series of 11 cases of vulvar DFSP that encompass the classic form, myxoid variant and tumors with fibrosarcomatous transformation. Our clinical, morphologic and immunohistochemical data are in line with prior reports of vulvar and extra-vulvar DFSP. *PDGFB* rearrangement is present and detectable by FISH in a majority of tumors that arise in the vulva, including variants and high grade tumors. These data support evaluation of *COL1A1-PDGFB* fusion transcript in diagnostically difficult cases in addition to providing genetic evidence of probable clinical response to targeted therapeutics in cases of locally advanced or metastatic vulvar DFSP.

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Figure 1.

Classic DFSP has a storiform architecture where a group of cells radiate from a shared center (A). These cells exhibit uniformly low grade cytologic features, are spindled in shape, contain elongated to wavy nuclei and low mitotic activity (B). Myxoid DFSP tends to lose the prototypical storiform architecture of classic DFSP, accentuating the tumor's vasculature, and mimicking a some myxoid genital stromal tumors (C). The tumor retains low grade features (D). Fibrosarcomatous DFSP has a dense herringbone or fascicular architecture (E) and cells with higher grade cytologic atypia and increased mitotic activity (F).

Figure 2.

CD34 is strongly and diffusely expressed by tumor cells in classic DFSP (A) while p53 expression is heterogeneous and low level (B). In contrast, fibrosarcomatous DFSP shows very limited CD34 positivity (C) and an increased degree of heterogeneous p53 reactivity (D). Fluorescence in situ hybridization using a break apart probe set to evaluate *PDGFB* status demonstrated rearrangement of the gene in a majority of tumors as evidenced by separation of red and green signals (E). A karyogram of a case of classic DFSP shows t(17;22)(q22;q13) and supernumerary ring chromosome formation (F).

Figure 3.
CD34 expression by classic DFSP is consistently diffuse and strong, making it a helpful marker in evaluating potential margin involvement in surgical specimens (A,B).

Table 1

Clinical and Pathologic Findings of Vulvar DFSP

Case	Patient Age (y)	Clinical Diagnosis	Tumor Size (cm)	Submitted Diagnosis	Final Diagnosis	PDGFB FISH	Initial Surgical Procedure	Margin Status	Additional Treatment and Follow Up
1	43	Vulvar abscess	5	Spindle cell neoplasm	Purely fibrosarcomatous DFSP	+	SE	+	Recent case
2	75	Vulvar mass	5	Neurofibroma	DFSP	-	SE	+	NED, 108 mos.
3	64	Sebaceous cyst	3	Neurofibroma	DFSP	+	SE	+	Local recurrence at 6 mos. treated by WLE x2 due to positive margins (final margins negative), NED, 1 mo.
4	49	Vulvar mass	4	N/A	DFSP	+	SE	+	WLE by Mohs micrographic surgery, NED, 1 mo.
5	49	Vulvar cyst	2	Spindle cell neoplasm	DFSP	+	SE	+	NED, 19 mos.
6	43	UNK	UNK	DFSP	DFSP	+	SE	+	UNK
7	51	Bartholin gland mass	5.7	Low grade dedifferentiated liposarcoma	Myxoid DFSP with fibrosarcomatous transformation	-	SE	+	WLE (negative margins); NED, 18 mos.
8	29	Vulvar mass	2.5	DFSP	Myxoid DFSP	+	SE	+	NED, 36 mos.
9	37	Vulvar mass	2.5	N/A	DFSP	+	WLE	+	Adjuvant radiotherapy; NED, 35 mos.
10	29	Vulvar mass	4	N/A	DFSP with fibrosarcomatous transformation	+	SE	-	NED, 21 mos.
11	42	Vulvar mass	6.3	N/A	DFSP	+	WLE	+	NED, 27 mos.

DFSP (dermatofibrosarcoma protuberans); SE (simple excision); WLE (wide local excision); NED (no evidence of disease); UNK (unknown)

Table 2

Immunophenotype of Vulvar DFSP

Case	Tumor	CD34	SMA	Desmin	p53
1	Purely fibrosarcomatous DFSP	1+, S	1+, S	0	3+, H
2	DFSP	3+, S	0	0	1+, H
3	DFSP	3+, S	0	0	1+, H
4	DFSP	3+, S	0	0	1+, H
5	DFSP	3+, S	0	0	1+, H
6	DFSP	3+, S	0	0	1+, H
7	Myxoid DFSP with fibrosarcomatous transformation	0 in FS; 2+, S in myxoid DFSP	1+, W in FS; 0 in myxoid DFSP	0	3+, H in FS; 1+, H in myxoid DFSP
8	Myxoid DFSP	2+, S	0	0	1+, H
9	DFSP	3+, S	0	0	1+, H
10	DFSP with fibrosarcomatous transformation	1+, S in FS; 3+, S in typical DFSP	0	0	3+, H in FS; 1+, H in typical DFSP
11	DFSP	3+, S	0	0	0

DFSP (dermatofibrosarcoma protuberans); FS (fibrosarcomatous transformation); 0 (negative); 1+ (expression in 1–33% cells); 2+ (expression in 34–66% cells); 3+ (expression in 67–100% cells); S (strong); H (heterogeneous); W (weak)