A retrospective cohort study of dermatofibrosarcoma protuberans at a large metropolitan academic center

To the Editor: Dermatofibrosarcoma protuberans (DFSP) is a rare, low-grade dermal mesenchymal malignancy that commonly presents as a slow-growing, asymptomatic nodule.[1,2] Although its metastasis is uncommon, the tumor tends to recur locally following incomplete excision.[3] The clinicopathologic features associated with poor outcomes include treatment with wide local excision (WLE), compared with that with Mohs micrographic surgery (MMS); positive histologic margins; the presence of a fibrosarcomatous change; age >50 years; a large tumor size (>5 cm); African American ethnicity; male sex; and a location on the head, neck, or extremity rather than on the trunk.[2-5] Given the rarity of DFSP, there is limited research characterizing it. We sought to determine the impact of clinicopathologic features on its recurrence after treatment.

This was a single-institution retrospective study of DFSP cases appearing in the electronic medical record of patients treated at an academic institution from 2010 to 2020. Patients who were aged ≥18 years with nonmetastatic, biopsy-proven DFSP were included. We collected all patient, tumor, and treatment variables. We used the 2-tailed *t* test, χ^2 test, and multivariate linear regression model for data analysis.

We identified 62 patients with DFSP (Table I). Most patients (94%) presented with primary DFSP rather than with recurrent DFSP, and most lesions were located on the trunk (50%) or extremities (37%) (Table II). The mean lesion duration prior to presentation was 8 years, and the lesions measured, on average, 4.0 × 2.4 cm^2. Most lesions were asymptomatic (58%) and slow growing, but 18% demonstrated rapid growth prior to surgery.

Of the 62 patients, 2 were lost to follow-up, whereas 60 had known recurrence data, obtained from medical records and/or via telephone follow-up. Of these 60 patients, 55 (92%) underwent MMS and 5 (8%) underwent WLE. No patient was referred for adjuvant treatment after surgery. Three patients experienced recurrence following treatment at our institution, yielding a 5% recurrence rate. Two of the 5 patients treated with WLE experienced recurrence, yielding a 40% recurrence rate. Of the 55 patients treated with MMS, 1 experienced recurrence, yielding a 2% recurrence rate. Upon follow-up, no patient was found to have died.

Table I. Patient demographics, clinical presentation, treatment, and outcomes

Variable	Mean (SD) or n (%)*
Patient demographics and clinical presentation	
Age	44.1 (13.4)
Male sex	19 (31%)
Current or former smoker	11 (18%)
Radiation to DFSP site	1 (2%)
On immunosuppressive medication	4 (7%)
Location	
Extremity	23 (37%)
Trunk	31 (50%)
Head and neck	8 (13%)
Size—larger dimension (cm)	4.0 (range: 1.1-13.0)
Size—smaller dimension (cm)	2.4 (range: 0.2-7.0)
Rapid growth	11 (18%)
Treatment and outcomes	
Treated with WLE	5 (8%)
Treated with MMS	55 (92%)
Number of stages if MMS	
1	17 (31%)
2	30 (56%)
3	7 (13%)
Mohs defect area (cm^2)	11.6 (14.5)
Time from biopsy to treatment (d)	49.5 (51.5)
DFSP recurred after treatment at UTSW	3 (5%)
Time to recurrence (y)	8.3 (range: 2.0-20.0)
Time from treatment to last appointment with dermatologist (mo)	8.6 (20.7)
Death	0 (0%)

DFSP, Dermatofibrosarcoma protuberans; *MMS*, Mohs micrographic surgery; *UTSW*, University of Texas Southwestern; *WLE*, wide local excision.
*Unless specified as range.

We identified several statistically significant associations between clinicopathologic features and recurrence after treatment using a univariate analysis. The recurrence rate following WLE was higher than that following MMS (40% vs 2%, respectively; $P = .00005$), consistent with prior studies. Lesions that recurred were more likely to exhibit rapid growth (100% vs 12%, $P = .0003$). Patients with recurrent lesions were more likely to have a history of skin cancer besides DFSP (33% vs 2%, $P = .004$). We did not find any significant associations between clinicopathologic features and recurrence in the

Table II. Demographic and clinical variables by tumor recurrence after MMS or WLE*

Variable	Mean (SD) or N (%)			
	Recurrent (n = 3)	Not recurrent (n = 57)	All (n = 60)	*P* value
Age (y)	51 (12.8)	44 (13.3)	44 (13.4)	.45
Male sex	2 (66%)	17 (30%)	19 (32%)	.27
Follow-up time (d)	333 (325.8)	258 (637.8)	257.2 (625.0)	.75
Larger dimension (cm)	7.5 (4.3)	3.8 (1.6)	4.0 (2.0)	.27
Smaller dimension (cm)	3.5 (1.8)	2.3 (1.6)	2.4 (1.6)	.38
Location				
Extremity	1 (33%)	22 (39%)	23 (38%)	Extremity vs other location: .89
Trunk	2 (66%)	28 (49%)	30 (50%)	Trunk vs other location: .68
Head and neck	0 (0%)	7 (12%)	7 (12%)	None—too few occurrences
Race				None—too few occurrences
Caucasian	1 (33%)	24 (42%)	25 (42%)	
African American	0 (0%)	7 (12%)	7 (12%)	
Asian	1 (33%)	7 (12%)	8 (13%)	
Hispanic	0 (0%)	4 (7%)	4 (7%)	
Repair type				5.2×10^{-5}
WLE	2 (66%)	2 (4%)	4 (7%)	
MMS	1 (33%)	53 (93%)	54 (90%)	
Unknown	0 (0%)	1 (2%)	1 (2%)	
None (deferred)	0 (0%)	1 (2%)	1 (2%)	
BMI	30.8 (8.0)	28.2 (10.0)	28.5 (8.1)	.70
Obesity	1 (33%)	14 (25%)	15 (25%)	.77
CHF, HTN, or stroke	2 (66%)	18 (32%)	20 (34%)	.32
Alcohol	2 (66%)	24 (42%)	26 (43%)	.53
Smoking				Current or former smoker vs never smoker: .43
Never smoker	3 (100%)	45 (79%)	48 (80%)	
Current smoker	0 (0%)	3 (5%)	3 (5%)	
Former smoker	0 (0%)	8 (14%)	8 (13%)	
Pregnancy at diagnosis	0 (0%)	2 (4%)	2 (3%)	.74
Autoimmune disease	1 (33%)	3 (5%)	4 (7%)	.07
Skin cancer history	1 (33%)	1 (2%)	2 (3%)	3.8×10^{-3}
Recurrent lesion upon treatment at UTSW	3 (100%)	1 (2%)	4 (6.7%)	1.3×10^{-10}
Rapid growth	3 (100%)	7 (12%)	10 (17%)	2.9×10^{-4}
Duration of lesion (mo)	172 (156)	91 (96)	96 (102)	.47
Symptomatic lesion	1 (33%)	18 (33%)	19 (33%)	.99
Time from biopsy to initial treatment	72 (62)	49 (51)	50 (52)	.59
DFSP type				.98
Classical	3 (100%)	55 (98%)	58 (98%)	
Fibrosarcomatous	0 (0%)	1 (2%)	1 (2%)	
Depth of invasion				.41
Deep	2 (66%)	2 (11%)	4 (19%)	
Superficial	1 (33%)	16 (89%)	17 (81%)	
CD34 + immunostaining	2 (100%)	43 (98%)	45 (98%)	.97

BMI, Body mass index; *CD*, clusters of differentiation; *CHF*, congestive heart failure; *DFSP*, dermatofibrosarcoma protuberans; *HTN*, hypertension; *MMS*, Mohs micrographic surgery; *UTSW*, University of Texas Southwestern; *WLE*, wide local excision.

*Statistical analysis of the comparison of recurrent versus nonrecurrent lesions was performed on 60 patients with known recurrence data in our full sample of 62 patients. Two patients had no known data on recurrence. A "deep" depth of invasion is considered beyond subcutaneous tissue, whereas a "superficial" depth of invasion is considered skin and subcutaneous tissue.

multivariate analysis. One lesion (2%) exhibited a fibrosarcomatous change and did not recur, limiting our ability to analyze this feature.

Our study was limited by its small sample size and single-institution nature, reducing its generalizability. Based on our study, one may consider WLE rather than MMS, rapid growth, and a history of other skin cancers as potential risk factors for recurrence, although these associations were found to be significant in the univariate analysis and not in the

multivariate analysis. Larger-scale studies are needed to better characterize the impact of clinicopathologic features on recurrence after surgery.

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Conflicts of interest

None disclosed.

REFERENCES

1. Asilian A, Honarjou N, Faghihi G, Saber M, Mozafarpoor S, Hafezi H. An experience of slow-Mohs micrographic surgery for the treatment of dermatofibrosarcoma protuberans: a long-term cohort study. *J Cosmet Dermatol*. 2020;19(10):2701-2705.
2. Brooks J, Ramsey ML. *Dermatofibrosarcoma protuberans*. StatPearls. Accessed January 24, 2021. https://www.ncbi.nlm.nih.gov/books/NBK513305/
3. Moore KJ, Chang MS, Weiss J, Olbricht SM, Hartman RI. Racial and ethnic differences in the surgical treatment of dermatofibrosarcoma protuberans: a retrospective cohort analysis. Preprint. Posted online August 11, 2021. *J Am Acad Dermatol*. https://doi.org/10.1016/j.jaad.2021.08.010
4. Kim M, Huh CH, Cho KH, Cho S. A study on the prognostic value of clinical and surgical features of dermatofibrosarcoma protuberans in Korean patients. *J Eur Acad Dermatol Venereol*. 2012;26(8):964-971.
5. Bowne WB, Antonescu CR, Leung DH, et al. Dermatofibrosarcoma protuberans: a clinicopathologic analysis of patients treated and followed at a single institution. *Cancer*. 2000;88(12):2711-2720.

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