Dermatofibrosarcoma Protuberans of the Vulva Treated Using Mohs Micrographic Surgery

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First described by Darier and Ferrand in 1924,[1] dermatofibrosarcoma protuberans (DFSP) is a rare, low-grade sarcoma with a tendency for recurrence. The overall annual incidence is 4.2 per million, or 0.1% of all cancers.[2] It tends to affect young to middle-aged adults and occurs more often in blacks than in whites, with no sex predilection. Dermatofibrosarcoma presents as a flesh-colored, violaceous, or reddish-brown nodule with persistent but indolent growth and is commonly located on the trunk. It rarely metastasizes, but when it does, it is most often to the lungs. Histopathologically, DFSP is a tumor consisting of spindled cells in a storiform pattern. It infiltrates the dermis, the subcutaneous fat, and even the fascia. Tumor cells stain positive for CD34 and negative for factor XIIIa.

Report of a Case

A 59-year-old woman noted a 6-year history of an asymptomatic, slowly enlarging mass on the right vulva. A dermatologist evaluated her, a biopsy was performed, and the findings were consistent with a diagnosis of DFSP. Her dermatologist referred her to our department for Mohs micrographic surgery (MMS) of the lesion.

On cutaneous examination, a 4- × 2-cm, ill-defined, pink, indurated, mobile plaque was present on the right labium major (Figure 1). No lymphadenopathy was noted. The lesion was removed with histologically clear margins using the Mohs technique in two stages (Figure 2). Immunostaining was not performed. The excision involved the superficial and deep fascial layers of the vulva and inner thigh.

The remaining defect measured 8 × 5 cm and involved the right labium major and the right upper inner thigh (Figure 3). A Burow's triangle was excised superiorly onto the right lateral suprapubic region, and a smaller Burow's triangle was excised inferiorly. The wound was then closed in a layered fashion by re-approximating the deep fascia, superficial fascia, dermis, and epidermis (Figure 4). At her 4-month follow-up, the patient continued to be tumor free and had no lymphadenopathy. Her wound exhibited an excellent cosmetic and functional outcome, with no evidence of deformity of the vulva (Figure 5). Her primary dermatologist recently evaluated her, and she remains tumor-free 2.5 years after her Mohs procedure.

Discussion

Dermatofibrosarcoma occurs on the trunk in 42% to 62% of cases, and cases involving the vulva are rare.[2,3] A total of 28 cases of vulvar DFSP have been reported in the literature.[3–24] An excellent review by Ghorbani and colleagues[4] in 1999 presented four new cases of vulvar DFSP and examined the first 18 reported cases. We revisited these cases along with six new cases reported in the interim (Table 1). Including the current case, most tumors occurred on the labia majora, with left side involvement being more common. The median patient age was 42. Limited demographic data were available, with eight

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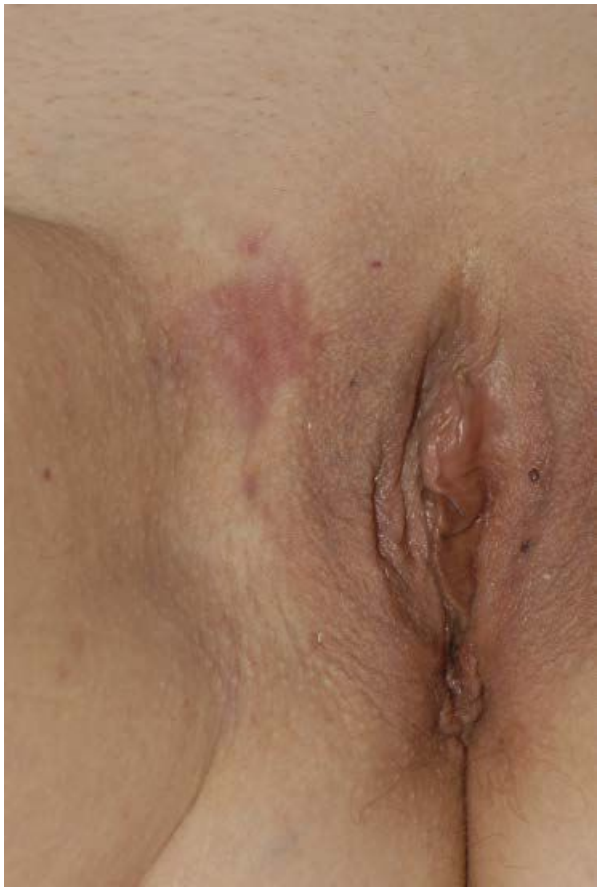

Figure 1. Preoperative photograph demonstrating a pink indurated plaque.

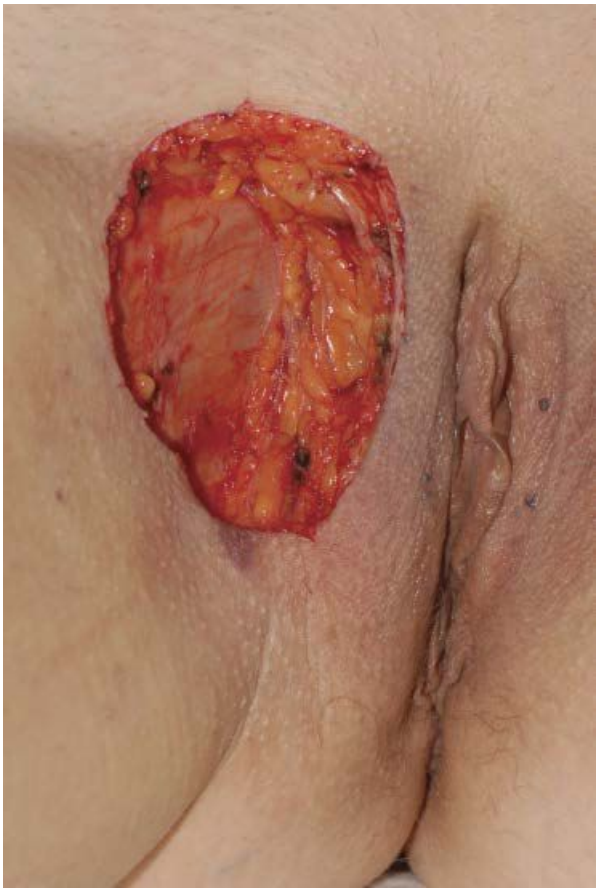

Figure 3. The surgical defect.

cases (including the current case) reporting race. Two African-American and 6 white women were documented. The average duration of tumor to definitive treatment was 45 months. The median size of the tumors was 4 cm in the largest dimension.

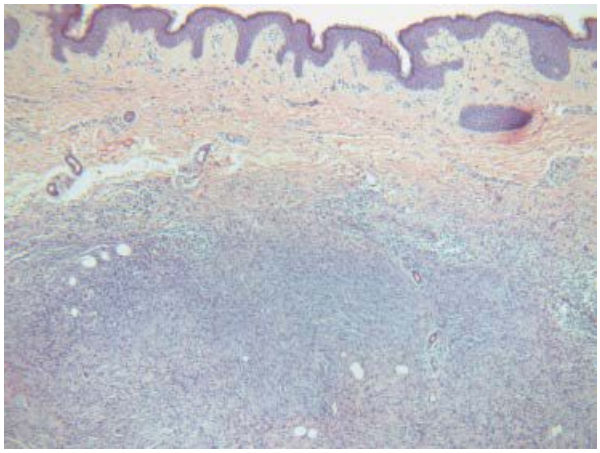

Figure 2. Mohs debulking layer. Dermal tumor composed of spindle cells extends to the subcutis.

Treatments ranged from simple excision to radical vulvectomy and radiotherapy. Three were treated with MMS (patients 27 and 28 in the Table and the current patient). Eight tumors recurred at least once; the most common treatment for recurrence was repeat local excision. Two tumors recurred after wide local excision (patients 5 and 9), and one patient had a recurrence after hemivulvectomy (patient 19). None of the three tumors treated with MMS recurred. The recurrence rate in patients treated with non-MMS modalities was 32%. One lung metastasis was reported in a patient treated first with partial radical vulvectomy and then wide local excision with radiotherapy (patient 18). One tumor enlarged rapidly during pregnancy.

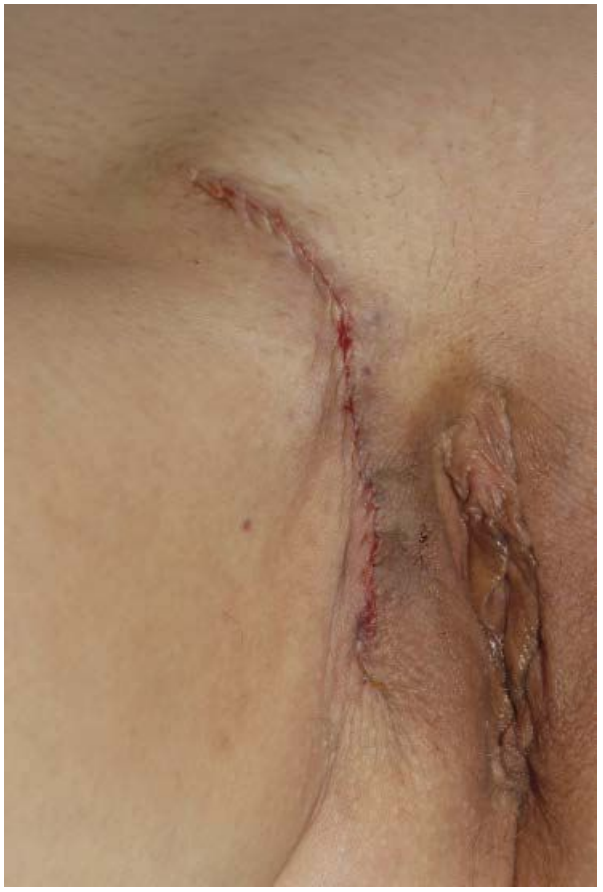

Figure 4. Repair of the surgical site.

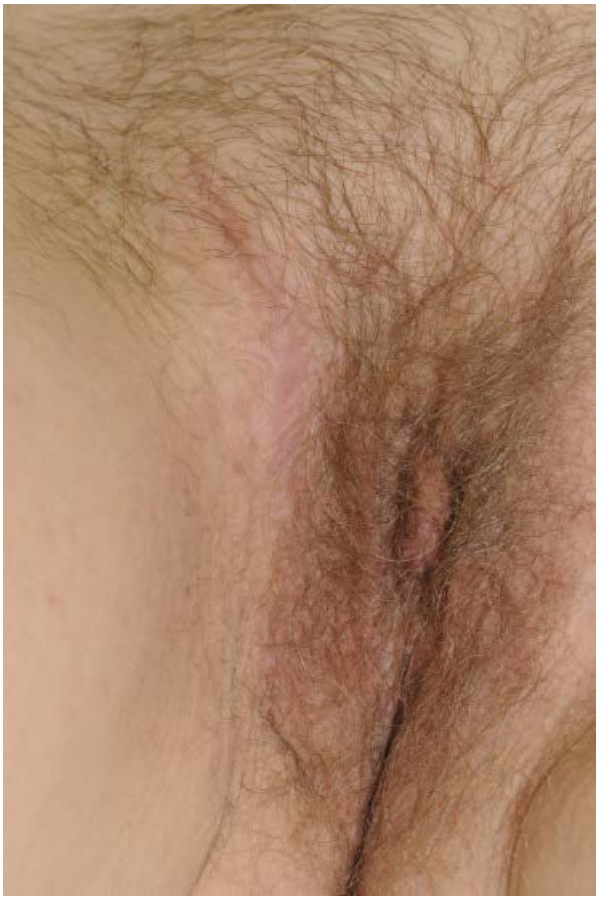

Figure 5. Healing at 4-month follow-up.

MMS is an effective treatment of DFSP.[3,24–27] In 1996, Gloster and colleagues[25] reviewed the experience at the Mayo Clinic in Rochester, Minnesota, along with the existing literature for treatment of DFSP. Dermatofibrosarcoma treated with MMS had an overall recurrence rate of 1.6%, versus 20% recurrence after wide excisions and 43% after conservative excisions. More recently, Thomas and colleagues[3] reviewed the experience with treatment of DFSP using MMS at the Geisinger Medical Center. Thirty-nine patients with DFSP were treated with MMS, and no tumor recurred over a mean follow-up of 39 months. Kimmel and colleagues[26] recently conducted a meta-analysis of 98 DFSP cases to review the relationship between clinically apparent tumor size and excision margins required for clearance. They determined only a weak relationship between tumor size and true histologic extent. Tumors of small to moderate size treated with wide local excision required 4-cm margins for 95% clearance. On the basis of these findings, the authors recommended excision with margin control to reduce the risk of recurrence while conserving tissue. This is especially important in certain anatomic sites such as the vulva.

Treating vulvar DFSP with MMS requires a trained staff and experienced surgeon. The frozen-section slides must be complete (e.g., no loss of adipose tissue) and free from freeze artifact. The nursing staff and surgeon should take extra time to comfort and reassure the patient. We have found that oral diazepam works well to alleviate patients' anxiety. A surgical table with stirrups is also helpful. As with surgery anywhere else on the skin, knowledge of local anatomy is important. The surgeon should

TABLE 1. Literature Summary

Study	*Patient*	*Age*	*Clinical Presentation*	*Duration*	*Site*	*Size, cm*	*Treatment*
Davos & Abell[5]	1	38	Nodule	NA	Labium major	3	WLE
Soltan[6]	2	83	Vulvar swelling, bloody discharge, ulcerated red nodule	10 months	L labium minor, major	5	LE
Agress et al.[7]	3	56	NA	NA	L labium major	NA	LE,* WLE
Bock et al.[8]	4	52	Enlarging vulvar swelling, pruritus	10 years	R mons	8	WLE
Barnhill et al.[9]	5	42	Firm nontender nodule	NA	R vulva lateral to clitoris	1	LE,* WLE, HV
Wrotnowski et al.[10]	6	58	NA	NA	NA	5	LE
Leake et al.[11]	7	37	Enlarging lesion	2 years	L mons, labium major	6.2	WRE
Leake et al.[11]	8	59	Nodule	NA	R labium major	5	LE,* PRV
Panidis et al.[12]	9	30	Enlarging nodule	4 months	R labium major	2	WLE,* RV
Aartsen & Albus-Lutter[13]	10	50	NA	NA	NA	1.2	RV
Nirenberg et al.[14]	11	41	Lump	NA	L labium major	8	WLE
	12	29	Mass	NA	R labium major	3.7	WLE
Alverez-Canas et al.[15]	13	58	Enlarging, firm painless mass	''Months''	L labium major	3.2	WLE
Karlen et al.[16]	14	36	Irritated lump	11 years	L labium major	5	WLE
Garcia et al.[17]	15	63	NA	NA	NA	NA	MMS
Chetty[18]	16	23	Vulvar swelling	6 months	R labium major	7	WLE
	17	29	Nodule	3 months	NA	3.5	LE
Soergel et al.[19]	18	47	Mass	8 months	L vulva, lung metastasis	3	PRV,* WLE + XRT, chemo
Ghorbani et al.[4]	19	47	NA	NA	L paraclitoral area	NA	HV,* WRE
	20	44	Enlarging mass	''Weeks''	L labium major	4	WRE
	21	66	Enlarging mass	3 months	Mons	1.5	WLE
	22	36	Enlarging mass	6 months	R labium	5	LE
Moodley & Moodley[20]	23	39	Vulval mass	NA	L labium major	12	WLE
Vanni et al.[21]	24	39	Firm nontender enlarging mass	1 years	Inferior vulva/perineum	6	WLE
Kholova et al.[22]	25	31	Recurrent nodule	10 years	L labium major	1.5	LE,* WLE + XRT
Ohlinger et al.[23]	26	36	Painless nodule	9 years	L vulva	2.8	LE,* WLE
Thomas et al.[3]	27	NA	NA	NA	NA	NA	MMS
Hancox et al.[24]	28	55	NA	7 months	R labium major	8	MMS
Current report	29	59	Enlarging mass	6 years	R labium major	4	MMS

*Recurrence.
chemo, chemotherapy; HV, hemivulvectomy; L, left; LE, local excision; MMS, Mohs micrographic surgery; NA, not available; PRV, partial radical vulvectomy; R, right; RV, radical vulvectomy; WLE, wide local excision; WRE, wide radical excision; XRT, radiotherapy.

allocate more time to obtain hemostasis because the vulvar region is highly vascularized, and wounds tend to ooze. The wound can be packed with saline-soaked 4- × 4-inch gauze and covered with gauze pads. Incontinence undergarments can be used in place of the patient's usual undergarments to apply additional pressure and prevent seepage of blood in cases of minor bleeding. Depending on the size of the tumor being treated, the tissue processing time may be lengthy, and the day's schedule may need to be adjusted to prevent a backlog of tissue specimens in the laboratory. The use of intraoperative immunostaining techniques may be helpful in interpreting the Mohs slides. We rely on pattern recognition and have not had recurrences in the limited number of cases treated over the past 5 years.

Conclusion

Dermatofibrosarcoma protuberans is a rare tumor, constituting only 0.1% of all malignancies. Vulvar DFSP is exceptionally rare, with only 27 previously reported cases. MMS is an effective treatment for DFSP, with a low rate of recurrence. Tumors of the vulva are within the scope of dermatologic surgery, and MMS is well suited for this anatomic region.

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