

Low Recurrence Rate after Surgery for Dermatofibrosarcoma Protuberans

A Multidisciplinary Approach from a Single Institution

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BACKGROUND. Dermatofibrosarcoma protuberans (DFSP) is a rare sarcoma with a propensity for local recurrence. Treatments with wide excision, Mohs surgery, and other approaches have been reported with widely variable local control rates. The objective of this study was to review the experience with a multidisciplinary approach employing wide excision and Mohs surgery selectively in the treatment of patients with DFSP at a single academic institution over the past 10 years.

METHODS. The records of 62 patients with 63 DFSP tumors who underwent wide excision, Mohs surgery, or a multidisciplinary combination approach from January 1991 to December 2000 were reviewed retrospectively. Primary endpoints included the ability to extirpate the DFSP lesion completely, the tumor recurrence rate, and the need for skin grafts or local tissue flaps. Additional objectives included defining surgical practice patterns at the authors' institution.

RESULTS. Sixty-three DFSP lesions were removed from 62 patients. At a median follow-up of 4.4 years, no local or distant recurrences were detected in any patient. Forty-three lesions were treated with wide local excision, 11 lesions were treated with Mohs surgery, and 9 lesions were treated with a combination approach. Ninety-five percent of lesions that were approached initially with wide local excision were cleared histologically. Two patients (5%) received postoperative radiation for positive margins after undergoing maximal excision. Eighty-five percent of lesions that were approached initially with Mohs surgery were cleared histologically. The remaining 15% of lesions subsequently were cleared surgically with a wide excision. DFSP lesions that were approached initially with Mohs surgery tended to be smaller. Patients with head and neck lesions most often underwent Mohs surgery or were treated with a multidisciplinary combination approach (87%).

CONCLUSIONS. Wide local excision with careful pathologic analysis of margins was found to have a very low recurrence rate and was used for the majority of patients with DFSP lesions at the authors' institution. Wide local excision, Mohs surgery, and a multidisciplinary combination approach, selected based on both tumor and patient factors, were capable of achieving very high local control rates in the treatment of DFSP. The evolution of a multidisciplinary approach has provided a level of expertise that no single individual could achieve for the treatment of the full spectrum of DFSP lesions at the authors' institution. *Cancer* 2004;100:1008-16. © 2004 American Cancer Society.

KEYWORDS: skin neoplasms, dermatofibrosarcoma protuberans (DFSP), Mohs surgery, neoplasm, local recurrence.

Dermatofibrosarcoma protuberans (DFSP) is a rare dermal soft tissue sarcoma that is characterized by progressive local growth and a propensity for extensive subclinical involvement and local

recurrence.¹ Wide excision with histologically negative margins is the cornerstone of treatment, but relatively high recurrence rates are described in the literature,¹⁻⁴ especially for DFSP lesions located on the head and neck.^{5,6} Despite the achievement of "negative" margins, it is believed that the high recurrence rate is due to the fact that large portions of the true margins are not evaluated by standard histologic processing, combined with the difficulty identifying the occult, finger-like projections that are characteristic of DFSP and are responsible for tumor recurrence. An extensive review of the literature published in 1996 found a mean recurrence rate of 18% (range, 0-60%) after treatment with wide local excision in 15 studies, with a total recurrence rate of 20% among all reviewed patients (100 recurrences of 489 reviewed patients).⁷ In contrast, a review of the mean recurrence rate in published series of patients who underwent Mohs surgery for DFSP was 0.6% (range, 0.0-6.6%). The total recurrence rate for Mohs surgery was 1.6% (1 recurrence of 64 reviewed patients).⁷ The limitations of available review articles are apparent: Many individual reports in large reviews are comprised of small, retrospectively reviewed cohorts of patients who were treated over many years with relatively short follow-up intervals. Lesion selection bias also may be present in some series, with the most aggressive lesions usually excised with wide local excision. Larger series of patients who were treated with modern approaches and were followed for an adequate length of time clearly are needed.

We previously described a technique for wide excision that was associated with excellent local control rates equivalent to the rates reported with Mohs surgery.⁸ As our experience with this disease has grown, we have focused increasingly on the proper selection of patients for wide excision or Mohs surgery and also have relied on the combination of both techniques when necessary. The purpose of the current study was to review the experience with Mohs surgery, wide excision, and a combination approach in the contemporary treatment of patients with DFSP at a single academic institution over the past 10 years. Primary endpoints included the ability to extirpate the DFSP lesion completely, the tumor recurrence rate, and the need for skin grafts or local tissue flaps. Additional objectives included defining surgical practice patterns at the study institution, where multidisciplinary management of cancer is standard practice.

MATERIALS AND METHODS

Approval for the current study was obtained from the University of Michigan Institutional Review Board for Medicine. Data were collected retrospectively from

the Cancer Registry and from medical records and were entered into deidentified research records. Sixty-nine patients who were seen at the study institution with DFSP from January 1991 through December 2000 were identified. Seven patients were excluded for the following reasons: four patients underwent surgery at an outside institution and received only postoperative care at the study institution, and it was determined that three patients had other histologic types of sarcoma and not DFSP (patients who had tumors with unequivocal evidence of fibrosarcomatous transformation from a preexisting DFSP were excluded from this review). The 62 remaining patients were separated into 3 treatment groups based on the surgical modality(s) used to treat the DFSP lesion: Mohs surgery alone, wide local excision alone, or a combination approach. The goal for each modality was a histologically negative surgical margin. If negative surgical margins could not be achieved despite maximal tissue excision, then postoperative radiation was used.

Mohs Surgery

The standard technique for Mohs surgery was performed as follows. The lesion/scar site was outlined. Local anesthesia was obtained using 1% lidocaine with epinephrine at a dilution of 1:400,000. All macroscopic tumor was excised/debulked in patients with remaining clinical lesions. Excision with 2-5-mm margins was made around the debulking defect. If only a biopsy scar was present (no clinical lesion), the scar was excised with 2-5-mm margins peripheral and deep to the initial excision/biopsy scar. Tissue was divided into multiple specimens, which were mapped and color coded for precise anatomic orientation, and sent to the Mohs histotechnician for horizontal frozen sectioning. The technique for processing of Mohs frozen sections differed from that used for standard frozen sections, with the production of high-quality slides that had complete tissue margin control, including adipose and fascia. Slides were interpreted by a Mohs surgeon with experience in histopathologic interpretation of DFSP. If any slides revealed positive margins, the patient then returned to the operative suite and underwent further excision of the precise positive area(s) with 2-5-mm margins. The process continued until tumor free margins were obtained. If the tumor extended deeply into muscle, then the procedure was terminated, and the patient was referred for wide excision under general anesthesia. Immunohistochemistry was not used routinely.

Wide Excision

The surgical technique for wide excision first involved accurately identifying the periphery of DFSP lesion

with careful inspection and palpation. Depending on the location of the lesion and the ability to achieve primary closure, a minimum margin of 1 cm and a margin not > 2 cm was measured and marked from the periphery of the lesion, creating an ellipse. A diamond-shaped incision was fashioned to incorporate the entire ellipse, so that the peripheral margins of the surgical excision formed straight lines, facilitating processing of the specimen for total circumferential pathologic evaluation. In some patients, intraoperative frozen sections were taken from suspect margins; if any margin was identified as positive on an intraoperative frozen section, then excision margins of at least 1 additional cm were redrawn at the positive margin. In all lesions, histopathologic analysis after fixation in formalin involved examination of the entire peripheral margin by "shaving" each diamond-edge peripheral margin of the resection and mounting it on one or more microscope slides. Any focus of DFSP identified in any edge section was considered to represent a positive margin requiring reexcision or other additional therapy. Horizontal sections of the deep margin were obtained routinely for complete evaluation of that margin. Finally, conventional transverse "bread-loaf" sections of the remainder of the specimen were taken to evaluate for the presence of residual neoplasm adjacent to and extending from the original biopsy site. Immunohistochemical staining with anti-CD34 antibodies was used to evaluate any equivocal margin sections. Resections always included the investing fascia of the underlying muscle and selectively included the muscle itself if the tumor encroached on or penetrated the fascia. Wounds were closed primarily whenever possible, avoiding extensive undermining of adjacent tissues. A homograft was applied to wounds that could not be closed primarily, followed by definitive skin grafting once permanent pathology revealed a histologically negative margin. Reexcision using the same technique was performed if margins were identified as positive on histopathologic evaluation of permanent sections, unless the location of the lesion was such that no further tissue could be excised without incurring prohibitive morbidity or cosmetic deformity.

Combination Approach

A multimodality combination approach was employed whenever it was suspected or proven that a single-modality approach was inadequate to achieve the two goals of complete tumor excision and satisfactory reconstruction of the defect. Examples include the following: 1) In patients who had evidence of tumor invasion deep to the fascia discovered during Mohs surgery, a wide reexcision taking small addi-

tional peripheral margins and more extensive deep margins was conducted using the technique for wide excision described above. 2) In patients who had positive deep margins despite maximal soft tissue excision by either Mohs surgery or wide excision, postoperative radiation was used after soft tissue reconstruction. 3) In patients who had tumor encroachment upon vital structures, such as the orbit or lip, the lesions were approached jointly with Mohs surgery and surgical subspecialist(s) with operative expertise in the involved anatomic region. 4) In patients with large DFSP lesions located in a cosmetically sensitive region, such as the breast, or with clinically very ill-defined boundaries, the concern was that wide excision either would require multiple iterations or would remove more normal tissue than was optimal. Therefore, a staged "square" technique was used first to define tumor free peripheral margins of the lesion using permanent section, total peripheral margin control.^{9,10} The "square" technique represents a staged approach. The purpose of the first stage was to define the peripheral margins by excision of a thin (2–3 mm) strip of peripheral tissue, usually starting 1 cm beyond the visible edge of the clinical lesion, using a 2-bladed scalpel with 2–3 mm spacers between the 2 blades. This stage was performed in a treatment room with local anesthesia. The thin peripheral strip wound was sutured closed. The excised tissue strip representing the circumferentially sampled peripheral margins was pinned to Styrofoam to prevent tissue rolling, placed in formalin, and processed using standard vertical permanent sections for complete peripheral margin examination. If it was found that any margins contained foci of tumor, then another strip around the area of positivity was excised. The process was repeated until all peripheral margins were free of tumor. Once tumor free peripheral margins were defined, excision of the central island was performed to obtain deep margin clearance, and the wound was reconstructed.

Clinical data regarding patient age, gender, clinical lesion size and location, prior treatment, details of the operative therapy, mode of wound closure, total number of procedures required for tumor extirpation, and wound closure type were recorded. The size of the postoperative defect was defined as the wound dimensions recorded immediately prior to definitive closure. Follow-up was calculated from the date of initial procedure at the University of Michigan until last contact. Follow-up was updated in March, 2003. The last contact with four patients (one patient in the Mohs surgery group, two patients in the wide excision group, and one patient in the multidisciplinary group) was at the initial postoperative follow-up appoint-

TABLE 1
Baseline Patient Characteristics Listed According to Surgical Treatment

Characteristic ^a	No. of patients			Total	P value
	Mohs surgery	Wide local excision	Multimodal therapy		
Presentation					0.69
Primary	10	34	7	51	
Recurrence	1	8	2	11	
Gender					0.45
Female	7	28	4	39	
Male	4	14	5	23	
Age					0.52
< 50 yrs	8	30	6	44	
> 50 yrs	3	12	3	18	
Average (yrs)	38	42	45	42	
Range (yrs)	6–58	21–83	21–61	6–83	

^a There were 63 dermatofibrosarcoma protuberans lesions identified in 62 patients.

ment; these patients were considered lost to follow-up. Independent variables were compared using the Fisher exact test or the chi-square test, whereas continuous variables were assessed with the Student *t* test, the Wilcoxon rank-sum test, or an analysis of variance.

RESULTS

General Characteristics

Sixty-three DFSP lesions were identified in 62 patients. One patient had synchronous lesions located on the left posterior shoulder and right flank. The average age was 42 years (range, 6–83 years). Thirty-nine patients (63%) were female and 23 patients (37%) were male. Fifty-one of 62 patients (82%) presented at the time of their initial diagnosis of DFSP, and 11 patients (18%) presented with a locally recurrent lesion after prior surgery. Forty-eight lesions (76%) were located on the trunk or extremity, and 15 lesions (24%) were located on the head and neck. Complete tumor extirpation was achieved by Mohs surgery alone in 11 patients, by wide excision alone in 42 patients (43 lesions), and with a multidisciplinary combination approach in 9 patients. Baseline characteristics are listed in Table 1.

Mohs Surgery

Definitive surgical therapy was attempted by Mohs surgery in 13 patients. There was evidence of tumor extension deep to the fascia in two patients who underwent further treatment with wide excision. Complete tumor extirpation was achieved by Mohs surgery in 11 patients. Ten patients presented with primary

lesions, and 1 patient had a recurrent lesion. Mohs surgery was used more commonly for head and neck lesions (seven patients) than for trunk or extremity lesions (four patients). DFSP lesions that were excised completely by Mohs surgery generally were smaller compared with the lesions that were treated with wide excision or the combination approach, with an average area of 5.3 cm². The operative defect after Mohs surgery was 21.7 cm². The average number of Mohs stages performed was 2.4, with a range of 1–5 stages. A subsequent surgical procedure was performed to reconstruct 5 of the 11 Mohs defects with a skin graft or local flap. One patient was lost to follow-up. There were no recurrences noted in the remaining 10 patients over a median follow-up of 5.2 years.

Wide Excision

Definitive surgical therapy was attempted by wide excision in 44 patients. Two patients had positive deep surgical margins despite maximal soft tissue excision and were treated further with postoperative radiation. Complete tumor extirpation was achieved by wide excision in 42 patients. One patient had two DFSP lesions, and each was excised completely. Thirty-four patients presented with a primary lesion, whereas 8 patients had a recurrent lesion. Wide excision was used more commonly for trunk and extremity lesions (*n* = 41 patients) than for head and neck lesions (*n* = 2 patients). The average size of DFSP lesions that were treated by wide excision was 14.8 cm². The operative defect after wide excision was 63.4 cm². A subsequent surgical procedure was performed to reconstruct 13 of 43 defects with a skin graft or tissue advancement flap. Two patients were lost to follow-up. No recurrences were noted in the remaining 40 patients over a median follow-up of 4.0 years.

Multidisciplinary Combination Approach

A multidisciplinary combination approach was used to treat nine patients. Multiple disciplines participated, including surgical oncology, Mohs surgery, plastic surgery, otolaryngology-facial plastic surgery, ophthalmology, oral-maxillofacial surgery, and radiation oncology. Seven patients presented with primary lesions, and two patients had recurrent lesions. The average size of the DFSP lesion was 10.7 cm². The operative defect after the combination approach was 50.3 cm².

Two patients with deeply infiltrative lesions that focally invaded underlying muscle were identified using a Mohs surgical approach. This level of invasion was not suspected preoperatively. The lesions were located on the upper back (2.5 cm × 4.5 cm) and the anterior thigh (3.0 cm × 2.5 cm). Mohs surgery estab-

lished clear peripheral margins, but positive deep margins through muscle were present. These patients underwent staged wide excision under general anesthesia with appropriate resection of the involved muscle, and the wounds were closed primarily.

Two patients had positive surgical margins after undergoing wide excision despite maximal attempts at complete excision. Their lesions were located on the cheek (4 cm × 6 cm) and the posterior scalp (2 cm × 2 cm). Both tumors extended to bone. After maximal soft tissue resection, the defects were reconstructed surgically followed by postoperative radiation. Both patients remained free of disease at last follow-up (7.4 years and 10.1 years, respectively).

Two patients presented with DFSP lesions that were recognized preoperatively to have encroached on vital head and neck structures. In both patients, Mohs surgery was used in concert with subspecialty surgeons to extirpate the tumor completely and to preserve tissue function maximally. The first patient had a 5 cm × 5 cm lesion involving the left upper lip and cheek with extension through the lip to the underlying mucosa. Peripheral margins were established with Mohs surgery followed by a through-and-through resection of the cheek and upper lip the next day to obtain clear deep margins. Definitive reconstruction was performed with a pedicle transfer of the temporalis muscle and placement of a bilobed thoracocervical fascial transposition flap. The second patient had a 2 cm × 3.5 cm DFSP lesion located on the left brow with focal extension onto the orbital septum and eyelid. In this patient, Mohs surgery and a specialist from the Ophthalmology Service established the peripheral and deep margins on the brow, forehead, and orbital septum. The Ophthalmology specialist resected the deep orbital septal and eyelid involvement in concert with the Mohs surgeon. Specialists from the Ear-Nose-Throat and Ophthalmology Services jointly reconstructed the soft tissue defect on the forehead and eyelid with a full-thickness skin graft.

Finally, three more patients had large, clinically ill-defined DFSP lesions located in cosmetically sensitive regions. The tumors were located on the scalp (5.5 cm × 7 cm), the breast (3 cm × 3 cm), and the anterior chest wall (10 cm × 3.5 cm). The *square* technique was used first to establish the extent of excision required to achieve negative peripheral margins prior to definitive wide excision.^{9,10} The wide excision was carried down to, and included, the superficial investing fascia on the breast and chest wall and the periosteum on the scalp.

A subsequent surgical procedure was performed to reconstruct five of the nine defects with a skin graft or tissue advancement flap. No patients were lost to

TABLE 2
Overall Final Management versus Planned Management for All Three Treatment Methods

Treatment method	No. of patients	
	Initial approach	Final treatment
Mohs surgery	13	11
Wide excision	44	42
Multimodal	5	9

follow-up. No recurrences were noted in this group of patients over a median follow-up of 5.4 years.

Comparison of Mohs Surgery, Wide Excision, and a Multidisciplinary Approach

Eleven of 13 lesions (85%) that were approached initially with Mohs surgery were cleared histologically. The remaining 2 lesions (15%) subsequently were cleared surgically with a wide excision. Forty-two of 44 lesions (95%) that were approached initially with wide local excision were cleared histologically. Two patients (5%) received postoperative radiation for positive margins after undergoing maximal soft tissue excision. For another five patients, it was planned at the outset to work in concert with Mohs and another service (two patients) and the square technique followed by wide excision (three patients). All five of those patients achieved tumor free margins. Final patient management (vs. planned management) is listed in Table 2.

Thus, complete DFSP tumor extirpation was achieved with Mohs surgery in 11 patients, with wide excision in 42 patients, and with a multidisciplinary combination approach in 9 patients. A comparison of results is listed in Table 3. The 15 head and neck DFSP lesions were approached more commonly with Mohs surgery (7 of 11 lesions) and with the multidisciplinary approach (6 of 9 lesions) compared with wide excision (2 of 43 lesions; $P < 0.01$). The 48 trunk and extremity lesions were approached more often with wide excision (41 of 43 lesions) compared with Mohs surgery (4 of 11 lesions) or the multidisciplinary approach (3 of 9 lesions; $P < 0.01$). DFSP lesions that were treated by Mohs surgery (5.3 cm²) were smaller compared with lesions that were treated with a multidisciplinary approach (10.7 cm²) or with wide excision (14.8 cm²; $P = 0.02$). Thus, the operative defect after Mohs surgery (21.7 cm²) was predictably smaller compared with the defects after either the multidisciplinary combination approach (50.3 cm²) or wide excision (63.4 cm²), although the percent difference between the initial size and postoperative defect (309% Mohs vs. 371% combination vs. 328% wide excision; $P = 0.69$) was similar

TABLE 3
Comparison of Surgical Treatments: Mohs Surgery, Wide Excision, and Multidisciplinary Approach

Characteristic	Mohs surgery	Wide local excision	Multimodal therapy	P value
Average size of DFSP lesions (cm ²)	5.3	14.8	10.7	0.02 ^a
Postoperative defect (cm ²)	21.7	63.4	50.3	—
Postoperative change in lesion size (%)	309	328	371	0.69
Head and neck location				
No. of patients	7/15	2/15	6/15	< 0.01 ^b
%	47	13	40	—
Trunk and extremity location				
No. of patients	4/48	41/48	3/48	< 0.01 ^b
%	8	86	6	—
Need for skin graft or tissue flap				
No. of patients	5/11	13/43	5/9	0.28
%	45	30	56	—
Recurrence of DFSP after treatment	0	0	0	—
Length of follow-up (yrs)	5.2	4.0	5.4	—

DFSP: dermatofibrosarcoma protuberans.

^a Statistical difference between Mohs surgery and wide local excision and between Mohs surgery and multimodal therapy.

^b Statistical difference between wide local excision and Mohs surgery and between wide excision and multimodal therapy.

(Table 3). The need for skin grafts or tissue advancement flaps was not statistically different between patients in the Mohs surgery group (5 of 11 patients; 45%), patients in the multidisciplinary approach group (5 of 9 patients; 55%), and patients in the wide excision group (13 of 43 patients; 30%; $P > 0.05$). No recurrences have been detected in any group with a mean 4.4 years follow-up.

DISCUSSION

DFSP lesions account for < 0.1% of all malignancies and for approximately 1% of all soft tissue sarcomas.^{11–13} The clinical behavior of DFSP is characterized by progressive local growth and a propensity for extensive subclinical involvement and local recurrence.¹ This neoplasm is most common in adults in their third to fifth decades¹⁴ but has been reported in all age groups, including children, infants, and newborns.^{15,16} Lesions are located most commonly on the trunk or proximal extremities but may develop anywhere on the skin.¹⁷ Clinically, these neoplasms usually present as a raised, indurated, asymptomatic plaque that may be any combination of blue, red, and brown or flesh colored.¹⁸ DFSP tumors usually are not fixed to deeper structures, unless they are long-standing or recurrent.¹³ This lesion rarely metastasizes, but

when it does, it usually is preceded by multiple recurrences.¹² DFSP spreads by a hematogenous route, with the lung the most common site of metastasis. Regional lymph nodes rarely are involved.^{14,19}

Microscopically, DFSP is highly cellular and is comprised of radially arranged spindle cells that demonstrate immunoreactivity to CD34.²⁰ The most salient histologic characteristic of DFSP is at the periphery of the tumor, where tentacle-like projections of neoplastic cells extend between normal collagen bundles, through adipose septae, and through fascia and muscle.^{7,21} The extensions cannot be detected macroscopically and can be difficult to appreciate histologically, because they may mimic normal adjacent dermal tissue and/or connective tissue septae.¹ Accurate histologic identification of the extent of the tumor can be hindered further because of fibrosis and other post-biopsy changes. Complete extirpation of DFSP, which is tantamount to cure because of the near-total absence of metastases, depends on histologic identification and excision of these finger-like projections that radiate out from the tumor mass. It is believed that the failure to eliminate these occult projections is the reason for the high recurrence rate of DFSP after patients undergo seemingly complete surgical excision.¹³

The recurrence rate after DFSP excision varies widely in the literature.⁷ Histologic subtype, high mitotic index, cellularity, size, location on the head and neck, and recurrent lesions are factors reportedly associated with higher recurrence rates.^{1,5,22} Clearly, the most important factor for local control is obtaining negative surgical margins.¹ The environmental framework that underlies our institutional approach to DFSP is multidisciplinary. The full spectrum of host and tumor factors and treatment options continues to evolve based on multidisciplinary sharing of knowledge, collegiality, and close clinical proximity with frequent communication and daily interaction.

Using our multidisciplinary environment and knowledge gained from a Mohs-focused histopathologic approach, our Surgical Oncology Service developed a wide local excision method with complete peripheral margin assessment.^{8,21} Our wide excision margin assessment and tissue processing are different from standard, bread-loaf tissue processing. The margin control described herein represents a more complete margin assessment (circumferential and deep), is more labor intensive for pathology, and is performed by a dermatopathologist with expertise and experience in DFSP. The technique employed in the patients who underwent wide excision involved excising a diamond-shaped surgical specimen.⁸ Each diamond edge was examined entirely for the presence of tumor (“shaved” peripheral margins) along with hor-

izontal sections of the deep margin. This approach facilitates complete peripheral margin evaluation and focused reexcisions directed at the site of positive margins if necessary. The deep margin consists of excision through fascia with a small amount of underlying muscle in most lesions. Intraoperative frozen sections occasionally were used to examine the most suspect peripheral margin as a guide during the initial excision under general anesthesia. Final margin assessment was obtained based on the permanent sections. Excision was continued until all margins were free histologically of tumor in 61 of 63 lesions. Negative histologic margins could not be obtained in two lesions despite maximal soft tissue resection; in those patients, postoperative radiation therapy was employed.

In this clinical series, there were no DFSP recurrences identified after wide excision, Mohs surgery, or combination treatment over an average follow-up of 4.4 years. Sixty-two patients were treated at a single institution over a 10-year period, representing what to our knowledge is one of the largest reported contemporary series on the surgical management of DFSP. Objective comparisons are made between the three treatment groups and are provided as a guide for the successful surgical management of DFSP developed at our institution. To our knowledge, our clinical approach reported herein is novel, fueled by our Cancer Center's commitment to a multidisciplinary approach to cancer of all types. The success in treatment of DFSP reported herein undoubtedly is the result of many factors. Perhaps chief among these is the commitment by multiple specialists to examine the advantages and disadvantages of varying approaches, each tailored selectively to each individual patient and DFSP lesion.

Based on our center's experience and surgical expertise, certain treatment trends have developed that are reflected in the data presented in the current series. The majority of patients with head and neck lesions underwent Mohs surgery or a combination approach. Small lesions located in a cosmetically sensitive region were treated with Mohs surgery alone; whereas large, cosmetically sensitive lesions were treated with a combination approach in effort to optimize the reconstruction. Based on anatomic location alone, it has been reported that head and neck DFSP lesions have the highest recurrence rate, believed to be due to the surgeon's reluctance to take a generous margin.^{5,22} The Mohs surgery technique is ideally suited for extirpation of tumors in these cosmetically sensitive regions. Mohs surgery is a proven method with extremely high local control rates and with maximal conservation of normal tissue for the treatment

of nonmelanoma skin malignancies.²³ Mohs surgery spares uninvolved tissue to the greatest degree possible due to the technique of tumor mapping and focused reexcisions.^{21,24,25} This approach uses a highly specialized frozen sectioning technique that provides accurate determination of the margins compared with traditional vertical pathology sectioning.⁷ Tumor mapping facilitates precisely aimed reexcisions, thus preserving normal tissue.²⁶

However, Mohs surgery does have limitations and complete excision with tumor free margins is not always attainable. Drawbacks to Mohs surgery for DFSP include the need for considerable training and a specialized team. Multiple stages usually are required. Particularly for larger lesions, the time and labor intensity involved to process a high number of Mohs frozen sections is disadvantageous. The average number of stages performed in the Mohs micrographic surgery group (comprised of smaller lesions than the wide excision group) was 2.4 stages, with a range of 1–5 stages. Approximately half of the Mohs surgery patients in the current series also required a secondary service to subsequently close the postoperative defect. Mohs surgery is performed with local anesthesia and therefore has limitations with respect to size for total amount of local anesthesia that can be used safely and patient comfort for deep lesions through fascia.²⁷ The reason Mohs surgery alone failed to achieve a negative margin in two patients was tumor extension into the deep tissues. These two patients were treated readily with wide excision and primary closure. Thus, our patient selection has evolved to employ Mohs surgery for patients with smaller DFSP lesions. Using these criteria, we were able to achieve the complete excision of DFSP lesions with Mohs surgery alone in 85% of patients over several hours under local anesthesia in a treatment room facility.

In contrast, larger lesions most commonly were approached with wide excision. Technically, the removal of bulkier lesions is accomplished more easily with wide excision in the operating room under general anesthesia. Wide excision also was favored in the majority of trunk and extremity lesions. DFSP lesions located on the trunk and extremity are approached easily with wide excision. Wide excision of DFSP is typically a short procedure, and general anesthesia greatly facilitates patient comfort. The majority of postoperative defects of the trunk and extremity also could be closed primarily, avoiding subsequent reconstructive procedures. Thus, our patient selection has evolved to use wide excision for extensive lesions and for lesions located on the trunk and extremity, given the high success in complete tumor extirpation.

During wide excision, the wound was closed pri-

marily with only minimal undermining (if any) or was covered with homograft if primary closure was not possible technically without more extensive undermining. After a final pathology review (including immunostaining for CD34 for equivocal results) and confirmation of negative margins, definitive wound closure was performed for wounds that were not closed primarily. This approach allows for directed reexcision should the final pathology reveal a positive margin, and it eliminates opening new tissue planes for tumor implantation. Using this approach, the initial margins of resection employed were never > 2.0 cm and usually measured 1.0–1.5 cm. Further excisions were performed only for positive margins. A negative margin could not be obtained in two patients in the wide excision group. Both of those lesions invaded the periosteum, and it was the deep margin that was limiting, not the peripheral margin. Instead of proceeding with a disfiguring radical procedure involving bone resection and reconstruction, postoperative radiation was used successfully as described in the literature.^{28–30}

The importance of not undermining widely or otherwise disturbing uninvolved planes deserves emphasis. One patient who initially underwent narrow excision at an outside institution was closed with bilateral rotation flaps, but the final pathology report demonstrated positive margins. The initial tumor was described as “small,” measuring < 1 cm. No clinical residual lesion was noted when the patient was referred for reexcision to achieve clear margins. Careful pathologic review clearly identified extension of tumor through the entire undermined tissue plane beneath the flaps, with no tumor noted in the overlying superficial adipose or dermal tissues.

Patients with DFSP lesions that encroached on vital structures or patients with large lesions in cosmetically sensitive locations were treated with a combination surgical approach. These patients were identified preoperatively, and a multidisciplinary plan was devised for tumor extirpation and soft tissue reconstruction. Mohs surgery was used in concert with surgical subspecialists to establish clear peripheral margins and to provide careful histopathologic assessment of the deep margins. The multidisciplinary approach that was used for the patient who had a brow-eyelid lesion preserved eyelid function maximally and avoided more radical surgery, such as an eye enucleation, that otherwise would have been required to completely resect the lesion, which extended into the orbital septum. Similarly, the multidisciplinary approach employed for the three patients who had large DFSP lesions located on the breast, scalp, and chest wall maximized tissue preservation, providing optimal

soft tissue reconstruction. However, it should be noted that coordination of the multiple surgical disciplines can be difficult, an increased number of operative procedures may be required, and the cost is significant for the combination approach. Thus, our patient selection is limited to DFSP lesions that encroach on vital structures or large DFSP lesions in cosmetically sensitive locations.

Wide local excision with careful pathologic analysis of margins appears to have a very low recurrence rate and is used for the majority of patients (68%) with DFSP lesions at our institution. Wide local excision, Mohs surgery, and a multidisciplinary combination approach with lesion and patient selection all appear to be efficacious in the treatment of DFSP. A multidisciplinary team approach with interdisciplinary education and collaboration has provided a level of expertise that no single individual could provide for the treatment of the full spectrum of DFSP lesions at our institution.

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