Annals of Surgical Oncology, **7**(9):696–704
Published by Lippincott Williams & Wilkins

Dermatofibrosarcoma Protuberans of the Head and Neck

Alexander Stojadinovic, MD, Howard M. Karpoff, MD, Cristina R. Antonescu, MD, Jatin P. Shah, MD, Bhuvanesh Singh, MD, Ronald H. Spiro, MD, Wilson Dumornay, MD, and Ashok R. Shaha, MD

Background: Dermatofibrosarcoma protuberans (DFSP) of the head and neck is a rare, locally infiltrative, low-grade sarcoma. This study defines the clinical behavior of DFSP, evaluates the role of frozen section analysis, and identifies factors that predict local control.

Methods: Hospital records and pathological slides were reviewed for 33 patients with pathologically confirmed head and neck DFSP treated at Memorial Sloan-Kettering Cancer Center between 1964 and 1999. Factors were analyzed by using Fisher's exact or χ^2 tests.

Results: For 21 primary and 12 recurrent patients, median age and tumor size at presentation was 39 years and 2.0 cm, respectively. Thirty-two (97%) patients were alive at a median follow-up of 82 months. Three patients recurred locally, all with smaller than 2-cm resection margins. Deep tumors were more likely to have a margin-positive resection than superficial lesions ($P = .03$). Gross margin 2 cm or more was a significant predictor of a negative histological margin ($P < .001$). There was a trend toward improved recurrence-free survival for tumors treated with wide ($\geq$2 cm) margin resection ($P = .059$). Accuracy, sensitivity, specificity, and false negative rates of frozen section were 80%, 43%, 100%, and 57%, respectively.

Conclusions: Wide margin resection of head and neck DFSP predicts negative histological margins and impacts favorably on local recurrence-free survival. Frozen section analysis does not assess resection margins accurately.

Key Words: Dermatofibrosarcoma protuberans—Head and neck—Sarcoma.

Dermatofibrosarcoma protuberans (DFSP), an indolent, low-grade sarcoma recognized for its progressive, locally infiltrative nature, accounts for less than 5% of adult soft tissue sarcomas and fewer than 1% of all malignant tumors of the head and neck.[1,2] It typically presents as an asymptomatic indurated plaque or firm, cutaneous nodule in early to middle adult life.[3–5] The most common site of origin is the skin of the trunk (50% to 60%) followed by the proximal extremities (20% to 30%) and the head and neck (10% to 15%).[6] The seemingly benign appearance of this tumor can be misleading to patients and physicians alike. For this reason, treatment frequently is delayed and all too often inadequate. The time interval from apparent onset to initial resection may exceed 10 years in more than 50% of cases.[4] The extremely rare presentation of DFSP of the head and neck is underscored by the fact that these tumors represent only 7% of head and neck sarcomas.[7]

It is generally agreed that the detailed clinicopathological description by Darier and Ferrand in 1924 established DFSP as a distinct disease process.[8] In 1925, Hoffman coined the term *dermatofibrosarcoma protuberans*, which remains the preferred appellation for this disease.[9] Taylor and Helwig emphasized the importance of a conspicuous microscopic architectural finding in their study of 115 cases: the arrangement of spindle-shaped tumor cells in a "cartwheel or whirligig" pattern.[3] This finding subsequently has been emphasized as diagnostically important and has been confirmed by other investigators as histologically specific for DFSP.[4] Despite its distinct microscopic presentation, the histogenesis of this tumor remains undefined.

Received February 14, 2000; accepted July 17, 2000.

From the Head and Neck Service, Department of Surgery (AS, HMK, JPS, BS, RHS, WD, ARS) and Department of Pathology (CRA), Memorial Sloan-Kettering Cancer Center, 1275 York Ave., New York, New York.

Presented at the 53rd Annual Meeting of the Society of Surgical Oncology, New Orleans, Louisiana, March 16–19, 2000.

Address correspondence to: Ashok R. Shaha, MD, Head and Neck Service, Memorial Sloan-Kettering Cancer Center, 1275 York Ave., New York, NY 10021; Fax: 212-717-3302.

The mainstay of treatment for DFSP is surgical resection. Resection with conservative margins has led to unacceptable local tumor control, with published local recurrence rates varying from 26% to 60%.[7,10–15] To improve local control after excision, many authors have recommended wide excision with more than 2 cm grossly uninvolved margin of skin and underlying deep fascia. This treatment approach has reduced local recurrence rates to a mean of about 20% (Table 1).

Microscopic examination of the tumor's periphery reveals multiple finger-like projections of neoplastic cells that can extend laterally or deeply into underlying fascia and muscle as much as 3 cm or more from the main lesion.[3,16] The high local recurrence rate after resection has been attributed to failure to excise these clinically inapparent projections of tumor.[17] Moreover, any micropathological sectioning technique that fails to examine all lateral margins and the deep aspect of the tumor inevitably will overlook these neoplastic extensions. This explains why the role of intraoperative frozen section analysis in the treatment of DFSP remains undefined. The present study was designed to review the Memorial Sloan-Kettering Cancer Center (MSKCC) experience with head and neck DFSP with specific focus on frozen section analysis and the impact of margin status on local recurrence.

TABLE 1. *Local recurrence rates after treatment for dermatofibrosarcoma protuberans*

Treatment	Author	Year	N	Follow-up (y)	Margin of excision	Recurrence rate (%)
Surgery—local excision						
	Pack & Tabah[17]	1951	39	0.5–20	Wide	18
	Taylor & Helwig[3]	1962	98	1–17	Wide	49
	Burkhardt et al.[4]	1966	21	>5	Wide	33
	McPeak et al.[5]	1967	82	3–15	>3 cm	10
	Longhin[11]	1967	44	1–11	Not stated	32
	Tamoney[25]	1971	12	1–30	Wide	25
	Bendix-Hansen et al.[22]	1983	7	5–13	≥3 cm	0
	Bendix-Hansen et al.[22]	1983	11	5–13	<3 cm	73
	Barnes et al.[13]	1984	15	1–23	Not stated	53
	Waldermann & Hagedorn[31]	1985	13	1–7	4–5 cm	23
	Petoin et al.[24]	1985	96	1–15	>4 cm	6
	Roses et al.[14]	1986	27	~7.3 (mean)	≥2 cm	24
	Roses et al.[14]	1986	21	~7.3 (mean)	<2 cm	41
	Rutgers et al.[20]	1992	14	2–28	>2 cm	0
	Mark et al.[7]	1993	15	9.5 (median)	Not stated	60
	Gloster et al.[55]	1996	39	0.1–14	2.5–5 cm	13
	Current series	2000	16	0.6–24	<2 cm	19
		2000	17	0.6–24	≥2 cm	0
Surgery—Mohs micrographic technique						
	Mikhail & Lynn[46]	1978	2	>5	Negative	0
	Mohs[47]	1978	7	>5	Negative	0
	Peters et al.[48]	1982	1	3.5	Negative	0
	Hess et al.[49]	1985	1	1.5	Negative	0
	Robinson[50]	1985	4	5	Negative	0
	Hobbs et al.[18]	1988	10	1.25–7.6	Negative	0
	Hobbs & Ratz[51]	1988	1	2	Negative	0
	Weber et al.[52]	1988	1	0.5	Negative	0
	Rockley et al.[44]	1989	1	1.5	Negative	0
	Goldberg & Maso[53]	1990	1	1.5	Negative	0
	Parker & Zitelli[54]	1995	20	0.25–8.75	Negative	0
	Gloster et al.[55]	1996	15	0.5–8	Negative	7
Surgery + radiotherapy						
	Marks et al.[56]	1989	7	1.5–5	Margin positive	14
	Suit et al.[58]	1996	12	6.7 (median)	Margin positive	17
			3	3.9–9.25	Margin ≤0.3 cm	0
	Ballo et al.[57]	1997	6	7.4 (mean)	Margin positive	0
			12	9.2 (mean)	Margin <3 cm	0
	Current series	2000	4	8–15	Margin positive	25
Radiotherapy alone						
	Marks et al.[56]	1989	3	2–2.75	Gross disease	0
	Mark et al.[7]	1993	1	3	Gross disease	0
	Suit et al.[58]	1996	3	7–9	Gross disease	0
	Ballo et al.[57]	1997	1	1.8	Gross disease	100

PATIENTS AND METHODS

The head and neck database was used to identify 26 patients with either primary or recurrent DFSP of the head and neck region who underwent resection for DFSP at MSKCC between 1982 and 1999. Seven additional patients treated between 1964 and 1984 were identified from the pathology database. Follow-up information was available for all patients through medical record review or patient contact by telephone or mail.

The head and neck region was defined as the region above the clavicles anteriorly, above the first thoracic vertebral spine posteriorly, and above the shoulder girdle laterally. A primary tumor was defined as a localized lesion previously untreated or biopsied (incisional or inadequate excisional biopsy) before definitive surgical therapy. Local recurrence was defined as clinically, radiologically, or pathologically evident tumor within or contiguous to the previously excised field 3 months or more after primary therapy.

Tumor size was defined as the maximum diameter of the tumor at operation or pathological analysis. Deep tumors were defined as those lesions that required bone or deep muscle resection to achieve a negative margin resection. Extent of resection was classified as wide local excision only with primary closure (WLE) or more extensive resections (WLE+) with or without underlying deep skeletal muscle, bone, or adjacent parotid gland, and local or free myocutaneous flap reconstruction. Surgical treatment decisions generally were based on several factors: (1) review of pathological material obtained from prior resections, (2) assessment of the adequacy of prior resection, (3) location of the tumor, and (4) estimate of surgical margins and the functional and cosmetic impact of re-excision. Resections included underlying bone, deep muscle, or adjacent parotid gland and local skin or free myocutaneous flap reconstruction to achieve a 2-cm or more grossly negative radial surgical margin. Achieving a 2-cm or more deep resection margin was not the objective of treatment, because such an aggressive approach is often not feasible in the head and neck region due to anatomic constraints. For deep margin of resection, the treatment aim was a negative deep histological margin. All patients were treated with surgical resection, and some received adjuvant radiation at the discretion of their surgeon. The technique used to deliver radiation therapy was external beam radiation therapy. Total adjuvant radiation dose ranged from 6000 to 6600 cGy.

Gross resection margins were obtained from pathology reports by subtracting the maximum tumor diameter from the lesser of the two measurements, length or width of the resected specimen, documented at the time of pathological specimen processing. For three of the earlier patients in the series, gross pathological description of the tumor was not available for review. In these cases, gross resection margins were derived from the dictated operative reports. Gross resection margins were classified as narrow (<2 cm) or wide (≥ 2 cm). A complete resection was defined as the absence of any gross residual disease at time of operation. No patient in this study had an incomplete resection that left gross residual disease behind.

Pathological record as well as histopathological slides were reviewed to confirm the histological diagnosis. Frozen section assessment of margin involvement was compared with permanent section evaluation of margins. Twenty patients had intraoperative frozen section analysis. Median number of intraoperative frozen sections was 3 (range, 1–20); typically we evaluated the deep margins and one or more lateral margins of resection. A microscopically negative margin was defined as tumor greater than 1 mm from the inked margin. A microscopically positive margin was defined as tumor present at the inked margin or less than 1 mm from it.

Classical DFSP was diagnosed when histological examination demonstrated that a tumor arose in the dermis that consisted of irregular, interwoven fibrocellular fascicles composed of uniform spindle-shaped cells and variable amounts of collagen, which produced a storiform appearance.[3–6] The fibrosarcomatous variant of DFSP (FS-DFSP) was diagnosed when there was histological evidence of high-grade fibrosarcomatous change in 5% or more of the lesion.[18] The fibrosarcomatous areas have a fascicular, herringbone growth pattern and display increased mitotic activity and cellularity, cytologic atypia, with necrosis or pleomorphism.[19]

Primary study endpoints included time to local recurrence and local recurrence-free survival. The following clinicopathological factors were correlated with study endpoints: (1) patient factors—patient age, sex, presenting symptoms, and time from symptom onset to presentation; (2) tumor factors—presentation status (primary vs. local recurrence), and tumor depth; (3) pathological factors—tumor size, histological subtype, and status of microscopic surgical margins (positive vs. negative); and (4) treatment factors—extent of surgery (WLE or WLE+), and gross resection margin (<2 cm vs. ≥ 2 cm).

Statistical Analysis

We obtained summary statistics by using established methods. Associations between categorical variables were evaluated with the Fisher's exact test. We tested the hypothesis by using the χ^2 test with Yates' correction when variable size or frequency was large enough to

justify its use. Local recurrence-free survival was calculated from the date of surgery. The rate of recurrence was estimated with the Kaplan-Meier method. The univariate influence of prognostic factors on study endpoints was analyzed by using the log-rank test. In all statistical analyses, $P < .05$ was considered statistically significant (JMP, SAS Institute, Inc., Cary, NC).

RESULTS

Our study population included 17 women and 16 men who ranged in age from 24 to 77 years (median, 39 years) at presentation. In 79% of patients, the tumor occurred between the ages of 24 and 50 years. The anatomical distribution of tumors by subsite included 14 scalp, 13 face, and 6 neck lesions. Most had noted a firm skin nodule (Fig. 1) that prompted them to seek medical attention after a time interval that ranged from 1 month to 60 years (median 3 years).

Seven patients received their initial surgical treatment at MSKCC, of whom five had diagnostic biopsies performed elsewhere. Fourteen patients were referred shortly after incomplete excision elsewhere, and 12 presented with recurrent disease after prior local excision elsewhere. Re-excision in the 14 patients with margin involvement after treatment elsewhere revealed residual disease in 13. None of these patients had clinical evidence of regional lymphatic or distant metastatic disease at the time of presentation.

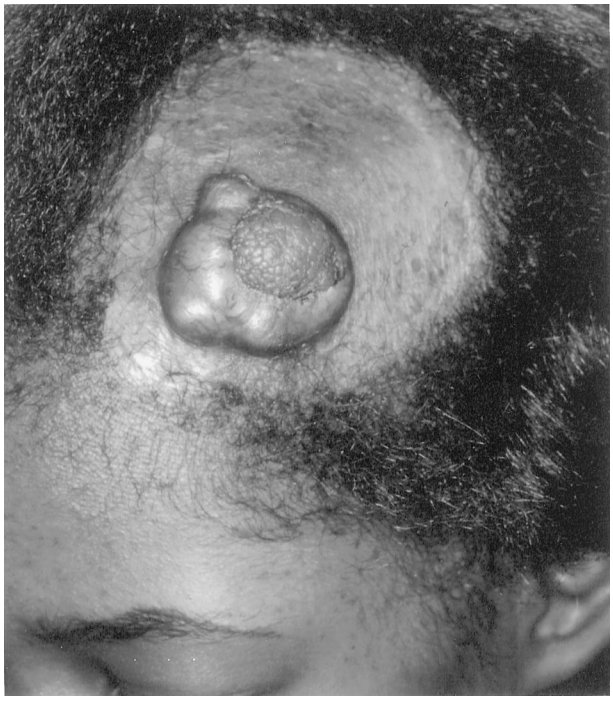

FIG. 1. A characteristic presentation for dermatofibrosarcoma protuberans of the head and neck region in a female patient with a slow-growing, painless, firm, solitary scalp nodule.

Median tumor size was 2.0 cm. For those who had their initial DFSP resection in our hospital, primary tumor size ranged from less than 1 cm to 4 cm. The findings in those patients seen after biopsy or incomplete excision included an indurated scar, an ill-defined subcutaneous nodule in a fibrotic wound, or a discrete firm subcutaneous nodule in proximity to the surgical wound. Of the 12 who presented with recurrence, one patient had microscopic residual disease in a subepidermal fibrotic plaque that involved the lateral canthus of the eye, and the remainder had firm circumscribed, single or multiple, subcutaneous nodules that ranged in size from 0.5 to 8.0 cm. Eight of the 12 patients with recurrent DFSP presented clinically with a palpable firm subcutaneous nodule in a fibrotic scar. The four largest lesions (3.5, 3.5, 6.0, and 8 cm) manifested as multinodular plaques. Pain, ulceration, or bleeding was not associated with any tumors. Twenty-nine patients had histological features consistent with classic DFSP, and 4 had FS-DFSP. Of the patients with FS-DFSP, two each presented with primary and locally recurrent tumors that ranged in size from less than 1 to 2.5 cm after incomplete excision elsewhere.

Patients who presented with primary DFSP were no more likely to have wide margin resection (≥2 cm) than those with recurrent disease ($P = .39$). The relative proportion of patients with negative microscopic margins did not differ between the primary and recurrent tumor groups ($P = .35$). There were 12 deep and 21 superficial tumors. Eight of 12 (67%) deep and 8 of 21 (38%) superficial DFSPs had less than 2 cm gross resection margins. Seven of 12 (58%) deep and 4 of 21 (19%) superficial DFSPs had positive microscopic margins after resection. Deep lesions were statistically more likely to have positive microscopic margin resections than superficial lesions ($P = .03$).

Ten patients had wide local excision with primary wound closure; seven had less than 2 cm gross resection margins, of whom three had negative and four had positive microscopic margins after resection. The remaining three of this group of 10 had wide resection margins resections (≥2 cm) and no microscopic margin involvement. The majority of patients had more extensive resections (23 of 30,70%) defined by wide local excision of the tumor that included parotidectomy or underlying bone resection, with skin graft, advancement flap, or free myocutaneous flap reconstruction. In this group of patients who underwent more extensive procedures, 14 (61%) had both wide (≥2 cm) gross and negative histo-

logical resection margins. Nine patients (39%) in the group that required more than wide local excision alone had less than 2 cm gross margin resections, of whom seven had positive microscopic margins. Resection of considerable tissue volumes was performed in pursuit of local tumor control. Median length, width, and depth of resected tissue specimen were 6.5 cm (range, 2.2–41.1 cm), 4.5 cm (range, 1.0–24.3 cm), and 1.2 cm (range, 0.4–7.5 cm).

No patient (0 of 17) treated with wide local excision (gross margin ≥2 cm) had margin involvement on permanent section analysis. In contrast, those tumors excised with less than 2 cm gross resection margins had positive microscopic resection margins in 69% (11 of 16) of cases. A gross resection margin greater than 2 cm was a significant predictor of a negative histological margin ($P < .001$). Of the three patients with local disease recurrence, all had smaller than 2 cm resection margins. One of these three patients had tumor-free histological margins after resection. There was a trend toward improved recurrence-free survival for DFSP treated with wide (≥2 cm) margin resection; however, the small number of recurrences in this study limits statistical analysis of the predictive effect of gross and microscopic margin on local disease control (Fig. 2).

Four patients in this series received adjuvant external beam radiation therapy (60–66 Gy): a 63-year-old woman with an incompletely excised scalp DFSP in whom satisfactory microscopic margins were not achieved after re-excision, who is alive with no evidence of disease after 8 years of follow-up; a 38-year-old woman with microscopic deep margin involvement after excision of scalp DFSP, who recurred 6 years later with aggressive locally recurrent disease, underwent major resection, and has been disease-free for 8 years; a 39-year-old woman with a deep locally recurrent scalp DFSP and microscopic margin involvement identified after wide excision who remained free of disease 5 years later; and a 44-year-old woman with a large, locally recurrent DFSP of the cheek located in proximity to the medial canthus of the eye treated with local excision with a grossly uninvolved 1-cm margin, but with identified tumor extension to the deep margin of resection. She remains alive with indolent, low-volume, locally recurrent disease, clinically unchanged after 12 years of follow-up.

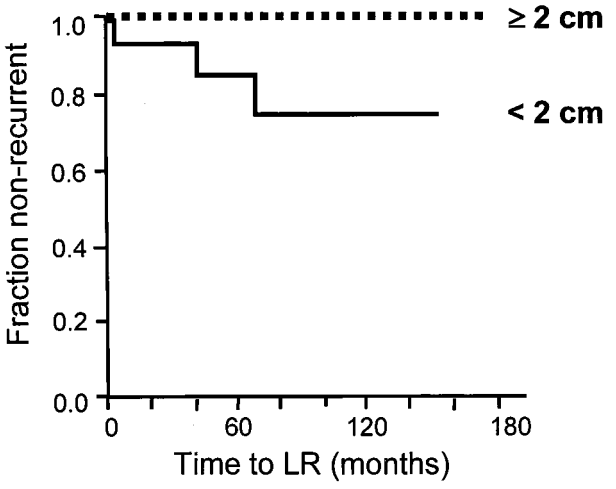

FIG. 2. Product-limit (Kaplan-Meier) local recurrence-free survival in patients with head and neck dermatofibrosarcoma protuberans after surgical treatment with 2 cm or more versus less than 2 cm gross resection margin. $P = .059$ with nonparametric long rank testing. Fraction nonrecurrent = recurrence-free survival.

Twenty patients had intraoperative frozen section evaluation: seven positive and 13 negative permanent sections. Four patients with positive and nine with negative microscopic margins on permanent section did not have frozen section analysis. Of the 17 margins reported as negative on frozen section, permanent section was later reported as positive in four (13 of 17, 76% true negatives). Permanent section confirmed margin involvement in the three patients with positive frozen section margins of resection (three of three, 100% true positives). Of the seven positive permanent section resection margins, frozen section correctly identified three (four of seven, 57% false negatives). Two of the four false negative results were due to specimen sampling error. The sensitivity, specificity, and positive and negative predictive values of frozen section analysis were 43%, 100%, 100%, and 76%, respectively. The false negative rate for frozen section analysis was 57%. Overall, the frozen section diagnosis was accurate in 16 of 20 patients (80%).

Median follow-up was 82 months (range, 7–284). Three patients (9%) had developed local recurrence, two with classic and one with FS-DFSP. No patient in this collective series had developed regional nodal or distant metastases. No patient had died of disease. One died free of disease 82 months after surgical treatment of DFSP. Of the 11 patients who had microscopic positive margins after resection, two had local recurrence. One of these 11 received primary adjuvant radiotherapy and three were treated with combined surgery and radiation for locally recurrent disease. Three of four patients treated with radiotherapy for microscopic margin involvement remain disease free. In addition, seven patients with positive microscopic margins who did not receive radiotherapy are also free of local recurrence.

DISCUSSION

DFSP represents less than 0.1% of all malignancies and 1% of head and neck soft tissue sarcomas.[17,20–22] Ten to 15% of all DFSPs originate in the head and neck region.[3,6,13,17] This disease commonly presents in adults in their third or fourth decade of life. The classic presentation for DFSP of the head and neck region is that of a slow-growing, painless, firm, solitary subcutaneous nodule[3,7,10,13,15,20,23–28] (Table 2).

The diagnosis of DFSP is confirmed only after microscopic evaluation. The multiformity of this tumor in conjunction with its paucity of symptoms contributes to the difficulty in correct diagnosis on the basis of clinical findings alone. Although growth of this tumor is predominantly horizontal, deep fascial, muscular, and bone invasion can be seen with neglected, long-standing, or locally recurrent lesions.[3,5,6,13,17,25,29]

DFSP typically is confined to the dermis and subcutaneous tissues, freely mobile on the fascia. Twelve of 33 (36%) patients had deep tumors, of which more than 50% demonstrated microscopic margin involvement on permanent section analysis. Deep lesions were statistically more likely to have positive microscopic margin resections than superficial lesions ($P = .03$).

Unlike its variable clinical presentation, DFSP has a consistent histological appearance that imparts a significant degree of histopathological specificity to this tumor. Taylor and Helwig identified the prominent histopathological features that reliably differentiated DFSP from other mesenchymal proliferations.[3] They identified a distinctive, invariable spindle cell arrangement with the tumor cells located radially about a central "hub" of collagen or small vascular space, producing a pattern like the spokes of a wheel or whirligig.[3]

Despite the reported proclivity for local recurrence after resection, DFSP rarely metastasizes to regional nodal basins.[33] Similarly, DFSP has low distant metastatic potential. Thirty-seven of 913 (4%) cases of DFSP had distant metastases[4,5,10–12,20,22,24,26,30–32,34–42]. One large study reported a 1% metastases rate; prognostic factors for distant disease failure included fibrosarcomatous variant of DFSP, positive microscopic margin, increased cellularity, high mitotic rate, age more than 50 years, and multiple prior recurrences.[19] In the current study, no patient developed clinically evident nodal or distant metastases.

The most significant factor that predicts outcome for DFSP is extent of resection. In a literature review of 317 patients who underwent conservative excision of DFSP, a mean local recurrence rate of 43% (range, 26% to 60%) was reported.[6] A similar analysis of 489 patients with "wide" or larger than 2 cm gross resection margins demonstrated a reduced rate of local recurrence (mean, 18%; range, 0% to 60%).[6] Adequate initial resection is important because multiple local recurrences predispose to distant disease spread.[4–6,20,43] Moreover, incomplete initial resection can lead to uncontrolled, potentially fatal, locally invasive recurrence.[5,44]

Prognosis is excellent for DFSP after adequate surgical clearance. Overall 5-year survival rates are 93% to 100%.[7] In this series, no patient has died of disease. Thirty-two (97%) patients are alive with a median follow-up of 7 years, which underscores the fact that this disease process primarily represents a local problem.

Treatment of choice for DFSP is surgical resection. Minimum gross resection margin to ensure local control is undefined, but most authorities would agree that a margin of normal tissue 2–3 cm from the gross tumor boundary including skin, subcutaneous fat, and underly-

TABLE 2. *Head and neck dermatofibrosarcoma protuberans—clinical features*

Clinical feature	Barnes et al. 1984 personal series	Barnes et al. 1984 literature review	Mark et al. 1993	Current series 2000
Number of patients	17	92	16	33
Sex (% males)	53	76	50	48
Age[a] (y)	31, 5–74	43, 10–78	36, 6–83	39, 24–77
Most common site	Face	Scalp	Scalp/forehead	Scalp/face
Duration of symptoms[a] (y)	2, 0.1–25	2, 0.1–30	Not stated	3, 0.1–60
Size[a] (cm)	2.0, 0.7–4.5	5.0, 0.5–21	Not stated	2.0, 0.1–8.0
Follow-up[a] (y)	5.7, 0.1–23	Not stated	9.5, 3–16.5	6.8, 0.3–23.7
Local recurrence (LR) rate	53%	73%	60%	9%
Time to LR $\leq$3 y	50%	75%	Not stated	33% (1/3)
LR after wide margin excision	Not stated	Not stated	0%	0%
Regional nodal or distant metastases	0%	0%	0%	0%

[a] Median and range.

ing fascia must be achieved to ensure adequate extirpation.[6] Historically, local recurrence rates have been higher after surgical treatment of DFSP of head and neck (50% to 75%) than for DFSP in other sites (Table 2). In the Mayo Clinic series of head and neck DFSP, local recurrence was significantly lower with wide (≥2 cm) than close (<2 cm) margins of resection.[45] The most significant predictor of recurrence was a resection margin less than 2 cm, although this was not statistically significant on χ^2 analysis.

In this series, a gross resection margin 2 cm or greater was a significant predictor of negative histological margin. No local recurrences developed after wide margin resection (≥2 cm). There was a trend toward improved local recurrence-free survival with wide margin resection, but few recurrences limited statistical analysis of the predictive effect of margin status on local disease control. One of the three local recurrences occurred after prior resections that had pathologically confirmed tumor free margins. Ten of 11 patients with microscopically positive margin resections remained disease free, seven of whom had no adjuvant radiotherapy, which suggests perhaps that microscopic margin involvement does not necessarily always predict local recurrence.

Wide surgical excision, including 2–3 cm margin of normal tissue measured from the palpable tumor boundary, is the preferred treatment for DFSP; however, this may not be readily achievable in the head and neck region. Maximizing local tumor control often entails removal of considerable tissue volumes that include underlying striated muscle and bone. In the majority of wounds, primary closure will not be feasible, and skin grafting or myocutaneous flap reconstruction will be required to cover the defect.

The amount of tissue excised to achieve 2 cm or more grossly negative surgical margin was substantial in many of our patients and required at times ambitious reconstructive attempts. Seventy percent of patients underwent major resections that included underlying bone, deep muscle, or adjacent parotid gland, in addition to local skin or free myocutaneous flap reconstruction. The low overall incidence of microscopic margin involvement and the infrequent recurrences in this group of patients suggest that our gross excision margins were adequate.

The role of intraoperative frozen section analysis in the treatment of DFSP is undefined. Frozen section analysis provided an accurate diagnosis of margin status in 80% of patients and a false negative rate of 57%. The proportion of patients with correctly identified margin involvement by frozen section is unacceptably low. We suggest that frozen section be used sparingly, and if it is to be employed at the time of operation, we recommend that sectioning techniques involve entire lateral and deep tumor margins to optimize the sensitivity of frozen section processing.

This study underscores the fact that wide resection is not always tenable, particularly on the face, where an extensive resection may entail significant functional and/or cosmetic deficits. To optimize local control of DFSP, some have applied Mohs micrographic surgery to treat this disease.[18,46–55] This technique allows precise histological mapping of all lateral and deep margins, which makes it possible to excise completely the peripheral pseudopod-like projections of neoplastic cells that may not be captured by standard histological vertical step sectioning, which can lead to a false-negative paraffin section interpretation.[6,16,55] The precise excision of DFSP with Mohs microsurgery has resulted in exceptionally high rates of cure; however, follow-up periods are relatively short in these studies (Table 1).

Mohs micrographic surgery may be particularly useful for head and neck surgery because it allows maximal conservation of uninvolved tissue where more than 2 cm gross resection margins are required to achieve local control of disease. The Mohs technique may permit smaller wounds and thereby allow primary closure without compromising margin control. Mohs micrographic surgery may become the treatment of choice for selected patients with DFSP. This technique, however, is very labor-intensive, is not readily applied to large tumors, and is of questionable value in recurrent tumors that are notoriously aggressive and more difficult to treat.

In the past, radiotherapy has had a limited role in the treatment of DFSP, because this treatment modality was thought to be ineffective.[3–6,17,22] Recently published data have demonstrated that radiotherapy is an effective adjunct to the surgical management of patients with DFSP and that radiation can be a viable primary therapeutic modality.[56–58] Actuarial 10-year local control and survival rates are 84% to 95% and 86% for adjuvant radiotherapy in the setting of positive or close postresection margins.[58]

The overall efficacy of radiotherapy for DFSP cannot be assessed definitively in light of the small overall patient numbers and lack of appropriate comparison groups. This series indicates that the majority of patients who have involved margins do as well without adjuvant radiotherapy as those who receive postoperative radiation. The indications for radiotherapy are evolving. The effectiveness of adjuvant radiotherapy in preventing local recurrence after margin positive excision remains unproven.[56,57] Radiotherapy may prove to be useful in situations where more limited resection would provide the patient with functional and/or cosmetic benefit, as in

the head and neck region.[57] Given the propensity of DFSP to recur despite seemingly adequate resection, some authors have advocated adjuvant radiation after re-excision of recurrent tumors to maximize the likelihood of eradicating the tumor.[57]

CONCLUSIONS

Head and neck DFSP is a rare, indolent tumor that presents a local problem. Regional nodal or distant metastases are exceedingly rare, and overall survival is excellent.

Deep lesions are significantly more likely to have involved microscopic margins after surgical resection than superficial head and neck tumors. Minimum gross resection margin to ensure local control is undefined. In this study, surgical resection with a gross margin of normal tissue 2 cm or greater from the gross tumor boundary uniformly ensured a negative histological margin. Wide margin excision had a favorable impact on recurrence-free survival. Intraoperative frozen section assessment of resected tumor margins is an inaccurate predictor of final resection margin status. Wide margin excision often requires resection of considerable volume of tissue that may have important functional or cosmetic implications.

Acknowledgments: Supported by grant CA47179 from the United States Public Health Service.

REFERENCES

1. Figueredo MTA, Marques LA, Campos-Filho N. Soft tissue sarcomas of the head and neck in adults and children: experience at a single institution and a review of the literature. *Int J Cancer* 1988;42:198–200.
2. Farhood AI, Hajdu SI, Shiu MH, Strong EW. Soft tissue sarcomas of the head and neck in adults. *Am J Surg* 1990;160:365–9.
3. Taylor HB, Helwig EB. Dermatofibrosarcoma protuberans: a study of 115 cases. *Cancer* 1962;15:717–25.
4. Burkhardt BR, Soule EH, Winkelmann RK, Ivans JC. Dermatofibrosarcoma protuberans: study of 56 cases. *Am J Surg* 1966;111: 638–44.
5. McPeak CJ, Crux T, Nicastri AD. Dermatofibrosarcoma protuberans: an analysis of 86 cases—five with metastases. *Ann Surg* 1967;166:803–16.
6. Gloster HM. Dermatofibrosarcoma protuberans. *J Am Acad Dermatol* 1996;35:355–74.
7. Mark RJ, Bailet JW, Tran LM, Poen J, Fu YS, Calcaterra TC. Dermatofibrosarcoma protuberans of the head and neck: a report of 16 cases. *Arch Otolaryngol Head Neck Surg* 1993;119:891–6.
8. Darrier J, Ferrand M. Dermatofibromes progressifs et recidivants ou fibrosarcomes de la peau. *Ann Dermatol Syphiligr (Paris)* 1924;5:545–62.
9. Hoffman E. Uber das knollentreibende fibrosarkom der haut. *Dermatol Ztschr* 1925;43:1–28.
10. Gentele H. Malignant fibrous tumors of the skin. *Acta Derm Venereol (Stockh)* 1951;31:91–132.
11. Longhin S. Considerations on the histopathologic diagnosis and treatment of the Darier-Ferrand dermatofibroma. *Ann Dermatol Syphiligr* (Paris) 1967;94:159–68.
12. Hadju SI. *Pathology of Soft Tissue Tumors*. Philadelphia: Lea & Febiger, 1979.
13. Barnes L, Coleman JA, Johnson JT. Dermatofibrosarcoma protuberans of the head and neck. *Arch Otolaryngol* 1984;110:398–404.
14. Roses DF, Valensi Q, LaTrenta G, Harris MN. Surgical treatment of dermatofibrosarcoma protuberans. *Surg Gynecol Obstet* 1986; 162:449–52.
15. Koh CK, Chung BK, Bury HPR, Path MRC, Wyatt EH. Dermatofibrosarcoma protuberans. *Int J Dermatol* 1995;34:256–60.
16. Hobbs ER, Wheeland RG, Bailin PL, Ratz JL, Yetman RJ, Zins JE. Treatment of DFSP with Mohs micrographic surgery. *Ann Surg* 1988;207:102–7.
17. Pack GT, Tabah EJ. Dermatofibrosarcoma protuberans: a report of thirty-nine cases. *Arch Surg* 1951;62:391–411.
18. Mentzel T, Beham A, Katenkamp D, et al. Fibrosarcomatous ("high grade") dermatofibrosarcoma protuberans: clinicopathologic and immunohistochemical study of a series of 41 cases with emphasis on prognostic significance. *Am J Surg Pathol* 1998;22: 576–87.
19. Bowne WB, Antonescu CR, Leung DHY, et al. Dermatofibrosarcoma protuberans (DFSP): a clinicopathological analysis of patients treated and followed at a single institution. *Cancer* (in press).
20. Rutgers EJ, Kroon BR, Albus-Lutter CE, Gortzak E. Dermatofibrosarcoma protuberans: treatment and prognosis. *Eur J Surg Oncol* 1992;18:241–8.
21. Simstein N, Tuthill RJ, Spesman EE, et al. Dermatofibrosarcoma protuberans: Case reports and review of the literature. *South Med J* 1977;70:487–9.
22. Bendix-Hansen K, Myhre-Jensen D, Haae S. Dermatofibrosarcoma protuberans: a clinico-pathological study of nineteen cases and review of the world literature. *Scand J Plast Reconstr Surg Hand Surg* 1983;17:247–52.
23. Brabant B, Revol M, Vergote T, et al. Dermatofibrosarcoma protuberans of the chest and shoulder: wide and deep excisions with immediate reconstruction. *Plast Reconstr Surg* 1993;92:459–62.
24. Petoin DS, Verola O, Banzet P, Dufourmentel C, Servant JM. Darier-Ferrand dermatofibrosarcoma protuberans: study of 96 cases over 15 years. *Chirurgie* 1985;111:132–8.
25. Tamoney HJ. Dermatofibrosarcoma protuberans: a review of twelve cases. *Conn Med* 1971;35:674–9.
26. Binkley GW. Dermatofibrosarcoma protuberans: report of six cases. *Arch Dermatol Syphiligr* 1939;40:578–94.
27. Gutierrez G, Opsina JE, De Baez NE, et al. Dermatofibrosarcoma protuberans. *Int J Dermatol* 1984;23:396–401.
28. Chattopadhyay TK, Singh MK, Aruna BH. Dermatofibrosarcoma protuberans: a clinicopathological study of ten cases. *Jpn J Surg* 1986;16:435–8.
29. Coard K, Branday JM, LaGranade L. Dermatofibrosarcoma protuberans: a 10-year clinicopathological review of an uncommon tumor. *West Indian Med J* 1994;43:130–3.
30. Przybora LA, Wojnerowicz C. Malignancy of dermatofibrosarcoma protuberans and report of 2 cases with lymph gland spread. *Oncologia* 1959;12:236–54.
31. Waldermann F, Hagedoorn M. Klinik und pathologie des dermatofibrosarcoma protuberans. *Z Hauktr* 1985;60:1886–94.
32. Kahn BB, Saxe NM, Gordon W. Dermatofibrosarcoma protuberans with lymph node and pulmonary metastases. *Arch Dermatol* 1978;114:599–601.
33. Brenner W, Schaeffeer K, Chhabra H, Postel A. Dermatofibrosarcoma protuberans metastatic to a regional lymph node. Report of a case and review. *Cancer* 1975;36:1897–1902.
34. Fisher ER, Hellstrom HR. Dermatofibrosarcoma with metastasis simulating Hodgkin's disease and reticulum cell sarcoma. *Cancer* 1966;19:1165–71.

35. Woolridge WE. Dermatofibrosarcoma protuberans. A tumor too lightly considered. *Arch Dermatol* 1957;75:132–5.
36. McGregor JK. Role of surgery in the management of dermatofibrosarcoma protuberans. *Ann Surg* 1961;154:255–8.
37. Adams JT, Saltzstein SL. Metastasizing dermatofibrosarcoma protuberans: report of two cases. *Am Surg* 1963;29:879–86.
38. Bezecny R. Lungenmetastasen beim dermatofibrosarcoma protuberans. *Arch Dermatol Syphiligr* 1933;169:347–53.
39. Penner DW. Metastasizing dermatofibrosarcoma protuberans: A case report. *Cancer* 1950;3:1083–6.
40. Whalen WP. Dermatofibrosarcoma protuberans. *Plast Reconstr Surg* 1963;31:461–6.
41. Bonnabeau RC, Stoughton WB, Armanious AW, et al. Dermatofibrosarcoma protuberans: a report of a case with pulmonary metastasis and multiple intrathoracic recurrences. *Oncology* 1974;29: 1–12.
42. Mendoza CB, Pinkus H. Dermatofibrosarcoma protuberans with metastases treated with methotrexate. *Am J Surg* 1970;120:119–21.
43. Connelly JH, Evans HL. Dermatofibrosarcoma protuberans: a clinicopathologic review with emphasis on fibrosarcomatous areas. *Am J Surg Pathol* 1992;16:921–5.
44. Rockley PF, Robinson JK, Magid M, et al. Dermatofibrosarcoma protuberans of the scalp: a series of cases. *J Am Acad Dermatol* 1989;21:278–83.
45. Gayner SM, Lewis JE, McCaffrey TV. Effect of resection margins on dermatofibrosarcoma protuberans of the head and neck. *Arch Otolaryngol Head Neck Surg* 1997;123:430–3.
46. Mikhail GR, Lynn BH. Dermatofibrosarcoma protuberans. *J Dermatol Surg Oncol* 1978;4:81–4.
47. Mohs FE. *Chemosurgery: Microscopically Controlled Surgery for Skin Cancer*. Springfield, IL: Charles C. Thomas, 1978.
48. Peters CW, Hanke CW, Pasarelle HA, Bennett JE. Chemosurgical reports: DFSP of the face. *J Dermatol Surg Oncol* 1982;8:823–6.
49. Hess KA, Hanke CW, Estes NC, et al. Chemosurgical reports: myxoid dermatofibrosarcoma protuberans. *J Dermatol Surg Oncol* 1985;11:268–71.
50. Robinson JK. Dermatofibrosarcoma protuberans resected by Mohs' surgery (chemosurgery). *J Am Acad Dermatol* 1985;12: 1093–8.
51. Hobbs ER, Ratz JL. Dermatofibrosarcoma protuberans of the hand: report of a case treated with Mohs micrographic surgery. *Cleve Clin J Med* 1988;55:252–6.
52. Weber PJ, Gretzala JC, Hevia O, et al. Dermatofibrosarcoma protuberans. *J Dermatol Surg Oncol* 1988;14:555–8.
53. Goldberg DJ, Maso M. Dermatofibrosarcoma protuberans in a 9-year-old child: treatment by Mohs micrographic surgery. *Pediatr Dermatol* 1990;7:57–9.
54. Parker TL, Zitelli JA. Surgical margins for excision of dermatofibrosarcoma protuberans. *J Am Acad Dermatol* 1995;32:233–6.
55. Gloster HM, Harris KR, Roenigk RK. A comparison between Moh's micrographic surgery and wide surgical excision for the treatment of dermatofibrosarcoma protuberans. *J Am Acad Dermatol* 1996;35:82–7.
56. Marks LB, Suit HD, Phil D, Rosenberg AE, Wood WC. Dermatofibrosarcoma protuberans treated with radiation therapy. *Int J Radiat Oncol Biol Phys* 1989;17:379–84.
57. Ballo MT, Zagars GK, Pisters P, Pollack A. The role of radiation therapy in the management of dermatofibrosarcoma protuberans. *Int J Radiat Oncol Biol Phys* 1998;40:822–7.
58. Suit H, Spiro I, Mankin HJ, Efird J, Rosenberg AE. Radiation in management of patients with dermatofibrosarcoma protuberans. *J Clin Oncol* 1996;14:2365–9.