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NCCN Clinical Practice Guidelines in Oncology (NCCN Guidelines®)

Dermatofibrosarcoma Protuberans

Version 1.2023 — December 8, 2022

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See NCCN Categories of Evidence and Consensus.

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Updates in Version 1.2023 of the NCCN Guidelines for Dermatofibrosarcoma Protuberans from Version 2.2022 include:

DFSP-1

- **Preliminary Workup:**
  - **Bullet removed: Complete skin exam**
  - **Second bullet, third sub-bullet revised: ~~Report evidence of fibrosarcomatous change or other high-risk features~~ *Debulking specimens from all excisions should be examined to identify fibrosarcomatous transformation of dermatofibrosarcoma protuberans (FS-DFSP).***
- **New pathway header added: DFSP confirmed.**
- **Additional Workup, new bullet added: Complete skin exam.**
- **Footnote d revised: If *areas of* ~~fibrosarcomatous~~ transformation *to fibrosarcoma or other sarcoma subtypes* ~~is found~~ *are identified,* multidisciplinary consultation for consideration of further treatment and surveillance is recommended. *Fibrosarcomatous transformation of DFSP (FS-DFSP) is associated with a metastasis risk of 15%–20%.* See the NCCN Guidelines for Soft Tissue Sarcoma for multimodal therapy and surveillance considerations *including CT of draining nodal basin and chest*. (Also on page DFSP-2)**

DFSP-2

- **Treatment, new bullet added: Consider neoadjuvant imatinib for unresectable/borderline resectable disease.**
- **Adjuvant Treatment, following Positive margins, option revised: Multidisciplinary consultaiton for consideration of *radiation therapy (*RT*)* ~~vs. other treatment options~~.**
- **Follow-up, new third bullet added: Consider MRI surveillance for deeply invasive disease or other concerns for recurrence.**
- **Footnote removed: MRI with contrast may be helpful to detect early recurrence in patients with high-risk lesions or delineate tumor extent when physical exam is insufficient or unreliable.**
- **Footnote j revised: When Mohs or other forms of PDEMA are utilized, RT is not recommended. When Mohs or other forms of PDEMA are not utilized, consider RT if margins are ~~<1 cm~~ *considered narrow by the treating physicians*...**

DFSP-A

- **Fourth bullet revised: Fibrosarcomatous transformation (FS-DFSP) is characterized by transition from storiform to a herringbone pattern, with a higher degree of cellularity, cytologic atypia, mitotic activity (>5/10 high-power fields [HPFs]), and frequent loss of CD34 immunostaining. *When CD34 is negative, other markers such as S100 should also be negative to rule out other spindle cell tumors*.**
- **Footnote removed: FS-DFSP should be noted when present as the metastatic risk is 15%–20% and the patient should be referred to a center with expertise in management of soft tissue sarcomas.**
- **New footnote 2 added: If areas of transformation to fibrosarcoma or other sarcoma subtypes are identified, multidisciplinary consultation for consideration of further treatment and surveillance is recommended. *FS-DFSP is associated with a metastasis risk of 15%–20%. The patient should be referred to a center with expertise in management of soft tissue sarcomas.* See the NCCN Guidelines for Soft Tissue Sarcoma for multimodal therapy and surveillance considerations including CT of draining nodal basin and chest.**

DFSP-B

- **Goal, first bullet revised: Every effort should be made to achieve clear surgical margins. Complete histologic surgical margin examination to include ~~review of~~ the entire excised peripheral and deep margin is recommended, whenever possible. Tumor characteristics include long, irregular, subclinical extensions. Debulking specimens from all excisions should be examined to identify ~~fibrosarcomatous transformation (~~FS-DFSP~~)~~ since this is associated with metastatic potential.**

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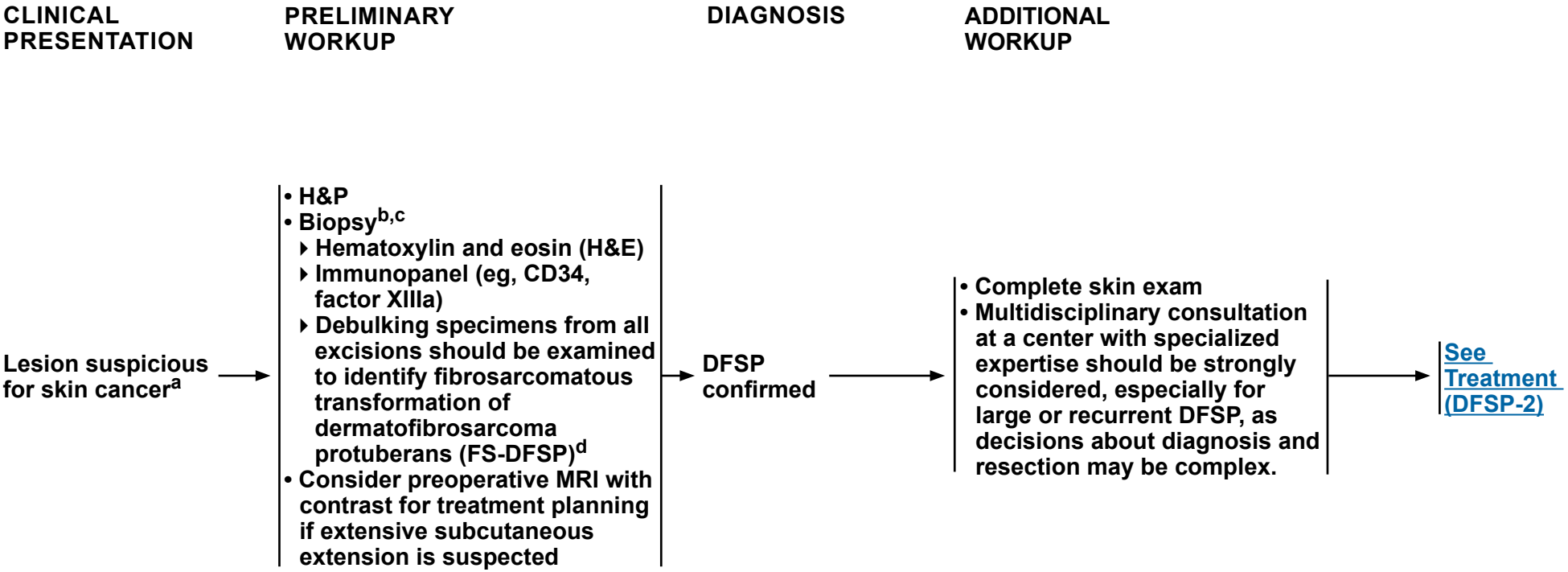


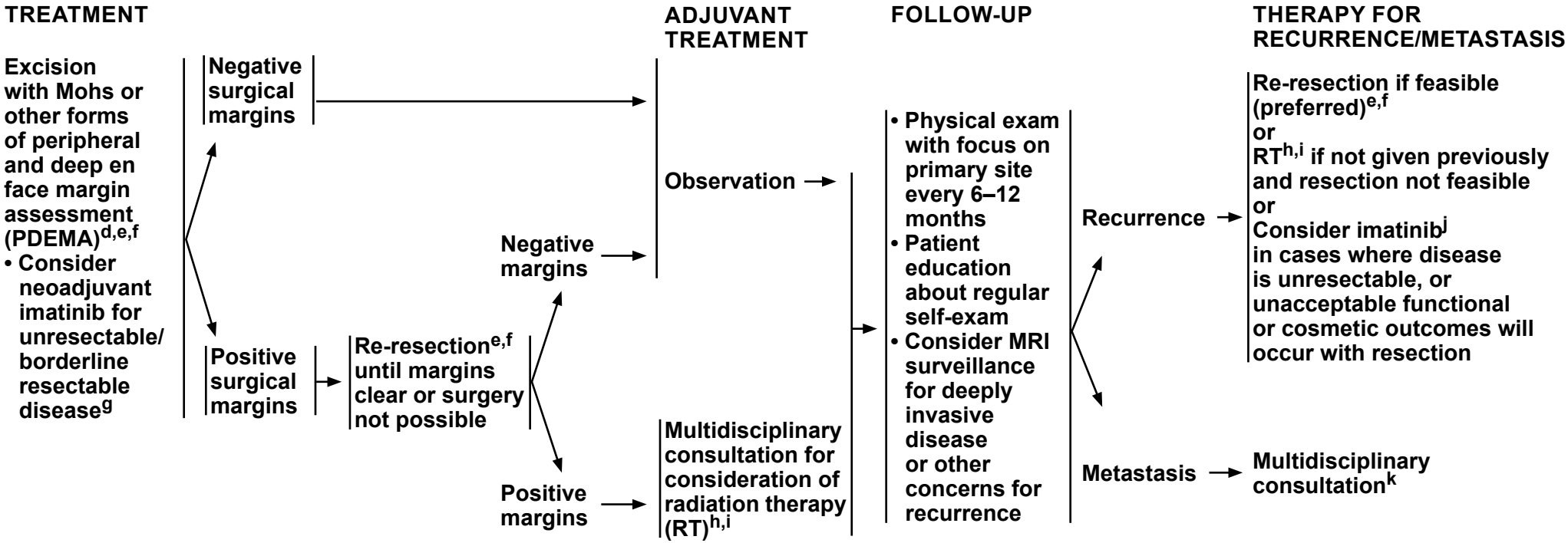
[a] For more information, see American Academy of Dermatology Association.

[b] This tumor is frequently misdiagnosed due to inadequate tissue sampling/superficial biopsy. Punch, incisional, or core biopsy, preferably including the deeper subcutaneous layer, is strongly recommended for sufficient tissue sampling and accurate pathologic assessment. If biopsy is indeterminate or clinical suspicion remains, rebiopsy is recommended. Wide undermining is discouraged due to the difficulty of interpreting subsequent re-excisions pathologically.

[c] See Principles of Pathology (DFSP-A).

[d] If areas of transformation to fibrosarcoma or other sarcoma subtypes is found are identified, multidisciplinary consultation for consideration of further treatment and surveillance is recommended. FS-DFSP is associated with a metastasis risk of 15%–20%. See the NCCN Guidelines for Soft Tissue Sarcoma for multimodal therapy and surveillance considerations including CT of draining nodal basin and chest.

Note: All recommendations are category 2A unless otherwise indicated.
Clinical Trials: NCCN believes that the best management of any patient with cancer is in a clinical trial. Participation in clinical trials is especially encouraged.



[d] If areas of transformation to fibrosarcoma or other sarcoma subtypes are identified, multidisciplinary consultation for consideration of further treatment and surveillance is recommended. FS-DFSP is associated with a metastasis risk of 15%–20%. See the NCCN Guidelines for Soft Tissue Sarcoma for multimodal therapy and surveillance considerations including CT of draining nodal basin and chest.

[e] The most commonly used form of PDEMA is Mohs. See NCCN Guidelines for Squamous Cell Skin Cancer - Principles of PDEMA Technique. When anatomic structures at the deep margin (eg, major vessels, nerves, bone) preclude complete histologic evaluation of the marginal surface via Mohs or other forms of PDEMA, Mohs or other forms of PDEMA should be used to evaluate as much of the marginal surface as feasible. Treatment considerations for non-visualized areas may be the subject of multidisciplinary discussion.

[f] If PDEMA is unavailable, consider wide excision. Wide undermining is discouraged prior to confirmation of clear margins due to the difficulty of interpreting subsequent re-excised margins, and the risk of concealing residual tumor below mobilized tissue. See Principles of Excision (DFSP-B).

[g] Consider neoadjuvant imatinib for patients in whom resection with negative margins may result in unacceptable functional or cosmetic outcomes. Ugurel S, et al. Clin Cancer Res 2014;20:499-510.

[h] See Principles of Radiation Therapy (DFSP-C).

[i] When Mohs or other forms of PDEMA are utilized and margins are negative, RT is not recommended. When Mohs or other forms of PDEMA are not utilized, consider RT if margins are considered narrow by the treating physicians. RT can be considered for treatment of positive margins if not given previously and further resection is not feasible.

[j] Navarrete-Dechent C, et al. JAMA Dermatol 2019;155:361-369.

[k] See NCCN Guidelines for Synchronous STAGE IV Soft Tissue Sarcoma (EXTSARC-5).

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PRINCIPLES OF PATHOLOGY[1]

- **Evaluation by a qualified physician with specific expertise in sarcoma/soft tissue pathology or dermatopathology is preferred (if available).**
- **The spindled cells arranged in a storiform or fascicular pattern are typically bland with minimal cytologic atypia.**
- **Immunohistochemistry for CD34 is mostly positive, and factor XIIIa negative.**
- **FS-DFSP[2] is characterized by transition from storiform to a herringbone pattern, with a higher degree of cellularity, cytologic atypia, mitotic activity (>5/10 high-power fields [HPFs]), and frequent loss of CD34 immunostaining. When CD34 is negative, other markers such as S100 should also be negative to rule out other spindle cell tumors.**
- **For equivocal lesions, consider fluorescence in situ hybridization (FISH), polymerase chain reaction (PCR), or conventional cytogenetics to detect t(17;22)(q22;q13), which is a hallmark of DFSP. Fusion of the collagen type I alpha 1 gene (*COL1A1*) at 17q22, with the platelet-derived growth factor Beta gene (*PDGFβ*) at 22q13, form the oncogenic chimeric fusion gene *COL1A1::PDGFβ*.**
- **Margin control during excision (see Principles of Excision [DPSP-B]) may occasionally be aided by H&E sections supplemented by CD34 immunohistochemistry.**

[1] Currently, no American Joint Committee on Cancer (AJCC) or College of American Pathologists (CAP) synoptic reporting is defined.
[2] If areas of transformation to fibrosarcoma or other sarcoma subtypes are identified, multidisciplinary consultation for consideration of further treatment and surveillance is recommended. FS-DFSP is associated with a metastasis risk of 15%–20%. The patient should be referred to a center with expertise in management of soft tissue sarcomas. See NCCN Guidelines for Soft Tissue Sarcoma for multimodal therapy and surveillance considerations including CT of draining nodal basin and chest.

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PRINCIPLES OF EXCISION

Goal:

- **Every effort should be made to achieve clear surgical margins. Complete histologic surgical margin examination to include the entire excised peripheral and deep margin is recommended, whenever possible. Tumor characteristics include long, irregular, subclinical extensions. Debulking specimens from all excisions should be examined to identify FS-DFSP since this is associated with metastatic potential.**

Surgical Approach: Mohs or Other Forms of PDEMA

- **See NCCN Guidelines for Squamous Cell Skin Cancer - Principles of PDEMA Technique.**
- **If Mohs or other forms of PDEMA are unavailable, consider wide excision.**
  - **Reconstruction should be delayed until clear margins have been verified to avoid the risk of translocating the tumor within the resection bed, thus making further margin assessments inaccurate.**

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PRINCIPLES OF RADIATION THERAPY

General Treatment Information

- **Adjuvant RT:**
  - **Positive Margins/Gross Disease**
    - ◊ **50–60 Gy for indeterminate or positive margins, and up to 66 Gy for positive margins or gross tumor (2-Gy fractions per day).**
    - ◊ **Fields to extend widely beyond surgical margins (eg, 3–5 cm) when clinically feasible.**
  - **Negative Margins**
    - ◊ **When Mohs or other forms of PDEMA are utilized, RT is not recommended.**
    - ◊ **When Mohs or other forms of PDEMA are not utilized, consider RT if margins are <1 cm.**
- **Recurrence/Metastasis:**
  - **RT if not given previously and further resection is not feasible; 50–60 Gy for indeterminate or positive margins, and up to 66 Gy for positive margins or gross tumor (2-Gy fractions per day).**
  - **Fields to extend widely beyond surgical margins (eg, 3–5 cm) when clinically feasible.**

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Clinical Trials: NCCN believes that the best management of any patient with cancer is in a clinical trial. Participation in clinical trials is especially encouraged.

ABBREVIATIONS

DFSP	**dermatofibrosarcoma protuberans**
FS-DFSP	**fibrosarcomatous transformation of DFSP**
PDEMA	**peripheral and deep en face margin assessment**
RT	**radiation therapy**

NCCN Categories of Evidence and Consensus	
Category 1	Based upon high-level evidence, there is uniform NCCN consensus that the intervention is appropriate.
Category 2A	Based upon lower-level evidence, there is uniform NCCN consensus that the intervention is appropriate.
Category 2B	Based upon lower-level evidence, there is NCCN consensus that the intervention is appropriate.
Category 3	Based upon any level of evidence, there is major NCCN disagreement that the intervention is appropriate.

All recommendations are category 2A unless otherwise indicated.

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Discussion

This discussion corresponds to the NCCN Guidelines for Dermatofibrosarcoma Protuberans. Last updated: January 31st, 2022.

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Overview

Dermatofibrosarcoma protuberans (DFSP) is an uncommon, low-grade sarcoma of fibroblast origin with an incidence rate of 4.1 to 4.5 cases per million persons per year in the United States.[1-4] A predilection for occurring in African Americans has been reported in one study.[3] Initial misdiagnosis, prolonged time to accurate diagnosis, and large tumor size at the time of diagnosis are common. However, it rarely metastasizes.[5] When metastasis occurs, it is typically in the lung, bone, or regional lymph nodes. Three-dimensional reconstruction of DFSP[6] has revealed tumors with highly irregular shapes and frequent finger-like extensions.[7] As a result, incomplete removal and subsequent recurrence are common without attention to full assessment of the peripheral and deep margin. The local recurrence rate for wide local excision (WLE) of DFSP in studies ranges from 10% to 60%, whereas the rate of development of regional or distant metastatic disease is only 1% and 4% to 7.4%, respectively.[8,9]

Literature Search Criteria and Guidelines Update Methodology

Prior to the update of this version of the NCCN Clinical Practice Guidelines in Oncology (NCCN Guidelines®) Dermatofibrosarcoma Protuberans, an electronic search of the PubMed database was performed to obtain key literature using the following search term: dermatofibrosarcoma protuberans. The PubMed database was chosen as it remains the most widely used resource for medical literature and indexes peer-reviewed biomedical literature.[10]

The search results were narrowed by selecting studies in humans published in English. Results were confined to the following article types: Clinical Trial; Guideline; Meta-Analysis; Randomized Controlled Trial; Systematic Reviews; and Validation Studies.

The data from key PubMed articles as well as articles from additional sources deemed as relevant to these guidelines and discussed by the panel have been included in this version of the Discussion section (eg, e-publications ahead of print, meeting abstracts). Recommendations for which high-level evidence is lacking are based on the panel's review of lower-level evidence and expert opinion.

The complete details of the Development and Update of the NCCN Guidelines are available at www.NCCN.org.

Evaluation

Histologically, DFSP typically presents as a storiform or fascicular proliferation of bland spindled cells that extends from the dermis into the subcutis.[11,12] Virtually all cases are CD34-positive and factor XIIIa-negative with rare exceptions.[13,14] Currently, no synoptic reporting is recommended. Workup for DFSP consists of history and physical (H&P) examination, complete skin examination, and biopsy. It should be noted that this tumor is frequently misdiagnosed due to inadequate tissue sampling resulting from shallow biopsy. As the superficial aspect of a DFSP may not be distinct from benign lesions, a punch or incisional biopsy that samples the subcutaneous layer, is strongly recommended. If a biopsy is indeterminate or clinical suspicion remains, rebiopsy is recommended.

In most cases, examination of hematoxylin and eosin-stained specimens by light microscopy results in an unequivocal diagnosis. However, differentiation of DFSP from dermatofibroma can be difficult at times. In such instances, staining with CD34,[14,15] factor XIIIa,[13,16] stromelysins 3,[17] and other immunomarkers such as nestin, apolipoprotein D, and cathepsin K,[18-20] might be useful. The NCCN Panel recommends that appropriate and confirmatory immunostaining be performed in all cases of suspected DFSP.

It is unclear whether the histologic features of a high mitotic rate or evidence of fibrosarcomatous (FS) change (FS-DFSP) have prognostic significance in DFSP.[21,22] Studies in the biomedical literature both support the connection between FS-DFSP and an increased risk of local recurrence, lower time to recurrence, and increased risk of metastasis,[23-31] and refute[32,33] this notion. A systematic review of 1422 patients with DFSP and 225 with DFSP-FS reported risks of local recurrence (29.8% vs. 13.7%), metastasis (14.4% vs. 1.1%), and death (14.7% vs. 0.8%) from the disease to be significantly higher in DFSP-FS versus DFSP.[34] The NCCN Panel recommends that FS change and other high-risk features such as deep lesions and high grade be noted in all pathology reports assessing this tumor. If FS transformation is found, multidisciplinary consultation for consideration of further treatment and surveillance is recommended. Clinicians should consult the NCCN Guidelines for Soft Tissue Sarcoma. Overall, as decisions about diagnosis and resection may be complex, multidisciplinary consultation at a center with specialized expertise should be strongly considered, especially for large or recurrent DFSP as it may optimize clinical and reconstructive outcomes.[35,36]

Treatment

Initial treatment of DFSP is surgical. Because of its proclivity for irregular and frequently deep subclinical extensions, every effort should be made to completely remove this tumor at the time of initial therapy. Excision with Mohs micrographic surgery (Mohs) or other forms of peripheral and deep en face margin assessment (PDEMA) is recommended over WLE. En face sectioning is preferred to prevent missing small foci of tumor. The most commonly used form of PDEMA is Mohs (Refer to NCCN Guidelines for Squamous Cell Skin Cancer – Principles of PDEMA Technique). When anatomic structures at the deep margin (eg, major vessels, nerves, bone) preclude complete histologic evaluation of the marginal surface via Mohs or other forms of PDEMA, Mohs and other forms of PDEMA should be used to evaluate as much of the marginal surface as feasible. A combination of PDEMA and WLE for the deep margin has been reported in the literature.[36] Treatment considerations for non-visualized areas may be the subject of multidisciplinary discussion. If PDEMA is unavailable, WLE can be considered. Wide undermining is discouraged prior to confirmation of clear margins due to the difficulty of interpreting subsequent re-excised margins, and the risk of concealing residual tumor below mobilized tissue. If initial surgery yields positive margins, re-resection is recommended whenever possible, with the goal of achieving clear margins.

Mohs or modified Mohs surgery, and traditional WLE with wider margins, which has been associated with higher tumor clearance and lower rates of recurrence,[37-39] are all methods to achieve complete histologic assessment. Studies examining outcomes of both Mohs and WLE have consistently reported lower recurrence rates for the former (0%–6.6% vs. 1.7%–30.8%) (Table 1).[40-49] The weight of the evidence supports the NCCN Panel's preference for Mohs using either frozen or permanent sections.[50-53] A large retrospective series of 204 patients with DFSP showed a very low local recurrence rate (1%) using WLE with total peripheral margin pathologic evaluation, underscoring the importance of meticulous pathologic margin evaluation with any surgical technique.[54] This notion was also supported by more recent studies.[55,56] It is recommended that any reconstruction involving extensive undermining be avoided. Tissue rearrangement, if necessary, should be delayed until negative histologic margins are verified to prevent displacing a potentially positive margin or hampering interpretation of re-excisions. If there is concern that the surgical margins are not clear when Mohs or PDEMA is not available, split-thickness skin grafting should be considered to monitor for recurrence.

Radiation therapy (RT) has occasionally been used as a primary therapeutic modality for DFSP,[57-59] but it is most beneficial as adjuvant therapy after surgery.[57-64] In a single-institution retrospective review of 53 patients treated with surgery and preoperative or postoperative RT, local control was 93% and actuarial overall survival was 98% at 10 years.[33] Another small patient series reported that 86% of patients treated with postoperative RT remained disease-free at a median follow-up of 10.5 years.[63] In a systematic review and meta-analysis of adjuvant RT for DFSP after WLE, the overall recurrence rate was reported to be 11.74%. Patients with positive/close margins had a recurrence rate of 14.23% whereas those with negative margins had no recurrence.[65] The NCCN Panel recommends that if a negative margin is achieved, no adjuvant treatment is necessary. When Mohs or other forms of PDEMA are used, RT is not recommended. When Mohs or other forms of PDEMA are not used, RT can be considered if margins are less than 1 cm. RT can be considered for the treatment of positive margins if not given previously and further resection is not feasible.

DFSP can be treated by targeted PDGF receptors. DFSP is characterized by a translocation between chromosomes 17 and 22 [t(17;22)(q22;q13)] resulting in the overexpression of platelet-derived growth factor receptor β (PDGFR β).[66-68] These findings suggest that targeting PDGF receptors may be an effective treatment for DFSP. In published results, imatinib mesylate, a protein tyrosine kinase inhibitor, has shown clinical activity against DFSP,[69-74] which has led to its approval by the U.S. Food and Drug Administration (FDA) for the treatment of unresectable, recurrent, and/or metastatic DFSP in adult patients. It is still unclear whether or not and the extent to which the COL1A1-PDGFB fusion gene dictates imatinib response.[70] In a recent systematic review that included patients receiving imatinib as monotherapy, adjuvant, or neoadjuvant therapy, complete response, partial response, stable disease, and progressive disease were reported in 5.2%, 55.2%, 27.6%, and 9.2% of patients, respectively.[70] The panel recommends neoadjuvant imatinib for patients in whom resection with negative margins may result in unacceptable functional or cosmetic outcomes. In the neoadjuvant setting, complete response, partial response, stable disease, and progressive disease rates were reported to be 7.1%, 50%, 35.7%, and 7.1%, respectively.[71]

Follow-up

Given the historically high local recurrence rates for DFSP, ongoing clinical follow-up with focus on the primary site every 6 to 12 months is indicated, with re-biopsy of any suspicious regions. Although metastatic disease is rare, a guided H&P and patient education about regular self-examination are recommended. For patients with high-risk features or who have undergone extensive surgery, additional imaging studies may be useful in detecting recurrence. MRI with contrast may be helpful to detect early recurrence in patients with high-risk lesions or delineate tumor extent when physical examination is insufficient or unreliable.

Recurrent tumors should be resected whenever possible. Adjuvant RT may be considered after surgery. For patients who are not surgical candidates, RT alone is an option if not given previously. Imatinib mesylate should be considered in cases where the disease is unresectable following multiple resections, or if unacceptable functional or cosmetic outcomes will occur with further resection.

In the rare event of metastatic disease, multidisciplinary consultation is recommended to coordinate treatment (Refer to NCCN Guidelines for Soft Tissue Sarcoma).

Table 1. Summary of Retrospective Reviews, Systematic Reviews, and Meta-Analyses Investigating Locoregional Recurrence Rates by Mohs Micrographic Surgery and Wide Local Excision for DFSP

Author - Year	*Study Type*	*Number of Tumors*	*Surgical Technique (n)*	*Locoregional Recurrence Rate (%)*	*Notes*
Gloster 1996[40]	Review (retrospective series)	15	Mohs	6.6%	Median follow-up 40 months
		39	WLE	10%	Median follow-up 36 months
Gloster 1996[40]	Review (literature search)	64	Mohs	1.6%	Total across 11 studies
		489	WLE	20%	Total across 15 studies
Dubay 2004[41]	Retrospective review	63	Mohs (11) WLE (43) Combination (9)	0%	Median follow-up 52.8 months
Paradisi 2008[45]	Retrospective review (retrospective series)	41	Mohs	0%	Mean follow-up 5.4 years
		38	WLE	13.2%	Mean follow-up 4.8 years
	Retrospective review (literature search)	463	Mohs	1.3%	Total across 30 studies
		1394	WLE	20.7%	Total across 31 studies
Meguerditchian 2010[42]	Retrospective review	20	Mohs	0%	Median follow-up 40.4 months
		28	WLE	3.6%	Median follow-up 49.9 months
Foroozan 2012[43]	Systematic review (non-randomized comparative studies)	90	Mohs	1.1%	Total across 4 studies
		174	WLE	6.3%	Total across 4 studies
Bogucki 2012[44]	Literature review	444	Mohs	1.1%	Total across 20 studies
		1443	WLE	7.3%	Total across 20 studies
Lowe 2017[46]	Retrospective review	67	Mohs	3.0%	Mean follow-up 4.8 years
		91	WLE	30.8%	Mean follow-up 5.7 years
Veronese 2017[47]	Review (retrospective series)	73	Mohs Tubingen	5.5%	Median follow-up 9 years
		62	WLE	8.1%	Median follow-up 4.7 years
	Review (literature search)	424	Mohs	0.94%	Total across 16 studies
		82	Mohs Tubingen	0%	Total across 3 studies
		1465	WLE	14.9%	Total across 30 studies
Malan 2019[48]	Meta-analysis	298	Mohs	2.7%	Median follow-up 5.32 years
		389	WLE	9.10%	Median follow-up 5.32 years
Durack 2021[49]	Retrospective review	97	Mohs	0%	Median follow-up 25.5 months
		362	WLE	1.7%	Median follow-up 25.5 months

WLE, wide local excision

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