National Comprehensive Cancer Network®

NCCN Clinical Practice Guidelines in Oncology (NCCN Guidelines®)

# Dermatofibrosarcoma Protuberans

Version 1.2025 — October 11, 2024

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**NCCN Categories of Evidence and Consensus:** All recommendations are category 2A unless otherwise indicated.

See NCCN Categories of Evidence and Consensus.

**NCCN Categories of Preference:** All recommendations are considered appropriate.

See NCCN Categories of Preference.

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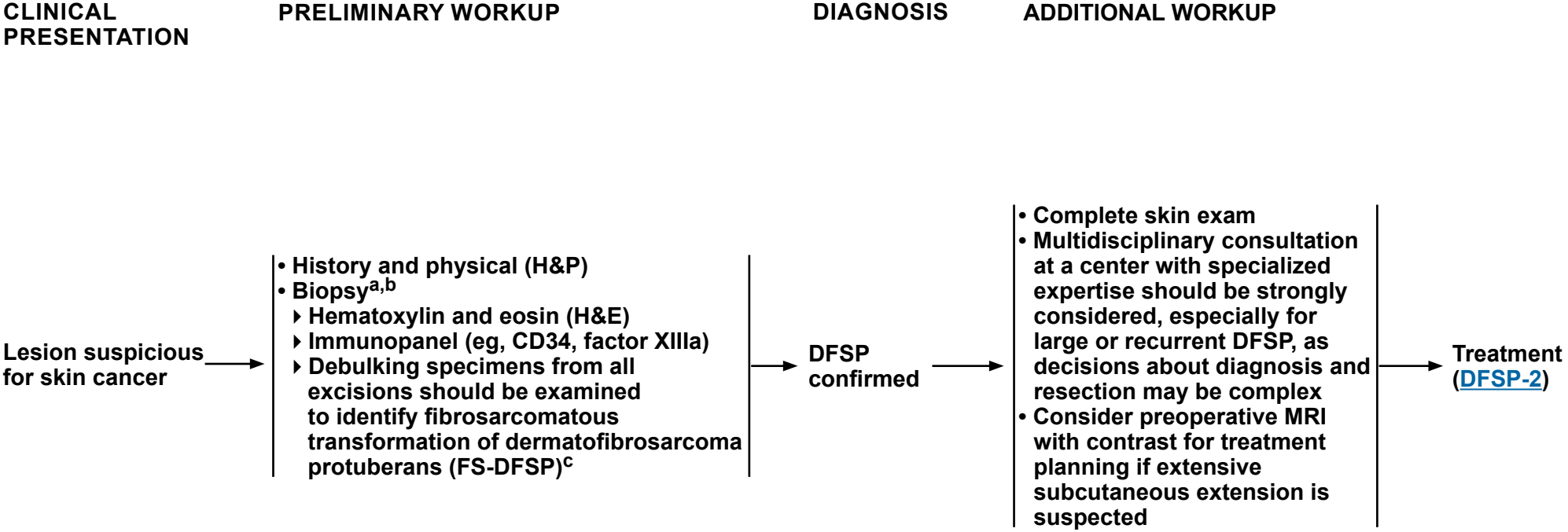
Updates in Version 1.2025 of the NCCN Guidelines for Dermatofibrosarcoma Protuberans from Version 1.2024 include:

DFSP-1
- Footnote c revised: . . .FS-DFSP is associated with a *higher* 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. (Also page DFSP-2)

DFSP-2
- Footnotes revised:
  - Footnote d: 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. (Also page DFSP-3)
  - Footnote e: 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 Surgery (DFSP-B). *Curtis KK, et al. J Natl Compr Canc Netw doi: 10.6004/jnccn.2024.7036*. (Also page DFSP-3)
  - Footnote h: 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. (Also page DFSP-3)

DFSP-A
- Footnote 2 revised: 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 *higher* 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.

DFSP-C
- General Treatment Information, Adjuvant RT, second sub-bullet:
  - First tertiary bullet revised: When Mohs or other forms of PDEMA are utilized *and margins are negative*, RT is not recommended.
  - Second tertiary bullet revised: When Mohs or other forms of PDEMA are not utilized, consider RT if margins are *considered narrow* ~~<1 cm~~ *by the treating physicians, RT not given previously, and further resection is not feasible*.




[a] 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.

[b] Principles of Pathology (DFSP-A).

[c] 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 higher metastasis risk. 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.

**Note: All recommendations are category 2A unless otherwise indicated.**

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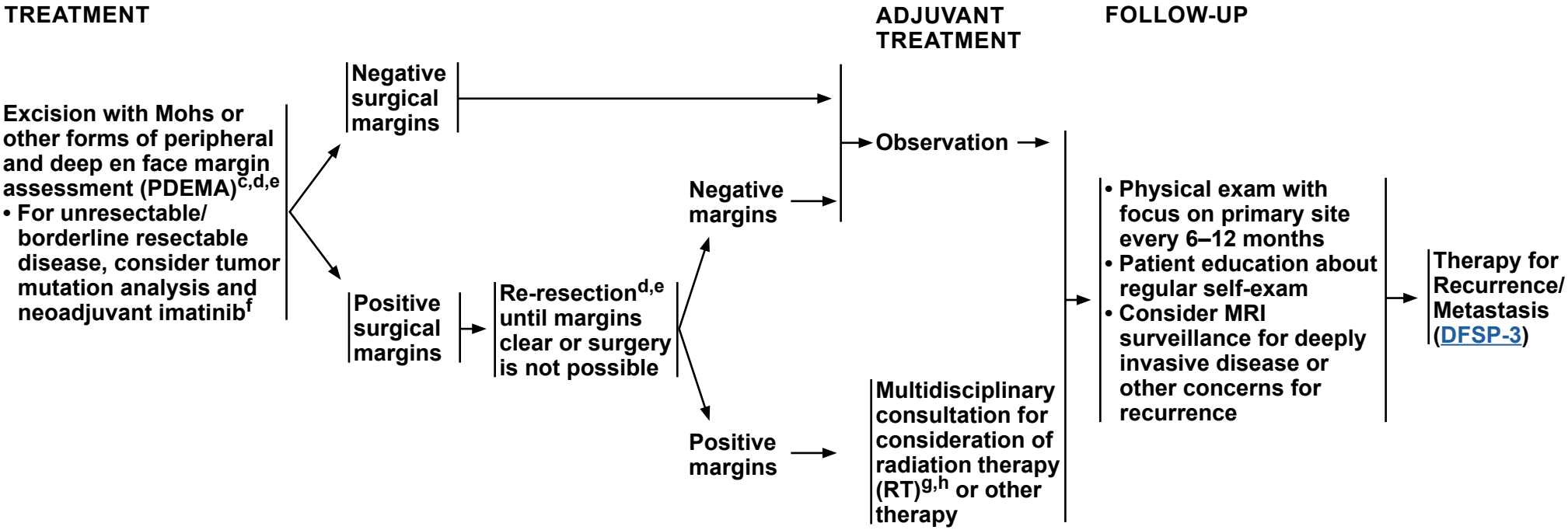


[c] If areas of transformation to fibrosarcoma or other sarcoma subtypes are identified, multidisciplinary consultation for consideration of further treatment and surveillance is recommended. FS-DFSP is associated with a higher metastasis risk. The patient should be referred to a center with expertise 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.

[d] 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, 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.

[e] 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 Surgery (DFSP-B). Curtis KK, et al. J Natl Compr Canc Netw doi: 10.6004/jnccn.2024.7036.

[f] 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.

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

[h] 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 if not given previously and further resection is not feasible.

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## THERAPY FOR RECURRENCE/METASTASIS

**Recurrence** → **Re-resection if feasible (preferred)[d,e]**
**or**
**RT[g,h] if not given previously and resection is not feasible**
**or**
**Consider imatinib[i] in cases where disease is unresectable, or unacceptable functional or cosmetic outcomes will occur with resection**

**Metastasis** → **Multidisciplinary consultation[j]**

[d] 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, 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.

[e] 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 Surgery (DFSP-B). Curtis KK, et al. J Natl Compr Canc Netwdoi: 10.6004/jnccn.2024.7036.

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

[h] 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 if not given previously and further resection is not feasible.

[i] Confirm tumor mutation with fluorescence in situ hybridization (FISH) for the translocation of *PDGRF*. Navarrete-Dechent C, et al. JAMA Dermatol 2019;155:361-369.

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

**Note: All recommendations are category 2A unless otherwise indicated.**

## 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 Surgery [DFSP-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 higher metastasis risk. 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 SURGERY

**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**

- **NCCN Guidelines for Squamous Cell Skin Cancer - Principles of PDEMA Technique**
- **If Mohs or other forms of PDEMA are unavailable, consider wide excision.[1]**
  - **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.**

[1] Farma JM, Ammori JB, Zager JS, et al. Dermatofibrosarcoma protuberans: how wide should we resect? Ann Surg Oncol 2010;17:2112-2118.

**Note: All recommendations are category 2A unless otherwise indicated.**

## 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 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 not given previously, and further resection is not feasible.**
- **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.**

**Note: All recommendations are category 2A unless otherwise indicated.**

## ABBREVIATIONS

| | |
|---|---|
| **CAP** | **College of American Pathologists** |
| **DFSP** | **dermatofibrosarcoma protuberans** |
| **FISH** | **fluorescence in situ hybridization** |
| **FS-DFSP** | **fibrosarcomatous transformation of DFSP** |
| **H&E** | **hematoxylin and eosin** |
| **H&P** | **history and physical** |
| **HPF** | **high-power field** |
| **PCR** | **polymerase chain reaction** |
| **PDEMA** | **peripheral and deep en face margin assessment** |

| NCCN Categories of Evidence and Consensus | |
|---|---|
| **Category 1** | Based upon high-level evidence (≥1 randomized phase 3 trials or high-quality, robust meta-analyses), there is uniform NCCN consensus (≥85% support of the Panel) that the intervention is appropriate. |
| **Category 2A** | Based upon lower-level evidence, there is uniform NCCN consensus (≥85% support of the Panel) that the intervention is appropriate. |
| **Category 2B** | Based upon lower-level evidence, there is NCCN consensus (≥50%, but <85% support of the Panel) 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.

| NCCN Categories of Preference | |
|---|---|
| **Preferred intervention** | Interventions that are based on superior efficacy, safety, and evidence; and, when appropriate, affordability. |
| **Other recommended intervention** | Other interventions that may be somewhat less efficacious, more toxic, or based on less mature data; or significantly less affordable for similar outcomes. |
| **Useful in certain circumstances** | Other interventions that may be used for selected patient populations (defined with recommendation). |

All recommendations are considered appropriate.

# Discussion

This discussion corresponds to the NCCN Guidelines for Dermatofibrosarcoma Protuberans. Last updated: October 11, 2024

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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, DFSP 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 4-7.4%, respectively.[8,9]

## Guidelines Update Methodology

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

## Literature Search Criteria and Guidelines Update Methodology

Prior to the update of this version of the NCCN Clinical Practice Guidelines in Oncology (NCCN Guidelines®) for Basal Cell Skin Cancer, an electronic search of the PubMed database was performed to obtain key literature using the following search term: basal cell skin carcinoma. 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; Practice Guideline; 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 as discussed by the panel during the Guidelines update have been included in this version of the Discussion section. Recommendations for which high-level evidence is lacking are based on the panel's review of lower-level evidence and expert opinion.

## Sensitive/Inclusive Language

NCCN Guidelines strive to use language that advances the goals of equity, inclusion, and representation.[11] NCCN Guidelines endeavor to use language that is person-first; not stigmatizing; anti-racist, anti-classist, anti-misogynist, anti-ageist, anti-ableist, and anti-weight biased; and inclusive of individuals of all sexual orientations and gender identities. NCCN Guidelines incorporate non-gendered language, instead focusing on organ-specific recommendations. This language is both more accurate and more inclusive and can help fully address the needs of individuals of all sexual orientations and gender identities. NCCN Guidelines will continue to use the terms men, women, female, and male when citing statistics, recommendations, or data from organizations or sources that do not use inclusive terms. Most studies do not report how sex and gender data are collected and use these terms interchangeably or inconsistently. If sources do not differentiate gender from sex assigned at birth or organs present, the information is presumed to predominantly represent cisgender individuals. NCCN encourages researchers to collect more specific data in future studies

and organizations to use more inclusive and accurate language in their future analyses.

## Evaluation

Histologically, DFSP typically presents as a storiform or fascicular proliferation of bland spindled cells that extends from the dermis into the subcutis.[12,13] Virtually all cases are CD34-positive and factor XIIIa-negative with rare exceptions.[14,15] Currently, no synoptic reporting is recommended. Preliminary workup for DFSP consists of history and physical (H&P) 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. Staining with CD34,[15,16] factor XIIIa,[14,17] and other immunomarkers such as stromelysins 3, nestin, apolipoprotein D, and cathepsin K,[18-21] might be useful in such instances. 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-DFSP) change have prognostic significance in DFSP.[22,23] Studies in the biomedical literature both support[24-32] the connection between FS-DFSP and an increased risk of local recurrence, lower time to recurrence, and increased risk of metastasis, and refute[33,34] this notion. A systematic review of 225 patients with FS-DFSP and 1422with DFSP 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 FS-DFSP.[35] Overall, FS-DFSP is associated with a metastatic risk range of 10 - 23.5%.[23,26,36] The NCCN Panel recommends that the debulking specimens from all excisions should be examined to identify fibrosarcomatous (FS) transformation of DFSP. If FS transformation or other sarcoma subtypes are found, multidisciplinary consultation for consideration of further treatment and surveillance is recommended. Clinicians should consult the NCCN Guidelines for Soft Tissue Sarcoma for multimodal therapy and surveillance considerations including CT of draining nodal basin and chest.

After DFSP confirmation, additional workup may include a complete skin examination as well as consideration of preoperative MRI with contrast for treatment planning if there is suspicion of extensive subcutaneous extension. 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.[37,38]

## 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).[39] 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, these surgical techniques 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.[38] 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. Additionally, tumor mutation analysis and neoadjuvant imatinib can be considered options for unresectable or borderline resectable disease. Consider neoadjuvant imatinib for patients in whom DFSP resection with negative margins may result in unacceptable functional or cosmetic outcomes.[40] 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, Radiation therapy (RT) is not recommended.

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,[41-43] 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%).[44-53]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 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 has occasionally been used as a primary therapeutic modality for DFSP along with other therapies,[57-59] but it is most beneficial as adjuvant therapy after surgery.[57-64] In a single-institution retrospective review of 53 patients with DFSP treated with surgery and preoperative or postoperative RT, local control was 93% and actuarial overall survival was 98% at 10 years.[34] Another small patient series reported that 86% of patients DFSP was 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 when Mohs or other forms of PDEMA are not used, RT can be considered if margins are deemed narrow by the treating physicians. 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 platelet-derived growth factor receptor (PDGFRs). DFSP is characterized by a translocation between chromosomes 17 and 22 [t(17;22)(q22;q13)] resulting in the overexpression of PDGFR β.[66-68] These findings suggest that targeting PDGFRs may be an effective treatment for DFSP. In published results, imatinib mesylate, a protein tyrosine kinase inhibitor, has shown clinical activity against DFSP,[40,69-73] 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 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] 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.[40]

## 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. Consider MRI surveillance for deeply invasive disease or other concerns related to recurrence.

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. It is recommended that the tumor mutation is confirmed with fluorescence in-situ hybridization (FISH) for *PDGRF* translocation.

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

 

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