Dermatofibrosarcoma Protuberans, Version 1.2025

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Abstract

Dermatofibrosarcoma protuberans (DFSP) is a rare cutaneous soft tissue sarcoma and affects an estimated 1,500 people annually in the United States. DFSP frequently exhibits extensive local infiltration. Initial treatment is through surgical excision, and care should be taken to ensure that negative margins are achieved to minimize recurrence. Although DFSP has a reported high rate of recurrence, metastasis is more uncommon. Fibrosarcomatous DFSP is an aggressive variant with an increased risk for local recurrence and metastasis. If achieving negative margins or resection is not feasible, radiation therapy or systemic treatment are options that may be considered by a multidisciplinary team. The NCCN Clinical Practice Guidelines in Oncology (NCCN Guidelines) outline recommended treatment options available for DFSP.

J Natl Compr Canc Netw 2025;23(1):e250001
doi:10.6004/jnccn.2025.0001

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 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 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”;

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"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 NCCN 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; antiracist, anticlassist, antimisogynist, antiageist, antiableist, and anti–weight biased; and inclusive of individuals of all sexual orientations and gender identities. NCCN Guidelines incorporate nongendered 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 examination and biopsy (Figure 1). It should be noted that this tumor is frequently misdiagnosed due to inadequate tissue sampling resulting from shallow biopsy. As

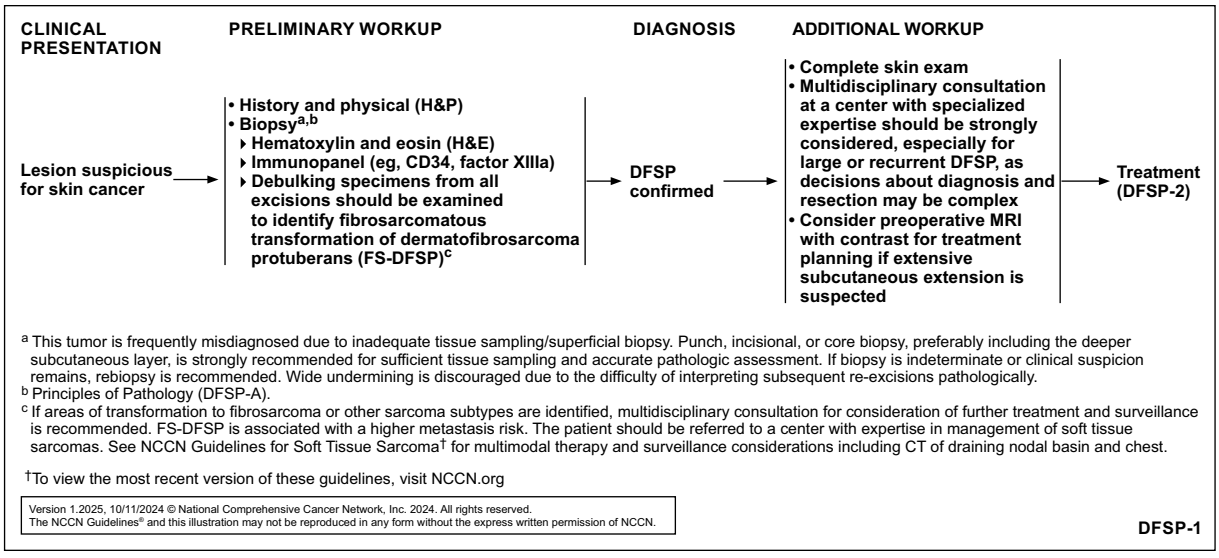


Figure 1. DFSP-1. NCCN Clinical Practice Guidelines in Oncology (NCCN Guidelines) for Dermatofibrosarcoma Protuberans, Version 1.2025.

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.

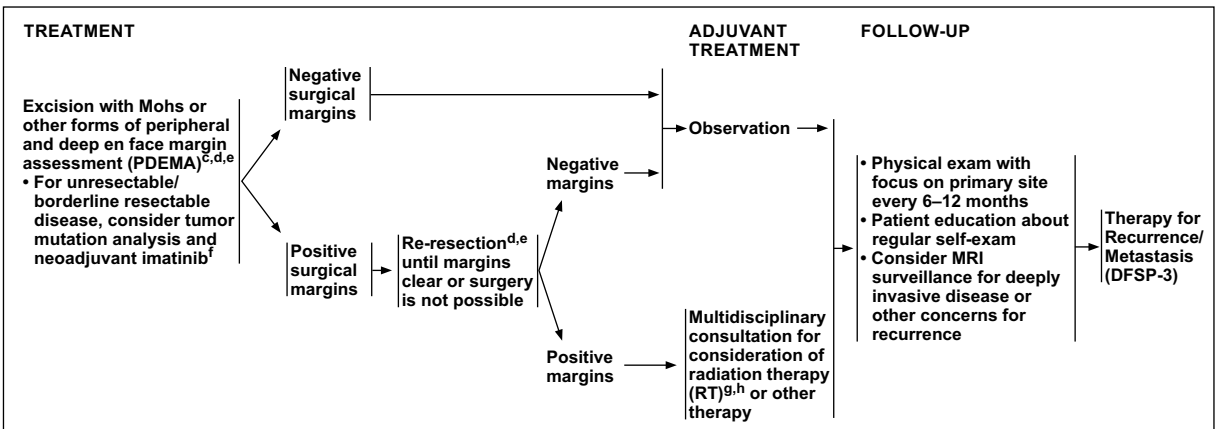
In most cases, examination of hematoxylin and eosin–stained specimens by light microscopy results in an unequivocal diagnosis. However, differentiation of DFSP from dermatofibroma can be difficult at times. Staining with CD34,[15,16] factor XIIIa,[14,17] and other immunomarkers such as stromelysins 3, 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.

Whether the histologic features of a high mitotic rate or evidence of fibrosarcomatous (FS-DFSP) change have prognostic significance in DFSP is unclear.[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 1,422 with 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 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 (available at NCCN.org) 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 and consideration of preoperative MRI with contrast for treatment planning if there is suspicion of extensive subcutaneous extension. Because 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 because it may optimize clinical and reconstructive outcomes.[37,38]

Treatment

Initial treatment of DFSP is surgical (Figure 2). Because of its proclivity for irregular and frequently deep subclinical extensions, every effort should be made to completely remove this tumor at 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 (See NCCN Guidelines for Squamous Cell Skin Cancer – Principles of PDEMA Technique, available at NCCN.org).[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 nonvisualized areas may be the subject of multidisciplinary discussion. If PDEMA is unavailable, WLE can be considered. Wide undermining is



[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.

†To view the most recent version of these guidelines, visit NCCN.org

DFSP-2

Figure 2. DFSP-2. NCCN Clinical Practice Guidelines in Oncology (NCCN Guidelines) for Dermatofibrosarcoma Protuberans, Version 1.2025.

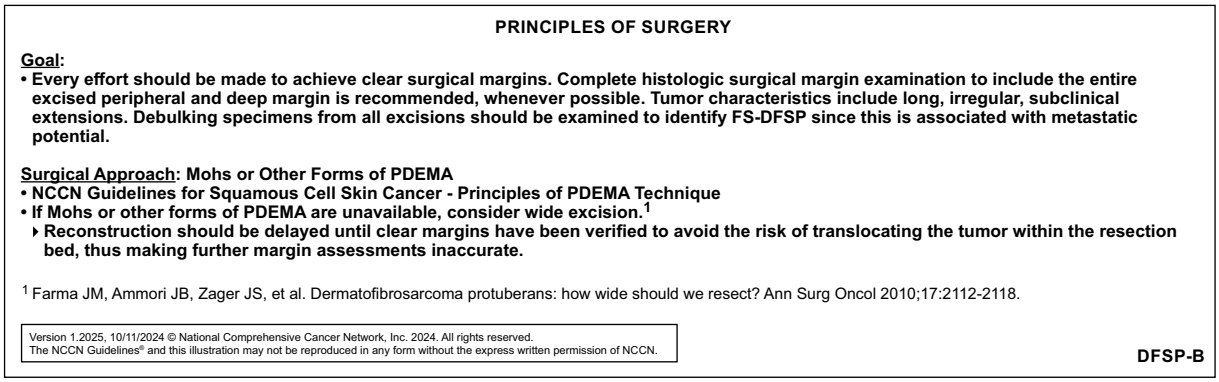


Figure 3. DFSP-B. NCCN Clinical Practice Guidelines in Oncology (NCCN Guidelines) for Dermatofibrosarcoma Protuberans, Version 1.2025.

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 is not recommended.

If initial surgery yields positive margins, reresection is recommended whenever possible, with the goal of achieving clear margins (Figure 3). 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 radiation therapy, local control was 93% and actuarial overall survival was 98% at 10 years.[34] Another small patient series reported that 86% of patients with DFSP treated with postoperative radiation therapy remained disease-free at a median follow-up of 10.5 years.[63] In a systematic review and meta-analysis of adjuvant radiation therapy 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, radiation therapy can be considered if margins are deemed narrow by the treating physicians. Radiation therapy can be considered for the treatment of positive margins if not given previously and further resection is not feasible (Figure 4).

DFSP can be treated by targeted platelet-derived growth factor receptors (Figure 5). 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.[66–68] These findings suggest that targeting platelet-derived growth factor receptors may be an effective treatment of DFSP. In published results, imatinib mesylate, a protein tyrosine kinase inhibitor, has shown clinical activity against DFSP,[40,69–73] which

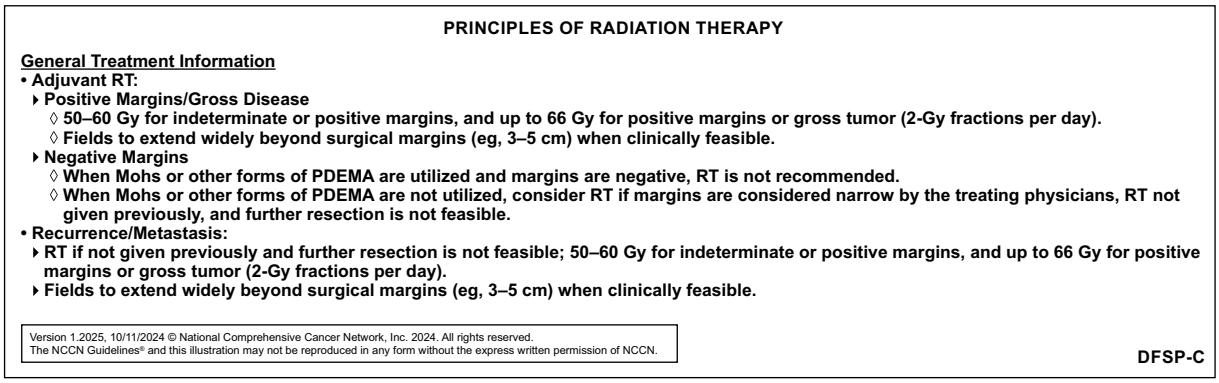


Figure 4. DFSP-C. NCCN Clinical Practice Guidelines in Oncology (NCCN Guidelines) for Dermatofibrosarcoma Protuberans, Version 1.2025.

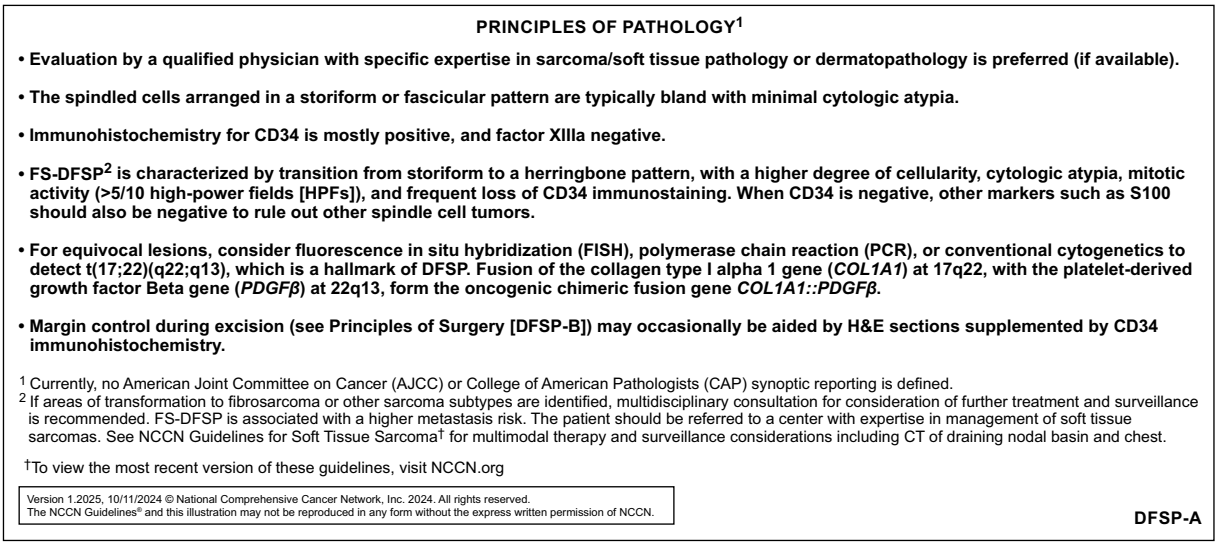


Figure 5. DFSP-A. NCCN Clinical Practice Guidelines in Oncology (NCCN Guidelines) for Dermatofibrosarcoma Protuberans, Version 1.2025.

has led to its approval by the US FDA for the treatment of unresectable, recurrent, and/or metastatic DFSP in adult patients. It is still unclear whether 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 rebiopsy of any suspicious regions. Although metastatic disease is rare, a guided history and physical 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 (Figure 6). Adjuvant radiation therapy may be considered after surgery. For patients who are not surgical candidates, radiation therapy alone is an option if not given previously. Imatinib mesylate should be considered in cases where the disease is unresectable after 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 for *PDGRF* translocation.

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

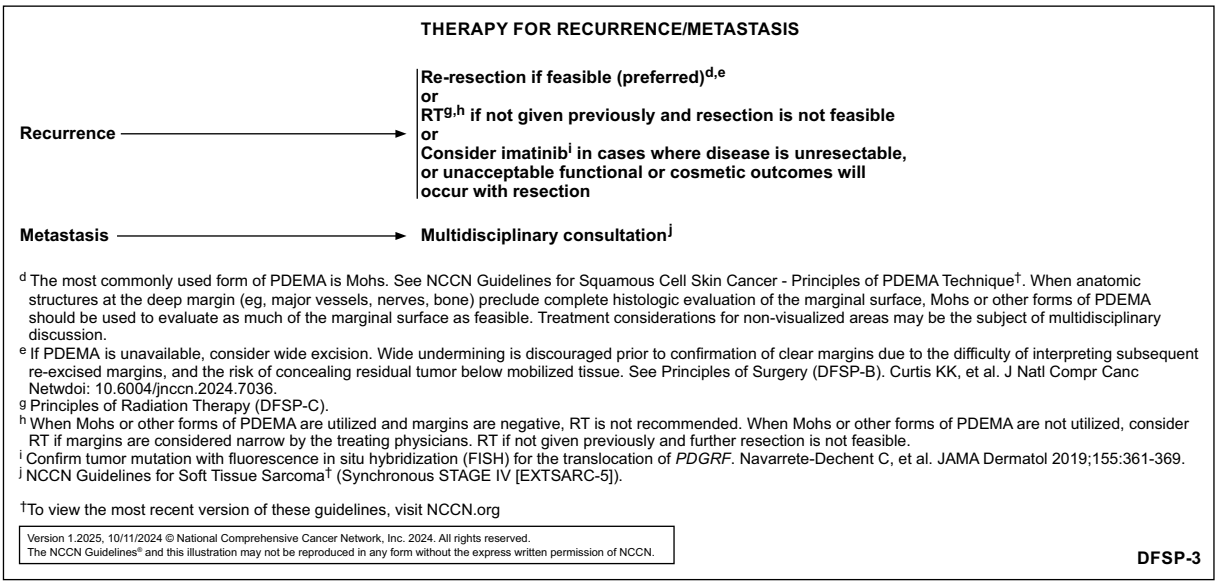


Figure 6. DFSP-3. NCCN Clinical Practice Guidelines in Oncology (NCCN Guidelines) for Dermatofibrosarcoma Protuberans, Version 1.2025.

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