

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.

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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 DFSP⁶ 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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To view disclosures of external relationships for the NCCN Guidelines panel, go to <https://www.nccn.org/guidelines/guidelines-panels-and-disclosure/disclosure-panels>

The complete and most recent version of these guidelines is available free of charge at NCCN.org.

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.

NCCN recognizes the importance of clinical trials and encourages participation when applicable and available.
Trials should be designed to maximize inclusiveness and broad representative enrollment.

PLEASE NOTE

The NCCN Guidelines® are a statement of evidence and consensus of the authors regarding their views of currently accepted approaches to treatment. Any clinician seeking to apply or consult the NCCN Guidelines® is expected to use independent medical judgment in the context of individual clinical circumstances to determine any patient's care or treatment. The National Comprehensive Cancer Network® (NCCN®) makes no representations or warranties of any kind regarding their content, use, or application and disclaims any responsibility for their application or use in any way.

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.

“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, antismisogynist, 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

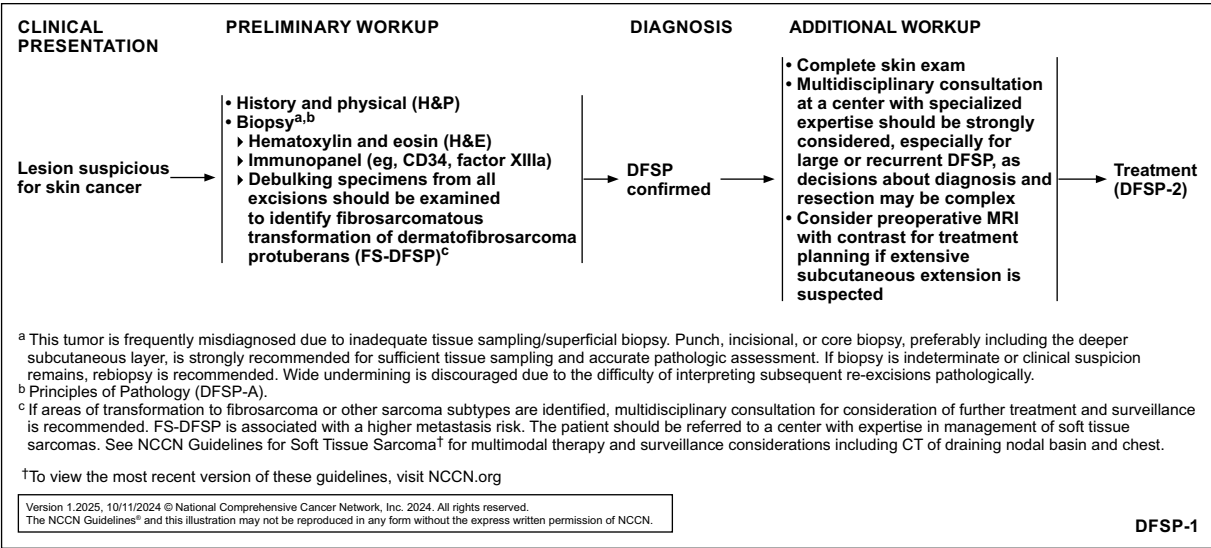


Figure 1. DFSP-1. NCCN Clinical Practice Guidelines in Oncology (NCCN Guidelines) for Dermatofibrosarcoma Protuberans, Version 1.2025.