Dermatofibrosarcoma Protuberans

Wide Local Excision Versus Mohs Micrographic Surgery

John T. Mullen, MD

KEYWORDS

• Dermatofibrosarcoma protuberans • Mohs surgery • Wide local excision
• Recurrence • Cutaneous sarcoma

KEY POINTS

- Dermatofibrosarcoma protuberans (DFSP) is a rare dermal soft tissue sarcoma characterized by a typically indolent clinical course.
- Complete surgical resection is the mainstay of treatment, and this is accomplished with either a standard wide local excision (WLE) or with Mohs micrographic surgery (MMS), as long as a comprehensive pathologic examination of the margins is completed before reconstruction of the defect.
- MMS is the ideal surgical approach for relatively small DFSPs in cosmetically sensitive areas (ie, face, scalp, or neck), where tissue preservation is critical to achieve optimal cosmetic and functional outcomes.
- WLE with a 1.0- to 1.5-cm margin width is the ideal approach for most DFSPs on the trunk or extremities, because it is likely to achieve complete tumor clearance with excellent cosmetic and functional outcomes in a single stage.

INTRODUCTION

Dermatofibrosarcoma protuberans (DFSP) is a rare dermal soft tissue sarcoma characterized by a typically indolent clinical course. Although transformation to a high-grade fibrosarcoma is possible, particularly in the case of recurrent DFSP, by far the greatest clinical challenge in the management of DFSP is achieving local control. Because DFSP arises in the dermis and invades radially through preexisting collagen bundles and deeply along connective tissue septae,[1] its extent of invasion is often difficult to clinically appreciate, and thus determining the appropriate width

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Division of Surgical Oncology, Massachusetts General Hospital, Harvard Medical School, 55 Fruit Street, Yawkey 7B, Boston, MA 02114, USA
E-mail address: jmullen@mgh.harvard.edu

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of the margins of resection is challenging. There has been vigorous debate in the literature as to the optimal surgical approach to these tumors, with some groups advocating for radical resection with defined margins of excision, and others advocating for Mohs micrographic surgery (MMS).

Ultimately, there is no "one size fits all" surgical technique for the treatment of DFSP. Indeed, the choice between wide local excision (WLE) and MMS for DFSP should be governed by the attainment of the following three goals: (1) to completely excise the tumor with negative margins, which is tantamount to cure, because these tumors very rarely metastasize; (2) to preserve function, optimize cosmesis, and minimize the morbidity of resection; and (3) to minimize the cost and inconvenience to the patient and the health care system at large. Both WLE and MMS are capable of achieving the first goal, such that the choice of surgical approach may be driven more by individual patient and tumor factors that make the attainment of the second and third goals important.

EPIDEMIOLOGY

DFSP is a rare sarcoma, accounting for less than 0.1% of all malignancies and between 2% and 6% of all soft tissue sarcomas,[2] although it is the most common sarcoma of the skin. Rouhani and colleagues,[2] in an analysis of the Surveillance, Epidemiology, and End Results database from 1992 to 2004, reported that the incidence of DFSP was 4.5 cases per million person-years and that DFSP comprised 18.4% of all cutaneous sarcomas during that time period. It typically occurs in the third and fourth decades of life, has an equal gender distribution, and has a higher incidence among black persons than seen for other cutaneous sarcomas.

CLINICAL PRESENTATION AND NATURAL HISTORY

DFSP most commonly presents as a slow-growing, asymptomatic nodular or plaque-like lesion that may have a violaceous or reddish brown appearance. The tumor has a hard consistency and is fixed to the dermis but is usually freely movable over the underlying fascia and muscle. Over time, which can vary from months to years, DFSP may grow radially to generate a larger plaque and vertically to generate multiple nodules within the plaque, from which its name "protuberans" is derived. The average tumor diameter is on the order of a few centimeters, although neglected tumors can grow to much larger sizes. DFSP can occur anywhere on the body, although the most common locations are the extremities, trunk, and head and neck. Diagnostic delays are common with these tumors, which are often mistaken as dermatofibromas, sebaceous cysts, lipomas, and scars.

Despite the frequent, long delays in diagnosis, distant metastasis is exceedingly rare, with a reported rate of 2% to 4%.[2,3] The main site of distant recurrence is the lung, via hematogenous tumor spread, and in nearly all cases metastases are preceded by multiple local recurrences or in the context of transformation to a higher-grade fibrosarcoma.[3] As with most other soft tissue sarcomas, DFSP rarely metastasizes to the regional lymph nodes, having been reported to occur in 1% or fewer cases.[4]

PATHOLOGY

Grossly, DFSP appears as a solitary, poorly circumscribed, gray-white mass that infiltrates the dermis and subcutaneous tissue (**Fig. 1**). Histologically, the tumor is composed of a dense, uniform array of cells with spindle-shaped nuclei embedded within collagen and may demonstrate a cartwheel or storiform pattern. Tumor cells