# Atrophic Dermatofibrosarcoma Protuberans: A Case Report and Reappraisal of the Literature

Daniel J. Sheehan, MD; Vandana Madkan, MD; William A. Strickling, MD; Christopher M. Peterson, MD

GOAL

To gain a thorough understanding of dermatofibrosarcoma protuberans (DFSP)

OBJECTIVES

Upon completion of this activity, dermatologists and general practitioners should be able to:

1. Explain the clinical presentations of DFSP.
2. Discuss the histology of DFSP.
3. Examine the treatment options for DFSP.

CME Test on page 251.

This article has been peer reviewed and approved by Michael Fisher, MD, Professor of Medicine, Albert Einstein College of Medicine. Review date: September 2004.

This activity has been planned and implemented in accordance with the Essential Areas and Policies of the Accreditation Council for Continuing Medical Education through the joint sponsorship of Albert Einstein College of Medicine and Quadrant HealthCom, Inc. Albert Einstein College of Medicine is accredited by the ACCME to provide continuing medical education for physicians.

Albert Einstein College of Medicine designates this educational activity for a maximum of 1 category 1 credit toward the AMA Physician's Recognition Award. Each physician should claim only that credit that he/she actually spent in the activity.

This activity has been planned and produced in accordance with ACCME Essentials.

Drs. Sheehan, Madkan, Strickling, and Peterson report no conflict of interest. The authors report no discussion of off-label use. Dr. Fisher reports no conflict of interest.

*Dermatofibrosarcoma protuberans (DFSP) is an uncommon cutaneous malignancy. Unusual presentations described as atrophic have been documented. A case of DFSP with both clinical and histologic atrophy is presented, and all cases purporting atrophy with this tumor are reviewed. Meaningful trends are extracted from this data. In addition, the imprecise use of the term* atrophic *in regard to DFSP is clarified. We maintain that the variant of atrophic DFSP that mimics atrophoderma or anetoderma, as in this case, is the rarest variant of atrophic DFSP. Atrophic DFSP should be in the differential for depressed lesions on the trunks of women or on the lower extremities of children.*

*Cutis.* 2004;74:237-242.

Accepted for publication June 29, 2004.

Drs. Sheehan and Peterson are from the Section of Dermatology, Medical College of Georgia, Augusta. Dr. Sheehan is Chief Resident and Dr. Peterson is Assistant Professor of Dermatology. Dr. Madkan is an intern, Department of Medicine, University of Maryland Medical System, Baltimore. Dr. Strickling is Assistant Chief, Dermatology Service, Dwight David Eisenhower Army Medical Center, Augusta.

Reprints: Christopher M. Peterson, MD, Section of Dermatology, Medical College of Georgia, 1004 Chafee Ave, Augusta, GA 30904 (e-mail: kmonroe@mail.mcg.edu).

Dermatofibrosarcoma protuberans (DFSP) is an uncommon cutaneous tumor of intermediate malignancy that is known to be locally aggressive.[1] It is thought to be of fibrohistiocytic origin and accounts for 1% of all soft tissue malignancies.[2] DFSP occurs in men and women with equal incidence, mostly on the trunk and less commonly on the proximal extremities, with a higher incidence in Caucasians. Most patients present between the second and fifth decades, though the disease can be noted in any age group and any anatomic location.[3]

Four clinical variants of early DFSP are thought to exist: confluent nodular lesions forming a sclerotic plaque, keloidlike sclerotic plaque, tumor, and atrophic plaque.[4,5] The usual clinical appearance of DFSP at an early stage is as a slow-growing, asymptomatic plaque or nodule. The tumor's gross pathology is of a gray-white mass involving the dermis and subcutis; histologically, it is a poorly circumscribed infiltrative spindle cell tumor. The spindle cells, which are $CD34^+$, are arranged in a storiform pattern described as a dense, poorly circumscribed, monomorphic cell proliferation arranged in fascicles that often reach and infiltrate the hypodermis.[6,7] The 3 recognized histologic variants of DFSP are fibrosarcomatous, plaquelike, and myxoid forms.[8] The infiltrating pattern accounts for the tumor's common recurrence after excision and the need for wide excision or Mohs micrographic surgery for curative removal.[2,3,9-11] One recent review of atrophic DFSP implicated chromosomal translocations or the formation of supernumerary ring chromosomes in the pathogenesis of DFSP.[12] These chromosomal events result in the fusion of the collagen type 1α1 gene with the platelet-derived growth factor-β chain gene. How this mutation could contribute to the atrophic phenotype is unknown.[12] This review documents only 25 cases of atrophic DFSP in the literature. We present a case involving a woman who had an atrophic DFSP treated successfully with Mohs micrographic surgery and review 35 cases in the literature regarding this rare presentation of DFSP.

## Case Report

A 41-year-old white woman presented with a 12-year history of a well-demarcated, scalloped, markedly depressed, reddish brown, slightly firm, 4×5-cm plaque on the right chest (Figure 1). On initial evaluation, the patient denied prior trauma to the area or any past manipulation of the lesion. The patient's preoperative clinical differential diagnosis included atrophoderma of Pasini and Pierini, anetoderma, morphea, and lipoatrophy.

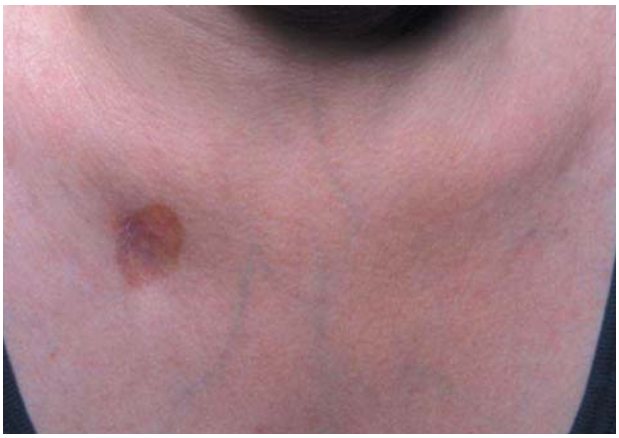

**Figure 1.** Reddish brown depressed plaque on the chest.

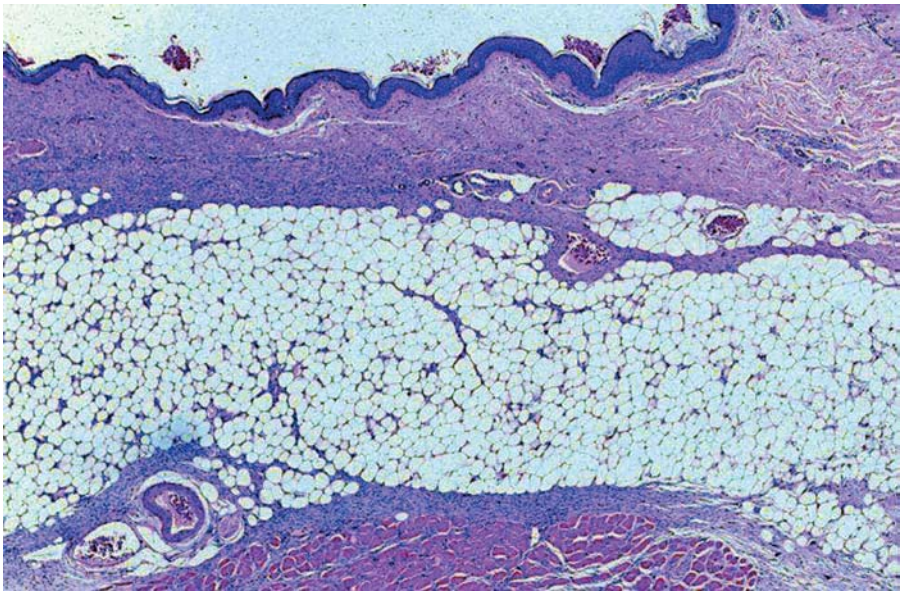

**Figure 2.** Dermal atrophy with infiltrative spindle cell neoplasm (H&E, original magnification ×4).

A punch biopsy of the lesion was performed. Results revealed a spindle cell neoplasm that was thought to be a dermatofibroma. The entire lesion was subsequently excised. Histopathology results of the excision specimen demonstrated atrophy of the dermis and a permeative spindle cell neoplasm with scattered multinucleated giant cells involving the dermis and underlying soft tissue with a few areas of a storiform growth pattern (Figure 2). The lesion also was diffusely immunoreactive for CD34 and extended to the margins of the specimen. A diagnosis of DFSP was made, and the patient was treated with Mohs micrographic surgery. Three stages of surgery were performed, and the final defect was 7.0×8.5 cm. The patient elected to allow the wound to heal by secondary intention. There was no evidence of recurrence 12 months after the procedure.

## Comment

The atrophic presentation of DFSP is the rarest variant of this infrequent neoplasm. Although this subject was recently reviewed, there are several other important issues in the literature that need to be clarified regarding this unusual phenotype.[12] For a lesion to be classified as atrophic, there should be both clinical and histologic evidence of atrophy. When the epidermis is atrophic, the result is shiny smooth skin clinically and flattening of the rete ridges histopathologically. Conversely, when the dermis or subcutaneous tissue is atrophic, the result is a loss in skin thickness and a corresponding loss of collagen bundles or subcutaneous fat histopathologically.[13]

Epidermal atrophy is commonly noted histologically in DFSP cases that present in the usual fashion.[14,15] Only rarely has DFSP presented as a clinically depressed lesion.[16-20] Even more rare is a documentation of dermal atrophy.[12,21] We report a case of DFSP in which the initial lesion was an atrophic depression of the skin lacking nodularity and histologically demonstrating significant dermal atrophy.

A number of cases have been reported describing atrophic presentations of DFSP (Table 1).[4,5,8,12,16-26] However, as Davis and Sanchez[8] noted, rarely does a so-called atrophic DFSP present as a visible depression in the skin.[16-21] Clearly, there is some discrepancy in the literature as to the nature of atrophy in relation to the clinical presentation of DFSP. Some authors have even speculated that the atrophic variant of DFSP is nothing more than an early presentation of the more usual DFSP prior to the formation of nodules.[4] However, the fact that our patient had her lesion for 12 years prior to treatment without the formation of a nodular component indicates that the atrophic variant of DFSP is a real entity.

Table 2 demonstrates the sex and anatomic location of the patients in the 35 reported cases of DFSP described as atrophic or morpheaform, including the present case. The majority (20/35) of cases have been in female patients, and the most common location in both sexes has been the trunk (24/35). The next most common location was the lower extremity (7/35).

Table 3 documents the age and anatomic location of the 9 cases clinically described as resembling lipoatrophy, atrophoderma or anetoderma, or that were significantly clinically depressed based on clinical photographs, ie, the cases most similar to the one we now present.[4,16,19-21,25] Three of the 9 cases were on the trunks of females aged 16 to 41 years. Interestingly, 4 of the remaining 6 cases were on the lower extremities of children aged 18 months to 16 years.

A number of conclusions can be drawn from this data. First, atrophic variants of DFSP tend to occur most often on the trunk, as do the usual variants of DFSP. However, the atrophic variants tend to occur more often in females as opposed to the standard presentation of DFSP, which occurs with equal frequency in males and females. In addition, a distinction exists in the presentation of atrophic lesions, which can have either a morpheaform or a more clinically depressed appearance, mimicking such disorders as anetoderma, atrophoderma, or lipoatrophy. We report such a case mimicking atrophoderma, and we contend that this is the rarest presentation of atrophic DFSP.

One final entity in the differential diagnosis of a depressed plaque on the trunk would be an atrophic dermatofibroma. The dermatofibroma, a well-known and benign cousin of DFSP, is often noted to have some dermal atrophy grossly with lateral pressure, described as a dimple sign.[27] As Requena and Reichel[28] pointed out, even when there is a central depression over a dermatofibroma without lateral pressure, no true loss of the dermis is seen histopathologically. As such, this central depression doesn't represent true dermal atrophy.[28] However, the dermatofibroma also can present as a depressed lesion demonstrating thinning of the dermis histologically, and this variety of dermatofibroma has been described as atrophic.[28-31] Although a recently published article recommended considering a diagnosis of atrophic dermatofibroma in the case of "atrophic, depressed lesions on the upper body of middle-aged women,"[31] we would maintain that atrophic DFSP also should be considered in the differential diagnosis.

Table 1.

## Reported Cases of Atrophic or Morpheaform DFSP*

| Reference | Age (Sex) | Location | Clinical Description | Histopathology |
|---|---|---|---|---|
| Martin et al[4] | 13 y (M) | Calf | 4 cm, atrophic | DFSP, CD34+ |
| | 11 y (F) | Thigh | Depressed, morpheaform | DFSP, CD34+ |
| | 16 y (F) | Calf | Hard, depressed | DFSP, CD34+ |
| | 18 mo (M) | Ankle | 6.5×3.5 cm, lipoatrophy | DFSP, CD34+ |
| | 3 y (F) | Periumbilical | 5×4 cm, atrophic | DFSP, CD34+ |
| | 21 y (M) | Epigastric | Multinodular, indurated | DFSP, CD34+ |
| Marini et al[5] | 16 (F) | Leg | Atrophic | DFSP, CD34+ |
| Davis and Sanchez[8] | 53 (M) | Clavicular | 7×5-cm, firm, exophytic nodule | DFSP, CD34+ |
| Young and Albertini[12] | 65 y (M) | Back | 2.5×3.0 cm hyperpigmented, atrophic | DFSP, CD34+, thinned dermis |
| Page and Assaad[16] | 21 y (F) | Back | 1×1.5 cm, anetodermalike | DFSP |
| | 27 y (F) | Back | 2×3 cm, atrophic | DFSP |
| Ashack et al[17] | 25 y (M) | Shoulder | 2.5×1.5 cm, violaceous, slightly depressed | DFSP |
| Annessi et al[18] | 16 y (F) | Periumbilical | 3×4 cm depressed, violaceous | DFSP |
| Chuan et al[19] | 24 y (F) | Infraorbital | Bluish, atrophic, Ota nevus | DFSP |
| Fujimoto et al[20] | 21 y (F) | Subclavicular | 2×3 cm, anetoderma vs morpheaform | DFSP, CD34+ |
| Teixeira et al[21] | 40 y (M) | Chest | 6 cm, depressed, orange-brown | DFSP, CD34+, thinned dermis |
| Zelger et al[22] | 40 y (F) | Periumbilical | Lymphocytoma | DFSP, CD34+ |
| | 55 y (F) | Shoulder | DFSP vs lymphoma | DFSP, CD34+ |
| | 42 y (F) | Groin | BCC vs scar | DFSP, CD34+ |
| Lambert et al[23] | 15 y (M) | Supraclavicular | 5×7 cm, morpheaform | DFSP |
| | 50 y (F) | Abdomen | 3×6 cm, depressed, firm | DFSP |
| | 37 y (M) | Chest | Morpheaform | DFSP |
| | 15 y (M) | Shoulder | 3×5 cm, morpheaform | DFSP |
| | 28 y (M) | Back | 2.5 cm, morpheaform | DFSP |
| McKee and Fletcher[24] | 7 y (F) | Arm | Flattened or atrophic pale to red-blue plaques | DFSP |
| | 14 mo (M) | Back | | DFSP |
| | 12 y (F) | Back | | DFSP |
| | 9 y (F) | Shoulder | | DFSP |
| | 6 y (M) | Shoulder | | DFSP |
| | 5 y (M) | Back | | DFSP |
| | 10 y (F) | Buttock | | DFSP |
| | 10 y (F) | Foot | | DFSP |
| Bouyssou-Gauthier et al[25] | 18 mo (M) | Ankle | 3.5×6.5 cm, morphea vs lipoatrophy | DFSP, CD34+ |
| See et al[26] | 72 y (F) | Back | 3×1.5 cm, atrophic, erythematous | DFSP, CD34+ |
| Present case | 41 y (F) | Chest | 4×5-cm scalloped, DFSP, markedly depressed plaque | CD34+, thinned dermis |

*DFSP indicates dermatofibrosarcoma protuberans; M, male; F, female; BCC, basal cell carcinoma.

Table 2.

**Atrophic or Morpheaform Dermatofibrosarcoma Protuberans by Sex and Anatomic Distribution**

| Sex | Head and Neck | Trunk | Upper Extremities | Lower Extremities | Buttocks or Groin |
|---|---|---|---|---|---|
| Female | 1 | 12* | 1 | 4 | 2 |
| Male | 0 | 12 | 0 | 3 | 0 |

*Includes the present case.

Table 3.

**Atrophic DFSP Cases That Either Were Reported to Resemble Lipoatrophy, Atrophoderma or Anetoderma, or Were Significantly Depressed*[4,16,19-21,25]**

| Sex | Head and Neck | Trunk | Upper Extremities | Lower Extremities | Buttocks or Groin |
|---|---|---|---|---|---|
| Female | 1 | 3† | 0 | 1 | 0 |
| Male | 0 | 1 | 0 | 3 | 0 |

*Includes the first, third, and fourth cases from Martin et al[4] and the first case from Page and Assaad.[16]
†Includes the present case.

One possibility to clarify the imprecision in the literature in relation to the clinical presentation of DFSP would be to eliminate protuberans from the name, thereby recognizing that some lesions that are histologically proven to be DFSP can present without nodularity and with epidermal or dermal atrophy. This change in the nomenclature was first proposed by Lambert et al[23] and was reiterated by Page and Assaad.[16] Perhaps a greater awareness that DFSP can present as atrophic lesions without nodules could lead to earlier diagnosis and decreased morbidity with smaller curative surgeries when the lesions are recognized at an earlier stage.

The atrophic variant of DFSP does not carry a different prognosis compared with the traditional variant; Mohs micrographic surgery is still the treatment of choice, providing a low rate of recurrence. Atrophic DFSP should be kept in the differential diagnosis for atrophic, depressed lesions, particularly those seen on the trunks of women or on the lower extremities of children.

## REFERENCES


1. Guillen DR, Cockerell CJ. Cutaneous and subcutaneous sarcomas. *Clin Dermatol.* 2001;19:262-268.
2. Nouri K, Lodha R, Jimenez G, et al. Mohs micrographic surgery for dermatofibrosarcoma protuberans: University of Miami and NYU experience. *Dermatol Surg.* 2002;28:1060-1064.
3. Goldberg DJ, Maso M. Dermatofibrosarcoma protuberans in a 9-year-old child: treatment by Mohs micrographic surgery. *Pediatr Dermatol.* 1990;7:57-59.
4. Martin L, Combemale P, Dupin M, et al. The atrophic variant of dermatofibrosarcoma protuberans in childhood: a report of six cases. *Br J Dermatol.* 1998;139:719-725.
5. Marini M, Saponaro A, Magarinos G, et al. Congenital atrophic dermatofibrosarcoma protuberans. *Int J Dermatol.* 2001;40:448-450.
6. Kutzner H. Expression of the human progenitor cell antigen CD34 (HPCA-1) distinguishes dermatofibrosarcoma protuberans from fibrous histiocytoma in formalin-fixed, paraffin-embedded tissue. *J Am Acad Dermatol.* 1993;28:613-617.
7. Aiba S. Dermatofibrosarcoma protuberans expresses CD34. *J Am Acad Dermatol.* 1994;30:508.
8. Davis DA, Sanchez RL. Atrophic and plaquelike dermatofibrosarcoma protuberans. *Am J Dermatopathol.* 1998;20:498-501.
9. Ratner D, Thomas CO, Johnson TM, et al. Mohs micrographic surgery for the treatment of dermatofibrosarcoma protuberans. *J Am Acad Dermatol.* 1997;37:600-613.

10. Gloster HM. Dermatofibrosarcoma protuberans. *J Am Acad Dermatol*. 1996;35:355-374.
11. Dawes KW, Hanke CW. Dermatofibrosarcoma protuberans treated with Mohs micrographic surgery. *Dermatol Surg*. 1996;22:530-534.
12. Young RJ, Albertini JG. Atrophic dermatofibrosarcoma protuberans: case report, review, and proposed molecular mechanisms. *J Am Acad Dermatol*. 2003;49:761-764.
13. Freedberg IM, Eisen AZ, Wolff K, et al. *Fitzpatrick's Dermatology in General Medicine*. New York, NY: McGraw-Hill; 1999:27.
14. Taylor HB, Helwig EB. Dermatofibrosarcoma protuberans: a study of 115 cases. *Cancer*. 1962;962;15:717-725.
15. McPeak CJ, Cruz T, Nicastri AD. Dermatofibrosarcoma protuberans: an analysis of 86 cases—five with metastasis. *Ann Surg*. 1967;166:803-816.
16. Page EH, Assaad DM. Atrophic dermatofibroma and dermatofibrosarcoma protuberans. *J Am Acad Dermatol*. 1987;17:947-950.
17. Ashack RJ, Tejada E, Parker C, et al. A localized atrophic plaque on the back. *Arch Dermatol*. 1992;128:547-552.
18. Annessi G, Cimitan A, Girolomoni G, et al. Congenital dermatofibrosarcoma protuberans. *Pediatr Dermatol*. 1993;10:40-42.
19. Chuan MT, Tsai TF, Wu MC, et al. Atrophic pigmented dermatofibrosarcoma presenting as infraorbital hyperpigmentation. *Dermatology*. 1997;194:65-67.
20. Fujimoto M, Kikuchi K, Okochi H, et al. Atrophic dermatofibrosarcoma protuberans: a case report and review of the literature. *Dermatology*. 1998;196:422-424.
21. Teixeira F, Devlin M, Hung N, et al. An atrophic plaque on the chest. *Aust Fam Physician*. 2002;31:359-360.
22. Zelger BW, Ofner D, Zelger BG. Atrophic variants of dermatofibroma and dermatofibrosarcoma protuberans. *Histopathology*. 1995;26:519-527.
23. Lambert WC, Abramovits W, Gonzalez-Sevra A, et al. Dermatofibrosarcoma non-protuberans: description and report of five cases of a morpheaform variant of dermatofibrosarcoma. *J Surg Oncol*. 1985;28:7-11.
24. McKee PH, Fletcher CDM. Dermatofibrosarcoma protuberans presenting in infancy and childhood. *J Cutan Pathol*. 1991;18:241-246.
25. Bouyssou-Gauthier ML, Labrousse F, Longis B, et al. Dermatofibrosarcoma protuberans in childhood. *Pediatr Dermatol*. 1997;14:463-465.
26. See ACY, Kossard SS, Murrell DF. Dermatofibrosarcoma protuberans presenting as an atrophic red plaque. *Eur J Dermatol*. 2001;11:147-149.
27. Odom RB, James WD, Berger TG. *Andrews' Diseases of the Skin: Clinical Dermatology*. Philadelphia, Pa: WB Saunders; 2000:773.
28. Requena L, Reichel M. The atrophic dermatofibroma: a delled dermatofibroma. *J Dermatol*. 1995;22:334-339.
29. Beer M, Eckert F, Schmoeckel C. The atrophic dermatofibroma. *J Am Acad Dermatol*. 1991;25:1081-1082.
30. Kiyohara T, Kumakiri M, Kobayashi H, et al. Atrophic dermatofibroma. elastophagocytosis by the tumor cells. *J Cutan Pathol*. 2000;27:312-315.
31. Hendi A, Jukic DM, Kress DW, et al. Atrophic dermatofibroma: a case report and review of the literature. *Dermatol Surg*. 2002;28:1085-1087.