BRIEF ARTICLE

A Case of a Large, Painful Dermatofibrosarcoma Protuberans Arising from a Traumatic Scar

Daniel L. Fischer, DO, MA[1], Graham Litchman, DO, MS[1]

[1]St. John's Episcopal Hospital, Far, Rockaway, NY

ABSTRACT

Here we report a case of a patient with dermatofibrosarcoma protuberans (DFSP) that presented originally to the emergency department with chief complaint of 'chest pain arising from a chest wall mass'. This case is unique for multiple reasons including the fact that the DFSP caused this patient severe pain that radiated from the site of the mass to the patient's back. The lesion originated from a traumatic scar that the patient had obtained following a car accident two years prior to presentation. In addition, the DFSP was significantly larger in size than what has typically been reported in the literature. This case illustrates that DFSP has the potential to present with multiple atypical features.

INTRODUCTION

Dermatofibrosarcoma protuberans (DFSP) is a rare, locally aggressive and seldom metastatic soft tissue tumor usually confined to the dermis and subcutaneous tissues.[1] The first case of DFSP was described in 1924 and termed by Hoffman in 1925.[2] It accounts for less than 0.1% of all malignant neoplasms and 1% of soft tissue sarcomas.[3] Diagnosis requires skin biopsy with histochemical analysis and mainstay of treatment is surgery. It is most commonly seen among adults in their thirties, with a predominance in blacks.[4] Most lesions are no more than 5 cm in diameter and occur spontaneously.[5] Here we describe a case of DFSP that arose within a preexisting scar and grew to much larger proportions than what is typically observed.

CASE REPORT

Our case involves a 35-year-old Caucasian man who presented to the emergency department for 10/10 sharp, squeezing chest pain originating at the site of a left chest mass. The mass had formed out of an area of scar tissue that had developed after a traumatic motor vehicle accident. However, the tissue had formed a mass and grew exponentially over the past two months (**Figure 1**). During this time the mass became increasingly painful causing intermittent episodes of stabbing pain, radiating to the back, lasting about 20 seconds, and recurring throughout the day. The patient had not sought medical help during this time due to lack of insurance.

Physical examination was remarkable for a large, multilobulated, highly vascular chest wall mass localized to the left sternal border measuring approximately 10 by 12 centimeters (cm) with well-demarcated margins. The lower portion of the mass was firm to palpation while the upper portion was

soft. The mass was also highly sensitive to touch. The differential diagnosis included arteriovenous malformation, soft tissue hemangioma, and soft tissue sarcoma.

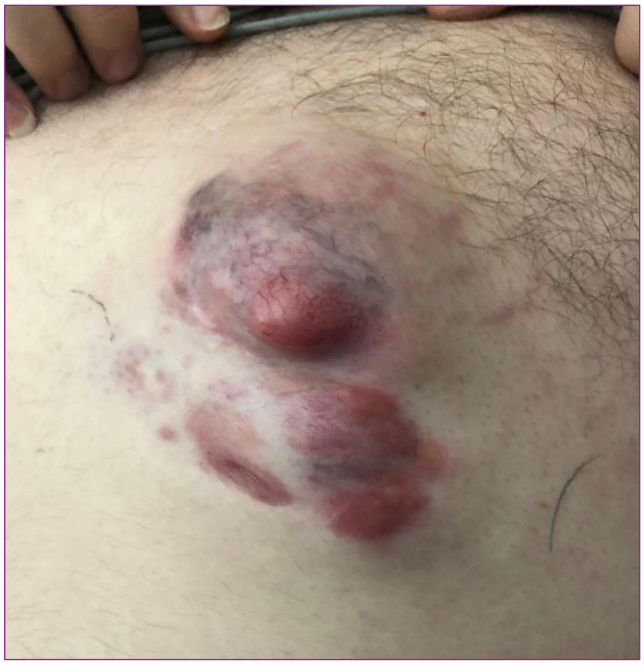

Figure 1. A large, highly vascular left-sided chest wall mass measuring 10 by 12 centimeters that was identified to be a DFSP on histology.

CT chest revealed an 11.3 by 8.8 by 6.3 cm left anterior chest wall mass located in the subcutaneous tissue with no evidence of muscle or bony invasion. MRI chest revealed multiple well-circumscribed, grape-like hypervascular masses in the subcutaneous soft tissue of the left anterior chest wall. There was predominant vascular supply from the intercostal and internal mammary chain vascular structures. No evidence of chest wall invasion was appreciated.

Ultrasound-guided core needle biopsy showed a relatively monotonous spindle cell proliferation with storiform architecture throughout. Immunohistochemistry showed positivity for CD34 and vimentin, and negative for S100, desmin, EMA, and CD68. Ki67 index was brisk and STAT-6 and Factor XIII were negative. The interpretation was read as spindle cell neoplasm of intermediate grade favoring DFSP.

DISCUSSION

Dermatofibrosarcoma protuberans typically presents as a slow growing, nontender plaque that with time develops an asymmetric and multinodular, red, yellow, or brown appearance.[1] DFSP most commonly appears on the trunk or extremities and less often on the head and neck but can appear almost anywhere.[3] Local recurrence following excision is fairly common.[4] With treatment, the prognosis is excellent (10-year survival rate of 99.1%).[4] DFSPs are typically asymptomatic which makes this case particularly unique as our patient was symptomatic with severe pain at the site of the lesion, and the prime reason for him seeking medical attention.

The cause of DFSP is not known but prior studies suggest that translocations between chromosomes 17 and 22 may play a role in their development. This translocation is thought to cause an upregulation of the platelet-derived growth factor beta polypeptide gene leading to cellular proliferation.[6] According to several studies, it is estimated that up to 10-20% of DFSP have arisen from prior trauma.[7-9] This was the case in our patient, as his malignancy had arisen from a scar that he had acquired after a motor vehicle injury two years prior to his presentation at our hospital.

Diagnosis of DFSP is confirmed via incisional or excisional biopsy. This can often be challenging if the lesion is highly vascular, as in our case. In this scenario, an MRI of the lesion was done to map the vascular regions and define tumor extension. This was followed by a core-needle biopsy without complication. Examination of hematoxylin

and eosin-stained specimens by light microscopy yields the diagnosis.[1] Pathology typically reveals monotonous spindle cell proliferation with storiform architecture throughout with positivity for CD34 and vimentin and negative staining for Factor XIII, S-100, CD68, and 5% positivity for Ki67.[10] This correlated with the histopathology findings in our patient.

When the diagnosis of a large DFSP is suspect, a multidisciplinary approach is recommended with tertiary center evaluation. The standard treatment for DFSP is surgical removal with Mohs micrographic surgery (MMS). This method is preferred over wide excision as it ensures complete removal of tumor margins and is appropriate in cases like DFSP in which the depth of extension is highly unpredictable. It also reduces the likelihood of tumor recurrence (1% with MMS; 7.3% with wide excision). In the rare scenario that DFSP has metastasized or is unresectable, chemotherapy with imatinib mesylate or radiation therapy may be used.[2] Close follow up is required post-treatment and evaluation ideally every 3-6 months due to the high recurrence rate of DFSP. Our patient was scheduled for treatment by MMS at a specialized surgical institute.

CONCLUSION

Although DFSP is a rare disease with definitive treatment and good prognosis, its high rate of misdiagnosis and potential for delay in management can lead to poorer outcomes and more complex treatment. It is important for primary care physicians, dermatologists, surgeons, and pathologists to be clinically suspicious of this tumor and its variations in presentation in order to diagnosis and treat early. Post-operative management is critical, as these lesions have a high rate of recurrence. Thus, physicians should pay close attention to patient history of prior DFSP and changing appearance of scar tissue in their clinical evaluation.

Conflict of Interest Disclosures: None

Funding: None

Corresponding Author:
Daniel Fischer, DO, MA
St. John's Episcopal Hospital
Far Rockaway, NY
Email: drdeefish@gmail.com

References:

1. Wu S, Huang Y, Li Z, Wu H, Li H. Collagen Features of Dermatofibrosarcoma Protuberans Skin Base on Multiphoton Microscopy. *Technol Cancer Res Treat*. 2018;17:1533033818796775.
2. Angouridakis N, Kafas P, Jerjes W, et al. Dermatofibrosarcoma protuberans with fibrosarcomatous transformation of the head and neck. *Head Neck Oncol*. 2011;3:5.
3. Harati K, Lange K, Goertz O, et al. A single-institutional review of 68 patients with dermatofibrosarcoma protuberans: wide re-excision after inadequate previous surgery results in a high rate of local control. *World J Surg Oncol*. 2017;15(1):5.
4. Kreicher KL, Kurlander DE, Gittleman HR, Barnholtz-Sloan JS, Bordeaux JS. Incidence and Survival of Primary Dermatofibrosarcoma Protuberans in the United States. *Dermatol Surg*. 2016;42 Suppl 1:S24-S31.
5. Archontaki M, Korkolis DP, Arnogiannaki N, et al. Dermatofibrosarcoma protuberans: a case series of 16 patients treated in a single institution with literature review. *Anticancer Res*. 2010;30(9):3775-3779.
6. Lemm D, Mügge LO, Mentzel T, Höffken K. Current treatment options in dermatofibrosarcoma protuberans. *J Cancer Res Clin Oncol*. 2009;135(5):653-665.
7. de Araujo RN, Marcarini R, Ferreira BDFM, Obadia DL. Dermatofibrosarcoma protuberans rapid growth after incisional biopsy. *Medicina Cutánea Ibero-Latino-Americana*. 2017;45(1):41-44.
8. Tanaka A, Hatoko M, Tada H, Kuwahara M, Iioka H, Niitsuma K. Dermatofibrosarcoma protuberans arising from a burn scar of the axilla. *Ann Plast Surg*. 2004;52(4):423-425.

9. Saeki H, Kadono T, Le Pavoux A, Mori E, Tamaki K. Dermatofibrosarcoma protuberans with COL1A1-PDGFB fusion transcript arising on a scar due to a previous drainage tube insertion. *J Dermatol*. 2008;35(10):686-688.
10. Yang H, Yu L. Cutaneous and Superficial Soft Tissue CD34+ Spindle Cell Proliferation. *Arch Pathol Lab Med*. 2017;141(8):1092-1100.