

Dermatofibrosarcoma protuberans with fibrosarcomatous transformation: A tale of unbridled expansion



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INTRODUCTION

Dermatofibrosarcoma protuberans (DFSP) is a malignant fibroblastic tumor with a low metastatic potential.¹ These tumors most often form a nodule or ill-defined dermal plaque with an infiltrating honeycomb pattern.¹ Although most tumors remain confined to the dermis, longstanding tumors can invade fascia, muscle, periosteum, and bone.¹ They are most commonly located on the trunk, followed by proximal extremities, then the head/neck area.² In the United States, the incidence is 4.2 per million people per year.³ The National Comprehensive Cancer Center classifies fibrosarcomatous change as a high-risk feature that necessitates more intensive postoperative monitoring and treatment.⁴ Histologically, DFSP has an infiltrative pattern of bland spindle cells in storiform fascicles.¹ Greater than 90% of DFSPs have a translocation, t(17;22) (q22;q13), giving a fusion gene of collagen type 1 α 1 (COL1A1) and platelet-derived growth factor β (PDGF β).⁵ A strong, diffuse expression of CD34 supports the diagnosis of DFSP, whereas immunohistochemistry will be negative for S100, smooth muscle actin and cytokeratins.⁶

CASE REPORT

A 32-year-old African-American woman transferred care to our institution from an outside hospital (OSH) for management of her biopsy-proven DFSP on the forehead and scalp (Fig 1). The patient reported the previously quiescent, longstanding flat scalp lesion that grew rapidly, with erosion of the overlying skin, in July 2017. Because of lack of insurance coverage, a core needle biopsy

Abbreviations used:

CCPDMA:	complete circumferential, peripheral and deep tumor margin assessment
CT:	computed tomography
COL1A1:	collagen type 1 α 1
DFSP:	dermatofibrosarcoma protuberans
FS-DFSP:	fibrosarcomatous dermatofibrosarcoma protuberans
MMS:	Mohs micrographic surgery
MRI:	magnetic resonance imaging
OSH:	outside hospital
PDGFB:	platelet-derived growth factor β
WLE:	wide local excision

at the OSH was delayed until February 2018. The biopsy found a cellular proliferation of spindle cells with mild-to-moderate cytologic atypia arranged in a fascicular growth pattern consistent with classic DFSP. Fibrosarcomatous change was not seen. Immunohistochemistry found tumor cells positive for CD34 with no significant staining for desmin, S100, epithelial membrane antigen, or transducin-like enhancer of split 1. Fluorescence in situ hybridization was positive for the COL1A1 and PDGF β translocation. She had a brain computed tomography (CT) scan and magnetic resonance imaging (MRI) performed in early 2018 at the OSH (Fig 2), which found a right frontal soft tissue mass without bony involvement. She did not have any significant medical history and no family history of head and neck cancers. She subsequently presented to the otolaryngology department at our institution for surgical management. The dermatology department was consulted to perform Mohs micrographic surgery (MMS) because complete circumferential,

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Fig 1. Anterior frontal scalp with exophytic and friable tumor with eschar on distal aspect after infiltration with local anesthetic immediately prior to Mohs excision. Speckled hypopigmentation is noticeable around the mid-stalk of the tumor with edema surrounding the base.

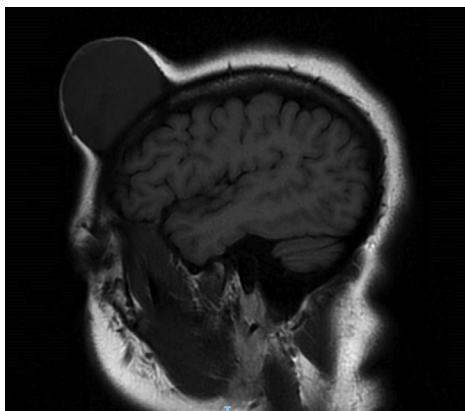


Fig 2. Brain MRI sagittal view (T1 flair propeller) shows a right frontal soft tissue mass measuring 4.7 cm × 4.6 cm × 5.9 cm without bony involvement and normal intracranial contents obtained on 02/06/2018 at OSH.

peripheral, and deep tumor margin assessment (CCPDMA) is associated with higher cure rates for DFSP compared with standard excision. Skepticism of Western medicine contributed to a delay in receiving definitive treatment until July 2018.

MMS was indicated given the subclinical infiltrative nature of DFSP and primary need for complete histologic margin evaluation. The preoperative tumor measured approximately 8 × 8 × 10 cm. MMS with a 2-cm margin found a tumor-free plane after 1 stage of excision. The final defect extending to the level of the bone measured approximately 10 × 10 cm. The otolaryngology department performed a parascapular fasciocutaneous free flap with microvascular anastomosis to repair the defect (Fig 3). Histopathology from the debulk specimen showed fibrosarcomatous DFSP (FS-DFSP; Fig 4), whereas the original core biopsy taken at the OSH did not.



Fig 3. Post-reconstruction photo of the defect shows a parascapular free flap with a wound drain in place.

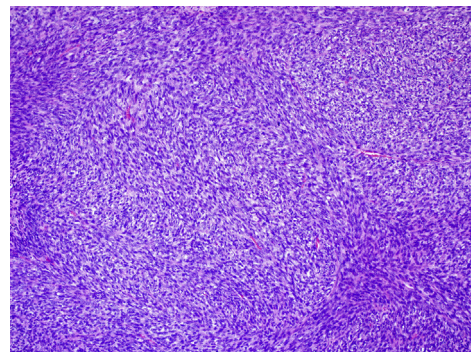


Fig 4. DFSP with fibrosarcomatous change shows long, sweeping fascicles of spindle cells intersecting at varied angles in a “herringbone” pattern. (Hematoxylin-eosin stain; original magnification, ×100.)

Multidisciplinary tumor board recommended imaging and adjuvant therapies. Baseline chest radiograph was normal. A staging CT (chest, abdomen, and pelvis) with contrast did not find evidence of metastatic disease. Imatinib mesylate, the tyrosine kinase inhibitor with activity against the PDGF receptor, would only be considered if the tumor were to recur or if she had metastatic disease. Given her younger age and high-risk features, radiation oncology recommended consideration of adjuvant radiation therapy, which the patient declined. In addition to routine postoperative surveillance via physical examination and chest radiography, quarterly CT imaging of the head was recommended to evaluate for local recurrence. Compared with MRI, contrast CT can better depict bony remodeling that could indicate recurrence when tumors approximate the cranium.

DISCUSSION

We report a case of a rapidly enlarging and morphologically impressive FS-DFSP on the scalp/forehead of a 32-year-old African-American woman.

Our case illustrates the potential for locally aggressive DFSP when untreated and highlights diagnostic pitfalls of small sample size for larger tumors.

A notable feature of this case was that the initial core needle biopsy obtained in February 2018 found classic DFSP. However, the giant tumor did not clinically correlate with the diagnosis of classic DFSP. History of rapid growth from a flat plaque to a 10-cm-tall tumor suggested fibrosarcomatous transformation had occurred. The entire giant tumor debulk removed at the time of Mohs excision was thus submitted for further histopathologic evaluation. Debulked DFSP specimens are suggested for histopathologic evaluation because of the variable accuracy of diagnostic tests for soft tissue sarcomas with regard to differentiating the various DFSP subtypes (core biopsy >incision biopsy >excision biopsy).⁷ Risk of local recurrence and metastasis is higher for FS-DFSP. Fibrosarcomatous DFSP features a 10% to 15% metastatic rate compared with 5% for a classic DFSP.¹ The lungs are a frequent site of metastasis, and surveillance by annual chest radiography is commonly recommended.¹

A key principle in the treatment of DFSP is obtaining clear surgical excision margins, as this reduces local recurrence and metastasis.⁸ Complete excision using CCPDMA methods such as the MMS technique yields the highest cure rates, especially for head and neck DFSP.⁴ A modified variant of the MMS technique using staged excisions over several days called *slow-MMS* represents another valid treatment option for DFSP.⁹ Local recurrence of DFSP is most common in the first 5 years after surgery.

Surgical management is first line for DFSP. The Mohs technique via CCPDMA improves clinical outcomes compared with standard wide local excision (WLE) by having better recurrence-free survival rates (1, 5, 10, and 15 years) and smaller postoperative defects.¹⁰ Margin recommendations for WLE have varied over time, making direct comparisons difficult for all data sets.¹⁰ Nonetheless, the preponderance of evidence favors MMS over WLE. Adjuvant radiation therapy may be indicated for margin-positive tumors that cannot be completely excised but is not typically recommended after negative surgical margins are achieved.¹ Imatinib mesylate is used for metastatic or inoperable disease. However, the 5-year progression-free survival rate of patients taking

imatinib mesylate to treat DFSP with fibrosarcomatous transformation is less than that of classic DFSP (33% vs 93%).¹¹ Once DFSP recurs, precise margin assessment is limited by scar tissue and disrupted noncontiguous growth. Surgery for primary DFSP offers the best chance for a long-term surgical cure.

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