

# Dermatofibrosarcoma Protuberans Arising at a Rho(D) Immune Globulin Injection Site

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*Dermatofibrosarcoma protuberans (DFSP) is a rare soft tissue tumor arising in the dermis. It is notorious for high rates of local recurrence despite its low metastatic potential. Although the etiology is unknown, DFSP often is considered to arise within scars and at sites of prior vaccination or trauma. Clinically, DFSP can be highly variable and mimic other soft tissue proliferations. We present a case of recurrent DFSP arising at the site of a Rho(D) immune globulin (RhIg) injection that was administered 7 years prior. We also discuss the diagnostic challenges of DFSP as well as the indolent and locally recurrent nature of the tumor. This case serves to remind dermatologists of the highly variable clinical appearance of DFSP as well as to warn against presumptive diagnoses of lesions that mimic keloids and hypertrophic scars.*

*Cutis.* 2012;90:233-234.

## Case Report

A 34-year-old woman presented with a small yellow growth on her right buttock that had slowly enlarged over the last 5 years. The lesion initially appeared as a mildly tender, 6- to 8-mm, erythematous nodule 2 years after the patient received a subcutaneous injection of Rho(D) immune globulin (RhIg) at the same site during pregnancy. The lesion was subsequently excised, and a pathology report indicated microscopic findings consistent with dermatofibroma. No immunohistochemistry was performed. The histology slides were not available for our review. Within 1 year, the nodule reappeared and continued to slowly enlarge. The patient did not seek additional medical care until the lesion became increasingly pruritic and

painful 4 years later. At that time, she presented to our clinic for treatment.

On physical examination, a firm, 2-cm, yellow plaque with a central scar was observed on the right upper lateral buttock (Figure 1). The location was consistent with the site of the RhIg injection administered 7 years prior. The patient had no lymphadenopathy. A punch biopsy of the plaque demonstrated a spindle cell proliferation throughout the entire dermis that extended into the subcutis. The cells were monomorphic and demonstrated minimal mitotic activity (Figure 2). The spindle cells stained positive with CD34 (Figure 3) and negative with factor XIIIa. The patient returned for staged excision with Mohs micrographic surgery, which resulted in complete margin control prior to closure. The patient is without complication or recurrence after 2 years.

## Comment

Our patient represents a previously misdiagnosed case of recurrent dermatofibrosarcoma protuberans (DFSP) arising 7 years after a RhIg injection. This case implicates RhIg injection as a previously undocumented etiology of DFSP and demonstrates the diagnostic and treatment challenges of this tumor.

Dermatofibrosarcoma protuberans is an uncommon mesenchymal sarcoma characterized by latency on



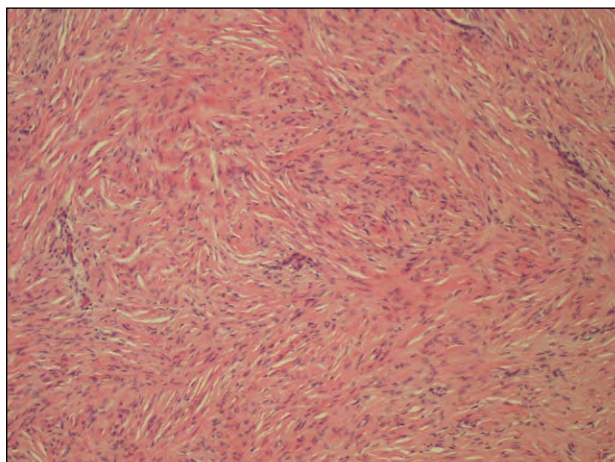
**Figure 1.** A recurrent yellow plaque with a central scar on the right upper lateral buttock prior to a second excision.

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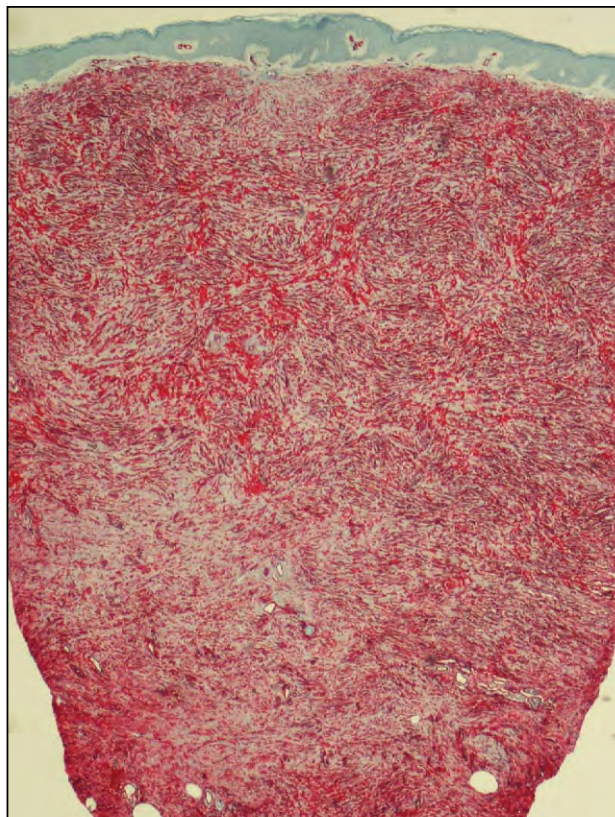
The authors report no conflict of interest.

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initial detection, slow infiltrative growth, low metastatic potential, and local recurrence if not adequately excised. The clinical appearance of DFSP is highly variable and often depends on the stage of the lesion. Usually, it is not until the late accelerated growth



**Figure 2.** Dermatofibrosarcoma protuberans characterized by spindle cell proliferation (fibroblasts) arranged in a storiform pattern. The cells were monomorphic, showed minimal mitoses, and extended throughout the entire dermis and into the subcutis (H&E, original magnification  $\times 100$ ).



**Figure 3.** Spindle cells characteristically stained positive with CD34 (original magnification  $\times 10$ ) and negative with factor XIIIa (not shown).

phase that the tumor ulcerates, bleeds, and/or becomes painful.<sup>1</sup> Definitive diagnosis must combine physical examination and histology. Often, DFSP may be misdiagnosed as a keloid, scar,<sup>2</sup> or dermatofibroma. Although keloids and scars can be differentiated with biopsy, the distinction between a dermatofibroma and a more benign-appearing DFSP may only be possible with immunohistochemistry, as in the case of our patient.

Microscopically, DFSP is characterized by a pattern of monomorphic, benign-appearing spindle cells arranged in a storiform architectural pattern.<sup>3</sup> The tumor border is difficult to distinguish because of irregular dendriticlike projections of neoplastic cells that infiltrate the surrounding tissue and can cause frequent reoccurrence after excision. With immunohistochemistry, DFSP generally stains positive with CD34 and vimentin and negative with factor XIIIa.<sup>3</sup>

The etiology of DFSP is unknown and often is considered to arise *de novo* in the dermis. Large case studies and reviews have found DFSP associated with a history of prior trauma from weapons and vaccination injections, repeated immunizations,<sup>4</sup> exposure to arsenic,<sup>5</sup> surgical scars, burns, acrodermatitis enteropathica, and intradermal nevi.<sup>6</sup>

### Conclusion

We report a rare case of DFSP following RhIg injection. This case reminds dermatologists of the highly variable clinical appearance of DFSP and warns against presumptive diagnoses of lesions that mimic keloids and hypertrophic scars.

*Acknowledgments*—We are indebted to Robert Pariser, MD, and Molly Smith, MD, both from Norfolk, Virginia, for their dermatopathologic analyses and expertise.

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