Case Report

Dermatofibrosarcoma Protuberans Masquerading as a Benign Scar on the Shoulder of a Caucasian Female

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Abstract

We report the case of a 62-year-old Caucasian female with an 11-year history of an asymptomatic and unchanging plaque on her left shoulder. Physical examination revealed a 2.5 cm round atrophic plaque with slight central nodularity and telangiectasias overlying a light-brown pigment network. The lesion was clinically most consistent with a scar from a prior surgical procedure. However, as there was no history of prior trauma at the site, a punch biopsy of the lesion was performed. Histopathological findings were suggestive of a neurofibroma, however, the possibility of a dermatofibrosarcoma protuberans was also considered. Immunohistochemical staining was performed which was negative for S100 and positive for CD34, consistent with DFSP. A larger repeat biopsy was then obtained, revealing findings consistent with dermatofibrosarcoma protuberans (DFSP). Fluorescence in situ hybridization (FISH) showed a PDGFB/COL1A1 fusion gene, confirming the diagnosis of DFSP. For patient tolerability, the sarcoma was completely excised via wide local excision under general anesthesia.

ABBREVIATIONS

DFSP: Dermatofibrosarcoma Protuberans; MMS: Mohs Micrographic Surgery; WLE: Wide Local Excision

INTRODUCTION

Dermatofibrosarcoma protuberans (DFSP) is a locally-aggressive soft tissue sarcoma that most commonly presents on the trunk of patients in the third to fourth decade of life. The sarcoma commonly presents as an asymptomatic, skin-colored to brown or pink indurated plaque however it may infrequently manifest as a scar-like or atrophic depression with telangiectasias. We herein report a case of a clinically nondescript scar-like plaque with initial sampling biopsy suggestive of a neurofibroma in which further immuno histochemical staining and deeper biopsy lead to the correct diagnosis of DFSP.

CASE PRESENTATION

A 62-year-old Caucasian woman with advanced dementia and mild mental retardation was accompanied to the dermatology clinic by her home health care provider for evaluation of a “mole” on her left forearm. While the forearm lesion was consistent with a seborrheic keratosis, upon routine full-body skin examination, a 2.5 cm round atrophic plaque with slight central nodularity and telangiectasias overlying a light-brown pigment network was noted on her left posterior shoulder (Figure 1). The shoulder lesion was clinically most consistent with a scar, such as that resulting from a prior shave biopsy or other minor surgical procedure. However, the patient’s home health provider was unaware as to whether a surgical procedure had ever been performed at the site and insisted that the lesion had been...
present and had not changed from the day she started providing care for the patient 11 years ago.

After obtaining informed consent from the patient’s state-appointed guardian, a 4mm sampling punch biopsy involving both lesional and adjacent normal skin was found to be histopathologically most consistent with a neurofibroma. However, with some suspicion of a possible dermatofibrosarcoma protuberans (DFSP), immunohistochemical staining was performed which was S100 negative and CD34 positive; with this staining pattern, DFSP could not be ruled out (Figures 2A and 2B). An excisional biopsy was subsequently performed and was interpreted by the same dermatopathologist to be DFSP; this diagnosis was supported by CD34 positive and factor XIII a negative immunohistochemical staining (Figures 3A and 3A). Fluorescence in situ hybridization (FISH) revealed PDGFB/COL1A1 rearrangement in 49% of the lesional cells (greater than 10% is considered a positive result), confirming the diagnosis.

**DISCUSSION**

DFSP is a locally-aggressive soft tissue sarcoma with a high propensity for local recurrence. It is a rare tumor with an incidence of 0.8 to 4.2 cases per million persons per year in the United States [1,2]. While a few cases of congenital and childhood-onset DFSP have been reported, it most commonly presents on the trunk (50-60%) and proximal extremities (20-30%) of patients in the third to fourth decade of life [1,3].

As was the case in our patient, DFSP in its early stages most commonly presents as an asymptomatic, skin-colored to brown or pink indurated plaque. Congenital and childhood-onset cases are particularly prone to manifesting as a scar-like or atrophic depression with telangiectasias [1,3]. These nonspecific lesions are frequently overlooked as scars and may go unrecognized until more distinctive features develop. DFSP lesions typically enlarge over months to years, evolving into firm, nodular, often violaceous fixed plaques, which may resemble keloid scars [3,4].

Histopathologically, DFSPs are composed of monomorphic dermal spindle cells in a “storiform” arrangement, which infiltrate the subcutaneous tissue in a “honeycomb” pattern. The nuclei are hyperchromatic, but tend to be monomorphic and the cells have a low mitotic activity. This is in contrast to a fibrosarcoma (DFSP-FS) which has increased nuclear pleomorphism compared with DFSP and a high mitotic rate. Thought to represent progression of DFSP, DFSP-FS typically has a poorer prognosis than DFSP.

The National Comprehensive Cancer Network (NCCN) recommends confirmatory immunostaining on all suspected DFSPs, allowing for differentiation from similar-appearing benign tumors and more importantly, from other malignant tumors such as fibrosarcoma, leiomyosarcoma, and undifferentiated/unclassified soft tissue sarcoma. The spindle cells of DFSP stain positively for CD34 and negatively for factor XIII, which results from the reciprocal chromosomal translocation t(17;22) and occurs in over 90% of DFSPs, providing further diagnostic guidance [4,5].

The plaque stage of DFSP may be misdiagnosed as a scar or a benign neoplasm, and thus the tumor may be either be overlooked or incompletely excised. As is illustrated in our case, if a lesion clinically resembles a scar but a history of trauma at the site cannot be confirmed, it is prudent to include DFSP in the differential diagnosis and perform a deep and wide biopsy for diagnostic confirmation. As in our case, a 4mm punch sampling biopsy was insufficient to confirm the diagnosis of DFSP. The importance of a large, deep biopsy is highlighted in this case, as insufficient sampling of a DFSP may resemble a benign spindle cell tumor such as a neurofibroma or a dermatofibroma. Our...
case also underscores the importance of immunohistochemical staining for CD34 and factor XIII a, as well as FISH studies to detect the PDGFB/COL1A1 fusion gene, if the diagnosis of DFSP is in question.

REFERENCES


