A Rare Case of Dermatofibrosarcoma Protuberans in the Lower Extremity

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Purpose

Dermatofibrosarcoma protuberans (DFSP) is a rare, locally invasive soft tissue sarcoma with high rates of recurrence and metastatic potential when not identified early and treated appropriately. There are few reports in the literature of this malignancy affecting the lower extremity. The present case study aims to highlight strategies for clinical identification and outline the treatment protocol to reduce complications following excision of this rare tumor.

Literature Review

DFSP occurs at a rate of 0.8-4.5 cases per million per year in the United States with most cases reported on the trunk followed by the upper extremities.[1-3] The peak age range reported for DFSP is 25-45 years old. A genetic component of this malignancy has been identified with over 90% of cases being associated with the characteristic translocation t(17;22) (q22;q13).[4]

DFSP presents early as an indurated nodular plaque with violet-red or blue margins, enlarging over months to years with asymptomatic progression to a raised firm nodular mass with surrounding telangiectasias.[5] If left untreated, the mass can become fixed or adhered to deeper surrounding structures and can progress to a large, protuberant mass.[6] Magnetic resonance imaging (MRI) often displays DFSP as a noncalcified, superficial, nodular mass arising from skin, with high T2 and low T1 signal intensities.[4] While MRI can be beneficial for diagnosis, biopsy and determination of soft tissue involvement is of urgent importance.

Despite the overall low potential for metastatic disease, high rates of local recurrence have been reported, likely due to the tumor's inherent microscopic projections. Residual tumor is present in 70% of surgical excisions when 1 cm margins are utilized, 20-40% with 2 cm margins, 9-15.5% with 3 cm margins, and 5% with 5 cm margins.[5] Surgical excision must take into account these microscopic projections and include wide negative margins to reduce risk of recurrence.[7]

Case Study

A 21-year-old otherwise healthy male presented with a several year history of a darkened, enlarging soft tissue mass on the medial ankle (Fig. 1). An MRI with and without contrast was obtained that demonstrated a 2.4 x 3.7 x 4.0 cm encapsulated superficial mass with a small region of capsule disruption (Fig. 2). The mass was hyperintense on T2 and STIR images and isointense to muscle on T1 images. These MRI features were thought to be most consistent with hemangioma by the interpreting radiologist. Based on continued enlargement and irritation in shoe gear, the patient elected to proceed with surgical excision. An initial excisional biopsy without orientation was sent for pathologic analysis which revealed DFSP with positive deep and peripheral margins (Fig. 5).

A wide re-excision was performed with 3 to 5 cm margins based on location to the Achilles tendon and medial malleolus (Fig. 3). A split-thickness skin graft was harvested from the patient's ipsilateral anterior thigh and was used to cover the deficit following the wide re-excision (Fig. 4). Following the re-excision, all margins were found to be clear of malignancy. Patient successfully healed the surgical site without wounding or infection. At final follow-up, 16 months post-excision, no evidence of local recurrence was noted.

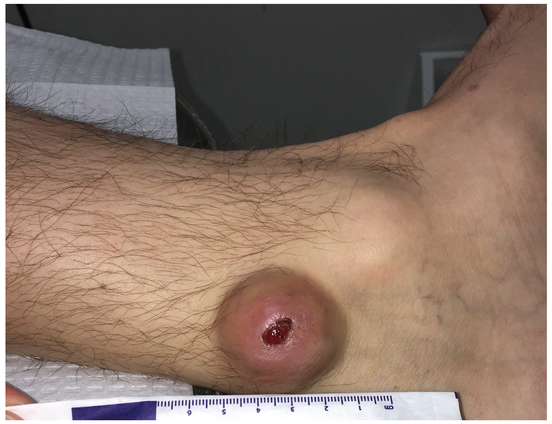

Figure 1: Clinical appearance of soft tissue mass at initial presentation.

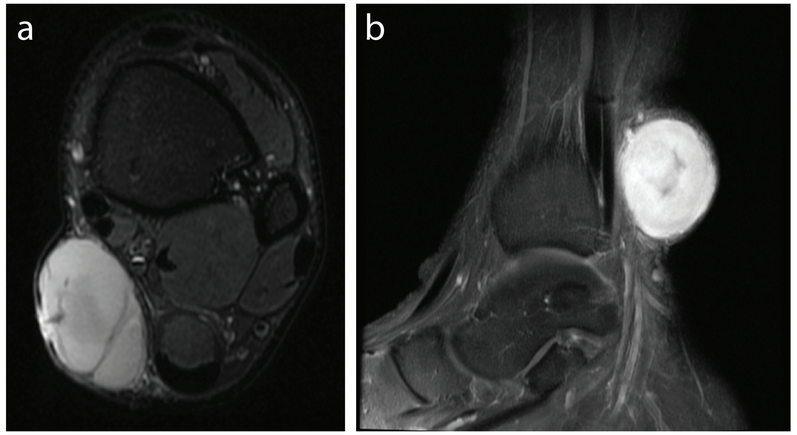

Figure 2: Solid encapsulated soft tissue mass demonstrated with a) axial T2 MRI and b) sagittal T1 MRI with contrast.

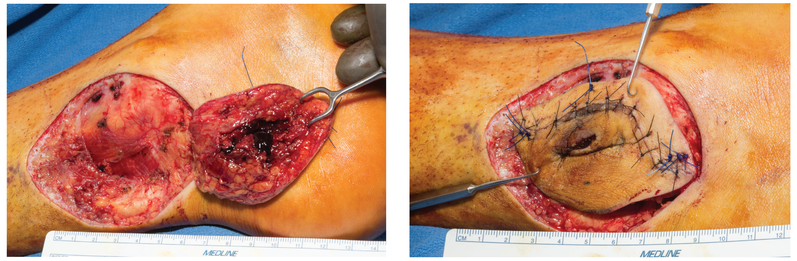

Figure 3: Intraoperative images of excision of lesion with wide margins.

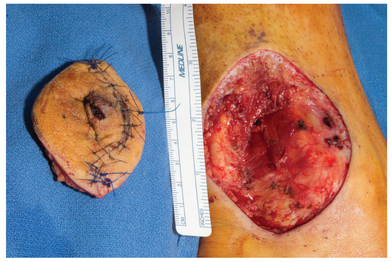

Figure 4: Intraoperative image demonstrating size of residual soft tissue deficit prior to skin graft placement.

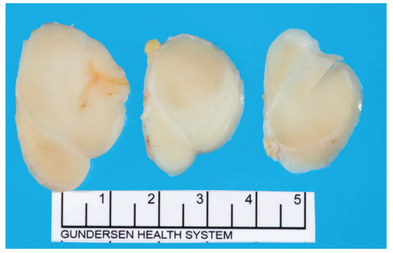

Figure 5: Cross sectional slices of soft tissue mass for pathologic analysis.

Analysis & Discussion

Since first being described by Taylor in 1890, DFSP has remained a rare malignancy accounting for less than 0.1% of all malignancies and 1.8% of all soft tissue sarcomas.[8, 9]

The patient presented here was 21 years old at the time of diagnosis and several years younger when the soft tissue mass initially appeared, making him younger than the peak age range of 25-45 years old.[2] The clinical course in this case study follows the typical pattern of a slow-growing mass of darkened discoloration that is nodular but non-painful. An MRI was ordered, but was non-specific, thus the gold standard of excisional biopsy was pursued for definitive diagnosis. The preferred treatment is excision of the lesion with wide margins, though Mohs surgery is becoming more popular with promising results.[2] In the present case, wide margins of 3 to 5 cm were obtained and required split thickness skin grafting to account for the remaining soft tissue deficit. Although rare, metastatic disease in DFSP is possible with the most common site being the lungs.[2] A formal consultation with Oncology is therefore recommended.

In conclusion, we present a rare case of DFSP of the lower extremity that was successfully identified, diagnosed and treated with the proper evidence-based treatment algorithm without evidence of local recurrence to date. We believe it is important to increase awareness of this locally aggressive soft tissue sarcoma which can mimic common soft tissue lesions and to highlight the proper treatment protocol to successfully prevent recurrence and minimize potential for metastatic disease.

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