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A RARE CASE OF DERMATOFIBROSARCOMA PROTUBERANS – CLINICAL INSIGHTS AND MANAGEMENT

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Abstract

Dermatofibrosarcoma protuberans (DFSP) is an uncommon, slow-growing, locally aggressive cutaneous soft tissue sarcoma of fibrohistiocytic origin. Despite its low metastatic potential, DFSP exhibits a high propensity for local recurrence, necessitating early and precise intervention. This report delineates a rare case of DFSP in a 35-year-old male presenting as an asymptomatic, progressive swelling on the left thigh, with a history of antecedent trauma. Diagnostic evaluation, including trucut biopsy and histopathological examination (HPE), confirmed the presence of spindle cells arranged in a storiform pattern, characteristic of DFSP. The patient underwent wide local excision with tumor-free margins, emphasizing the critical role of surgical precision in management. This case underscores the diagnostic challenges, histopathological nuances, and therapeutic imperatives associated with DFSP, while also reviewing contemporary literature to enhance clinical understanding.

INTRODUCTION

Dermatofibrosarcoma protuberans (DFSP) is a rare dermal mesenchymal malignancy, accounting for approximately 1% of all soft tissue sarcomas and less than 0.1% of all malignancies [1]. First described by Darier and Ferrand in 1924, DFSP typically manifests in young to middle-aged adults, with a slight predilection for males [2]. The tumor predominantly arises on the trunk (42-72%), followed by the proximal extremities (20-30%) and head-neck region (10-16%) [3].

DFSP is characterized by a recurrent chromosomal translocation t(17;22)(q22;q13), resulting in the fusion of the collagen type 1 alpha 1 (COL1A1) gene with the platelet-derived growth factor beta (PDGFB) gene [4]. This fusion oncogene drives tumorigenesis via constitutive activation of the PDGFB signaling pathway, promoting fibroblast proliferation [5]. Clinically, DFSP presents as an indolent, skin-colored to erythematous plaque that gradually evolves into nodular protuberances, often misdiagnosed as benign lesions initially [6].

Although metastasis is rare (<5%), local recurrence rates range from 10-50% following inadequate excision, underscoring the need for meticulous surgical planning [7]. This report presents a classic case of DFSP, highlighting its clinical, histopathological, and therapeutic dimensions, while integrating evidence-based insights from contemporary literature.

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Case Report

Clinical Presentation
A 35-year-old male with no significant comorbidities presented to our outpatient department with an 8-month history of a slowly enlarging, painless swelling on the anterolateral aspect of the left thigh. The patient recalled sustaining blunt trauma to the same site four years prior but reported no other contributory history, such as fever, weight loss, or constitutional symptoms.

Physical Examination

- **Inspection:** A solitary, 8x7 cm ovoid swelling with irregular surface contours and a pinkish-purple hue was noted (Figure 1).
- **Palpation:** The lesion was firm, non-tender, and mobile, with no associated warmth or fluctuance. Regional lymph nodes were non-palpable.

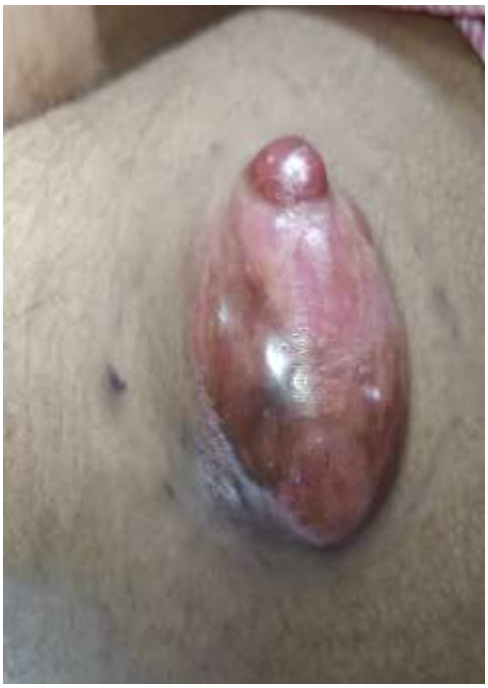

Fig 1- External appearance of DFSP

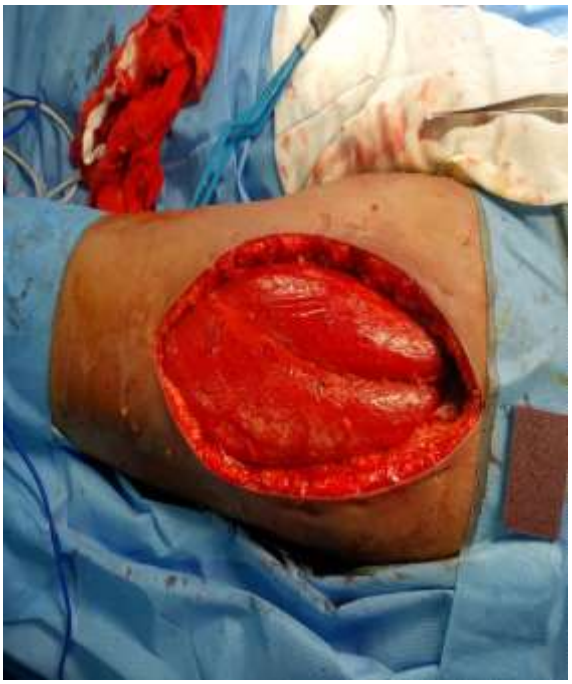

Fig 2- Wide Local excision

Diagnostic Workup

1. **Trucut Biopsy:** Preliminary findings suggested a spindle cell neoplasm, necessitating further histopathological evaluation.
2. **Histopathological Examination (HPE):** Microscopic analysis revealed:
    1. Stratified squamous epithelium with focal ulceration.
    2. A tumor composed of monotonous spindle cells arranged in a storiform (cartwheel) pattern.
    3. Cells exhibited ovoid to elongated nuclei, moderate eosinophilic cytoplasm, and increased mitotic activity (5-10 mitoses/10 HPF).
    4. Tumor infiltration into subcutaneous adipose tissue was evident, with no necrosis.
    5. Surgical margins were free of tumor involvement.
    6. Immunohistochemistry (IHC) showed diffuse CD34 positivity, corroborating the diagnosis of DFSP (Figure 2).

Differential Diagnosis
The clinical and histologic mimics included:

1. Dermatofibroma (benign fibrous histiocytoma)
2. Schwannoma
3. Cutaneous neurofibroma
4. Solitary fibrous tumor

Management
The patient underwent **wide local excision** with 3 cm lateral margins and deep fascial resection, followed by primary closure. Postoperative recovery was uneventful.

Follow-Up
The patient was advised regular clinical examinations every 6 months for 5 years to monitor for recurrence.

DISCUSSION

Epidemiology and Pathogenesis
DFSP has an estimated annual incidence of 0.8–4.5 cases per million, with a peak occurrence in the third to fifth decades of life [8]. Over 90% of DFSPs harbor the *COL1A1-PDGFB* fusion, which upregulates PDGFB-dependent signaling, fostering tumor proliferation [9].

Clinical and Histopathological Features
DFSP typically begins as an asymptomatic plaque, progressing to multinodular lesions with a "protuberant" morphology [10]. Histologically, the storiform pattern of spindle cells infiltrating subcutaneous fat is pathognomonic. CD34 immunopositivity aids in distinguishing DFSP from dermatofibroma (typically CD34-negative) [11].

Therapeutic Strategies

1. **Surgery:** Wide local excision (2-4 cm margins) remains the gold standard. Mohs micrographic surgery is an alternative for cosmetically sensitive areas [12].
2. **Medical Therapy:** Imatinib, a tyrosine kinase inhibitor, is FDA-approved for unresectable, recurrent, or metastatic DFSP, targeting the PDGFB pathway [13].

Prognosis and Recurrence
The 5-year recurrence rate is <5% with adequate excision but escalates to 20-50% with positive margins [14]. Long-term surveillance is imperative, as recurrences may manifest decades post-treatment [15].

CONCLUSION

This case exemplifies the indolent yet locally aggressive nature of DFSP, emphasizing the importance of early diagnosis, histopathological corroboration, and radical excision to mitigate recurrence. Molecular insights into the *COL1A1-PDGFB* fusion have revolutionized targeted therapy, offering hope for advanced cases. Continued research into novel biomarkers and adjuvant therapies is warranted to optimize outcomes.

REFERENCES

1. Paramythiotis D, Bangeas P, Kofina K, Karakatsanis A, Michalopoulos A. Aggressive angiomyxoma of the perineum: a case report and review of the literature. Hippokratia. 2016;20(1):80-3.
2. Llombart B, Serra-Guillén C, Monteagudo C, López-Guerrero JA, Sanmartín O. Dermatofibrosarcoma protuberans: a comprehensive review and update on diagnosis and management. Semin Diagn Pathol. 2013;30(1):13-28.
3. Rutkowski P, Van Glabbeke M, Rankin CJ, Ruka W, Rubin BP, Debiec-Rychter M, et al. Treatment of advanced dermatofibrosarcoma protuberans with imatinib mesylate: results of a multicenter phase II study. J Clin Oncol. 2010;28(10):1772-9.
4. Gloster HM Jr. Dermatofibrosarcoma protuberans: a review of 163 cases. Dermatol Surg. 1996;22(3):263-7.
5. **Paradisi A, Abeni D, Rusciani A, et al.** *Dermatofibrosarcoma protuberans: wide local excision vs. Mohs micrographic surgery.* Cancer Treatment Reviews. 2008;34(8):728–736. A highly cited comparative analysis of surgical approaches.