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CASE REPORT

A clinical case report on dermatofibrosarcoma protuberans

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Abstract

This case report highlights the diagnostic challenges and therapeutic considerations when dermatofibrosarcoma protuberans (DFSP) present in atypical locations, such as the head and neck. A 46-year-old Hispanic male presented with a large, rapidly growing, protruding mass on the left medial forehead, initially diagnosed through ultrasound as a benign cyst. The lesion exhibited a significant increase in size over 5 months. Physical examination revealed a firm nodular mass without signs of inflammation or lymphadenopathy. The lesion was excised for biopsy, and histopathological analysis confirmed a diagnosis of DFSP, characterized by spindle cells arranged in a storiform pattern. Immunohistochemical staining revealed strong CD34 positivity, with negative markers for desmin, factor XIIIa, and SOX10. The tumor exhibited an infiltrative pattern, extending into the skeletal muscle. The patient was referred for wide local excision due to the complexity of achieving clear margins in a cosmetically sensitive area such as the forehead. DFSP most commonly occurs on the trunk and proximal extremities, with less frequent involvement of the head and neck. This case is notable for its atypical location on the forehead, making surgical excision particularly challenging due to the need for margin control while preserving facial esthetics. Initial misdiagnosis as a benign cyst highlights the diagnostic difficulty of DFSP in uncommon locations. The tumor's rapid growth and deep infiltration into skeletal muscle are further complicated management, requiring careful surgical planning to prevent a recurrence. Long-term follow-up is essential due to the high recurrence rate of DFSP when margins are inadequate.

Keywords: Dermatofibrosarcoma protuberans; CD34; Spindle cell; Cutaneous sarcoma; Local recurrence; Head and neck tumor; Wide local excision; Skeletal muscle invasion

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1. Introduction

Dermatofibrosarcoma protuberans (DFSP) is a rare and enigmatic soft-tissue tumor characterized by its insidious growth and remarkable local invasiveness. The tumor

typically invades the dermis, subcutaneous fat, and, in rare cases, deep layers such as the muscle and fascia.[1] Accounting for < 1% of all soft-tissue sarcomas, DFSP poses diagnostic challenges owing to its clinical subtleties and histological mimicry with other dermatopathological entities. The exact cause of DFSP is ambiguous; however, studies have suggested the chromosomal translocation t(17;22)(q22;q13), which results in the fusion of *COL1A1* and *PDGFB* genes, ultimately promotes tumor growth by overproducing platelet-derived growth factor (PDGF).[1,2] DFSP clinically manifests as a firm, indurated plaque or nodule, most commonly presenting on the trunk or proximal extremities.[3] The annual incidence rate of DFSP is between 0.8 and 4.5/million persons. The typical age at diagnosis is between 20 and 50 years, but DSFP may present at any age, especially if congenital.[4] Studies have also suggested that DFSP most commonly presents in males than females (57% vs. 43%) and occurs most frequently in black patients.[4,5] DFSP has an overall excellent prognosis when effectively treated, with a 10-year survival rate of 99%.[1] Those with lesions located on the head or neck, which accounts for roughly 10 – 15% of cases, tend to have a worse prognosis due to unique surgical challenges or inadequate surgical margins.[6]

DFSP is diagnosed through skin biopsy.[6] Histologically, the tumor is composed of spindle-shaped cells arranged in a storiform pattern, often infiltrating surrounding tissues.[7] Upon immunohistological staining, one of the most notable markers for DFSP is the strong and diffuse positivity for CD34.[7,8] DFSP also stains strongly for vimentin, consistent with its mesenchymal origin; however, vimentin is not a specific marker, as it is expressed in nearly all sarcomas and other spindle cell neoplasms.[8] Additional characteristic immunohistological features include negativity for desmin, factor XIIIa, CD163, and D2-40, which are markers more commonly seen in dermatofibroma.[8] Although considered an intermediate-grade malignancy with a low probability of metastasis, DFSP is characterized by its propensity for local recurrence.[1] The optimal treatment for DFSP is Mohs micrographic surgery or, alternatively, wide local excision (WLE) of the tumor. Radiation treatment has also proven to reduce the risk of recurrence and may be considered after surgery.[9] In circumstances of unresectable, recurrent, or metastatic DFSP, the Food and Drug Administration-approved chemotherapeutic drug imatinib mesylate can serve as a possible treatment option.[1,10] Imatinib is a tyrosine kinase inhibitor that specifically targets and blocks PDGF receptors, which are overexpressed in DFSP due to the *COL1A1-PDGFB* fusion gene. By inhibiting PDGF signaling, imatinib can lead to significant tumor shrinkage, offering a promising therapeutic approach for patients with advanced or metastatic DFSP who may not be candidates for surgery.[10] Here, we present a case of a 46-year-old Hispanic male affected by DFSP on the left forehead with infiltration extending to the skeletal muscle. According to our current knowledge of DFSP, this is a rare occurrence, as described in scientific literature.

2. Case presentation

A 46-year-old Hispanic male presented to our clinic with a large, protruding mass located on the left medial forehead. The patient reported he first noticed the lesion 5 months before his visit and noted a significant increase in size during the preceding months. The patient denied any recent fever, weight loss, night sweats, chills, and any pain or discomfort at the site. Upon physical examination, a firm nodular mass was noted with no signs of localized redness or heat. In addition, there was no evidence of cervical or axillary lymphadenopathy. The patient reported no significant personal medical history, and pertinent family medical history was non-contributory.

A soft-tissue ultrasound of the left forehead was performed and revealed an egg-shaped cyst of 1.5 × 0.6 mm in size (Figure 1A), showing increased transmission through the posterior wall. The lesion was excised and sent for pathologic analyses (Figure 1B). Upon histological examination, cellular spindle cell proliferation was described. Hematoxylin-eosin (H&E)-stained sections of the spindle cells showed fusiform nuclei with tapered ends, fine chromatin, and eosinophilic cytoplasm. In addition, the neoplastic cells were arranged in fascicles oriented in a uniform storiform pattern. Sections showed scattered mitoses of normal configuration and no histological evidence of biological transformation. On immunohistological stains, the tumor cells showed positivity for CD34 antigen and were negative for desmin, factor XIIIa, and SOX10 (Figure 2A and B). Vimentin staining was not performed. Per pathology, the soft-tissue excised represents DFSP, grade 1/3. The tumor is centered in the dermis and subcutis, has an infiltrative pattern, and extends into the skeletal muscle. The patient was referred to a surgical oncologist for appropriate imaging and WLE of the lesion with negative margins.

3. Discussion

DFSP is a rare, slow-growing cutaneous sarcoma that arises from the dermis and subcutaneous tissue, accounting for <0.1% of all malignancies.[11] While DFSP has a low metastatic potential, its locally invasive nature poses significant clinical challenges. The tumor is notorious for high recurrence rates, particularly following incomplete excision.[12] Therefore, early diagnosis and appropriate treatment are critical to prevent local invasion

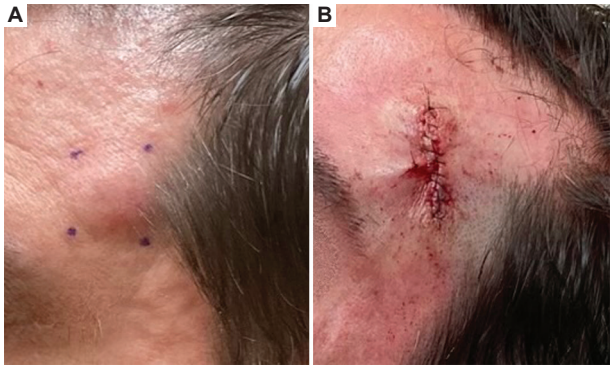

Figure 1. Macroscopic appearance of the lesion. (A) A dome-shaped, protuberant mass located on the left medial forehead is consistent with dermatofibrosarcoma protuberans and is marked for excisional biopsy. (B) Post-excisional biopsy site of the previously identified facial mass, pending histopathological identification.

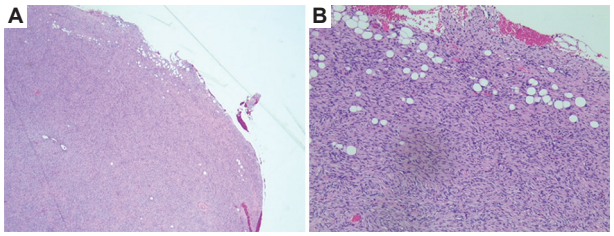

Figure 2. Immunohistochemical staining of a biopsied skin specimen reveals cellular spindle cell proliferation centered in the dermis and subcutis, with an infiltrative pattern extending into the skeletal muscle. H&E staining shows that the specimen are positive for CD34 antigen and negative for desmin, factor XIIIa, CD163, and D2-40, as observed under magnifications of ×40 (A) and ×100 (B).
Abbreviation: H&E: Hematoxylin and eosin.

and recurrence. This case provides a unique clinical presentation of DFSP for several reasons.

One notable aspect is the tumor's location on the left forehead, which is relatively uncommon, as DFSP most often appears on the trunk and proximal extremities. Lesions on the head and neck account for only 10 – 15% of cases.[3,13] Tumors in this region present unique challenges both in diagnosis and surgical management due to the proximity to critical facial structures. Achieving adequate surgical margins is difficult, as it necessitates balancing oncologic safety with preserving both cosmetic and functional outcomes.

Another significant point in this case is the initial misdiagnosis. The lesion was originally identified as a benign cyst, which highlights the difficulty in diagnosing DFSP, especially in atypical locations. This emphasizes the importance of considering DFSP in the differential diagnosis of any firm, slow-growing, cutaneous mass. In addition, the lesion's rapid growth within 5 months – while DFSP is typically slow-growing – illustrates that in some cases, DFSP can exhibit aggressive progression, especially in cosmetically sensitive areas such as the face.[1,11]

Upon H&E staining, spindle cells showed fusiform nuclei with tapered ends, fine chromatin, and eosinophilic cytoplasm. The neoplastic cells were arranged in fascicles in a storiform pattern, characteristic of DFSP.[11] Sections showed scattered mitoses of normal configuration with no evidence of biological transformation. Immunohistochemically, the tumor cells were positive for CD34, a hallmark of DFSP,[11,15] whereas they were negative for desmin, factor XIIIa, and SOX10.

The absence of desmin helps to rule out muscle-derived tumors such as leiomyosarcoma,[14] whereas the lack of factor XIIIa staining aids in distinguishing DFSP from dermatofibroma, which is typically factor XIIIa-positive.[15] Similarly, the negative SOX10 stain excludes tumors of neural crest origin, such as neurofibroma.[16] Thus, the immunohistochemical profile of CD34 positivity, combined with the absence of these markers, supports the diagnosis of DFSP. However, what makes this case particularly challenging is the extent of the tumor's infiltration into the underlying skeletal muscle. While DFSP is generally locally invasive, deeper invasion into the muscle is relatively rare and adds complexity to the surgical approach.[2,17] This deep tissue involvement necessitates a more extensive surgical excision, as incomplete removal significantly increases the risk of recurrence.[12] Ensuring clear margins becomes especially critical in cases like this, where the tumor has extended beyond its typical boundaries into deeper structures.

Mohs micrographic surgery is typically preferred for DFSP in cosmetically sensitive areas due to its high margin accuracy and tissue-sparing properties.[12] However, due to the tumor's skeletal muscle involvement, WLE was considered necessary in this case to ensure complete removal. After the excisional biopsy performed at our center showed extending margins on pathology, the patient was referred to a surgical oncologist for definitive excision and to manage the deeper tissue involvement.

DFSP is notorious for local recurrence, particularly when margins are inadequate. Recurrence rates can be as high as 50% in cases with compromised margins. Even with clear margins, the tumor's ability to infiltrate surrounding tissues, such as muscle, increases the risk of recurrence.[12] Long-term follow-up is critical, especially during the first 3 years post-surgery when the risk of recurrence is highest. Imaging techniques, such as magnetic resonance imaging or ultrasound, may be used to assess deep tissue involvement, which may not be easily detected through physical examination alone. After 3 years, annual follow-up is recommended, though continued vigilance

remains necessary, particularly in patients with deeper tissue involvement or those requiring extensive excisions. Patients should be educated on self-monitoring for any new or recurrent growths at the surgical site.[18-20]

For our patient, long-term surveillance will be scheduled every 6 months postoperatively for the first 3 years, followed by annual check-ups. Imaging will be performed as necessary to monitor for recurrence, and the patient will be educated on the importance of self-examination.

4. Conclusion

The current case illustrates the importance of considering DFSP in the differential diagnosis for rapidly growing, asymptomatic cutaneous masses, even when initial imaging suggests benign pathology. Early diagnosis, accurate histopathological assessment, and appropriate surgical management are critical to prevent recurrence and minimize morbidity. In cosmetically sensitive areas such as the face, striking a balance between adequate margin control and preservation of function and symmetry remains a key challenge. Surgical excision in these regions must account for potential impacts on facial expression, eyelid closure, speech, and oral competence. As demonstrated in this case, WLE with negative margins is essential, and ongoing advances in targeted therapies may further improve outcomes for patients with more advanced or recurrent diseases.

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Conflict of interest

The authors declare they have no competing interests.

Author contributions

Conceptualization: Isabella Gianina Raffa, Christoper Mendez, Jose E. Mendez
Formal analysis: All authors
Investigation: Isabella Gianina Raffa, Christoper Mendez, Jose E. Mendez
Methodology: All authors
Writing – original draft: All authors
Writing – review & editing: All authors

Ethics approval and consent to participate

Verbal informed consent was obtained from the patient prior to his participation in this study.

Consent for publication

Verbal consent has been obtained from the patient to publish his data.

Availability of data

Not applicable.

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