Section: General Surgery

Case Report

DERMATOFIBROSARCOMA PROTUBERANS OF THE ANTERIOR CHEST WALL- A CASE REPORT

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ABSTRACT

Dermatofibrosarcoma protuberans (DFSP) is a rare, low-to-intermediate grade fibroblastic neoplasm of dermal origin characterized by locally aggressive behavior and a high propensity for recurrence if inadequately excised. It accounts for less than 0.1% of all malignancies and approximately 1% of soft tissue sarcomas. Although metastasis is rare (<5%), the infiltrative growth pattern with tentacle-like projections into surrounding tissue necessitates meticulous surgical management.

We report the case of a 36-year-old male who presented with a slowly progressive plaque over the left anterior chest wall for five years. The lesion was initially misdiagnosed as tinea incognito. Clinical examination revealed a well-defined infiltrative plaque measuring 10 × 8 cm with nodular components, confined to the dermis and subcutaneous tissue. Punch biopsy demonstrated spindle cell proliferation arranged in a storiform pattern. Immunohistochemistry showed strong CD34 positivity and S-100 negativity, confirming DFSP. Contrast-enhanced CT scan revealed tumor confinement to superficial planes without deep muscle or bony invasion.

The patient underwent wide local excision (WLE) with adequate margins followed by split skin graft reconstruction. Histopathology confirmed tumor-free margins with a low Ki-67 index (6%). Postoperative recovery was uneventful, with excellent graft uptake and no recurrence to date.

This case emphasizes the importance of early recognition, histopathological confirmation, immunohistochemistry, and adequate surgical margins in achieving optimal outcomes. Long-term surveillance remains essential due to the risk of late recurrence.

INTRODUCTION

Dermatofibrosarcoma protuberans (DFSP) is a rare cutaneous soft tissue sarcoma first described by Darier and Ferrand in 1924. It is classified as an intermediate-grade fibroblastic tumor with low metastatic potential but marked local aggressiveness. The incidence is approximately 4–5 cases per million person-years, making it an uncommon entity in surgical practice.[1]

DFSP most commonly arises on the trunk (approximately 50% of cases), followed by proximal extremities and the head and neck region.[2] It typically presents in early to middle adulthood, with a slight female predominance. Epidemiological data suggest a higher incidence among Black patients compared to White populations.[3]

Pathogenetically, over 90% of DFSP cases harbor a characteristic chromosomal translocation t(17;22)(q22;q13), resulting in fusion of the COL1A1 and PDGFB genes.[4] This fusion gene leads to constitutive overexpression of platelet-derived growth factor beta (PDGF-β), promoting autocrine tumor growth. This molecular hallmark has therapeutic implications, particularly in unresectable or metastatic cases where targeted

therapy with imatinib has demonstrated significant response rates.[5]

Clinically, DFSP begins as a slow-growing, indurated plaque that may remain stable for years before developing nodular or protuberant components. The lesion is often misdiagnosed as benign conditions such as dermatofibroma, keloid, morphea, or fungal infection, resulting in delayed diagnosis.[6]

Histopathologically, DFSP is characterized by monomorphic spindle cells arranged in a storiform (cartwheel) pattern infiltrating the dermis and subcutis. A classic "honeycomb" infiltration into subcutaneous fat is frequently observed.[7] Immunohistochemically, DFSP shows strong and diffuse CD34 positivity, which serves as a diagnostic hallmark, while S-100 negativity helps exclude neural or melanocytic lesions.[8]

The cornerstone of management is complete surgical excision with negative margins. Historically, wide local excision (WLE) with 3–5 cm margins has been recommended. However, recurrence rates following conventional excision have ranged from 20% to 50% when margins are inadequate.[9] Mohs micrographic surgery (MMS) offers superior margin control with recurrence rates below 3%.[10]

Although metastasis is rare, fibrosarcomatous transformation (FS-DFSP) represents a high-risk variant associated with increased metastatic potential and poorer prognosis.[11] Therefore, accurate histopathological classification is critical.

We present a case of DFSP of the anterior chest wall that was initially misdiagnosed and subsequently managed successfully with wide local excision and split skin grafting.

MATERIALS AND METHODS

Study Design: Single case report.

Patient Information

A 36-year-old male mechanic came to the OPD with chief complaints of progressive skin lesion on the left anterior chest wall. Initially a small papule which was gradually enlarged to current size (10 × 8 cm) not associated with pain or discharge.

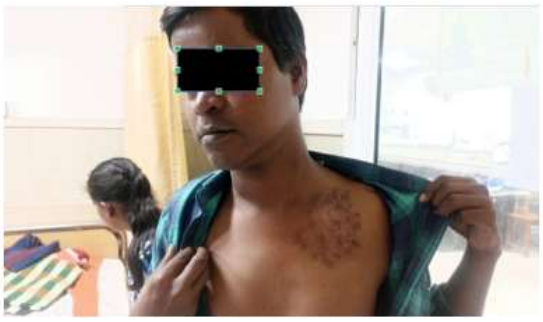

Figure 1: Preoperative clinical photograph showing a hyperpigmented nodular plaque over the left anterior chest wall

Clinical Evaluation

A single, well-defined plaque measuring 10 × 8 cm is present on the left anterior chest wall. The lesion shows both hyperpigmented and hypopigmented areas with a few nodular components. The surface is smooth and non-ulcerated, with no tenderness, discharge, or local warmth. No axillary lymphadenopathy is noted. The lesion appears confined to the skin, dermis, and subcutaneous tissue, and is not fixed to the underlying muscle or bone.

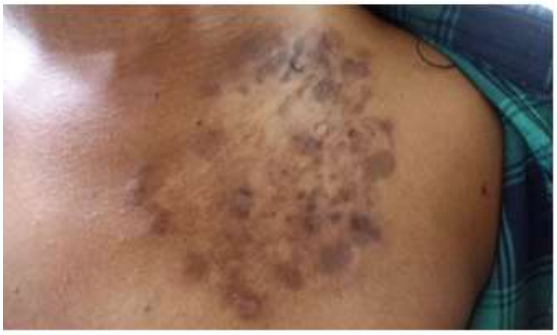

Figure 2: Clinical presentation of dermatofibrosarcoma protuberans.

Diagnostic Workup

1. Histopathology

Punch biopsy was performed under local anesthesia. Histopathological examination shows spindle cell proliferation infiltrating the dermis and subcutaneous tissue, arranged in a storiform pattern, with mild cellular atypia.

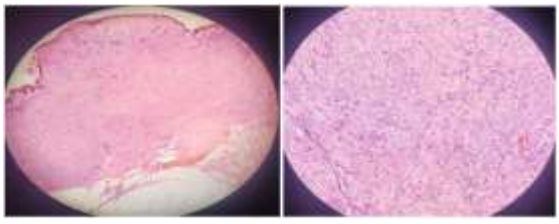

Figure 3: Histopathological features of dermatofibrosarcoma protuberans.

The following markers were assessed: **CD34, S-100, Ki-67, and β-catenin**.

Table 1: Immunohistochemistry (IHC) Results

Marker	Result	Interpretation
CD34	Positive	Diagnostic for DFSP
S-100	Negative	Excludes neural origin
Ki-67	5%	Low proliferation

2. Radiological Evaluation

Contrast-enhanced CT (CECT) of neck and chest to assess:

CECT of the Neck and Chest shows focal thickening in the left infraclavicular region measuring approximately 1.2 cm. The tumor is confined to the superficial planes, involving the dermis and subcutaneous tissue. No enhancing nodular component is identified. There is no evidence of invasion into the underlying fat, pectoralis muscle, or the bony thoracic cage.

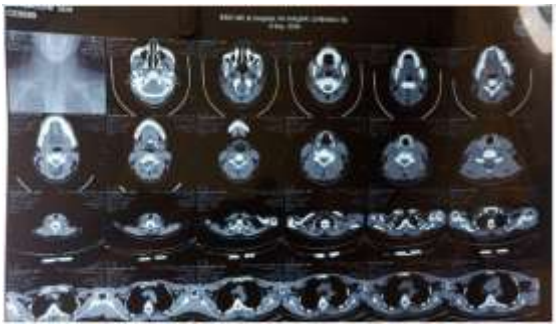

Figure 4: Radiological evaluation of tumor extent.

RESULTS

Surgical Procedure

The tumor is confined to the dermis and subcutaneous tissue, with all surgical margins negative for tumor involvement. The achieved margins are as follows: superior margin – 3.5 cm, lateral margin – 3.4 cm, inferior margin – 2.5 cm, and deep margin – 1.9 cm. The revised medial margin is also tumor-free.

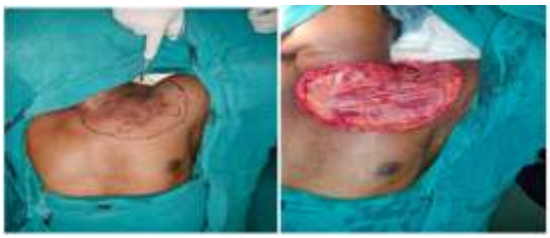

Figure 5: Intraoperative images of surgical management.

Histopathology

Postoperative Follow-Up

The patient had an uneventful postoperative recovery. There was greater than 90% graft take, with no evidence of wound infection or necrosis. Surgical margins were clear, and no recurrence has been observed to date. The patient remains under periodic follow-up.

Figure 6: Postoperative outcome following surgical excision and grafting.

DISCUSSION

DFSP is a unique soft tissue sarcoma distinguished by high local recurrence and low metastatic potential. The presented case highlights several classical features: slow progression, superficial confinement, and CD34 positivity.

The average delay in diagnosis ranges from 3–6 years due to its benign appearance.[6] Our patient similarly experienced a five-year delay secondary to misdiagnosis as fungal infection.

The infiltrative growth pattern with microscopic extensions explains the historically high recurrence rates after narrow excision.[9] Bowne *et al.* demonstrated that recurrence rates drop to below 10% when margins exceed 3 cm.[9] Mohs micrographic surgery provides optimal margin control, with recurrence rates as low as 1–3% [10].

The fibrosarcomatous variant is associated with increased mitotic activity and metastatic potential.[11] Fortunately, our case showed a low Ki-67 index and absence of fibrosarcomatous transformation, indicating favorable prognosis.

Imatinib, targeting PDGFB receptor activation, has shown a 60% response rate in advanced disease.[5] However, surgery remains the mainstay of treatment in localized tumors.

Long-term follow-up is essential, as recurrence has been reported up to 20 years post-excision.[12] Regular clinical surveillance is recommended.

Overall survival remains excellent, with 10-year survival rates exceeding 99% in classic DFSP.[13]

CONCLUSION

Dermatofibrosarcoma protuberans is a rare but locally aggressive cutaneous sarcoma that requires early recognition and appropriate surgical management. Although metastasis is uncommon, inadequate excision leads to significant recurrence

risk. This case underscores the importance of histopathology and immunohistochemistry in establishing diagnosis, particularly CD34 positivity and S-100 negativity.

Wide local excision with adequate margins remains the cornerstone of treatment. In our patient, margin clearance exceeding 2 cm resulted in successful disease control with no recurrence to date. Reconstruction with split skin grafting provided satisfactory cosmetic and functional outcomes.

Radiological imaging plays an essential role in assessing depth and operability. Molecular understanding of PDGFB fusion has expanded therapeutic options, especially for advanced cases.

Long-term surveillance is mandatory due to potential late recurrence. Multidisciplinary management ensures optimal outcomes.

Limitations

- Single case report.
- Short duration of follow-up.
- Molecular confirmation (COL1A1-PDGFB fusion) not performed.
- No comparison with Mohs surgery.

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