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CASE STUDY OF DERMATOFIBROSARCOMA PROTUBERANS AND ITS MANAGEMENT

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ABSTRACT

Background: Dermatofibrosarcoma Protuberans (DFSP) is a rare, low-to-intermediate grade soft tissue sarcoma of dermal origin characterized by slow growth, local aggressiveness, and a high propensity for local recurrence despite its low metastatic potential. It commonly presents as a painless, indurated cutaneous plaque that gradually progresses into nodular or protuberant masses, most frequently affecting the trunk and proximal extremities. Diagnosis is established through histopathological examination demonstrating spindle cell proliferation in a storiform pattern, supported by immunohistochemical positivity for CD34. Surgical excision remains the cornerstone of treatment, with wide local excision or Mohs micrographic surgery providing optimal local control through complete margin clearance. In cases of unresectable, recurrent, or metastatic disease, targeted therapy with imatinibmesylate has shown significant clinical benefit due to the characteristic COL1A1-PDGFB fusion gene associated with DFSP. Long-term follow-up is essential because of the risk of local recurrence, particularly in cases with inadequate surgical margins. Early diagnosis, appropriate surgical management, and vigilant surveillance are critical in achieving favorable outcomes and minimizing morbidity associated with this uncommon neoplasm.

KEYWORDS: Dermatofibrosarcoma protuberans, DFSP, soft tissue sarcoma, wide local excision, Mohs micrographic surgery, imatinib, local recurrence.

INTRODUCTION

Dermatofibrosarcoma Protuberans (DFSP) is a rare, locally aggressive mesenchymal neoplasm arising from the dermis and subcutaneous tissue. First described by Darier and Ferrand in 1924, DFSP accounts for less than 0.1% of all malignant neoplasms and approximately 1–6% of all soft tissue sarcomas. Despite its low incidence, DFSP is clinically significant because of its infiltrative growth pattern, tendency for extensive local invasion, and high rate of local recurrence when inadequately treated. However, distant metastasis is uncommon, occurring in less than 5% of cases, usually following multiple recurrences or fibrosarcomatous transformation.

The tumor typically affects young and middle-aged adults between the second and fifth decades of life, although it can occur at any age. There is no strong gender predilection. The trunk is the most commonly involved site, followed by the proximal extremities, head, and neck region. Clinically, DFSP often presents as a slow-growing, painless cutaneous plaque or nodule that may remain stable for several years before progressing into a multinodular protuberant mass. Because of its indolent course and nonspecific appearance, the lesion is frequently misdiagnosed as a benign dermatological condition such as a keloid, dermatofibroma, lipoma, epidermoid cyst, or hypertrophic scar, resulting in delayed diagnosis and treatment.

Histopathologically, DFSP is characterized by a dense proliferation of uniform spindle-shaped cells arranged in a storiform or cartwheel pattern with infiltration into the surrounding adipose tissue, producing the characteristic “honeycomb” appearance. Immunohistochemically, tumor cells demonstrate strong positivity for CD34 and are typically negative for factor XIIIa, aiding in differentiation from other spindle cell tumors. Molecular studies have identified a characteristic chromosomal translocation t(17;22)(q22;q13), resulting in fusion of the COL1A1 and PDGFB genes. This genetic alteration leads to constitutive activation of platelet-derived growth factor beta (PDGFB) signaling and plays a central role in tumorigenesis.

The mainstay of treatment for DFSP is complete surgical excision with histologically negative margins. Wide local excision and Mohs micrographic surgery are considered the treatment modalities of choice due to their ability to achieve superior local control while minimizing recurrence. In advanced, unresectable, recurrent, or metastatic cases, targeted therapy with imatinibmesylate has emerged as an effective treatment option by inhibiting

PDGFB receptor signaling. Radiotherapy may also be considered in selected patients with positive margins or when complete surgical excision is not feasible.

Given the rarity of DFSP and its propensity for local recurrence, awareness among clinicians and surgeons is essential for early diagnosis and appropriate management. This article aims to discuss the clinical presentation, diagnostic approach, histopathological features, and current management strategies of Dermatofibrosarcoma Protuberans, highlighting the importance of complete surgical excision and long-term follow-up in achieving optimal patient outcomes.

MATERIALS AND METHODS

Case presentation

Chief Complaints

A 37 years old female patient visited OPD of our hospital with lesion at Right scapular region since 3 weeks, pain at lesion site since 3 weeks, Bloody discharge from the lesion since 2 days.

History of present illness

Approximately since 3weeks the patient developed a lesion at right scapular region with pain and bloody discharge from the lesion, for which she visited our OPD for immediate surgical intervention.

History of past illness

Other than the present complaints, the patient had surgical history of LSCS 5 years ago with no medical history.

Personal and family history

The patient was a housewife. Diet: Mixed (vegetarian + non-vegetarian), Habits: No addictions. Allergies: No known drug allergies. Family history: Mother died due to CA breast.

Physical examinantion

General condition: Fair, afebrile P- 78/min

BP-130/80 mmHg

SpO_2- 98% on room air

RR-18/Min

CNS: conscious and oriented

CVS: S1 S2 normal

RS: AEBE clear

Abdomen: Soft, Bowel sounds present, motion passed

Pallor: Mild seen at conjunctiva

Icterus/ Lymphadenopathy: Not seen.

Local examination (Fig. 1.):

On Inspection :Swelling at right scapular region approximately 2×3cm, Blood oozing from the swelling site, Blackish discoloration noted over the swelling site, Shape of the lesion being irregular On palpation: Tenderness was present, Consistency- hard, Fluctuation test- Negative.

Laboratory examinations

Hb- 8.2 gm/Dl

RBC- 4.29 million/mm^3

WBC- 9020/mm^3

Platelets- 4.47 lakh/mm^3

Management

Excision of lesion at Right scapular region was performed under Local Anaesthesia.(Fig. 2)

The Specimen was sent for Histopathology Examination.

Histopathology Report

It showed lesion composed of spindle cells arranged in a storiform pattern.

Moderated increase in mitotic activity.

Diagnosis: Spindle cell neoplasm

S/O **Dermatofibrosarcoma protuberans**

The patient was further adviced for PET CT Scan.

REPORT

Post operative inflammation in operative bed weakly metabolic enhancing lesion in inferior quadrant of left breast measuring 13mm Further WIDE LOCAL EXCISION was planned and performed.

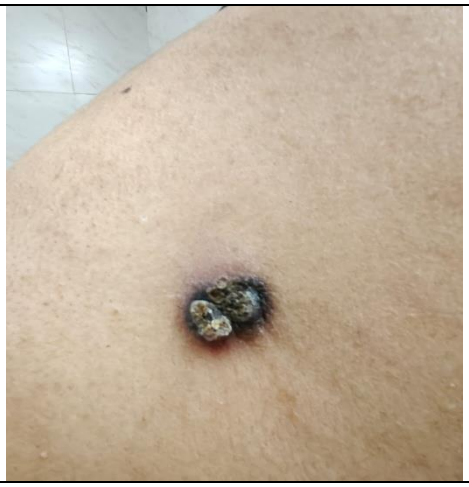

Figure 1: Pre op.

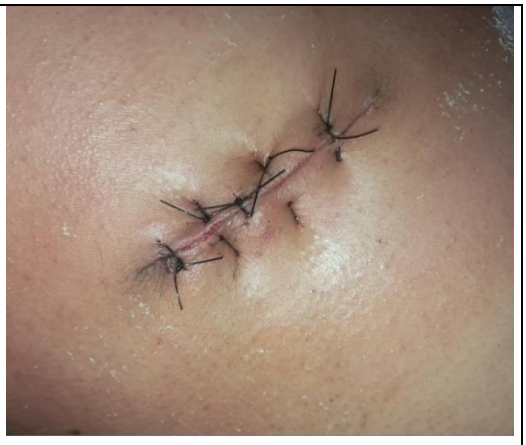

Figure 2: Post op.

DISCUSSION

Dermatofibrosarcoma Protuberans (DFSP) is a rare soft tissue sarcoma characterized by a paradoxical biological behavior: although it exhibits low metastatic potential, it is highly infiltrative and associated with a significant risk of local recurrence. The rarity of the tumor, coupled with its slow growth and often innocuous clinical appearance, frequently leads to delayed diagnosis. Many patients initially present with a long-standing painless cutaneous lesion that may be mistaken for benign conditions such as lipoma, dermatofibroma, epidermoid cyst, keloid, or hypertrophic scar. Consequently, diagnosis is often established only after progressive enlargement or histopathological examination following excision.

The hallmark of DFSP is its infiltrative growth pattern. Histologically, tumor cells extend beyond clinically visible margins through finger-like projections into the surrounding dermis, subcutaneous tissue, fascia, and occasionally underlying muscle. This microscopic extension explains the historically high recurrence rates observed following simple excision or inadequate surgical margins. Recurrence rates of up to 50–60% have been reported after limited excision, emphasizing the importance of achieving complete tumor clearance.

Histopathological examination remains the gold standard for diagnosis. The characteristic storiform arrangement of spindle cells and honeycomb infiltration of subcutaneous fat are important diagnostic features. Immunohistochemical staining plays a crucial role in differentiating DFSP from other spindle cell neoplasms, with strong CD34 positivity being a distinguishing characteristic. Advances in molecular pathology have further improved diagnostic accuracy through identification of the COL1A1-PDGFB fusion gene, which is

present in the majority of cases and contributes to tumor development through activation of platelet-derived growth factor signaling pathways.

Surgical excision remains the cornerstone of treatment. Wide local excision with margins of 2–3 cm has traditionally been recommended to reduce the likelihood of recurrence. However, because of the tumor's irregular microscopic extensions, Mohs micrographic surgery has gained increasing acceptance as an effective treatment modality. Mohs surgery allows complete circumferential and deep margin assessment while preserving uninvolved tissue, resulting in lower recurrence rates and improved cosmetic outcomes, particularly in anatomically sensitive regions such as the head and neck.

In patients with unresectable, recurrent, or metastatic disease, targeted therapy with imatinibmesylate has significantly altered management strategies. The drug specifically inhibits PDGFB receptor signaling associated with the characteristic chromosomal translocation of DFSP, leading to tumor regression in many patients. Imatinib may also be utilized as neoadjuvant therapy to reduce tumor size and facilitate surgical resection. Nevertheless, surgery remains the definitive treatment whenever feasible.

Although distant metastasis is uncommon, occurring in a small percentage of patients, the risk increases in cases demonstrating fibrosarcomatous transformation. Fibrosarcomatous DFSP represents a more aggressive variant characterized by increased cellular atypia, higher mitotic activity, and a greater propensity for both recurrence and metastasis. Identification of this variant is therefore important for prognostication and treatment planning.

Long-term surveillance is essential because local recurrence may occur several years after initial treatment. Regular clinical examination and imaging when indicated are recommended, particularly during the first five years following surgery when recurrence is most likely to occur. Early detection of recurrent disease allows timely intervention and improves overall outcomes.

In conclusion, DFSP is an uncommon but clinically important soft tissue sarcoma that requires a high index of suspicion for diagnosis. Complete surgical excision with negative margins remains the primary treatment strategy, while advances in molecular diagnostics and targeted therapies have improved outcomes in advanced disease. Careful long-term follow-up

is necessary due to the persistent risk of local recurrence, highlighting the importance of multidisciplinary management in achieving optimal patient care.

CONCLUSION

Dermatofibrosarcoma Protuberans (DFSP) is a rare, locally aggressive soft tissue sarcoma with a high propensity for local recurrence but low metastatic potential. Early diagnosis, accurate histopathological evaluation, and complete surgical excision with negative margins are essential for optimal outcomes. Long-term follow-up remains crucial for the timely detection and management of recurrence.

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