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CASE REPORT

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Rare case of massive inguinal dermatofibrosarcoma protuberans: A case report

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Key Clinical Message

DFSP is a cutaneous soft tissue sarcoma. A 35 year old male patient presented with DFSP in the inguinal region which is a rare soft tissue sarcoma which usually presents in the torso, occurring very rarely in the inguinal region. Hence in case of any swelling in the inguinal region, DFSP should be in differential diagnosis.

KEYWORDS

dermal sarcoma, dermatofibrosarcoma protuberans, fibrosarcomatous transformation, soft tissue sarcomacutaneous soft tissue sarcoma, surgical considerations

1 | INTRODUCTION

Dermatofibrosarcoma protuberans (DFSP) is a rare type of skin cancer that originates from cells in the dermis, the middle layer of the skin. It typically manifests as a slow-growing, firm plaque that can be violet-red or blue in color.[1] In advanced stages, DFSPs may grow larger and develop protrusions or ulcers.[2] Most DFSP cases involve a specific chromosomal translocation, t(17;22) (q22;q13).[1] The incidence of DFSP is low, with only about one case per million people each year. It represents 1%–6% of all soft tissues sarcomas and 18% of cutaneous soft tissue sarcomas. In pakistan DFSP accounts for less 0.1% of all cutaneous malignancies.[3] While recent studies indicate no gender bias in its prevalence, the condition is predominantly found in middle-aged men.[1,4] Histopathologically, DFSP is characterized by a neoplasm with an indistinct border, featuring spindle-shaped cells arranged in a storiform pattern within short, interwoven fascicles.[5] DFSP most frequently appears on the trunk and proximal extremities. Rarely being reported on pubic and inguinal region. This case is reported due to its rarity.

2 | CASE HISTORY/ EXAMINATION

A 35 years old male from Pakistan, with a previous history of tuberculosis presented at the hospital with a chief complaint of a mass in the right inguinal region along with abdominal pain. The patient said that the size of the swelling had been steadily rising over the period of 8 years. There was no past history of weight loss or any family history of cancer.

During the physical examination, a large, hard tumor was appreciated to be about 25 cm in size located in the biinguinal area of the patient with origin from the right inguinal area (Figure 1). The mass was firm and non tender;

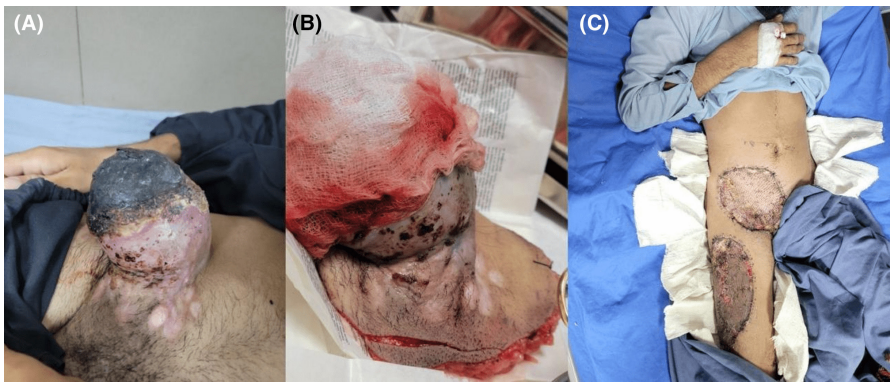

FIGURE 1 (A) Initial presentation of the dermatofibrosarcoma in the inguinal region that the patient presented with. (B) Excised mass of dermatofibrosarcoma after surgical excision. (C) Patient after the skin grafting was done.

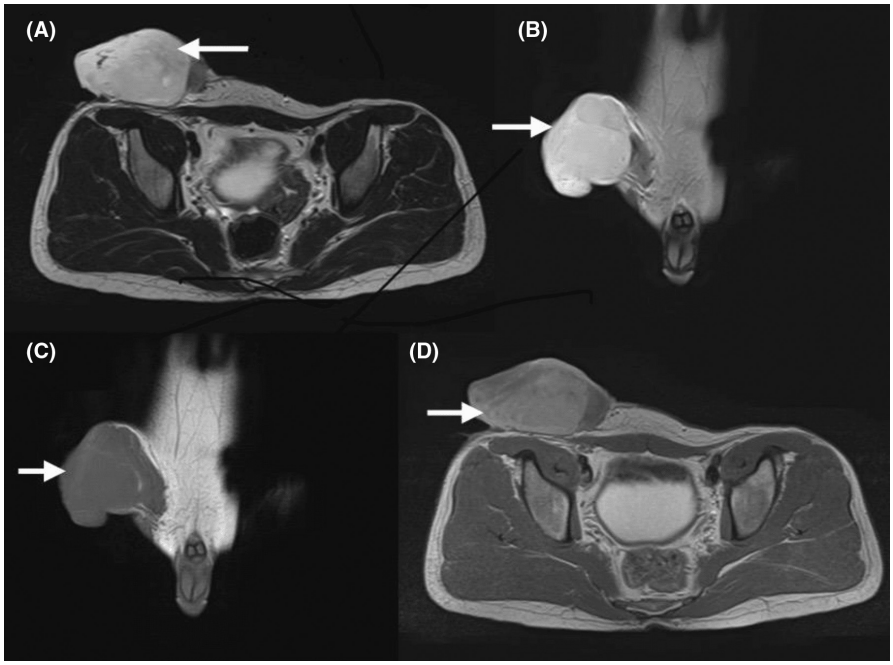

FIGURE 2 (A) (axial T2W), (B) (coronal T2W), (C) (coronal T1W) and (D) (axial post contrast T1W fat sat) showing a large, lobulated, exophytically growing T1 hypointense (in (C)) and T2 hyperintense (A, B) lesion in right inguinal region showing heterogenous post contrast enhancement (in D).

no ulceration of the skin or infection over the mass was observed. The patient was also suffering from localized abdominal pain.

3 | METHODS (DIFFERENTIAL DIAGNOSIS, INVESTIGATIONS, AND TREATMENT)

An MRI (Figure 2) was performed on which a large lobulated abnormal MR signal intensity with circumscribed lesion was noted in the right inguinal region growing exophytically and producing significant extraneous bulge on overlying skin with adjacent thickening of pelvic wall. The lesion displayed hypointense signals on T1w, hyperintense on T2w and STIR levels and lesion displayed heterogeneous post contrast enhancement.

An excision biopsy was done and viewed under a microscope. The tissue showed spindle cell proliferation with areas of hypocellularity. The cells were arranged in the form of sheets with focal fasciculation and storiforming (Figure 3). Immunohistochemically, spindle cells were found positive for CD 34 antigen.

4 | CONCLUSIONS AND RESULTS (OUTCOME AND FOLLOW-UP)

The mass was surgically excised and initially a full thickness skin graft was taken from the right thigh region, which did not heal properly, so was replaced by split thickness skin graft taken from the left thigh region which was successful (Figure 1). After which, the patient was discharged and sent back home.

5 | DISCUSSION

DFSP is a rare form of soft tissue sarcoma, typically located on the trunk and upper limbs.[1]

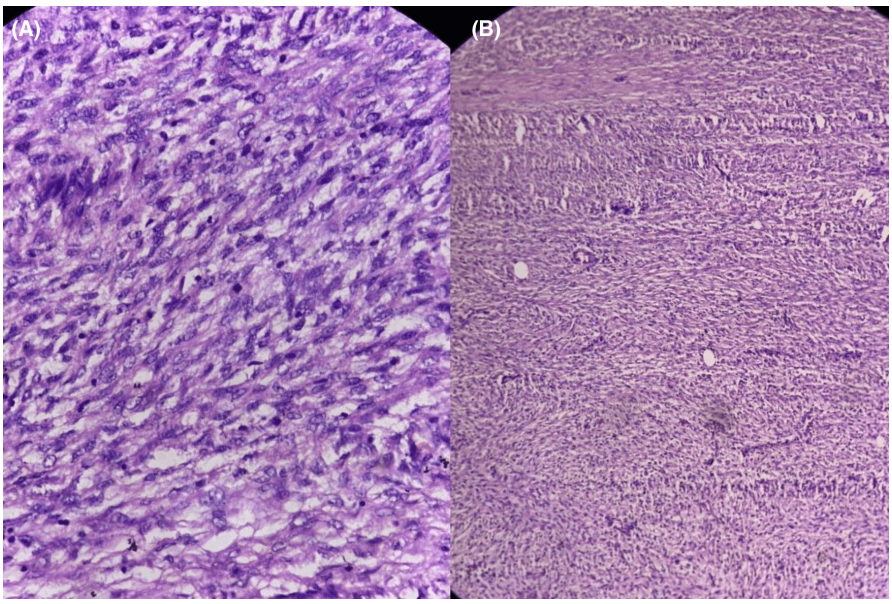

FIGURE 3 (A) Round to ovoid tumor cells with bland nuclei (40x). (B) Storiform pattern (10x).

It is a slow growing tumor but is highly likely to recur because it tends to extensively invade subcutaneous tissue, fascia, and underlying muscle, often spreading in a pseudopod-like manner.[6] The case of giant dermatofibrosarcoma in inguinal region exist as a gap in literature.

More than 90% of dermatofibrosarcomas cases, irrespective of their histological pattern, are linked to a distinctive reciprocal t(17;22) (q22;q13) translocation. This genetic alteration fuses the COL1A1 gene (collagen type 1 alpha 1) with the PDGFB gene (platelet-derived growth factor beta).[7]

Several studies have also proposed a link between the overexpression of the PDGFB gene and the development of DFSP. This overexpression is secondary to the translocation between chromosome 17 and 22.[8]

DFSP usually appears as a slow growing dermal nodule, often with a violaceous coloring.[9]

Distinguishing DFSP from other neoplasms such as dermatofibroma, fibrosarcoma, leiomyosarcoma, undifferentiated or unclassified soft tissue sarcoma, or atypical fibroxanthoma can be challenging for clinicians.[1] In rare instances, DFSP may exhibit regions with high-grade fibrosarcomatous features, such as more than 5 mitoses per 10 high-power fields (HPF), a fascicular growth pattern, increased cellularity, and atypical cells. When these high-grade areas make up more than 5% of the tumor, the lesion is classified as fibrosarcomatous.[10]

Accurate diagnosis requires a thorough assessment of clinical presentation and morphological features, supported by biopsy. Microscopic examination usually reveals a uniform proliferation of spindle-shaped cells arranged in a storiform pattern, often encircling and entangling subcutaneous fat to create a honeycomb-like appearance. In difficult cases, molecular techniques like FISH and RT-PCR can detect gene rearrangements and fusion transcripts in formalin-fixed, paraffin-embedded tumor tissues.[11,12]

Mohs Micrographic surgery (MMS) is often the preferred method for treating DFSP, especially for tumors located in delicate skin areas or those that have recurred. In this approach, tissue layers are removed one at a time and inspected microscopically during the surgery to determine the extent of tumor spread.[13]

Metastatic DFSP should be managed with adjuvant radiation therapy or chemotherapy regimens, either single-agent or multi-agent, typically used for treating sarcomas.[14]

DFSP typically has a positive survival outlook, even in cases with lymph node involvement or distant metastases.[15]

DFSP are tumors which have a high rate of recurrence occurring late in 24% to 90% cases. Factors linked to increased recurrence rates include the histological subtype, cellular density, tumor size, its location on the head and neck, and a high mitotic rate.(6)The recurrence-free survival rates for DFSP are 86% at 5 years and 76% at 10 years.[14]

However, mortality specific to DFSP is significantly higher in patients with grade III tumors or those with tumors measuring 10 cm or larger.[15]

AUTHOR CONTRIBUTIONS

Abdul Ahad Riaz: Conceptualization; investigation; writing – original draft; writing – review and editing. **Faisal Naseer:** Conceptualization; data curation; resources; writing – original draft. **Linta Malik:** Conceptualization; project administration; visualization; writing – original draft. **Allahdad Khan:** Conceptualization; methodology; project

administration; writing – original draft; writing – review and editing. **Aseel Kamal:** Conceptualization; methodology; visualization; writing – original draft. **Mudasira Habib:** Methodology; project administration; supervision; writing – original draft.

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CONFLICT OF INTEREST STATEMENT

We have no conflict of interest.

DATA AVAILABILITY STATEMENT

Data available on request from the authors.

CONSENT

Written consent from the patient was taken.

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