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CASE REPORT PEER REVIEWED | OPEN ACCESS

Interesting case of dermatofibrosarcoma protuberance

Agil Selvam, Pallavi Kothe, Vinoth M, Arcot Rekha, Jaya Ganesh

ABSTRACT

Introduction: Dermatofibrosarcoma protuberans (DFSP) is an uncommon soft tissue neoplasm of intermediate to low-grade malignancy. It is a locally aggressive tumor with a high local recurrence but rarely has metastasis. It is a cutaneous malignancy that arises from the dermis and invades deeper tissue (e.g., fat, fascia, muscle, and bone).

Case Report: A 45-year-old male patient presented to the surgery outpatient department with complaints of swelling in the lateral aspect of the left thigh since six months, insidious in onset, gradual in progression. It was associated with pain on walking, sudden increase in size over the last three months. Examination showed swelling of 8 × 5 × 3 cm in anterolateral aspect of left thigh, mobile, mixed in consistency with no peripheral vascular deficit. Ultrasound imaging showed a well-defined isoechoic lesion in subcutaneous plane in anterolateral aspect of left thigh, probably sarcomatous lesion. Trucut biopsy revealed features of spindle cell sarcoma probably fibroblastic. Histopathology examination revealed features of dermatofibrosarcoma protuberance.

Conclusion: In the differential diagnosis of nontraumatic leg pain, tumors, particularly dermatofibrosarcoma should be considered. Magnetic resonance imaging (MRI) is diagnostic and complete surgical resection is curative where recurrence rate is less.

Keywords: COL1A1-PDGFB, DFSP, Morphea-like, Storiform

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INTRODUCTION

Dermatofibrosarcoma protuberance is an uncommon cutaneous malignancy. It is soft tissue neoplasm that invades deeper tissues. The cellular origin is fibroblastic, histiocytic, or neuroectodermal though some doubt still exists regarding its cell of origin. It is known to invade fat, fascia, muscle, and even bone. It arises from the dermis and has intermediate to low grade malignancy [1].

It has various presentations ranging from indurated plaque to firm, irregular mass. Color of the skin over the nodule varies from flesh to reddish brown. Plaque may present as morphea-like, atrophic, sclerotic, or violaceous and may progress to ulcerate. Telangiectasia may also be an accompanying feature over the nodule.

CASE REPORT

A 45-year-old male patient presented to the surgery outpatient department with complaints of swelling in the lateral aspect of the left thigh since six months, insidious in onset, gradual in progression. It was associated with pain on walking, sudden increase in size over the last three months. Examination showed swelling of 8 × 5 × 3 cm in anterolateral aspect of left thigh, mobile, mixed

in consistency with no peripheral vascular deficit as described in Figure 1A and B. Ultrasound Imaging showed a well-defined isoechoic lesion in subcutaneous plane in anterolateral aspect of left thigh, probably sarcomatous lesion. Trucut biopsy revealed features of spindle cell sarcoma probably fibroblastic.

Imaging: MRI of the left thigh as described in Figure 2A–C revealed evidence of well-defined signal intensity soft tissue mass noted involving lateral aspect of left thigh. Perilesional fat plane was maintained. No evidence of intramuscular extension was seen. Lesion of 6 × 5.8 × 3.5 cm displaying hypointensity in T1 and hyperintensity in T2. Metastatic work-up was negative.

The patient was taken up for surgery with a clinical diagnosis of a soft tissue sarcoma. The mass was excised with a 1 cm margin and the fascia was intact. The ensuing defect was covered with a split skin graft as described in Figure 3A–C. Histopathology showed spindle cells in a storiform to whorled pattern with abundant eosinophilic cytoplasm as described in Figure 4A–D. There was infiltration of subcutaneous fat with a honeycomb pattern suggestive of DFSP, PT_2 FNCLCC grade I (positive for CD34).

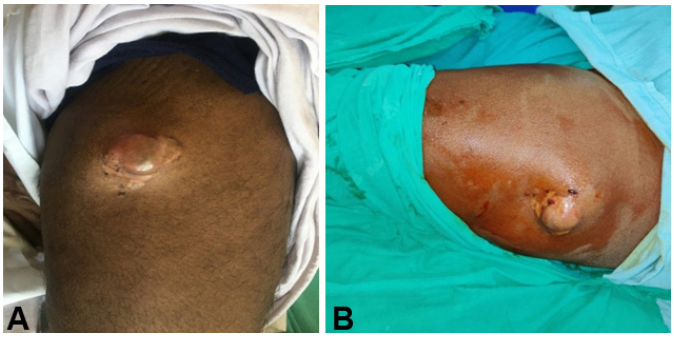

Figure 1: (A) and (B) Showing preoperative images of the swelling.

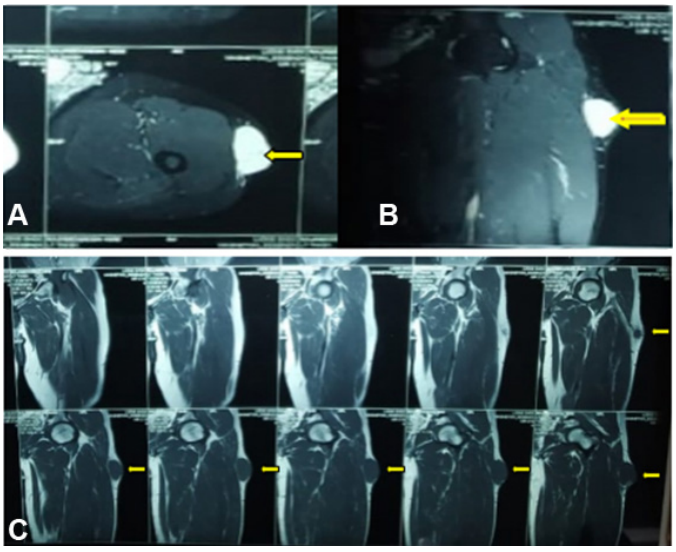

Figure 2: (A)–(C) Magnetic resonance imaging showing about 6 × 5.8 × 3.5 cm lesion with no intermuscular plane seen in coronal and sagittal plane shows perilesional fat plane (arrow).

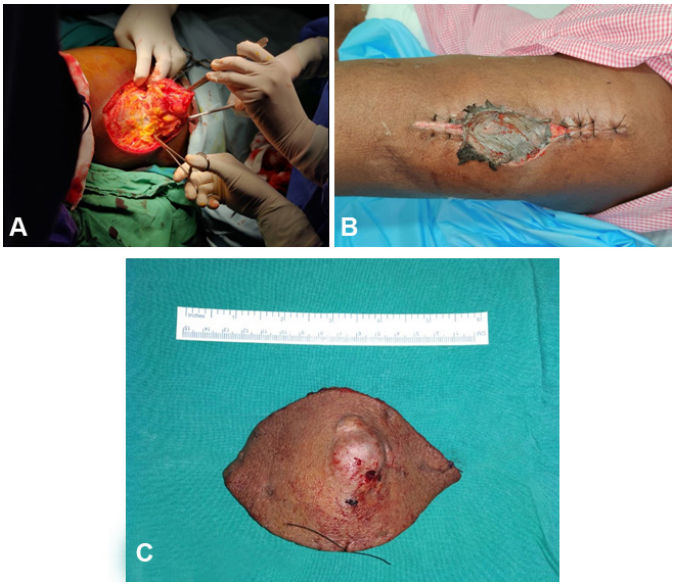

Figure 3: (A)–(C) Excision of the tumor completely, complete enucleation of tumor with Scandinavian sarcoma group (SSG) and specimen in toto.

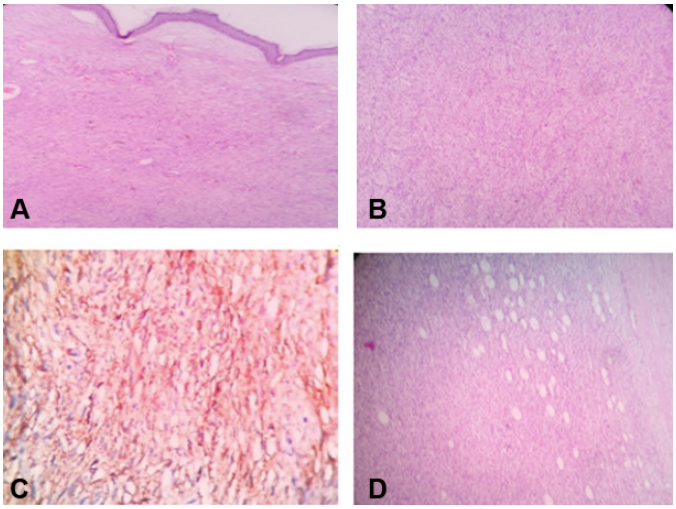

Figure 4: Histopathological examination and staining. (A) H&E staining epidermis with a tumor in dermis composed of spindle cells. (B) Bland spindle cells with elongated nuclei in a prominent storiform or cartwheel appearance. (C) Immunohistochemically strong and diffuse expression of CD34. Linearly oriented tumor strands that infiltrate the subcutaneous fat in "honeycomb pattern." (D) Higher magnification demonstrating tumor cells infiltrating the fatty tissue.

DISCUSSION

Dermatofibrosarcoma protuberance is an uncommon cutaneous malignancy. It is soft tissue neoplasm that invades deeper tissues. The cellular origin is fibroblastic, histiocytic, or neuroectodermal though some doubt still exists regarding its cell of origin. It is known to invade fat, fascia, muscle, and even bone. It arises from the dermis and has intermediate to low grade malignancy [2–5].

Cellular origin of the tumor is fibroblastic, histiocytic, or neuroectodermal. Pluripotential progenitor cells may

be the origin of DFSP because they have the capacity to differentiate into all three cell types.

Dermatofibrosarcoma protuberance pathogenesis is still under study. It is not known to be hereditary or familial though chromosomal aberrations are found [6]. One of the most common etiologies is trauma wherein previous history of burns, surgical scars, or vaccinations have contributed to its cause.

It has been reported in persons of all races, and no racial predilection seems to exist in previous reports. Studies reveal equal sexual distribution or a slight male predominance. It usually occurs in adults aged 20–50 years. Rarely, DFSP has been reported in newborns and elderly individuals (80 years) [7].

In early stages, tumor is mobile. In advanced stage with deeper invasion into fascia, muscle, bone mobility becomes restricted. It is most common in trunk (42–72%) followed by proximal extremities (16–30%), and rarely above neck or breast according to studies. Metastatic work-up includes ultrasonography (USG)/chest X-ray, and MRI.

A skin biopsy is essential for definitive diagnosis of DFSP.

1. Plaque type—spindle-shaped nuclei of slender tumor cells are distributed uniformly in collagen stroma with sparse mitotic figures in microscopic studies.
2. Nodular type—It is known to show storiform pattern and cartwheel pattern of atypical cells and fibrous tissue. Cellular atypia is higher in nodular type.
3. Pigmented variant also known as Bednar tumor—the melanin-containing dendritic cells are scattered between the neoplastic spindle-shaped cells.
4. Juvenile form (giant cell fibroblastoma), cleft-like pseudo-vascular spaces are lined by multinucleated cells. The intervening tumor may have loose hypocellular areas and areas that resemble mature DFSP.

CD34 immunohistochemistry study is necessary for differentiating DFSP from normal stromal cells, dermatofibroma (positive for factor XIIIa) and identification of positive surgical margins. It shows moderate to strong staining in tumor cells.

Staging for DFSP is as discussed below:

Staging system	
Stage I	Primary tumor, localized disease
Stage II	Lymph node metastasis
Stage III	Distal metastasis

Radiation therapy is used when tumor margins are positive in histopathological examination. In case of insufficient evidence of clear margins intraoperatively, postoperative adjuvant radiotherapy is advised. It reduces the risk of recurrence. The dose of radiotherapy ranges from 50 to 70 Gy [8]. Postradiation complications are low. Chemotherapy is rarely used.

Molecular directed therapy is under research for routine application as treatment with PDGFB-PDGFR-beta signaling pathway plays a pivotal role in pathogenesis of DFSP and proliferation of tumor cells.

Imatinib mesylate has been found to have significant therapeutic value in the treatment of DFSP. Imatinib is a potent and specific inhibitor of several protein-tyrosine kinases, including the platelet-derived growth factor (PDGF) receptors. It is indicated for the treatment of adult patients with unresectable, recurrent, and/or metastatic DFSP. The recommended oral dose is 800 mg/d. Neoadjuvant imatinib therapy for DFSP has been proposed in recent studies [9].

In case of recurrent DFSP or locally advanced tumor, imatinib plays a role to decrease tumor load, promote apoptosis, and reduce extent of surgery.

Excision is the primary treatment wherein Mohs micrographic surgery is the treatment of choice. It has the advantage of less tissue removal and complete margin assessment over the other options. Dermatofibrosarcoma protuberans has high recurrence rate after standard surgical excision [10]. This is due to infiltrating growth pattern of the tumor which extends beyond the clinical margins of the tumor.

It has to be kept in mind during resection of tumor a margin of more than 3 cm is attained in all planes including deeper fascia if needed for appropriate treatment. For inoperable, metastatic, recurrent tumors, medical therapy is advised where in imatinib mesylate is effective.

Recent Advances: DFSP usually has fusion genes COL1A1-PDGFB [11], COL1A2-PDGFB [12], COL6A3-PDGFD, and EMILIN2-PDGFD. Incomplete resection, fibrosarcomatous transformation increased age, male sex, and tumor size are associated with worse overall survival. Cytogenetic studies identified a supernumerary ring chromosome t (17:22) q (22:13) [12, 13].

CONCLUSION

These dermatofibrosarcoma protuberances are less frequent in the lower extremities according to the literature. In the differential diagnosis of nontraumatic leg pain, tumors, particularly dermatofibrosarcoma should be considered. Magnetic resonance imaging is diagnostic and complete surgical resection is curative where recurrence rate is less.

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Author Contributions

Agil Selvam – Conception of the work, Design of the work, Acquisition of data, Analysis of data, Interpretation of data, Drafting the work, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Pallavi Kothe – Conception of the work, Design of the work, Acquisition of data, Analysis of data, Interpretation of data, Drafting the work, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Vinoth M – Design of the work, Drafting the work, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Arcot Rekha – Conception of the work, Design of the work, Acquisition of data, Analysis of data, Interpretation of data, Drafting the work, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Jaya Ganesh – Analysis of data, Interpretation of data, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Guarantor of Submission

The corresponding author is the guarantor of submission.

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Consent Statement

Written informed consent was obtained from the patient for publication of this article.

Conflict of Interest

Authors declare no conflict of interest.

Data Availability

All relevant data are within the paper and its Supporting Information files.