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Case Report

From subtle to striking: A rare case of dermatofibroma protuberans and its clinical journey ☆,☆☆

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ABSTRACT

Dermatofibrosarcoma protuberans (DFSP) is a rare type of soft tissue sarcoma, which is slow-growing. It arises from the dermal, subcutaneous layer and is locally aggressive. It commonly affects adults between 20 and 50 years old, with a slight male predominance. This case report emphasizes the case of a 35-year-old male patient who presented with a large, firm, nodular mass on his left upper arm that had been progressively increasing in size over 3 years. On inspection, the lesion measured approximately 13 cm in diameter, was large and lobulated with overlying red-brown discolouration. The patient reported movement discomfort, and mild itching was noted. Diagnostic evaluation was advised, including X-ray, Ultrasound, Computed Tomography and Magnetic resonance imaging. A provisional diagnosis of soft tissue sarcoma was made. An excisional biopsy was performed, and histopathological analysis demonstrated a storiform pattern of spindle-shaped cells infiltrating the dermis and subcutis, consistent with dermatofibrosarcoma protuberans. The patient underwent wide local excision with clear margins to prevent recurrence, and no metastasis was detected. Dermatofibrosarcoma protuberans is characterized by a high rate of local recurrence but a low risk of metastasis. Early diagnosis and complete surgical excision are treatment options for good prognosis. This case underscores the importance of diagnosing and treating Dermatofibrosarcoma protuberans without delays.

Introduction

Dermatofibrosarcoma protuberans (DFSP) is a rare low-grade malignant cutaneous neoplasm. It arises from dermal and subcutaneous tissue elements due to the proliferation of its fibroblasts. Dermatofibromas are typically asymptomatic but

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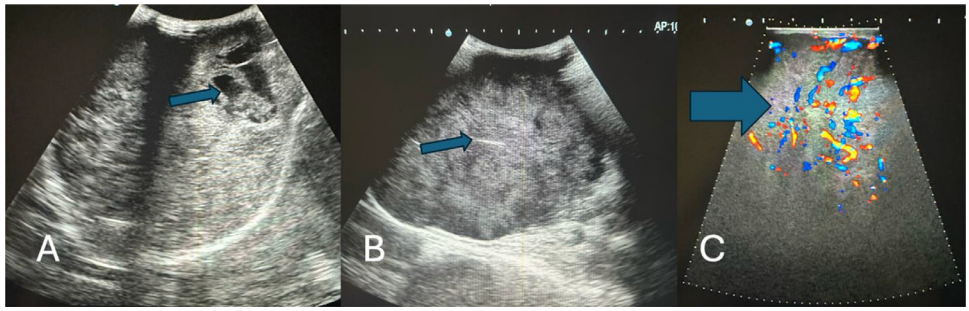

Fig. 1 – Grey scale B mode ultrasound image shows an ill defined heterogeneously hypoechoic mass lesion (A) was identified within the subcutaneous tissues of the medial aspect of the left arm with few cystic and calcific areas (A, B). The lesion was taking vascularity on Doppler and was suggestive of a soft tissue tumor (C).

may sometimes cause pain, tenderness, or itchiness if larger. It has predominance in the trunk and proximal extremities. Dermatofibrosarcoma protuberans, derived from dermal fibroblasts, was initially called keloid sarcoma [1].

This rare soft tissue sarcoma was initially described in 1890 by Taylor. Dermatofibrosarcoma protuberans lesions are generally seen as red-violet plaques with surrounding areas of telangiectasia. Lesions may be larger and nodular in late stages. Dermatofibrosarcoma protuberans lesions can be fixed to the dermis but move freely over deeper-lying tissue [2]. The size of the tumor depends essentially on the duration of growth. At onset, the tumor is generally small, ranging from 1 to 5 cm, but sizes greater than 20 cm have been reported. WHO has classified dermatofibrosarcoma protuberans as a fibrous, fibro-histiocytic, or histiocytic tumor [3].

Dermatofibrosarcoma protuberans are primarily diagnosed through histopathological analysis. Radiological imaging with X-ray, Ultrasound, Computed Tomography, and Magnetic resonance imaging is crucial in knowing deeper extensions. The treatment for localized dermatofibroma protuberans typically involves complete surgical excision of the lesion. Conventional surgery with wide margins (greater than 3 cm) or Mohs micrographic surgery can be performed [4].

We present a rare and unique case report highlighting dermatofibroma protuberans, discussing its clinical presentation, histopathological findings, radiological insights and management strategies. Through this case, we aim to enhance the understanding of this variant and underscore the importance of differential diagnosis in dermatological practice

Case report

Please expand on the patient's medical and surgical history, family history, initial presentation of the illness, laboratory results including normal values, management approach, and outcome.

A 35-year-old male residing in Southeast Asia came to the surgery outpatient department with complaints of a progressively enlarging mass on the left arm over the course of 2 years. The patient reported a small nodule on his left arm, initially asymptomatic, which had gradually increased in size. No significant pain, but cosmetic and occasional discomfort was noted. On physical examination, a firm, nontender nodular mass measuring approximately 13 cm in diameter was palpated on the left arm. The overlying skin was smooth but stretched due to the mass. There were no signs of ulceration or regional lymphadenopathy.

The patient had no known history of chronic illnesses such as diabetes, hypertension, tuberculosis, or autoimmune disorders. He denied any history of previous surgeries, major trauma, or radiation exposure. There was no record of prior hospital admissions for similar complaints or other systemic diseases. The patient reported no known family history of malignancies, connective tissue disorders, or genetic syndromes associated with soft tissue tumors.

To evaluate the patient's overall health and surgical candidacy, baseline blood tests were performed. Hematological findings indicated a hemoglobin level of 14.5 g/dL (reference range: 13.0-17.0 g/dL), white blood cell count of 9,600/mm^3 (reference range: 4,000-11,000/mm^3), and a platelet count of 275,000/mm^3 (reference range: 150,000-450,000/mm^3). Inflammatory markers included an erythrocyte sedimentation rate (ESR) of 16 mm/hr (reference range: 0-20 mm/hr) and a C-reactive protein (CRP) level of 6 mg/L (reference range: $<$10 mg/L). Kidney function assessment showed a serum creatinine level of 1.1 mg/dL (reference range: 0.7-1.2 mg/dL), while liver function tests were within the normal limits. Coagulation studies, including prothrombin time (PT), international normalized ratio (INR), and activated partial thromboplastin time (aPTT), were also within normal parameters. Additionally, tumor markers such as lactate dehydrogenase (LDH) and carcinoembryonic antigen (CEA) were within standard limits, with no significant elevation of specific sarcoma markers.

The patient was referred to the Radiology Department for an imaging workup. X-ray examination of the medial aspect of the left arm revealed a soft tissue mass with no evidence of calcification or underlying bony involvement. Greyscale B mode ultrasound image shows an ill-defined heterogeneously hypoechoic mass lesion (A) was identified within the subcutaneous tissues of the medial aspect of the left arm with few cystic and calcific areas (A, B). The lesion was taking vascularity on Doppler and was suggestive of a soft tissue tumor (C) (Fig. 1). Axial nonenhanced computed tomography scan of soft tissue (A) and bone window (B) and scanogram (C) of

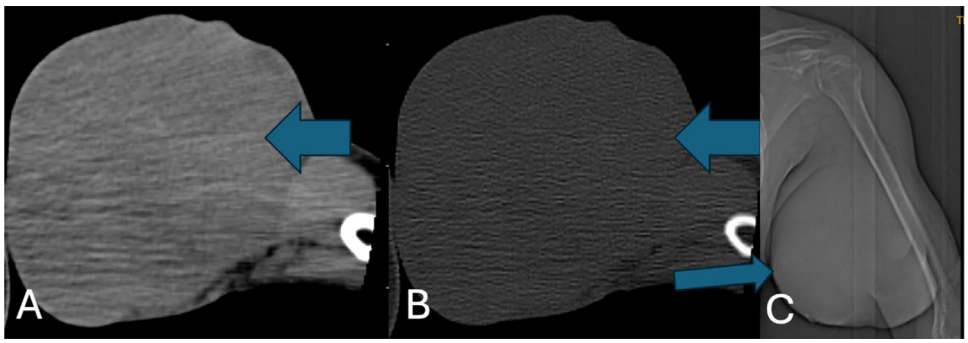

Fig. 2 – Axial nonenhanced computed tomography scan of soft tissue (A) and bone window (B) and scanogram (C) of the left arm demonstrated a well-defined, heterogeneous density mass lesion within the subcutaneous tissue with few hypodense cystic/necrotic areas, without invasion into deeper muscle layers.

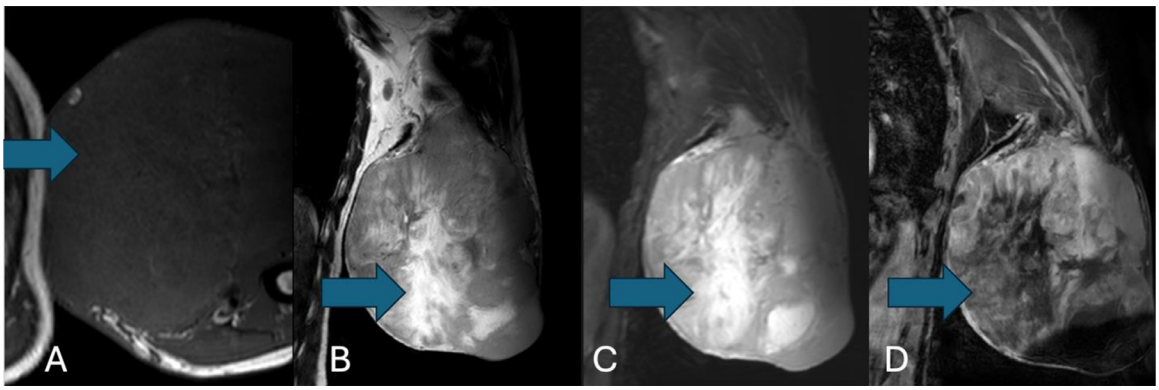

Fig. 3 – Magnetic resonance imaging of the left arm of the lesion shows a large lobulated heterogeneous soft tissue lesion with few nonenhancing areas of necrosis noted in the superficial plane of the medial aspect of the arm region on the left side. The lesion shows T1 hypointense (A), T2/STIR hetero-intense with few hyperintense areas within (B, C) with areas of blooming within the lesion. It shows post contrast enhancement (D). It measures approximately 115 x 153 x 127 mm. Diffuse subcutaneous edema with intervening streaks of fluid is noted in the thigh region. The underlying muscles appear normal. The head, greater and lesser tubercle and shaft of the left humerus show normal contour, morphology and marrow signal. No fracture or focal lesion.

the left arm demonstrated a well-defined, heterogeneous density mass lesion within the subcutaneous tissue with few hypodense cystic/necrotic areas, without invasion into deeper muscle layers (Fig. 2). Magnetic resonance imaging of the left arm of the lesion shows a large lobulated heterogeneous soft tissue lesion with few nonenhancing areas of necrosis noted in the superficial plane of the medial aspect of the arm region on the left side. The lesion shows T1 hypointense (A) and T2/STIR hetero-intense with few hyperintense areas within (B, C) with areas of blooming within the lesion. It shows post-contrast enhancement (D) (Fig. 3). It measures approximately 115 × 153 × 127 mm. Diffuse subcutaneous edema with intervening streaks of fluid is noted in the thigh region. The underlying muscles appear normal. The head, greater and lesser tubercle and shaft of the left humerus show normal contour, morphology and marrow signal. No fracture or focal lesion.

Based on the clinical presentation and imaging findings, a provisional diagnosis of soft tissue sarcoma was made. The patient was scheduled for surgical excision. Wide local excision of the mass was performed with a latissimus dorsi flap. The resected specimen was sent for histopathological examination.

Microscopic analysis of the excised tissue was done. The given section stained with Haematoxylin and Eosin (H&E) (high power view: 40x) (A, B, C, D, E, F) shows an infiltrative tumor with alternate hypocellular and hypercellular areas of spindle cells with high degree of nuclear pleomorphism, hyperchromasia, atypical mitosis and intervening blood vessels (Fig. 4). Histological features suggest a malignant mesenchymal tumor, consistent with dermatofibrosarcoma protuberans (DFSP). Immunohistochemical staining showed positivity for CD34, which further confirmed the diagnosis.

The patient's postoperative course was uneventful. Wound healing progressed without signs of infection or necrosis. He was advised on regular follow-up due to the potential for local recurrence, which is common in dermatofibrosarcoma protuberans despite adequate surgical margins.

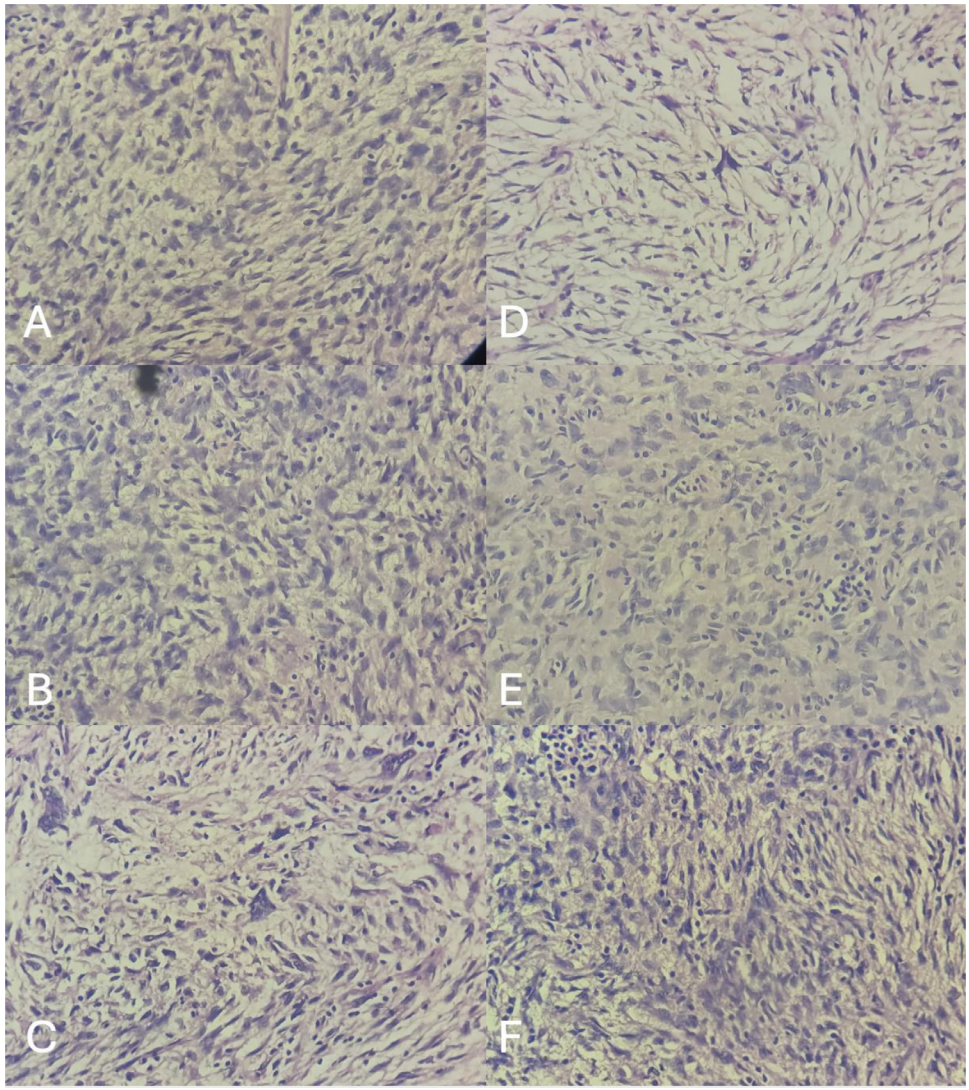

Fig. 4 – The given section stained with Haematoxylin and Eosin (H&E) (high power view: 40x) (A-F) shows an infiltrative tumor with alternate hypocellular and hypercellular areas of spindle cells with high degree of nuclear pleomorphism, hyperchromasia, atypical mitosis and intervening blood vessels. Histological features are suggestive of a malignant mesenchymal tumor.

Discussion

Dermatofibrosarcoma protuberans represents a superficial, low-grade, locally aggressive sarcomatous neoplasm characterized by the proliferation of spindle-shaped fibroblastic cells. As a relatively rare neoplastic entity and locally invasive cutaneous tumor, it is distinguished by elevated local recurrence rates while exhibiting a diminished propensity for metastasis. Dermatofibrosarcoma protuberans are generally noted as an asymptomatic plaque or nodule with a purple or pink hue, accompanied by a clinical history marked by gradual yet continuous growth [5].

Dermatofibrosarcoma protuberans (DFSP) possess the potential to infiltrate deeper anatomical structures, including adipose tissue, fascia, skeletal muscle, and osseous tissue. This neoplasm is recognized as the most prevalent fibrohistiocytic tumor, accounting for approximately 1.8% of the entirety of soft tissue sarcomas and constituting 0.1% of the overall cancer incidence. The incidence of this condition is estimated to range from 0.8 to 5 cases per million individuals annually [4].

From a clinical perspective, it is characterized as a neoplasm exhibiting slow proliferation, classified as a low- to intermediate-grade malignant sarcoma, and it is predominantly observed in adults of middle age, between 25 and 45 years of age. Histopathologically, it is comprised of coherent spindle cell fascicles that exhibit a storiform arrangement, featuring numerous variants and demonstrating robust widespread CD34 immunoreactivity [6]. Dermatofibrosarcoma protuberans lesions can be raised, fixed to the skin, and 75% displaced over deep planes [7]. Dermatofibrosarcoma protu-

berans predominantly manifests on the trunk and proximal limbs. It has a higher prevalence in the male population compared to females [2].

Ultrasound typically shows a heterogeneously hypoechoic matrix with few hypoechoic patches. Tiny echogenic foci (<0.5 mm) within the lesion can be noted. It is common to observe posterior acoustic enhancement. On color Doppler imaging, the majority of tumors exhibit moderate vascularity. The location of the lesion, its relationship to underlying structures, and architecture can be evaluated using computed tomography or magnetic resonance imaging. There is modest to moderate hyper vascularity. Enhancement observed on computed tomography scans obtained after contrast material injection [8]. Enhancement is often homogeneous after contrast injection and infrequently heterogeneous. A single, largely well-defined mass lesion in the subcutaneous area may also be seen on computed tomography. Bigger lesions (>5 cm) could have intratumoral nonenhancement zones, which might be indicative of cystic and necrotic intratumoral degeneration. On T1 sequences, the lesion is typically iso-intense to muscle, hyperintense on T2 sequences, and does not exhibit any signal drop-out on fat-suppressed sequences. Magnetic resonance imaging is useful in its function of locating and determining the degree of the lesion. Postcontrast enhancement is noted in the lesions. Regional lymphadenopathy and distant metastases are uncommon. However, localized invasion and infiltration are frequent [9].

Standardized treatment is surgical with wide local excision, ensuring negative margins of 3-5 cm from the tumor periphery. For osseous involvement, removal of the periosteum or bony segment is done. Recurrence can be prevented by adequate resection of margins. After excision, a local skin flap, skin graft, or myocutaneous flap can be done. An alternative to extensive surgical resection is Mohs micrographic surgery, widely regarded as the preferred treatment modality for dermatofibrosarcoma protuberans. Adjuvant therapy with imatinib mesylate is done for unresectable, recurrent and metastatic cases. Imatinib exerts its effects by inhibiting the tyrosine kinase activity of PDGF, which has proven effective. Postoperative radiotherapy is also advocated. The integration of conservative excision with adjuvant radiotherapy has been shown to result in a diminished local recurrence rate of 5% [10].

Ethics approval and consent to participate

Written consent taken.

Consent for publication

Written consent taken.

Availability of data and material

N/A.

Author contributions

DN and PHP was involved in providing clinical details of the patient. RP discussion on the pathology. SD accumulated the results of the patient's radiological investigations. AC and RK was involved in collecting images and formatting data. All authors have read and approved the manuscript.

Patient consent

Informed and written consent was obtained from the patient.

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