

REVIEW

Dermatofibrosarcoma protuberans (DFSP). A narrative review of the literature and current guidelines for its management

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Dermatofibrosarcoma protuberans (DFSP) is a slow growing low grade malignant tumour with a high rate of local recurrence and lower rate of metastasis and effecting patients between the 3rd and 5th decade. It accounts for less than 1% of all soft tissue sarcomas and can equally involve the trunk and extremities. It usually presents in the subcutaneous tissue but can grow deeper if increases in size. Diagnosis is usually with biopsy and is confirmed with its typical histological appearance of uniform spindle cells arranged in a storiform or honeycomb pattern with minimal cytologic atypia and low mitotic activity and shows diffuse positivity for CD34 on immunohistochemistry. The standard treatment after staging involves wide excision to achieve clear margins hence avoiding risk of local recurrence, which may be reduced with the use of adjuvant radiotherapy (RT), however, its use should be considered after discussion in multidisciplinary team meetings for select cases. The use of RT is more pronounced in larger/ unresectable or metastatic disease cases along with the use of Tyrosine kinase inhibitors (TKI) which has shown very promising results, particularly with Imatinib in cases with PDGFB mutations. The reported overall survival at 10 years following appropriate surgical treatment with localized DFSP is around 90%. The aim of this current review is to highlight the clinical features of DFSP, its surgical management and adjuvant treatments along with the current management guidelines.

Keywords: Dermatofibrosarcoma protuberans, DFSP, Sarcoma, wide local excision, Tyrosine kinase inhibitors.

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Introduction

This slow growing low to intermediate grade malignant cutaneous soft tissue sarcoma was named as DFSP in 1925 by the renowned German dermatologist, Dr. Eric Hoffman¹. It usually effects middle aged patients, but has been reported in all age groups, and can involve any part of the body but mostly extremities and the trunk and presents less than 1% of all soft tissue sarcomas. They effect men more than women and is more common in the darker skin population than white races^{2,3}. Its exact incidence in Pakistan, however, cannot be reported because of the lack of any national tumour registries or referral centres⁴. They can vary in size from sub centimeter to larger or recurrent lesions of varying sizes. They are usually painless lesions and can eventually cause symptoms depending on its location and size due to compression of surrounding structures. Radiological diagnosis is usually by magnetic resonance imaging (MRI) and for complete staging a computerized tomography scan (CT scan) or 18F-

fluorodeoxyglucose-positron emission tomography/computed tomography (18F-FDG PET/CT) is required^{5,6}. A punch or excision biopsy is required to confirm the histological diagnosis which shows spindle cell neoplasms in a characteristic honeycomb pattern with abundant myxoid stroma and increased cellularity and CD34 positivity on immunohistochemical staining⁷.

Standard treatment involves surgery in the form of wide local excision with attainment of R0 margins or Moh's micrographic surgery in certain cases⁷. In larger or recurrent lesions, wide local excision followed by soft tissue coverage with pedicled or free flaps is required (Figure 3A-E). In some advance cases involving the limbs, ablative surgery may be required where absolute indications for such are met⁸. They carry a very high risk of local recurrence (10-60%) and in certain cases, they can metastasize to various organs including lungs, lymph nodes, brain and adjacent structures (1-7.4%)^{9,10}. In cases of inoperable or multiply recurrent lesions, adjuvant treatments

such radiotherapy and more recently targeted therapy in the form of Tyrosine kinase inhibitors is utilized¹¹. The reported overall survivorship in non-metastatic disease is around 75% at 10 years¹².

This review article will highlight the clinical features, diagnostic work up along with surgical and medical management in keeping with the current available best clinical practices for this often misdiagnosed and difficult to treat soft tissue sarcoma.

Clinical Features

DFSP originates from the dermis and has a tendency to invade the surrounding tissues and manifests itself in the form of a solitary or a multi lobulated, painless, slow growing cutaneous lesion^{13,14}. They can present in various pathologic forms including a nodule, plaque, atrophic plaque or large lump and can be congenital or acquired, with acquired variety due to genetic mutations in chromosomes 17 & 22¹⁴. These lesions can be of varying sizes and can at times be static for years which can lead to delayed presentation. The superficial lesions can be red-brown to bluish nodular in appearance and may exhibit irregular margins¹⁵. Due to these varied presentations, they can often be missed, mis-treated surgically leading to positive margins excision or missed diagnosis delaying further treatment and compromising outcomes¹⁶. Most DFSP's present in the truncal lesion, followed by extremities and head neck^{17,18}.

Clinical examination along with the lesion must always include the local lymph nodes for involvement to document the staging of the disease. The lack of clinical awareness due to these varied presentations and rare occurrence of DFSP along with the absence of referral centers are also contributory causes of these guarded outcomes.

Diagnosis

Radiology & Staging

Ultrasound can be utilized as a screening investigation, which demonstrates a predominantly hypoechoic lesion with tail like projections seen in the periphery (Figure 1). Mild increased signal on Doppler imaging can be seen in some cases. Ultrasound is also helpful in performing image guided biopsy particularly in cases where the lesion is overlying or close to important structures.

MRI is the investigation of choice both for primary and recurrent lesions demonstrating the size, actual depth of the lesion and its association with important structures such as the neurovascular bundle and tendons. These are low to isointense to skeletal muscle on T1 and demonstrate high signal on fluid sensitive sequences. Peripheral appendage like extensions along the dermis can be seen on MRI. These demonstrate

homogenous or heterogeneous enhancement on post contrast images^{5,19}.

CT scans and 18F-FDG PET/CT can be used for staging of the disease along with evaluation of local lymph nodes, whereas the later can also be used to evaluate treatment response^{12,20}.

Biopsy & Histology

Diagnosis of DFSP requires a well performed biopsy in the form of a punch biopsy for smaller lesions and a trucut biopsy or excision biopsy for larger lesions. In cases of a recurrent or larger lesion in close proximity to important neurovascular structures, an ultrasound guided biopsy is advised. On gross microscopy, DFSP are dermal based spindle cell neoplasms which shows diffuse infiltration into the subcutaneous tissue and extension towards the overlying dermis. These tumor cells infiltrate the subcutaneous adipose tissue in a characteristic honeycomb or storiform pattern showing hypo cellular spindle cells embedded within abundant myxoid stroma and characterized by increased cellularity and intersecting fascicles arranged in a herringbone architecture (Figure 2A-D).

Immunohistochemistry panels will show strong and diffuse expression of CD34 which may be reduced or even lost in almost half the cases of DFSP with fibrosarcomatous transformation. CD 34 positivity is also noted in certain other soft tissue lesions including fibromyxomas, fibromas and Kaposi's sarcoma. DFSP will typically show negative expression for S100 and Factor XIIIa^{21,22}. DFSP's also demonstrate a morphological heterogeneity with multiple variants including myxoid, giant cell, sclerotic, pigmented and fibrosarcoma components. Except for fibrosarcoma, these variants do not have any significant clinical variations¹⁷.

Treatment

DFSP should be treated in a local referral center in a tumour board setting with all expertise on board wherein the best treatment can be offered keeping in view the recent most guidelines^{23,24}. This multidisciplinary approach has led to improved oncological and reconstructive outcomes leading to reduced risk of local recurrence and improving functional. However, treatment should be commenced only after complete local and systemic staging of the disease.

Surgery

Complete and wide surgical excision of DFSP remains the mainstay of treatment and adequate surgical excision guarantees lower risk of local recurrence and subsequent systemic disease. Various surgical techniques have been reported in the literature for treatment of DFSP, but Mohs micrographic surgery (MMS) is still considered to be the ideal form. This technique, involves microscopic examination of each removed tissue for cancer cells with main difference from conventional surgical excision being the different pathological processing of the lesion but offering equal results to it with

minimal surgical morbidity²⁵. This, however, can be tedious and not available in all centers. With conventional surgery, aim should be to achieve Ro margins with a minimum of 2 cm circumferentially, where possible¹⁵. In order to reduce the risk of positive margins, fresh frozen specimen sectioning should be utilized where available. This is more important because of the finger like projections of DFSP in surrounding tissues. The choice of surgical procedure along with expertise, is usually dependent upon the size and location of tumour, where a DFSP of small size and in the head and neck region would be best dealt with MMS.

In a study by Snow et al., they reported a 7% risk of local recurrence at 5 years after MMS²⁶. They suggested that adequate local control be provided by a further repeat MMS if required. The efficacy of MMS and wide excision in achieving clear margins has been reported to be equal in various studies, however, Meguerditchian et al., have reported increased incidence of positive margins with conventional surgery^{27,28}. It is worth noting, that all the studies propose a long term follow up, usually longer than or up to 10 years, due to reported incidence of delayed local recurrence.

Radiotherapy

The role of radiotherapy is well established in the management of DFSP and has been used and described in various studies both in the neoadjuvant and adjuvant settings. Various studies have reported a lower risk of local recurrence with its use. In a systematic review by Chen et al., the authors proposed that adjuvant radiotherapy may be considered for all patients even with a negative margin to reduce the risk of local recurrence²⁹. This effect was more pronounced in cases of large or recurrent tumors where wider excision with negative margins would lead to significant surgical morbidity. Radiotherapy is also recommended in cases of positive margins excision wherein further surgical resection may not be possible or will lead to significant morbidity (e.g. Amputation), to reduce the risk of local recurrence. This decision regarding the use of radiotherapy should be undertaken in a specialized tumour board setting to weight the benefits against risks.

Medical Treatment

As DFSP shows over expression of platelet derived growth factor receptors (PDGFR's) due to genetic translocation between chromosomes 17 & 22, targeting the PDGFR's may be an effective treatment option for DFSP. Imatinib, the tyrosine kinase inhibitor has been used in the medical management of DFSP and has shown some early fair response, particularly in recurrent, inoperable and/ or metastatic DFSP cases^{30,31}.

Conclusion

DFSP is a rare soft tissue sarcoma with a high rate of local recurrence and usually mismanaged. Treatment should be offered in specialized referral units in a multidisciplinary

setting with surgery being the mainstay of treatment. Radiotherapy may be of benefit in the adjuvant setting to reduce the risk of local recurrence in negative margins cases and should be offered in locally advanced cases where surgical morbidity may outweigh benefit, and in inoperable cases for local control. The role of Imatinib continues to evolve as medical treatment, with some early fair results. Long term follow up is advised according to locally agreed protocols to monitor for any delayed local recurrence.

Conflict of interest: None.

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Figures

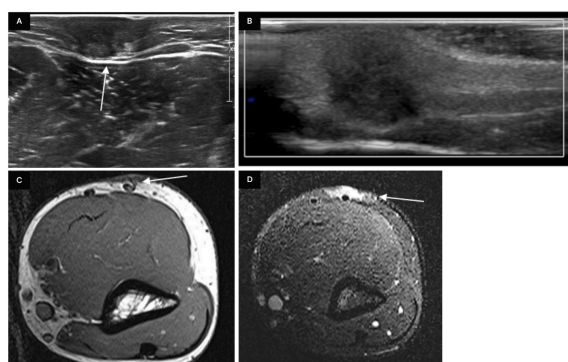


Figure 1A-1D: Ultrasound and MRI features of a subcutaneous DFSP on the volar aspect of antecubital fossa region.

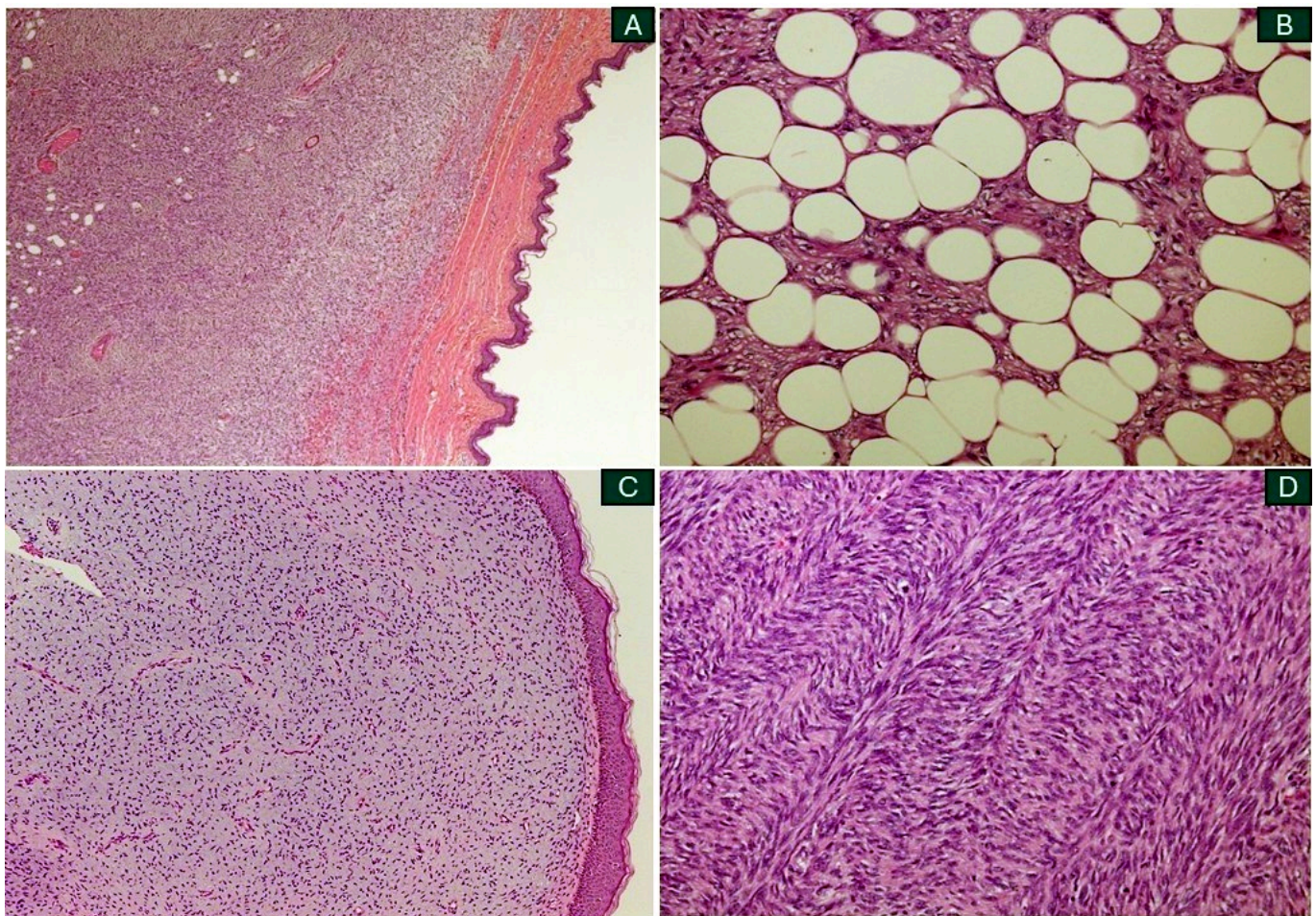


Figure 2:(A) Low-power view demonstrating a dermal-based spindle cell neoplasm with diffuse infiltration into the subcutaneous tissue and extension toward the overlying dermis. (B) Tumor cells infiltrating the subcutaneous adipose tissue in a characteristic honeycomb pattern. (C) Myxoid dermatofibrosarcoma protuberans (DFSP) showing hypocellular spindle cells embedded within abundant myxoid stroma. (D) Fibrosarcomatous transformation characterized by increased cellularity and intersecting fascicles arranged in a herringbone architecture.



Figure 3 (A-E): Third time recurrent DFSP on volar aspect of right forearm, with wide local excision followed by resection of defect with free Latissimus dorsi flap from contra lateral chest wall.