

## Original Article

# SURGICAL MANAGEMENT OF DERMATOFIBROSARCOMA PROTUBERANS

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**Background:** Dermatofibrosarcoma protuberans (DFSP) is a rare, low to intermediate grade cutaneous sarcoma, with recurrence rates as high as 60%.

**Material & Methods:** 11 patients were treated over a period of 5 years in Sheikh Zayed Hospital Lahore.

**Results:** The tumors presented as asymptomatic nodule in 63.6% cases. Average size of tumor at the time of presentation was 8.5x6.6cm. Wide Local Excision (more than 3 cm) was done to reduce the chances of recurrence. Reconstruction of the excised lesions was carried out in the same sitting after frozen section examination of the tumor and margin clearance. 10 patients (90.1%) were free from recurrence after a follow up of 24 months.

**Conclusion:** Dermatofibrosarcoma protuberans is a rare, indolent tumor that presents as a local problem. Wide excision of margins has a favorable impact on recurrence free life interval. Frozen section analyses, margin clearance and reconstruction should be carried out during one operation. Fibrosarcoma variant of DFSP may behave more aggressively and radiation therapy may have a role in controlling recurrence of this sub group of tumor. We advocate wide local excision as an effective measure to control local recurrence of the indolent tumor.

**Keywords:** Dermatofibrosarcoma protuberans; Wide Local Excision; Recurrence; Soft tissue

## Introduction

Dermatofibrosarcoma protuberans (DFSP) is a variant of soft tissue sarcoma of the skin that originates in the dermis and accounts for less than 1% of skin tumors.<sup>1</sup> It was first described by Darrier and Ferrand<sup>2</sup> in 1924 but currently accepted term was coined by Hoffman<sup>3</sup> when he reported three cases in 1925. The cellular origin of DFSP is not clear at this time. Evidence exists that supports the cellular origin being fibroblastic histiocytic or neuroectodermal. It is known for its locally aggressive growth and high rate of local recurrence.<sup>4</sup> DFSP is characterized by its progressive, locally infiltrative behavior, and if left untreated, these tumors continue to grow slowly, with invasion into surrounding tissues like fat, fascia, muscle and bone including neurovascular bundles.<sup>5</sup> The time interval from apparent onset to initial resection may exceed 10 years in more than 50% of cases.<sup>6</sup>

Primary therapy for DFSP is surgical excision. Inadequate resection of margins with conservative intent leads to very high recurrence, reported rates being 26-60%.<sup>4,5</sup> To improve local control after excision many authors have recommended wide excision with more than 2 cm grossly involved margin of skin and underlying deep fascia. In present paper we discuss our experience with

dermatofibrosarcoma protuberans along with literature review. Study also recommends wide excision as it effectively reduces the local recurrence.

## Material and Methods

The study was carried out at the Department of Plastic and Reconstructive Surgery, Shaikh Zayed Hospital, Lahore from January 2005 to December 2009. All the patients with diagnosis of DFSP on histological findings and confirmation with CD34 tumor marker were included in the study. Demographic data of the patients, site and size of the tumor, presenting features, time since noticing the lesion, tumor size, tumor recurrence, surgical findings, excision margins and mode of reconstruction were recorded. All the lesions were excised with gross resection margins of 3 cm and excision of one layer deep to the involved area. Frozen sections were sent for tumor clearance before reconstruction. The excised tissues were sent for frozen section margin clearance and later on for histological evaluation and immunohistochemical staining for CD34 tumor marker assessment. Radiotherapy was administered to those cases that had close clearance margins or patients with DFSP-FS variant of the tumor. All these cases were followed up every three months for first two years. They were advised follow up visits every six months for next two

years. Cases that needed radiotherapy were referred only after their suture lines had healed and flaps were stable and healthy. All the patients were regularly checked up for any recurrence at the follow up visits.

**Results:**

A total of 11 cases were treated during five years. The mean age of patients at the time of surgery was 37.7 years (range 17-53 years). The sex ratio was 5.5:1 (9 males: 2 females). The most common site of tumor was the trunk (n=5), followed by neck and extremities (3 cases). Sixty-three percent (63.6%) of the tumors presented as nodules. Two patients (18.2%) presented with limb movement and other two (18.2%) presented with the previously excised tumor. The tumors were mistakenly diagnosed as hypertrophic scars. The size of tumors ranged from 1.2x1.2x1.2 cm (average 8.7x6.6 cm). Four patients were referred after incomplete excision/ recurrence at other institutions. The average repeat development of lesion and seeking medical advice was 4½ years (range 1-10 years). Various reconstruction methods were used for wound coverage. Split skin grafting (Fig. 1), coverage with pedicle flap (Fig. 2), ray amputation in one case (9.1%) was used in one case. Two patients (Fig. 3) presented with a variant of the tumor (18.2%). Two patients had less aggressive variant (27.3%) received wide excision, 2 with DFSP-FS and 2 with DFSP-FS and shown involvement of muscle. The microscopic analysis of resected tumor mean follow up was 27 months (range 6-60 months). Ten patients were disease free whereas one patient, diagnosed as DFSP-FS, presented with tumor recurrence and lung metastasis within three months after primary excision in the subclavicular region. The patient died one month in the radiotherapy department.

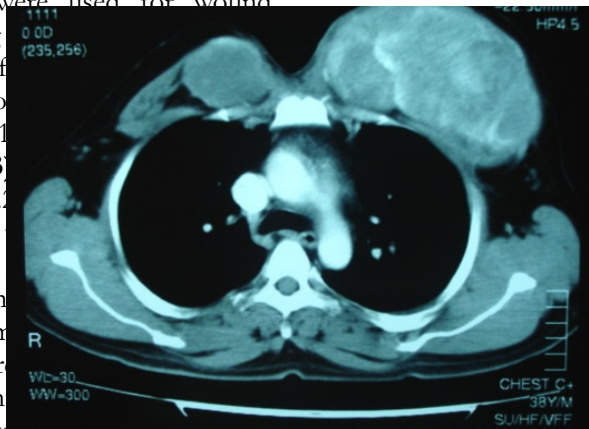
**Patient 1:**

A 35 years old male was referred with a painless multi-lobulated swelling 8x6 cm on left side of chest and progressively increasing in size over 5 years (Fig 1a). Incisional biopsy showed DFSP. Microscopic features: DFSP, fibrosarcoma protuberans. Microscopic features: DFSP, swelling without infiltration of muscle. Excision of tumor with wide resection of 3 cm of macroscopically clear margins

and excision of pectoralis major muscle (Fig 1b). The defect was covered with split skin graft after confirmation of margin clearance with frozen section (Fig 1c). Final fixed section showed 3 mm deep margin clearance. The patient was free of tumor at 40 months follow up.



**Figure 1a**



**Figure 1b**



**Figure 1c**

**Patient 2:**

A 45 years female presented with multiple non tender swellings in right inguinal region, measuring 10x6 cm, for the last 4 years (**Fig 2a**). Wide local excision with 3 cm margins and reconstruction with pedicle tensor fascia lata flap was done. The patient was tumor free after 30 months of follow up (**Fig 2b**).



**Figure 2a**



**Figure 2b**

**Patient 3:**

A 16 years old male presented with recurrent DFSP-FS on the nape of neck three months after primary excision at another hospital (**Fig 3a**). Wide Local Excision with 3 cm margins was done. Frozen section revealed involvement of superior margins. Re-excision with further 1cm margin was done (**Fig 3b**). The wound was covered with split thickness skin graft (**Fig 3c**). The patient was also given radiation therapy postoperatively. The patient was disease free for last 18 months and was under regular



**Figure 3a**



**Figure 3b**



**Figure 3c**

**Discussion**

Dermatofibrosarcoma protuberans represents less

**Table-1:** Sociodemographic and tumour related factors of studied patients

Age	Sex	Size of lesion	Site of lesion	Reconstruction	Histopathological Analysis
16	M	2×2cm	Nape of neck	Ststg	DESP:FS
28	M	12×6cm	R. Subscapular region	L.D mycutaneous flap	DESP
35	M	10×15cm & 8×10cm	Chest	Ststg	DESP
53	M	7×6cm	R. Infradavicular region	LD mycutaneous flap	DESP:FS
33	F	1.2×1.2cm	Right ring finger	Ray amputation	DESP
42	M	7×3cm	L. Gluteal region	L.D myocutaneous flap	DESP
45	F	10×6cm	L. Forehead	TFL myocutaneous flap	DESP
23	M	10×7cm	R. Submandibular region	Stsg	DESP
29	M	3×2cm	DESP:FS	Stsg	DESP
27	M	7×5cm	R. Thigh posteromedial aspect	Stsg	DESP
51	M	3×4cm	L. Subcostal region	Local transposition flap	DESP

region where 10-15% of these lesions originate.<sup>7</sup> The diagnosis of DFSP is confirmed after microscopic evaluation and CD34 tumor marker. The multifocality of the tumor in conjunction with its paucity of symptoms contributes to the difficulty in correct diagnosis on the basis of clinical findings alone. Although growth of the tumor is predominantly horizontal, deep facial, muscular and bone invasion can be seen with neglected, long-standing or locally recurrent lesion.<sup>8,9</sup> In our series, the mean time interval between the onset of the lesion and its presentation was 4 ½ years which corroborates to the observations by Alexander Stojadinovic.<sup>1</sup> Approximately sixty three percent patients in our study presented with asymptomatic nodules while similar results were observed by Hin-Lun Liu<sup>10</sup> who have mentioned 62.5% asymptomatic patients. The late presentation leads to larger size of tumor and therefore requires use of different modalities to cover the post-ablative soft tissue defects. In none of our cases primary closure was possible. This is in contrast to study by Hin-Lun Liu et al where the authors could achieve primary closure in 40.6% cases.<sup>10</sup> However, they reported one month time interval between the noticing of the growth and reporting for treatment in contrast to 4½ years (range 1-10 years) in present study. This very fact speaks of the importance of early diagnosis of the disease as it is going to dictate the tumor size and later reconstruction modality. This also demands of increased level of suspicion for this lesion to have an early diagnosis and treatment.

The most significant factor that predicts outcome

for DFSP is extent of resection. In a literature review of 317 patients who underwent conservative excision of DFSP, a mean local recurrence rate of 43% was reported.<sup>7</sup> A similar analysis of 489 patients with “wide” or larger than 2 cm resection margins demonstrated a decreased rate (43%) of local recurrence.<sup>7</sup> Incomplete initial resection can lead to uncontrolled, potentially fatal, locally invasive recurrence; therefore adequate initial resection is important because multiple local recurrences predispose to distant disease spread.<sup>11</sup> Overall 5-year survival rates are 93% to 100%.<sup>4</sup> In our series, 10 patients (90.9%) were alive after a mean follow up of 24 months. The only patient who died after recurrence and metastasis had the fibrosarcoma variant of the DFSP. Above all he had a margin free histopathological report. Seven of the patients had a tumor free interval of more than 36 months while the 3 had less than 3 years of follow up. The duration of follow up is important because 50-75% tumor recurrence is noted within 3 years after operation.<sup>4</sup>

Mohs micrographic surgery (MMS) may be a useful option for maximum conservation of uninvolved tissue. The documented local recurrence rate after MMS ranges from 0-7%.<sup>12</sup> This technique allows precise histological mapping of all lateral and deep margins that may not be captured by standard histological vertical step sectioning. This can lead to false-negative paraffin section interpretation.<sup>7,12</sup> Large and recurrent tumors may need exorbitant

margins.<sup>13</sup> Surgery requires experienced Mohs surgeons, histo-pathologists and laboratory technicians. This facility is not available in most institutions of the country. We have to rely on wide local excision, which has given comparable international results in our series.

## Conclusion

Dermatofibrosarcoma protuberans is a rare, indolent tumor that presents as a local problem. Wide excision of margins has a favorable impact on recurrence free life interval. Frozen section analyses,

margin clearance and reconstruction should be carried out during one operation. Fibrosarcoma variant of DFSP may behave more aggressively and radiation therapy may have a role in controlling recurrence of this sub group of tumor.

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