

Dermatofibrosarcoma protuberans: Role of wide local excision

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Abstract

Objectives: The main objective of the present study was to study the outcome of surgical treatment of dermatofibrosarcoma protuberans. **Materials and Methods:** This study included 45 patients both retrospective and prospective from December 1995 to December 2010. **Results:** Out of 45 patients, 30 were males and 15 females with the male to female ratio of 2:1. Mean age of presentation was 38.4 + 13.2 years. Commonest mode of presentation was raised firm multinodular lesion with fixity to overlying skin. Site distribution was 42.22% trunk, 57.88% extremities and head and neck. None of the patients had lymph node involvement. All patients underwent wide local excision. On histological examination, 8 patients had positive margins. Overall recurrence rate was 22.22%. (please clarify what is the difference between the rate of recurrence following surgery and the overall recurrence rate) Only 2 patients developed metastasis to lungs in the course of their follow-up. Out of 45 patients, 35 remained recurrence free over a varying period of 5 months to 13 years (mean 68 months). Ten patients developed one or more local recurrences. Average time from initial treatment to recurrence was 32 months. All patients with recurrent tumors were subjected to salvage treatment, i.e., re-excision. Average recurrence-free period was 36 + 44 months within a mean follow-up of 68 months. **Conclusion:** Because of the potential of local recurrence, therapy for DFSP should be directed toward adequate local excision of the primary lesion. Minimal resection should include a surrounding margin, comprising 3-cm margin of normal skin and removal of underlying deep fascia. Compromising on margins invites higher chances of local recurrence.

Key words: Dermatofibrosarcoma protuberans, local recurrence, resection margin, dermatofibrosarcoma protuberans

Introduction

Dermatofibrosarcoma protuberans (DFSP) is a clinico-pathological entity which the general surgeon may encounter infrequently but, with which it would be well to be acquainted with, since the ultimate success of surgical management of this lesion is often determined by the initial treatment.^[1] Earliest description of DFSP was by Taylor in 1890. It is a rare cutaneous tumor constituting < 0.1% of all malignancies. It occurs between 20 and 40 years of age.^[2] It has been reported to involve many body surfaces, mainly trunk, followed by extremities and less commonly the head and neck.^[3] The neoplasm appears spontaneously as a pea-sized cutaneous nodule. Subsequently, the tumor grows more rapidly into a boss-like or mushroom-like growth.^[4]

Multiple theories have been put forward for the origin of the DFSP. Darrier originally attributed it to an unknown parasite. Persistent irritation by a button of cloth is mentioned by Hoffman as a presumptive cause of tumor in different series. Some studies reported a history of trauma

in 13% cases. Hereditary influence was not found to be significant in the origin of DFSP.

Growth originates in dermis. It is grayish-yellow and firm in consistency.^[5] Tumor cells are mainly spindle in shape and arranged in compact bundles. It contains fibroblasts with prominent storiform pattern (cartwheel appearance).^[2] 85-90% of all DFSPs represent low-grade tumors.^[3] Despite their locally aggressive behavior, they rarely metastasize to regional lymph nodes or the viscera. Probability of regional and distant metastasis is < 5% postoperatively.^[6]

The recommended standard form of treatment for DFSP is wide local excision of tumor-bearing area, including the subjacent fascia and margin of apparently normal tissue in all planes.^[3] Most authorities would suggest a margin of 2-3 cm of normal tissue from the gross tumor boundary with a three-dimensional resection that includes skin, subcutaneous tissue, and the underlying investing fascia.^[2,4,8,14]

Adjuvant radiotherapy administered either before or after surgery may significantly reduce the risk of local recurrence in patients who have or who are likely to have close or positive margins. Sometimes, surgical management of DFSP can be difficult. Problems arise because this locally aggressive tumor is often misdiagnosed as a benign lesion.^[7] Even in metastatic tumors, local resection should be considered.^[8] The present study was designed to review the clinical experience and outcome of the surgical treatment of this tumor.

Materials and Methods

The present study was a retrospective and prospective study on all cases of DFSP treated in the Department of Plastic and Reconstructive Surgery, SKIMS, Srinagar,

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from December 1995 to December 2010. Retrospective period was from December 1995 to May 2006 and prospective study started from May 2006 to December 2010. Thirty five patients belonged to retrospective and ten patients belonged to prospective group. In the retrospective group, the case records of all patients diagnosed as DFSP who had been treated in the department were analyzed for relevant data. Patients in retrospective group were contacted on telephone and most of them were called for personal interview regarding any postsoperative problems, any recurrence, and information regarding any additional treatment received. Clinical information, surgical therapy, pathological details, and follow-up information were also reviewed from case files.

Results

The DFSP patients constituted 3.00% of all skin tumors admitted and treated in the Department of Plastic and Reconstructive Surgery during the study period [Table 1].

Males were found to be more commonly affected than females. There were 30 (66.66%) males and 15 (33.33%) females with a male to female ratio of 2:1. The patients in the present study were aged between 7 years and 65 years. Mean age of presentation was 38.4 ± 13.2 years. Likewise, peak incidence was observed in third decade of life (33.33%). There was only one patient below 10 years of age [Table 2].

Patients presented within an average duration of 29.3 ± 8.6 months after noting the lesion. Shortest and longest duration of presentations were 1 month and 12 years, respectively [Table 3].

In majority of patients, 27 (60.00%) primary lesions were between 5 and 10 cm in size. Five lesions were more than 10 cm in size [Table 4].

Commonest mode of presentation was a raised firm multinodular lesion (84.44%) with fixity to overlying skin seen in the majority of the cases [Figures 1 and 2]. Ulceration was associated with primary lesion in 8 (15.55%) cases, dull pain was seen in 4 (8.88%) cases, reddish-to-bluish discoloration of skin in 4 (8.88%)

patients, lesions were fixed to underlying structures in 3 (6.66%) patients, discharge was seen in 2 (4.44%) patients, and fungation in 2 (4.44%) patients [Table 5].

Majority of patients, 19 (42.22%) in the present series, had lesions confined over the trunk. Rest of the lesions were distributed as 16 (35.55%) on lower extremities [Figure 3],

Table 1: Proportion of DFSP cases to skin tumors

	No.
Total no. of patients with skin tumors	1500
Total no. of patients with dermatofibrosarcoma protuberans	45
Percentage of dermatofibrosarcoma protuberans to skin tumors	3.0

DFSP=Dermatofibrosarcoma protuberans

Table 2: Distribution of cases as per age

Age in years	No. of patients	Percentage
<10	01	2.22
11-20	04	8.89
21-30	15	33.34
31-40	09	20.00
41-50	09	20.00
51-60	04	8.89
61-70	03	6.66
Total	45	100

Table 3: Distribution of cases as per duration of presenting complaint

Duration	No. of patients	Percentage
<6 months	18	40.00
6-12 months	07	15.57
1-2 years	06	13.33
2-3 years	06	13.33
3-5 years	03	06.66
>5 years	05	11.12
Total	45	100.00

Table 4: Distribution of lesions as per size

Size	No.	Percentage
<5 cm	12	26.66
5-10 cm	27	60.00
>10 cm	06	13.33
Total	45	100.00

Table 5: Distribution of cases as per presentation and associated clinical features

Presentation of lesion*	No. of patients	Percentage
Protuberant nodules (multinodularity)	38	84.44
Fixity to overlying skin	27	60.00
Ulceration	08	17.77
Pain	04	08.88
Reddish to bluish discoloration of skin	04	08.88
Fixity to underlying structures	03	06.66
Discharge	02	04.44
Fungation	02	04.44

*More than one feature was present in many of the patients



Figure 1: DFSP lesion on upper chest

6 (13.33%) on upper extremities, 3 (6.66%) had lesions over head and neck, and 1 (2.22%) on external genitalia [Figure 4] [Table 6].

Reconstructive soft-tissue procedures included split thickness skin grafting and pedicled flaps in 37 (82.22%) and 3 (6.66%) patients respectively [Figures 5-7]. In 5 (11.11%) patients, wounds could be closed by primary approximation after mobilizing the wound margins [Figure 8, Table 7].

Width of the resection margin in most of the instances varied from 2-3 cm. Out of 45 patients, 2 (4.44%) patients in the course of follow-up developed lung metastasis [Table 8].

Of the 45 patients studied, 35 (77.77%) remained recurrence free over a period of 12 months to 14 years (median 68 months). 10 patients (22.23%) had developed one or more local recurrences [Figure 9], but there was no instance of regional metastasis to lymph nodes. Average time from initial treatment to recurrence was 32 months. All those patients with locally recurrent disease were subjected to salvage treatment, i.e., re-excision. Out of 10 patients with local recurrence, 7

had a single recurrence, 1 had two recurrences, and 2 had more than two recurrences. These 2 patients with more than two recurrences ultimately developed lung metastasis and succumbed to their disease. Of these

Table 6: Distribution of lesions as per anatomical locations

Anatomical location	No.	Percentage
Head and neck	03	06.66
Anterior trunk	12	26.67
Posterior trunk	07	15.56
Upper extremities	06	13.33
Lower extremities	16	35.56
External genitalia	01	02.22
Total	45	100.00

Table 7: Reconstructive surgical procedures

Procedure	No. of patients	Percentage
Wide local excision with split thickness skin grafting	37	82.22
Excision with pedicled flap	03	6.66
Excision with primary closure	05	11.11



Figure 2: DFSP lesion lower abdomen



Figure 3: DFSP lesion posterior aspect of right thigh



Figure 4: DFSP lesion on glans penis



Figure 5: Postoperative picture after excision of tumor in patient shown in Figure 1 [split thickness skin graft applied]



Figure 6: Postoperative picture after excision of tumor in patient shown in Figure 2 [split thickness skin graft applied]



Figure 7: Postoperative picture after excision of tumor in patient shown in Figure 3 [split thickness skin graft applied]



Figure 8: Postoperative picture after excision of tumor in patient shown in Figure 4 [defect closed primarily]



Figure 9: Recurrent DFSP lesion right lower abdomen and inguinal area

Table 8: Metastasis

Metastasis	No. of patients	Percentage
Patients without distant metastasis	43	95.55
Patients with lung metastasis	02	04.44
Total	45	100.00

10 patients, 8 patients had positive margin at the time of first excision.

Initial recurrence was observed within 28 months of surgical excision in 7 patients. In rest of the 3 patients, recurrence was observed after prolonged time period ranging from 3 to 8 years. In general, subsequent recurrences became obvious in shorter periods of time after treatment.

In the present series of cases, lesions were multinodular in 38 (84.44%) patients; rest of the lesions were of varying shape. Size ranged from 1 to 20 cm. Six patients who had larger size > 10 cm lesions had involvement of deeper structures. In the present study, none of the patients had lymph node involvement on cut section, the tumor had a grayish-yellow color and firm consistency.

Histopathology of the resected tumor revealed presence of spindle cells arranged in interwoven pattern and radiating from a focal point with whorl formation (cartwheel pattern) [Figure 9]. Tumor cells were mainly spindle in shape and arranged in compact bundles. Tumor was without a capsule in all the cases. Lateral margin was positive for tumor cells in eight instances (17.77%). Four of these patients were subjected to re-excision to achieve negative margin; in rest of the four patients re-excision could not be done because of proximity to vital structures. None of the patients had a fibrosarcomatous element in their specimens.

In most of the cases, the tumor had a uniform histological appearance with cartwheel pattern being the most consistent findings.

Discussion

There were 66.66% males and 33.33% females in the present series. This corresponds well with the sex distribution as reported by other authors.^[1,4,8,9]

In the present series, peak age of incidence was observed in third decade of life, the mean age of presentation being 38.4 + 13.2 years irrespective of sex. Only 11.11% patients

were below 20 years and 35.55% were above 40 years of age. Minimum age was 7 years and maximum 65 years.

Our observation fully corresponds with the observation of Taylor and Helwig, Brenner *et al.*, and Jambhekar and Chinoy.^[10-12] In about half of the patients in our series, age was between 20 and 40 years. This corresponds with the observation made by McGregor and McPeak *et al.*^[2,8]

In the present series, the duration of lesions at the time of first presentation was < 2 years in 68.88% cases. The minimum duration of illness was 1 month and maximum duration was 7 years. Median duration of swelling in our study was 7 months. This does not correspond well with the series reported by Taylor and Helwig in which about half the patient had duration of illness < 5 years and half had duration of illness > 5 years.^[10] Similarly, in a series by Burkhardt *et al.*, duration of illness was > 10 years in about 50% of the cases.^[13]

As far as the duration of illness is concerned, the observations made by above authors do not correspond with those of present series, but it is certain that the tumor is slow growing and the time of initial appearance and the time when patient seeks medical attention may range from months to years, depending upon the location of tumor, level of literacy, and availability of health care facilities. The decline in the duration of the illness in our series is probably due to freely available medical facility, increased literacy, and increase in the familiarity with the disease.

The total number of cases of DFSP patients that reported in the study period was 45. This constituted about 3% of all skin tumors admitted and treated in the Department of Plastic and Reconstructive Surgery during this time period. In our study, DFSP occurred at the rate of 3.46 cases per year. The number of cases per year in our study is more than that reported by authors like Burkhardt *et al.*, Roses *et al.*, and Jambhekar and Chinoy.^[3,11,14]

This apparent increase in the number of cases per year may be due to the fact that people are now better educated, more health conscious, and economically better placed than before, and hence, they try to seek or report for medical advice for this neoplasm more often than earlier times.

In the present series of cases, lesions were multinodular in 38 (84.44%) patients, rest of the lesions were of varying shape. Taylor and Helwig noticed in their series that most common presentation was raised firm multinodular tumor fixed to overlying skin.^[13] In a series of cases by Phelan and Juado, in all instances, tumor was nodular and bosselated and fixed to overlying skin. The preponderance of the nodular lesions noticed by various workers mentioned above is in perfect agreement with our findings. Majority of the lesions in the present series were free from the underlying structures McPeak *et al.*^[2] in a series of cases reported that in 8.13% of the cases of DFSP, underlying rib, clavicle, and other osseous structures were involved.^[2] Our observations correspond well with that of the McPeak *et al.*

In our series of 45 cases of DFSP, history of trauma was elicited in 5 (11.11%) patients. History of trauma prior to development of lesions was elicited in 20.5% cases in a series by Pack and Tabah.^[4] Taylor and Helwig observed prior history of trauma to the site of lesions in 16.5% of patients.^[10] Our findings correspond well with those of Pack and Tabah, but some authors mention no history of trauma in their case series like Brenner *et al.* and Jambhekar and Chinoy.^[11,12] Trauma has infrequently been incriminated as a possible factor influencing the growth of the tumor. But from the above-mentioned and present series, the relationship between trauma and tumor seems to be only coincidental. It could have served to attract the patient's attention to a previously existing mass. Though mentioned that trauma could have served as a trigger mechanism in stimulating the neoplasm to appear in a person already predisposed to its development, we conclude that injury as an etiological factor played no significant role in our series of cases.

In the present series only, 4 (08.88%) patients had history of pain. Pain was in most of the cases dull in type. In a series of 39 patients of DFSP by Pack and Tabah, the patients presented with painless swelling slowly increasing in size for a number of years. Pain and/or tenderness was reported in 28.57% of patients in a series by Burkhardt *et al.*^[13] Incidence of pain in our series is somewhat lower compared to that of Burkhardt *et al.*^[13] This may be because most of the patients now report earlier and are now more health conscious. Our observation of history of pain do not correspond with the observation of Pack and Tabah.^[4] This variation may be because in some instances, patients may experience discomfort by reason of location or size or because of some injury to the tumor.

In the present series, 39 (86.66%) cases of DFSP were less than or equal to 10 cm in maximum diameter. To some extent our findings are consistent with those of the Taylor and Helwig, Burkhardt *et al.*, and Jambhekar and Chinoy.^[10,12,13] We conclude that our study had lesions of variable size and shape ranging from 1 cm single nodular lesion to that of 20 cm lesion having multinodularity.

In the present study, none of the patients had lymph node involvement and this finding is concordant with that of Taylor and Helwig and Phelan and Juado who also did not report any lymph node metastasis. McPeak *et al.* in their study found that in no instance lymph node metastasis was found when groin and axillary dissection was performed in continuity with excision of primary tumor, i.e., clinically and grossly enlarged nodes had no evidence of metastasis on microscopy.^[2,10,15] This reinforces the concept that lymphatic spread does not generally occur in these tumors.

Trunk was the commonest site of DFSP in present series (42.22%), followed by lower extremities (35.55%), upper extremities (13.33%), and head and neck (6.66%). This observation is in agreement with the observation made by other authors, viz. Pack and Tabah, Taylor and Helwig, and Burkhardt *et al.*^[4,10,13]

In the present study, the majority of cases, 37 (82.22%) patients, underwent wide local excision and split thickness skin grafting (STSG) of raw area. Width of the resection margin in most of the instances varied from 2-3 cm. Five patients (11.11%) with smaller lesions did not require any reconstruction and were closed primarily after mobilization of edges. In a series of cases by Taylor and Helwig, the most frequently employed procedure was wide local excision after including the subjacent fascia and a margin of apparently normal tissue in all planes. Enucleation of tumor was performed on 30% of patients.^[10] McGregor in his series of 10 cases of DFSP performed radical surgical excision in most of the instances and observed if tumor is small, wide local excision, and primary closure without the aid of skin grafting is possible.^[8] Phelan and Juardo in his study pointed out that the microscopic tumor spreads from main body of the tumor and probably accounts for the high incidence of local recurrence and stressed the necessity for wide local excision of this tumor.^[15]

McPeak in their series of 86 cases of DFSP stressed that excision is inadequate unless a margin of at least 3 cm of normal appearing tissue and underlying deep fascia is included in the resection.^[2] Roses *et al.* presented a series of 50 cases of DFSP. Treatment in all the instances was excision with removal of margin of normal skin from the gross confines of the neoplasm or of the site of the biopsy to underlying fascia.^[14] Only 5 patients required skin grafts for complete closure. Majority of wound excisions were closed primarily.

In our series, majority of the lesions, 37 (82.22%) patients, were treated with excision and primary skin grafting in accordance with McGregor.^[8] The margin of excision in our series ranged from 2 to 3 cm in accordance with Pack and Tabah, McPeak, and Roses *et al.* We conclude that treatment of choice for DFSP is radical surgical excision, as recommended by other authors. If the tumor is small, wound can be generally closed primarily without skin grafting. Skin grafting should be used to close a bigger defect after radical excision of tumor.

In the present study, all the lesions were treated by wide local excision. Overall recurrence rate was 22.22% (10 patients out of 45) at local site. Recurrence rates as reported by Pack and Tabah and Taylor and Helwig were 20.5% and 49%, respectively.^[4,10] Bukhardt *et al.* reported recurrence rate of 33% in patients who had initial operation at their clinic.^[13] In a series of 27 patients of DFSP by McPeak *et al.*, recurrence rate was 11.11%. Roses *et al.* reported a recurrence rate of 32% (41% when resection margin was < 2 cm and 24% if resection margin exceeded 2 cm). Jambhekar and Chinoy reported an overall recurrence rate of 25%.^[12] Lindner *et al.* reported 2.5-3 cm resection margin improved local control of disease. We also recommend 2-3 cm or more resection margins wherever feasible to improve the local control of the disease.^[17]

Overall recurrence rate in our series corresponds well with that of Pack and Tabah and Jambhekar and Chinoy.^[4,12]

However, recurrence rate in our series was lower as compared with the recurrence rate reported by Taylor and Helwig, Bukhardt *et al.*, and Roses *et al.*, but it was higher when compared with the recurrence rate of McPeak *et al.*^[2,10,13,14] Rutgers *et al.* in a review study had a recurrence rate of 50% decreasing to 13% after adequate wide excision.^[18]

Patients undergoing wide excision with margins more than or equal to 3 cm were found to have lower recurrence rate compared with those margins 1.5-2 cm.^[19] Reason for lower recurrence rate in present series as compared to that reported by Taylor and Helwig may be because of wider margin of resection more than or equal to 2-3 cm in our patients and better facilities for reconstruction. Lesser margins often invites higher rates of recurrence as observed by Roses *et al.*^[10,14]

From the above findings, it is clear that the recurrence rate can be low or expected to fall if tumor is excised early and with adequate margins. Recurrence rate can also be reduced by referring such patients to a tertiary care hospital where the margin of the resection can be adequate irrespective of the anatomical site and where facilities for closure of the defect with skin grafting/flaps are available.

In the present study, none of the patients had metastasis either regional or metastatic on presentation. However, 2 patients of recurrent lesions developed lung metastasis in the course of their disease. Common features about these patient were that lesions were large > 10 cm, involving deeper structures, with positive postoperative margins and had recurred locally multiple times even after wide local excision and postoperative radiotherapy.

The recurrent lesions were associated with higher chances of more recurrence and distant metastasis. Out of three lesions on head and neck, 2 patients had recurrence because lesser resection margins were possible due to surrounding structures. In Farmas^[20] recent study of 204 patients of DFSP, two local recurrences were reported in head and neck, may be due to conservative approach in dealing with DFSP lesions in this area resulting in inadequate local control, as was revealed in our study. Most of the authors like McGregor, Roses *et al.*, and Khatri *et al.* observed in their respective case series that DFSP rarely metastasizes to regional or distant sites.^[8,14] McPeak and Jambhekar and Chinoy observed that histologically documented metastasis was reported in 5.81% and 6%, respectively, of all DFSP cases.^[17] Our findings are consistent with those of McGregor, McPeak *et al.*, and Khatri *et al.*^[2,8]

It is clear from the above discussion that the recurrence rate after DFSP excision varies widely in literature presumably depends on histological subtype, mitotic index, margin of resection, and multiple previous recurrences. Most important factor among these for local control is obtaining a negative surgical margin. Although radiotherapy may be used in patients with positive margins, there is little

or no benefit. Wide local excision with careful pathological analysis of margin appears to have very low recurrence and should be used as standard care for patients with DFSP. Confirmation of negative margins by frozen section, wherever possible should always be done before definitive wound closure is performed because it allows for directed re-excision should final pathology reveal a positive wound margin.

Conclusion

From the analysis of 45 patients of DFSP operated over a period from December 1995 to December 2010 in the Department of Plastic and Reconstructive Surgery SK Institute of Medical Sciences, following observations and conclusions were drawn.

These patients constituted 3.0% of all skin tumors patients, 30 (66.66%) were males and 15 (33.33%) were females with a male to female ratio of 2:1. Average age for patients with DFSP was 38.4 ± 8.25 years. The mean duration of disease at the time of first presentation was 29.3 ± 8.6 months. Commonest mode of presentation was a multinodular lesion (62.22%) with fixity to overlying skin. Trunk was the commonest site of DFSP 19 (42.22%), followed by lower extremities in 16 (35.55%), upper extremities in 6 (13.33%). Average size of lesion (DFSP) was 38.8 ± 48.3 cm². None of the patients of DFSP had regional lymph node involvement on presentation or on follow up. Most constant microscopic histopathological feature of lesions was cartwheel or whirling pattern of tumor cells. Overall local recurrence rate after treatment was 22.22%. Two patients (4.44%) of DFSP developed lung metastasis in the course of their follow up and succumbed to the disease. The average recurrence-free period was 63 ± 44 months within a mean follow-up of 68 months.

From the above observations, we conclude that DFSP behaves like a locally infiltrating neoplasm. Despite their locally aggressive behavior, distant metastasis occurs with extreme rarity. Because of this, it is reasonable that therapy should be directed towards adequate local excision of the primary lesion. Minimal resection should include a surrounding margin of at least 2-3 cm and removal of underlying deep fascia is essential. Adequate resection will need a skin graft/skin flap replacement in nearly every instance which is possible in a tertiary care setup only. Compromising on margins always invites higher chances of local recurrence.

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