The Surgical Management of Dermato Fibro Sarcoma Protuberance

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Abstracts: Background: Dermatofibrosarcoma protuberance is relatively uncommon low to intermediate grade malignant neoplasm with characteristic cytogenetic features. It constitutes less than 0.1% of all malignant neoplasms. Although metastasized rarely, it is locally aggressive tumour with high recurrence rate. It is a one of the rare type of law grade sarcomas that occur s anywhere in the body, usually arise from trunks and extremities. It rarely arises from abdominal wall. **Objectives**: The main objective of the present study was to study the outcome of surgical treatment of dermatofibrosarcoma protuberance. **Methods**: This study included 4 patients all retrospective and prospective from September 2008 to December 2013. They were subjected to detailed history taking and examination with relevant investigations and were subjected to surgery. They were followed up for at least 36-month period to assess for any recurrence. **Results and Interpretation**: Out of 4 patients, all were males. Mean age of presentation was 32 years. Site distribution was 25% trunk, 50% extremities and 25% neck. All patients underwent wide local excision. At the end of follow-up period of up to 3 years after surgery Overall recurrence rate was nil. 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. **Conclusion**: DFSP behaves like a locally infiltrating neoplasm. Despite their locally aggressive behaviour, 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. Compromising on margins always invites higher chances of local recurrence. [Bhadreshwara K NJIRM 2014; 5(4):98-101]

Key Words: Dermatofibrosarcoma protuberance, wide local excision, Recurrence.

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Introduction: Dermatofibrosarcoma protuberance (DFSP) is a clinico-pathological entity which the general surgeon may encounter infrequently but, with which it would be well acquainted with, since the ultimate success of surgical management of this lesion is often determined by the initial treatment. [2] Earliest description of DFSP was by Taylor in 1890. It is a rare cutaneous tumour constituting < 0.1% of all malignancies. It occurs between 20 and 40 years of age. [3] It has been reported to involve many body surfaces, mainly trunk, followed by extremities and less commonly the head and neck. [4] The neoplasm appears spontaneously as a pea-sized cutaneous nodule. Subsequently, the tumour grows more rapidly into a boss-like or mushroom-like growth. [5]

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 tumour 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 greyish-yellow and firm in consistency. [6] Tumour cells are mainly spindle in shape and arranged in compact bundles. It contains fibroblasts with prominent storiform pattern (cartwheel appearance). [3] 85-90% of all DFSPs represent low-grade tumours. 4 Despite their locally aggressive behaviour, they rarely metastasize to regional lymph nodes or the viscera. Probability of regional and distant metastasis is < 5% postoperatively. [7]

The recommended standard form of treatment for DFSP is wide local excision of tumour-bearing area, including the subjacent fascia and margin of apparently normal tissue in all planes. [4] Most authorities would suggest a margin of 2-3 cm of normal tissue from the gross tumour boundary with a three-dimensional resection that includes skin, subcutaneous tissue, and the underlying investing fascia. [3, 5, 9, 10]

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 tumour is often misdiagnosed as a benign lesion. [8] Even in metastatic tumours, local resection should be considered. [9] The present study was designed to review the clinical experience and outcome of the surgical treatment of this tumour.

Material and Methods: In present study 4 cases of Dermatofibrosarcoma protuberance has been studied at teaching institute in department of surgery. The present study was a retrospective and prospective study on all cases of DFSP from 2008 to December 2013. We describe study of 4 cases of Dermatofibrosarcoma protuberance treated with wide local excision and of which three were covered with primary split thickness skin grafting.

Result: Out of 4 patients, all were males and no females with the male to female ratio of 1:0. Amongst all, 2 patients were presented with recurrence.Mean age of presentation was 32 years. Commonest mode of presentation was raised firm multinodular lesion with fixity to overlying skin. Site distribution was 25% trunk, 50% extremities and 25% neck. None of the patients had lymph node involvement. All patients underwent wide local excision. In 3 patients wound was covered with primary split thickness skin grafting while in 1 patient wound primary skin suturing was done. On histological examination, no patients had positive margins. At the end of follow-up period of up to 3 years after surgery Overall recurrence rate was nil.

Figure 1 :
Recurrent Dermatofibrosarcoma Of Thigh

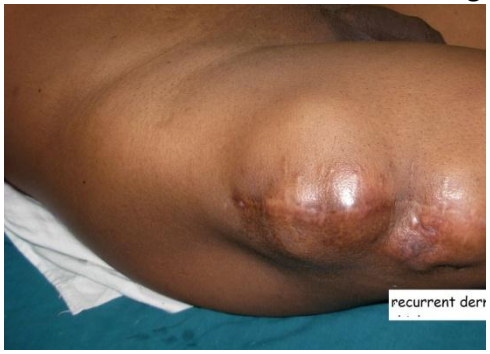

Figure 2: Dermatofibrosarcoma on Anterior Abdominal Wall- A Rare Site.

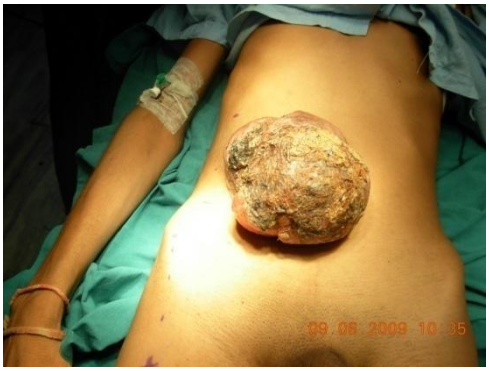

Table 1: Comparison of Age, Site, Mode of Presentation And Skin Coverage

CASE	AGE (YRS)	SITE	PRESENTATION	COVERAGE
1	40	RT.THIGH	RECURRENT	STG
2	30	RT.THIGH	PRIMARY	STG
3	28	NAPE OF NECK	RECURRENT	PRIMARY SUTURING
4	30	ANTERIOR ABDOMINAL WALL	PRIMARY	STG

Discussion: In the present series, peak age of incidence was observed in third decade of life, the mean age of presentation being 32 years. Our observation fully corresponds with the observation of Taylor and Helwig, Brenner *et al*., and Jambheker and Chinoy. [11, 12, 13]

In the present series, the duration of lesions at the time of first presentation was < 2 years in 75% cases. The minimum duration of illness was 18 month and maximum duration was 3 years. Median duration of swelling in our study was 19.5 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. [11] Similarly, in a series by Burkhardt *et al*., duration of illness was > 10 years in about 50% of the cases. [14] 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 tumour is slow growing and the time of initial appearance and the time when patient seeks medical attention may range from months to years.

Majority of the lesions in the present series were free from the underlying structures our observations correspond well with that of the McPeak *et al*.

We conclude that our study had lesions of variable size and shape ranging from 5 cm single nodular lesion to that of 18 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 Juardo who also did not report any lymph node metastasis. This reinforces the concept that lymphatic spread does not generally occur in these tumours.

Lower limb was the commonest site of DFSP in present series (50%), followed by trunk (25%), and neck (25%). This observation is not in agreement with the observation made by other authors, viz. Pack and Tabah, Taylor and Helwig, and Burkhardt *et al*. [5, 11, 14]

In the present study, the majority of cases, 3 (75%) 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. 1 patient (25%) 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. McGregor in his series of 10 cases of DFSP performed radical surgical excision in most of the instances and observed if tumour is small, wide local excision, and primary closure without the aid of skin grafting is possible. [9] Phelan and Juardo in his study pointed out that the microscopic tumour spreads from main body of the tumour and probably accounts for the high incidence of local recurrence and stressed the necessity for wide local excision of this tumour. [15]
In our series, majority of the lesions, 3 (75%) patients, were treated with excision and primary skin grafting in accordance with McGregor. [9] 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 tumour 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 tumour.

In the present study, all the lesions were treated by wide local excision. Overall recurrence rate was nil at local site. Recurrence rates as reported by Pack and Tabah and Taylor and Helwig were 20.5% and 49%, respectively. [5, 11] 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. [16]
Rutgers *et al*. in a review study had a recurrence rate of 50% decreasing to 13% after adequate wide excision. [17]

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. [18] 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. Lesser margins often invites higher rates of recurrence as observed by Roses *et al*. [11, 15]

From the above findings, it is clear that the recurrence rate can be low or expected to fall if tumour 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.

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.

Conclusion: From the above observations, we conclude that DFSP behaves like a locally infiltrating neoplasm. Despite their locally aggressive behaviour, 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. Compromising on margins always invites higher chances of local recurrence.

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