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Original research article

Recurrent Dermatofibrosarcoma Protuberans (DFSP) Of Chest Wall: Analysis Of 10 Cases

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ABSTRACT

Aim: Dermatofibrosarcoma Protuberans (DFSP) is a rare locally aggressive spindle cell soft tissue neoplasm with high rate of local recurrence and rare distant site metastasis. Our study aimed to analyze the pattern of recurrence, best possible surgical treatment and factors preventing its recurrence.

Methods: We studied prospectively 10 patients of recurrent DFSP attending to Gujarat Cancer Research Institute & M.P. Shah Regional Cancer Hospital, Ahmedabad, India from January to December 2015. All patients evaluated clinically, pathologically and radiologically. We also analysed oncologic and aesthetic outcome after a minimum follow up period of 12 months.

Results: 8 males and 2 females (M:F=4:1), with average age of 44.8 years (range 16-75), presented with recurrent chest wall DFSP - 06 patients with 1st recurrence; 02 patients with 2nd recurrence; 01 patient with 3rd recurrence and 01 patient was not actually recurrent rather with positive previously excised mass margin. The average duration of recurrence was 14.6 months (range 6-36 months). The average size of the tumour was 58.2cm2 (range 5×3-15×12 cm2) located over superior presternal (05 cases), parasternal (03 cases), axilla (01 case) and breast (01 case). 01 patient had lung metastasis since the previous surgery for which he took Imatinib Mesylatedefaultly . All 10 patients underwent wide local excision of mass including scars with a minimum margin of 3 cm in all directions. 08 patients reconstructed with local myocutaneous flap. Based on histopathology, 01 patient required postoperative adjuvant radiotherapy. After minimum follow up of 12 months all are oncologically and aesthetically well.

Conclusion: Dermatofibrosarcoma Protuberans is a locally aggressive neoplasm notorious for recurrence. Accurate diagnosis and wide local excision with at least 3 cm margin with reconstruction at the time of first surgery as well as proper histopathologically directed adjuvant radiotherapy or chemotherapy can prevent recurrence.

Keywords: Dermatofibrosarcoma Protuberans (DFSP), Imatinib Mesylate

Introduction

Dematofibrosarcoma protuberans (DFSP) accounts for approximately 6% of all soft tissue sarcomas and it typically presents as a small superficial soft tissue mass.[1] Due to its infiltrating growth pattern, the tumor tends to extend far beyond the clinical margin. This explains the high recurrence rate of 20-15% after excision.[2] Ultrasonography is considered an additional valuable method of monitoring this disease.[3,4]

The most frequent presenting location of DFSP is the skin of the trunk (50-60%).[3] DFSP is a relatively uncommon neoplasm of the deep dermis and subcutaneous tissue with low-grade malignant potential. DFSP is a locally aggressive tumor with a high recurrence rate. Because of its infiltrating growth pattern, the tumor tends to extend far beyond the clinical margin. This explains the high recurrence rate of 15-20% after excision. Most recurrences of DFSP are detected within 3 years of primary excision.[2] The histologic subtype, a high mitotic index, the cellularity and size, a location on the head and neck, and recurrent lesions are factors that are reportedly associated with higher recurrence rates. When the surgical margins are inadequate or conservative, the recurrence rates increase.[2,5]

DFSP usually occurs in young to middle-aged patients but can present in all age groups.[6] It is commonly found on the trunk, however, it can also develop in the extremities, head or neck. DFSP demonstrates local infiltrative growth but seldom metastasizes distally.[7] DFSP is divided histopathologically into classical and non-classical types.[8] Classical-type DFSP typically forms a radial or storiform pattern, with the cancer tissue extending into the subcutaneous fat and forming a honeycomb-like structure.[9] Atypical DFSP comprises at least 10 subtypes, of which the most common include pigment type, mucus type, and sarcoma type.[7] Fibrosarcomatous DFSP (FS-DFSP) is also an atypical DFSP subtype, with high rates of recurrence and metastasis.

The main problem of DFSP is not metastasis, which rarely occurs, but local aggression with a high recurrence rate. Surgery is the preferred treatment of DFSP; three-dimensional histology surgery is a new concept. During the procedure, a narrow lateral strip (1–1.5 cm wide) is excised around the perimeter of the tumor border and then a horizontal slice is excised from the bottom of the tumor. Both the strip and the slice are sent for routine pathologic examination. The procedure will not stop until all surgical margins are tumor-negative.

Our study aimed to analyze the pattern of recurrence, best possible surgical treatment and factors preventing its recurrence. **Materials And Methods**

It was a prospective study of 10 cases of recurrent chest wall tumour presented at Gujarat Cancer Research Institute & M.P shah cancer hospital, Ahmedabad from January 2015 to December2015.

All patients were evaluated clinically, radiologically and pathologically. Already diagnosed cases were reviewed. Detailed history regarding previous surgeries, number and duration gap of

recurrence as well as any previous adjuvant treatment were taken. One patient had lung metastasis at previous surgery for which he took imatinib default. Wide local excision with a grossly 3 cm tumour free margin all around was done and flaps were used to cover the defect.

Immunohistochemical staining was diffusely positive for CD34 and negative for CD117. Mib-1 (Ki-67) index was approximately 15- 20%. Based on postoperative histopathology, patients were judged for the need of adjuvant treatment. We analyzed the pattern of recurrence best possible surgical treatment as well as aesthetic outcome after a minimum follow up of 12 months.

Statistical Analysis

All the data were analyzed using SPSS package (Stata, version 26.0 SPSS INC, Chicago, IL, USA) for windows. The data were presented as descriptive statistics for continuous variables and percentage for categorical variables and was subjected Chi-square test, t test & Anova test. Other values were represented in number, proportions (%) and mean ± SD.

Results

Table 1: Gender Distribution

Gender	**N %**
Male	8 (80%)
Female	2 (20%)

Out of 10 patients of recurrent DFSP, 08 were males and 02 females with an average age of 44.8 years (range 16-75).

Table 2: Recurrence

Recurrence	**N %**
1st recurrence	6 (60%)
2nd recurrence	2 (2%)
3rd recurrence	1 (1%)
Positive previously excised margin	1 (1%)

06 patients presented with 1st recurrence, 02 patients with 2nd recurrence, 01 patient with 3rd recurrence and 01 patient was not actually recurrent rather with positive previously excised margin. The average duration gap for development of recurrence was 14.6 months (range 6-36).

Table 3: Size of lesion in cm^2

Size of lesion	**cm2**
Minimum	15
Maximum	180
Average	58.2

Size and locations of the lesions were variable with average size was 58.2cm^2 (range 5×315×12cm^2).

Table 4: Location of tumour

Location of tumour	N %
Upper presternal	5 (50%)
Presternal	3 (30%)
Axilla	1 (10%)
Breast	1 (10%)

In the present study, tumour was located at upper presternal in 05 cases, at parasternal in 03 cases, at axilla in 01 case and in the breast in 01 case.

Table 5: Postop pathology

Postop pathology	N %
R0 resection	9 (90%)
R1 resection	1 (10%)

Final histopathology showed R0 resection in 09 cases while R1 resection in 01 patient who then sent for adjuvant radiotherapy.

Discussion

It was first described in 1924 by Darier and Ferrand as a distinct cutaneous disease entity called progressive and recurring dermatofibroma.[10,11] Hoffman termed it DFSP in 1925. DFSP is slightly more common in males and is seen most often in adult life.

The initial lesion of DFSP can be misdiagnosed for keloid due to the similar appearance, therefore prompt diagnosis is vital for the treatment of this malignant tumour. This fact, coupled with the rarity of DFSP and diagnostic delay, often leads to inadequate initial resection. In our study, though documentation not available, only resection would have been done in the absence of prompt diagnosis that lead to recurrence.

Bowne WB et al reported the risk of a local recurrence is as high as 50 percent with simple excision, and it is even higher if the margins are positive.[12] For the 34 patients who developed a recurrence after surgical resection (21%), the median time to local recurrence was 32 months. In this study, the mean time of recurrence is 14.6 months which may be described due to previous surgical positive margins as per this study.

Brabant B et al showed that DFSP has characteristically high and varying recurrence rate ranging 10-80%.[13] Among the various treatment modalities, surgery is the mainstay of treatment by wide excision with a safety margin of 3 cm, including the underlying fascia.

Roses et al[14] studied 50 consecutive patients of DFSP patients and concluded that 2 cm or lesser than 2 cm are associated with local recurrence rate of 41% and surgical margins more than 2 cm are associated with local recurrence rate of 24%. Five year recurrence free survival at surgical margins at <1cm,≥=1cm,≥=2cm and ≥=3cm are 59%, 66%, 70%, and 80% respectively.

Rutger et al[15] described the overall recurrence rate is about 50%; after adequate wide excision, 13%. Recurrent tumour is safely treated by wide re- excision and the recurrence rate is then 12%.

Stivala A et al[16] reported that radiation therapy (RT) may be recommended if adequate wide excision may result in major cosmetic defect or if the tumour margins are positive. Postoperative adjuvant RT may reduce the risk of recurrence when clear surgical margins are equivocal.

In our study, similar to the literatures, all the lesions were treated by wide local excision with minimum margin of 3 cm. and no recurrence is noted in any case at least after 12 months which is supported by these studies. One patient with R1 resection advised for adjuvant radiotherapy.

Regional and distant recurrences are infrequent. In Rutger et al study 1% was reported to have regional lymph node metastases and 4% distant metastases, principally in the lung. The prognosis after appearance of regional or distant recurrence is bad. The role of radiotherapy in the management of this tumour is unclear. Elective lymph node dissection is not advised. Imatinib mesylate has been approved by the FDA and used in the adjuvant treatment in patients with unresectable, recurrent, or metastatic DFSP with objective response rate approaching 50%.[17]

One patient in our study had lung metastasis since previous resection and took Imatinib defaultly. He again prescribed Imatinib after local curative resection but there was no reduction in the size of metastasic lesion after 10 months post surgery and remains static. No elective lymph node dissection done in any case in this study. This is also in accordance with these literatures.

Mansour KA et al[18] described massive chest wall resection with immediate reconstruction has been shown to be safe and aesthetically effective. We also did the immediate reconstruction after resection by PMMC flap and aesthetically it was good in most cases.

Conclusions

DFSP is rare locally aggressive neoplasm notorious for recurrence. Correct diagnosis is vital in management. Wide local excision with at least 3 cm margin all around with reconstruction at the time of initial surgery and proper histopathologically directed adjuvant treatment are essentially preventive for recurrence.

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