Outcome of wide local excision in dermatofibrosarcoma protuberans and use of radiotherapy for margin-positive disease

Raashid Hamid, Adil Hafeez, Mohd A. Darzi, Inaam Zaroo, Altaf Rasool, Haroon Rashid

ABSTRACT

Purpose: Wide local excision (WLE) is the preferred treatment of dermatofibrosarcoma protuberans (DFSP). The aim is to achieve negative margins. We followed the impact of radiotherapy used postoperatively for both margin-negative and margin-positive DFSP tumors. Materials and Methods: Outcome of treatment of 36 patients of DFSP treated at our hospital was assessed. Thirty patients received radiotherapy postoperatively and six patients received radiotherapy alone. The maximum dimension of the lesion was 15 cm². Patients were followed up for varying periods of time for any recurrence. Results: 10-year actuarial local control rate was determined. Local control was realized in six patients who were treated with radiotherapy alone. 30 patients were treated by radiotherapy and surgery. Out of these 30 patients, there were 6 local failures (failure rate 10%). Actuarial control rate was 82%. The failures were among patients who had positive margins. Conclusion: Radiotherapy is effective, and it decreases the recurrence rate in the treatment of DFSP. It is especially helpful in margin-positive disease. This appears true for patients treated with radiotherapy alone or radiotherapy used postoperatively.

Key words: Dermatofibrosarcoma protuberans, local recurrence, radiotherapy, wide local excision

INTRODUCTION

Dermatofibrosarcoma protuberans (DFSP) is a locally aggressive but slow-growing tumor of dermis. It was first described by Darrier and Ferrand histopathologically in 1924. But the name was proposed in 1925 by Hoffman. Local growth of DFSP following surgical resection occurs in 20.5% as reported by Pack and Tabah (1951) and 49% Taylar and Helwing (1962). Bukhardt, et al. (1906) reported recurrence rate of 33% in patients who had initial surgery at their clinic.[1-3] This failure rate is reduced to approximately 13% following resection with wide margins. Successful treatment outcome depends almost entirely on the achievement of local control, as regional and distant metastasis is rare. We assessed the result of 36 patients of DFSP managed by radiation alone or combined with surgery at SKIMS (Sher-i-kashmir Institute of Medical Sciences), Srinagar, Jammu and Kashmir (India).

MATERIALS AND METHODS

This was a retrospective and prospective study of 36 cases of DFSP admitted and treated at the Departments of Plastic Surgery and Radiotherapy, SKIMS, Srinagar, between December 2000 and December 2010. Records of all patients diagnosed as DFSP and treated in the department were analyzed. For the prospective arm, detailed history-taking, physical and local examination was done.

Pathological diagnosis

Fine needle aspiration cytology and incisional biopsy was done of all lesions. In surgical patients, the resected lesion was subjected to histopathological examination and diagnosis confirmed [Figure 1].

Treatment methods

Surgical resection with 2.5-3 cm healthy margin was done in 30 patients with the aim of achieving a negative margin [Figures 2-4]. All patients were subjected to radiotherapy postoperatively. The other six patients (three with recurrent lesions, one with lesion close to the eye, and two with lesions <4 cm) were treated with radiotherapy alone. All patients received external beam radiation therapy with total dose ranging from 65 to 70 Gy.
Follow-up of patients
All the patients were followed up for any systemic or local recurrence. Follow-up interval was six monthly for first five years and then yearly for the next five years.

Statistical analysis
The Kaplan–Meier product limit method was used to estimate actuarial local control probabilities.

RESULTS

Baseline data
Thirty patients were treated with surgery and radiotherapy, six patients were treated with radiotherapy alone. Three patients had recurrent lesions, one had a lesion close to the eye, and two had smaller lesion <4 cm. The anatomic sites of the lesions are detailed in Table 1. Age of patients ranged from 7 years to 65 years. The mean age of patients was 38.4 ± 13.7 years. Peak incidence was in the third decade of life [Table 2]. The ratio of males to females was 5:2. In the majority of patients,
Table 1: Anatomical distribution of lesions

| Anatomical location       | n |
|---------------------------|---|
| Head and neck             | 03 |
| Anterior trunk            | 12 |
| Posterior trunk           | 08 |
| Upper extremities         | 06 |
| Lower extremities         | 06 |
| External genitalia        | 01 |
| Total                     | 36 |

Table 2: Demographic features of patients

| Age group in years | No. of male patients | No. of female patients | N |
|--------------------|----------------------|------------------------|---|
| <15                | 01                   | 0                      | 01 |
| 16-30              | 03                   | 02                     | 05 |
| 31-45              | 12                   | 04                     | 16 |
| 46-60              | 09                   | 03                     | 12 |
| >60                | 01                   | 01                     | 02 |

Table 3: Size of lesions

| Size (cm) | n |
|-----------|---|
| <05       | 6 |
| 5-10      | 21 |
| >10       | 09 |
| Total     | 36 |

The size of tumor ranged from 2 to 20 cm [Table 3]. The greatest dimension was <5 cm in six patients. Three patients who had recurrence after initial surgery elsewhere were treated with radiotherapy at our hospital [Figures 5-8].

DISCUSSION

In this study, 30 patients of DFSP were treated by WLE followed by radiotherapy. Six patients were treated by radiotherapy alone. The overall recurrence rate was 19.44% (7 of 36 patients). Recurrence rate among patients who received both surgery and radiotherapy was 20%, and recurrence rate in patients who received radiotherapy as the sole form of treatment was 33.33%. Recurrence rate as reported by Pack Tabah (1951) and Taylor and Holwing (1962) was 20.5% and 49%, respectively. Burkhardt BR. (1966) reported recurrence rate of 33% among DFSP patients who had initial...
surgery at their clinic.[3] In a series of 27 patients of DFSP by McPeak, et al. (1969), recurrence rate was 11.11%. [5] Roses, et al. (1986) reported a recurrence rate of 32% (41% when resected margin was <2 cm and 24% when the resection margin exceeded 2 cm). [6]

The overall recurrence rate in our series is lower when compared to the recurrence rate reported by Taylor and Helwig (1962), Burkhartd et al. (1966), and Roses et al. (1986), but higher when compared to the recurrence rate of McPeak et al. (1967). [2,3,5,6] The reason for lower recurrence rate in our study when compared with that reported by Taylor and Helwig may be because of wide margin of resection 2-3 cm in our patients and better facilities for reconstruction. Lesser margins invite a higher rate of recurrence, as observed by Roses et al. (1986). [6]

From the above findings, it is clear that the recurrence rate can be reduced if tumor is excised early and resected by WLE. Referring patients to a tertiary care centre where WLE irrespective of anatomical site could be offered along with closure of the defect with skin grafting/flap can also diminish recurrence rates.

There are few references of the role about radiotherapy in the management of DFSP. At Ontario Cancer Institute, 19 patients of DFSP were managed by surgery and radiation as described by O’Sullivan et al. (1995). [7] Margins were positive in 18 patients. There were two local failures. These patients underwent salvage surgery.

Radiotherapy in the dose of 50–60 Gy is effective in preventing local regrowth of DFSP following margin-positive resection. Zeong Mig Chang et al. published an article in 2004 (Chiv Journal of Radiation oncology) [8] on the role of radiotherapy in treatment of DFSP. They concluded that postoperative radiotherapy can decrease the local recurrence rate in DFSP. In our study also the use of postoperative radiotherapy has demonstrated a decrease in the local recurrence rate of DFSP. We recommend WLE of the skin tumor followed by radiotherapy in patients with margin-positive disease.

REFERENCES

1. Pack GT, Tabah EJ. Dermato-fibrosarcoma protuberans: A report of thirty nine cases. AMA Arch Surg 1951;62:391-411.
2. Taylor HB, Helwig EB. Dermatofibrosarcoma Protuberans: A study of 115 cases. Cancer 1962;15:717-25.
3. Burkhartd BR, Soule EH, Winkelmann RK, Ivin JC. Dermatofibrosarcoma protuberans: Study of fifty six cases. Am J Surg 1966;11:638-43.
4. Eisen RN, Tallini G. Metastatic dermatofibrosarcoma protuberans with fibrosarcomatous change in the absence of local recurrence: A case report of simultaneous occurrence with a malignant giant cell tumor of soft parts. Cancer 1993;15:462-8.
5. McPeak CJ, Cruz T, Nicastri AD. Dermatofibrosarcoma Protuberans: An analysis of 86 cases – Five with metastasis. Ann Surg 1967;100:303-16.
6. Roses DF, Valensi Q, LaTrenta G, Harris MN. Surgical treatment of Dermatofibrosarcoma Protuberans. Surg Gynecol Obstet 1986;162:449-52.
7. O’Sullivan B, Catton C, Bell R, Fornasier V, Cummings B, Hao D, et al. Treatment out come in dermatofibrosarcoma protuberans referred to aradiation oncology practice. Int J Radiat Oncol Biol Phys 1995;289:s1.
8. Sun LM, Wang CJ, Huang CC, Leung SW, Chen HC, Fang FM, et al. Dermatofibrosarcoma protuberans: Treatment results of 35 cases. Radiother Oncol 2000;57:175-81.