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ORIGINAL ARTICLE

Dermatofibrosarcoma protuberans: A 10-year experience

Tzu-Ching Chan, Cheng-Jung Wu, Seng-Feng Jeng*

Department of Plastic Surgery, E-Da Hospital, Kaohsiung, Taiwan

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KEYWORDS
dermatofibrosarcoma protuberans (DFSP);
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Summary *Background:* Providing soft-tissue coverage after wide excision of dermatofibrosarcoma protuberans (DFSP) is always challenging; according to the literature, a skin graft is often chosen as the first option. However, possible suboptimal functional and cosmetic results have been noted.
Aims and objectives: We present our 10-year experience using pedicled or free flaps for reconstruction after a wide excision of DFSP to provide adequate soft-tissue augmentation and enhanced esthetic results.
Materials and methods: This was a retrospective study comprising 14 DFSP patients who were treated between February 2003 and December 2013. All patients underwent a wide excision with a 3-cm safe margin with immediate reconstruction, and a negative deep margin confirmed with a frozen section. The reconstruction method included nine pedicled perforator flaps and five free perforator flaps. All patients received adjuvant radiotherapy after surgery.
Results: The peak incidence that occurred in this series was the highest in patients younger than 30 years. None of the 14 patients exhibited recurrence, and the mean follow-up time was 30 months. Half of the patients exhibited DFSP distributed over the trunk; the patients in the series were predominantly male. A total of 13 flaps were successful except for one pedicled flap that failed from venous congestion; we used it in a salvage procedure by using full-thickness skin graft for coverage.
Conclusion: A wide excision with a 3-cm margin of safety with immediate reconstruction is a reliable method for DFSP resection. Initiating adjuvant radiotherapy might reduce the chance of local recurrence. To minimize the complications of skin graft, pedicled, or free flaps provide superior functional outcome, soft-tissue augmentation, and esthetic results.

Conflicts of interest: The authors have no conflicts of interest to declare.

* Corresponding author. Department of Plastic Surgery, E-Da Hospital, Number 1, Yi-Da Road, Jiao-Su Village, Yan-Chao District, Kaohsiung City 824, Taiwan.
E-mail address: jengfamily@hotmail.com (S.-F. Jeng).

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1. Introduction

Dermatofibrosarcoma protuberans (DFSP) is a rare dermal mesenchymal tumor first described by Taylor in 1890; it accounts for $< 0.1\%$ of all cutaneous tumors with an annual incidence of approximately 4.2 cases per million per year in the United States.[1] Although DFSP can be found in all parts of the body, the trunk is the most common site (up to 50% of all cases); it is most common in middle-aged males. DFSP is locally aggressive and rarely metastasizes distally or to regional lymph nodes postoperatively. The pathogenesis remains unclear, and no hereditary influence has been identified. According to the guidelines of The National Comprehensive Cancer Network, the standard treatment involves a wide excision with a safety margin to adjacent fascia. Local recurrence remains challenging; adjuvant radiotherapy can be administered after a wide excision to reduce the risk of local recurrence for patients with close or positive margins. An oral tyrosine kinase inhibitor, imatinib, has been approved for inoperable tumors, local recurrences, and metastatic diseases. Skin graft has been widely used after oncologic resections in most studies, but it is associated with complications such as scar contracture, graft ulceration or loss after radiation,[2] movement restriction, and poor esthetic outcomes.

In this paper, we present a series of 14 DFSP patients who underwent a wide excision and immediate reconstruction with a perforator flap for providing excellent soft tissue coverage as well as functional and esthetic outcomes.

2. Materials and methods

In a retrospective study between February 2003 and December 2013, 14 patients were treated in E-da Hospital Department of Plastic Surgery. All patients underwent a wide excision with a 3-cm deep safe margin to investing fascia followed by immediate reconstruction. A frozen section of deep margin was sent to pathologist immediately after excision of the tumor to confirm a free margin. The reconstruction methods included pedicled and free perforator flaps, depending on the specific location and defect size; all patients received adjuvant radiotherapy 4 weeks after wound healing. All patients received a total radiation dose ranging from 45 Gy to 50 Gy after the wound healed during a 5-week period, as suggested by a radiologist. All patient data, clinical information, surgical therapies, pathology results, and follow-up information were reviewed from case files.

3. Results

The mean age of 14 patients was 41.29 ± 14.01 years (range 18—62 years), and males were more dominant than females at a ratio of 9:5. The peak incidence occurred at approximately the age of 30 years in five patients in this series. Seven patients exhibited DFSP in the trunk, four patients in the head and neck, and three patients in the lower and upper extremities (see Table 1).

Because of the possible unfavorable esthetic results and movement restrictions after scar contracture resulting from suboptimal outcomes, we did not consider skin graft as an option of reconstruction.[3] Immediate reconstruction with a pedicled perforator flap was the first option. If no suitable regional flap is available, a free anterolateral thigh (ALT) perforator flap is a versatile option to provide adequate soft-tissue augmentation and re-create anatomical structures after excision. We performed nine pedicled flaps and five free flaps for soft-tissue defect coverage.

No recurrence of the tumor occurred after surgery in our study, regardless of whether the patients exhibited a primary tumor, local recurrence, or inadequate excision. The mean follow-up time was 30 months (range 18—68 months).

Two postoperative complications occurred in 14 patients. Patient 5 suffered from wound dehiscence after radiotherapy but healed completely through secondary intension. The propeller perforator flap in Patient 9 exhibited a total loss because of venous congestion, and a salvage full-thickness skin graft from the congested flap was performed to cover the defect. An ALT flap was used in six cases; four were free flaps, and two were pedicled ALT flaps. Primary closure of the donor site was achieved in four patients; the other two required skin graft.[4] All the pedicled perforator flaps of the donor site achieved primary closure except in Patient 9, for which skin graft was used to cover the donor site. No debulking or revision procedures were necessary for patients with pedicled or free flaps; patients were satisfied with the surgical and esthetic outcomes.

4. Case reports

4.1. Patient 4

A 55-year-old female patient exhibited a 3 cm $\times$ 2 cm mass over her left lower leg for 3 years. An incisional biopsy revealed that it was DFSP. After a wide excision of the tumor with a 3-cm margin, the frozen section appeared margin free. A pedicled medial sural perforator flap (9 cm $\times$ 8 cm) was elevated to cover the defect, and the donor site was closed through skin graft. She received postoperative radiotherapy 4 weeks later. No local recurrence was detected during a 27-month follow-up period. The cosmetic result was acceptable (see Fig. 1).

4.2. Patient 12

A 42-year-old female patient exhibited a scalp tumor for 4 years. She underwent excision at an outside clinic and pathology proved it was DFSP with inadequate resection. She underwent a wide excision of the tumor and the outer cortex of the scalp bone. Immediate reconstruction with a free ALT flap (10 cm $\times$ 8 cm) was performed. Because of a bulked flap in the thigh, we performed immediate thinning of the ALT flap. The recipient vessel was the right superficial temporal vessel. The postoperative course was uneventful. The flap sufficiently tolerated the radiotherapy well, and no debulking procedure was required. The patient was satisfied with the cosmetic result, after a 24-month follow-up period (see Fig. 2).

Table 1 Patient profile and results.

Patient no./age (y)/sex	Tumor location	Margin	Tumor origin	Reconstruction method	Tumor size/ skin paddle (cm)	Complication	Follow-up time (mo)	Recurrence
1/30/F	Middle lower abdomen	Free	Primary	Pedicled abdominal flap	3 × 1/X	None	31	None
2/18/M	R't thigh	Free	Local recurrence	Pedicled R't ALT flap	3 × 2/9 × 5	None	21	None
3/38/M	Right lower abdomen	Free	Primary	Pedicled L't ALT flap	5 × 3/20 × 10	Donor site skin graft	43	None
4/55/F	L't lower leg	Free	Primary	Pedicled L't medial sural a. flap	3 × 2/9 × 8	Donor site skin graft	27	None
5/30/M	Left upper back	Free	Inadequate excision	Pedicled L't dorsal scapular a. flap	3 × 3/12 × 9	dehiscence after R/T →secondary healing	31	None
6/50/F	R't upper back	Free	Local recurrence	Pedicled L't thoracodorsal a. flap	5 × 4/22 × 10	None	26	None
7/27/M	R't lateral chest	Free	Primary	Pedicled R't lateral thoracodorsal a. flap	3 × 3/16 × 8	None	23	None
8/30/F	R't superior breast	Free	Primary	Pedicled R't LD myocutaneous flap	4 × 2/12 × 6	None	18	None
9/60/M	L't shoulder	free	Primary	Pedicled L't thoracoacromial a. flap	3 × 3/10 × 8	Flap failure → salvage FTSG	21	None
10/31/M	L't upper back	free	Primary	Free R't trapezius flap	7 × 3/18 × 9	None	33	None
11/55/M	Scalp	free	Primary	Free L't ALT flap	2 × 1/6 × 6	None	68	None
12/42/F	Scalp	free	Inadequate excision	Free L't ALT flap	4 × 2/10 × 8	None	24	None
13/50/M	Scalp	free	Primary	Free L't ALT flap	4 × 3/15 × 8	None	42	None
14/62/M	Left cheek	free	Primary	Free L't ALT flap	4 × 3/12 × 10	Donor site skin graft	18	None

a = arterial; ALT = anterolateral thigh; F = female; FTSG = full thickness skin graft; LD = latissimus dorsi; L't = left; R't = right; M = male; R/T = radiotherapy.

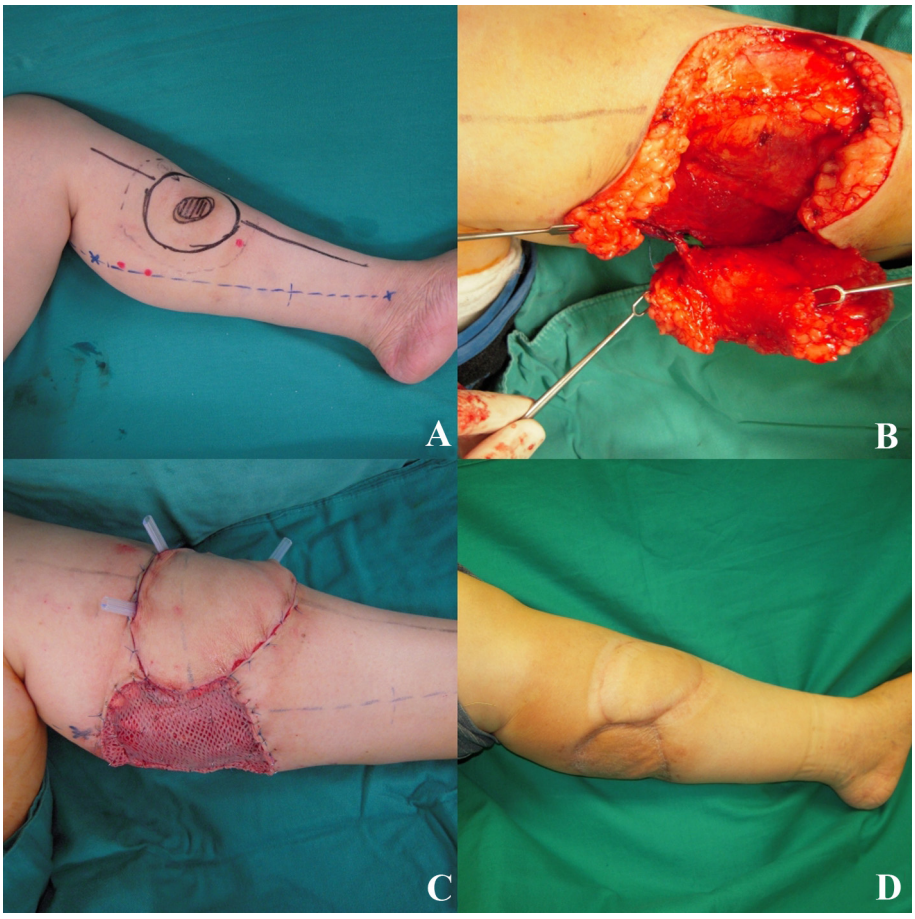

Figure 1 Patient 4 is a 55-year-old female. (A) A 3 cm × 2 cm lesion over the left medial lower leg is shown. (B) A 9 cm × 8 cm pedicled medial sural flap. (C) The donor site is covered with split-thickness skin graft. (D) A good esthetic result with minimal scar contracture over the skin graft area is seen.

5. Discussion

DFSP is a rare dermal mesenchymal tumor but the most common cutaneous sarcoma. Although DFSP can be found in all parts of the body, the trunk is the most common site. In our study, in half of the patients DFSP was prevalent in the trunk; 23.1% exhibited DFSP in the head and neck, and 15.4% and 7.7% in the lower and upper extremities, respectively. It affected males between the ages of 20 years and 50 years; the mean age in our series was 41.29 years. Initially, the lesion appears as a small nodule or plaque-like object, but it grows rapidly into a mushroom-like shape, typically yellow-brown in color. DFSP is characterized by a locally aggressive behavior that can involve surrounding tissues including fascia, muscle, and bone. No muscle or bone involvement was observed in our series. McPeak et al[5] observed only 8.13% of underlying bone involvement for DFSP. DFSP rarely metastasizes distally or to regional lymph nodes postoperatively; the lung is the primary site for distant metastasis.

The pathogenesis of DFSP remains unclear and no hereditary influence has been discovered. Several studies have proposed that a history of trauma or persistent irritation can be a presumptive cause of tumor occurrence.[6] Recent molecular studies have found that a chromosomal translocation involves 17q22; 22q13, related to a fusion of the genes *COL1A1* and *PDGFβ*. The fusion protein binds to the PDGFβ receptor then stimulates the emergence of DFSP.[8] Although a history of trauma was proposed as the cause of DFSP, only Patient 9 exhibited such history.

DFSP spreads cutaneously or subcutaneously, but magnetic resonance imaging and ultrasound can provide only limited information regarding this. Gross inspection is not sufficient and requires an incisional or excisional biopsy for confirmation. Diffuse infiltration of a spindle shape, CD43 positive fibroblast arranged in a storiform pattern can be observed through histologic study. The peripheral extensions of tumor cells have a bland appearance and make it difficult to determine the margin histologically.

According to the 2012 National Comprehensive Cancer Network guidelines, the recommendation for treatment involves complete surgical excision to adjacent fascia with immediate reconstruction.[9] According to the literature, the margin of safety varies between 1 cm and 5 cm. Based on Mohs micrographic surgery (MMS), an incision of 1-2cm margin and immunostaining were performed, and repeated resection was performed if the margin was identified as positive. MMS can be superior in soft-tissue preservation, but it has not been proven to reduce local recurrence.[10] Although recurrence can occur at surgical margins, we have observed cases of recurrence beyond the original lesion described in previous studies. We performed a wide

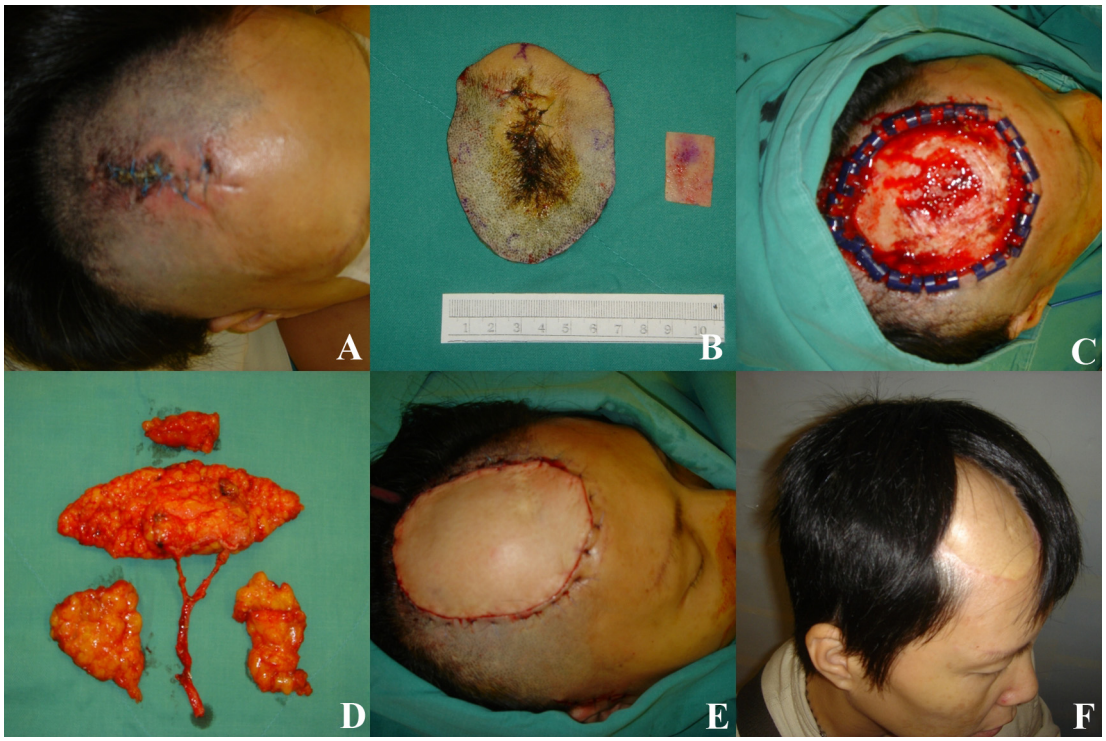

Figure 2 Patient 12 is a 42-year-old female. (A) A 4 cm × 2 cm lesion after inadequate excision at another hospital is shown. (B, C) Wide excision with a 3-cm margin and removed outer cortex. (D) A 10 cm × 8 cm free anterolateral thigh flap with immediate defatting. (E) Anastomosis to superficial temporal vessels. (F) Excellent soft-tissue augmentation and contour is seen.

excision with a 3-cm safe margin and deep to fascia, which has been recommended in numerous studies. The microscopic spread of the tumor may contribute to a high local recurrence. No recurrence was observed in this study. Rutgers et al[11] observed a 50% recurrence rate, which was reduced to 13% after an adequately wide excision. We sent specimen from the deep margin for frozen section for all cases in this series. The data from the frozen section were not comparable to a pathological report, but indicated whether the resection was adequate. An invasion of DFSP can affect subcutaneous tissue, muscle, deep fascia, and bone. We performed a wide excision deep to the investing fascia, which is often less than 3 cm deep. To cover the muscle and the exposed underlying bone, we used flap reconstruction instead of skin graft to maintain functionality and prevent disability. DFSP is clinically benign but pathologically malignant; we performed resection again if recurrence occurred. We did not use skin graft as a reconstructive option in this study because of its esthetic and functional disadvantages. However, in cases with an unfavorable general condition, lesion sites without functional restriction, and in easily concealed areas, skin graft should be considered an option because of several advantages such as a quick sighting of recurrence, easy postoperative care, lower cost, shorter surgery time, and minimal chance of reconstructive failure.

Local recurrence remains challenging; adjuvant radiotherapy can be administered after a wide excision to reduce the risk of local recurrence in patients with close or positive margins. DFSP is radiosensitive and disposed to excellent local control.[12] radiotherapy is recommended as a systemic adjuvant treatment to reduce the risk of local recurrence.[13] Adjuvant radiotherapy has been shown to enable effective control of local recurrence even for positive margins. For patients who are not candidates for surgical resection, radiotherapy is recommended in the National Comprehensive Cancer Network guidelines. The chance of radiation toxicity is low, but the benefit must be weighed against the risk.[12]

Although radiotherapy can be used for a positive margin or to reduce local recurrence, the most crucial factor in preventing recurrence is obtaining a negative margin through a wide excision. Castle et al[12] stated that 93% of patients with a history of multiple recurrence and positive margins, combining surgical excision and adjuvant radiotherapy, exhibited a 10-year actuarial local control. We send resected tumor to a pathologist for frozen section to confirm a negative margin before wound coverage and allowed superior local control and reduced the recurrence rate.

The fusion gene *COL1A1-PDGFβ* resulting from translocation t(17:22) is present in > 90% of DFSPs. Inhibitors of this pathway may become a nonsurgical option in the future. The oral tyrosine kinase inhibiter, imatinib, has exhibited a favorable response rate in unresectable and metastatic DFSP studies.[4] Although imatinib should not be used to reduce the surgical margin, the current National Comprehensive Cancer Network guideline recommends it for unresectable diseases, positive surgical margins, recurrent diseases in critical functional and cosmetic areas, and metastatic diseases.[10]

There is no standard staging for DFSP. The primary tumor can be considered Stage I, regional lymph node metastasis as Stage II, and distant metastasis as Stage III.[9] Although

DFSP rarely distally metastasizes and the reported 5-year survival is > 99%, most patients with metastatic disease die within 2 years. The most crucial prognostic factor is to obtain a negative surgical margin. Lesions > 10 cm, involving a deeper structure, positive margin, and multiple recurrences share a higher risk of local recurrence and distant metastasis. The overall survival rates for both 5 years and 10 years were 98% with a negative margin excision and adjuvant radiotherapy.[12,14]

According to previous studies, numerous surgeons have used skin graft after tumor resection. Scar contracture, hyperpigmentation, ulceration, and partial graft loss have often occurred after adjuvant radiotherapy.[15] Although skin graft tolerated the adjuvant radiotherapy after complete healing, we still had to wait for 8 weeks after the operation for improved results.[16] Nevertheless, the loss of skin graft is considerably increased after a single dose of ≥ 25 Gy, even after complete healing within 4 weeks.[2] Numerous surgeons do not advocate skin graft for oncologic reconstruction if adjuvant radiotherapy is required. With the refinement of surgical techniques, it is possible to use regional or free flaps to improve the esthetic results and functional outcomes for oncologic reconstruction with a high success rate.[17] Nearby pedicled perforator flaps can reconstruct the defect with anatomically similar tissue to resolve the color mismatch and volume deficiency of the skin graft. Rarely, perforator flaps require a debulking procedure and an excellent esthetic outcome can be achieved. Faster wound healing allows patients to initiate radiotherapy 4 weeks after an operation, compared with 8 weeks after having undergone a skin graft. Perforator flaps provide an adequate volume and superior resistance to radiotherapy-induced contracture, ulceration, and partial loss, and also eliminate secondary surgeries. Although > 80% of the ALT flap donor sites could be primarily closed, we did not achieve primary closure in three patients. Overall, the donor sites with a flap width-to-thigh circumference ratio of < 16% can be primarily closed.[17] Skin graft was used to cover the donor sites in those three patients. Furthermore, skin graft was implanted on the donor site and did not result in any restriction of movement after healing. An ALT flap recently became the prime-example flap for soft-tissue reconstruction and could be the first choice for free-tissue transfer. To improve donor-site esthetics, a remainder island ALT flap from the distal portion, which is based on a retrograde pedicle, can be used to close in a V-Y advancement manner.[17] Up to 16% of the circumference can be primarily closed for improvement of the scar. It can be harvested as myocutaneous or fasciocutaneous flaps according to the defect requirement, has long and reliable pedicles for microanastomosis or even for transfer as a regional flap, has a primary closed donor site with minimal morbidity, and the scar can be easily hidden underneath clothing.[3]

6. Conclusion

A wide excision with a 3-cm margin of safety with immediate reconstruction is a reliable method for DFSP resection. To minimize the chance of local recurrence, a negative margin should be confirmed through a frozen section prior to wound coverage. Adjuvant radiotherapy can be initiated to reduce the local recurrence to 4 weeks with perforator flaps compared with a skin graft. In addition, perforator flaps offer superior resistance to radiation-induced skin ulceration and provide adequate tissue to prevent restriction of movement through contracture; furthermore they provide excellent functional outcomes and esthetic results.

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