


Reconstruction options in recurrent dermatofibrosarcoma protuberans: A scoping review

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Abstract

Background: This review will outline the evaluation, diagnosis, and management of dermatofibrosarcoma protuberans and emphasizes multidisciplinary role of nurses, plastic surgeons and radiation oncologist in this recurrent metastatic lesion. It pinpoints affected population at risk, clinical features, and reconstruction options. No analytical research has been done in this area.

Material and Methods: A scoping review of patients of DFSP who underwent reconstruction after excision of tumors was performed in the Department of Burn and Plastic Surgery, AIIMS Rishikesh. It used a five framework approach. A review of 85 similar cases reported in the literature have been scrutinized in relation to the reconstruction options, sites of the tumor, margins of excision and recurrence.

Results: 85 full length English studies were included out of the 445 cases found in Pubmed and related search engines to reveal various reconstructive options in reconstruction of DFSP defects. Present scoping review identifies free anterolateral thigh flap to be useful in 7 review articles followed by propeller flaps in 3 isolated case reports. 2 cases of free latissimus dorsi flap were used for reconstruction of abdominal defects.

Conclusion: All patients should undergo a strict screening protocol where the health personnel can play a crucial role by educating parents on the follow up and report new lesions as early as possible. All operated tumor patients can be given safety tips and education on care and risks after reconstruction with skin flaps or skin grafting. A multidisciplinary approach between the surgeon, nurse and radiation oncologist is needed for effective management of these lesions.

Keywords

Recurrence, dermatofibrosarcoma protuberans, margin

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Introduction

Dermatofibrosarcoma protuberans (DFSP) is a relatively rare malignancy of the skin. It is a locally aggressive tumor with a high recurrence rate. It arises from the dermis and invades deeper tissues like fat, fascia, muscle, or bone. DFSP is the most common type of cutaneous sarcoma, with an incidence of 0.8 to 5 cases per million populations per year. It most commonly occurs on the trunk (42–72%), followed by the proximal extremities (16–30%). It is most commonly seen in adults.¹

Demartofibromasarcoma protuberans was coined by Hoffman in 1925.² It is difficult to identify this lesion in children.³ *Postoperative* radiotherapy reduces the risk of

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recurrence in DFSP.⁴ The precise aetiology of the lesion is unknown but it has been linked to translocation of chromosomes 17 and 22 leading to alteration of the platelet-derived growth factor gene. DFSP most commonly affects adults between the ages of 30–50 years, with a significantly higher annual incidence rate among women and blacks compared with whites.

DFSP constitutes approximately 7% of cases in the head and neck.

The lumpy nodules cannot be seen and felt because of slow progression and therefore diagnosis is delayed for years. Later it may form a new atrophic and sclerotic plaque over the trunk or abdominal wall mimicking a hypertrophic scar or keloid.⁵

The local recurrence rates can be reduced by Mohs surgery.⁶

The recurrence rates are higher in patients with fibrosarcomatous variety and the degree of cancer increases with multiple recurrences.⁷ The histologic hallmark of this lesion is a storiform proliferation of spindle cells in the deep dermis. The COL1A1-PDGFB fusion gene is seen in 85–90% of the cases. Histologically, the epidermis overlying DFSP shows thinning and may be separated from the neoplasm by a Grenz zone. Dermatofibrosarcoma protuberans start as a nodule or ill-defined dermal plaque that infiltrates the subcutis or sometimes skeletal muscle with a characteristic “honeycomb” pattern. The epidermis is usually not involved. The immunohistochemical analysis reveals CD34 positivity that suggests dendritic cell histogenesis. Myoid nodules are more frequently seen in fibrosarcomatous variant of DFSP.

Melanin pigment is seen in the Bednar variant.

Trauma is the inciting factor in the majority of these neoplasms. Chromosomal aberrations play a pivotal role in their pathogenesis. The cytogenetic analysis has revealed a reciprocal translocation $t(17;22)(q22;q13)$ or supernumerary ring chromosome derived from $t(17;22)$. These produce a fusion between the genes encoding COL1A1 and PDGFB. CD 34 is positive in DFSP indicating a dendritic histogenesis. This positivity is lost in tumors that undergo fibrosarcomatous transformation. PDGF Beta is a potent mitogen for mesenchymal cells.

It frequently presents as an indolent, nontender cutaneous nodule and is most commonly seen on the trunk and proximal extremities. There is an initial slow growth phase making it difficult to differentiate from benign dermatofibroma. The studies have reported a high incidence of DFSP among men. The peak age incidence is between 25 and 45 years, but it has been reported in infants. Tissue sparing is relevant in children as wide defects are created after excision similar to other case series in the literature.⁸ Clear pathological margins are achieved with wider resections as seen in other studies conducted by various others.⁹

DFSP may arise from a pre-existing trauma to the skin, such as vaccination, tattoos, radiation, or burns. It carries a 2–5% risk of metastasis. The fibrosarcomatous changes are associated with increased rates of recurrence and poor survival. Tumors often enter a rapid growth phase where they start to infiltrate vertically and become fixed to structures of the deep subcutaneous tissue and to fascial planes. Rapid tumor enlargement with high mitotic figures and atypia heralds the onset of fibrosarcomatous transformation. In the differential diagnosis, these tumors mimic hypertrophic scars or benign soft tissue tumors without any specific symptoms. A high index of suspicion is seen in tumors that exhibit aggressive behaviour, spreading within the dermis, subcutaneous tissue, and ultimately into muscles. Fibrosarcomatous transformation of dermatofibrosarcoma protuberans (FS-DFSP) is considered to be an intermediate-grade neoplasm with a slightly increased risk of distant metastasis. The $t(17;22)$ translocation is also seen in myxoid and pigmented subtypes and fibrosarcoma arising in DFSP. Bednar tumor is cytogenetically identical to regular dermatofibrosarcoma. Chromosome painting with FISH shows the supernumerary ring chromosome to be composed of discontinuous, interwoven sequences from chromosomes 17 and 22. Prompt and definite reconstruction options can be viable options to treat this disease. The purpose of this scoping review was to identify evidence detailing the reconstruction options, margins of excision and recurrence in such tumors.

Methods

This is a scoping review of patients of *Dermatofibrosarcoma protuberans* who underwent reconstruction after excision of the tumors. A review of 85 similar cases reported in the literature have been outlined and discussed in relation to the reconstruction options out of a total of 445 cases in PubMed, Ovid and Embase.

A five-stage framework developed by Arksey and O'Malley was adopted to identify the research question; sort out relevant studies; select the studies for review; charting the data and display results based on inclusion criteria.

A thorough literature search has been performed in articles describing the reconstruction options after excision of DFSP. The choice of reconstruction was based on the site, size and reconstructive ladder followed in the armamentarium.

Inclusion criteria included articles encompassing reconstruction options after excision of tumors with margins of resection and recurrence.

Exclusion criteria were articles without evidence of reconstruction, not written in English and articles with abstracts.

The lead author (VM) and one co-collaborator (AP) independently selected articles based on the inclusion and

exclusion criteria. Discrepancies in article selection were resolved through discussion between reviewers. Inclusion/exclusion decisions were recorded.

Data extracted from each reviewed article included the following variables i.e. study design, margins of resection, recurrence, metastasis and intervention.

Results

The PRISMA flow diagram (Table 1) throws light on the total literature review. 448 unique records were identified. 3 articles were excluded because they were not written in English. 85 were found to meet inclusion criteria for analysis. 8 isolated case reports using free flaps were reported in this review. 77 reviews found tumor to be localised over the trunk, margins of excision to be 3 cm and recurrence free survival rate to be 94%. REGESMOHS study also reiterated site of tumor to be trunk and pointed out that recurrent tumors had deeper invasion and needed more MMS sessions.

One patient with post-mastectomy DFSP occurring after breast irradiation and reconstruction was treated by autologous fat transfer. Table 2 outlines the review articles based on surgeries performed after creation of defects and subsequent reconstruction by flaps.

Wider resection margins were significantly correlated with more reconstructions. Choice between MMS and wide local excision is based on cosmesis, preservation of function and decrease in morbidity and recurrence. Frozen section in the intraoperative period can help reduce the need for repeat surgeries.

The choice of reconstruction after creation of defect in the abdomen can be closed by local thoracoepigastric flaps. Free flaps is an alternative option where microsurgical services are available in the form of free latissimus dorsi or anterolateral thigh flaps.

Local flap options can be in the form of rotation or propeller flaps based on defined tissue loss, size of defect, location, tissue components and sensibility requirements. Familiarity with the great variety of flaps and microsurgical expertise such composite defects can be reconstructed after adequate patient counselling. Defects in the shoulder and arm can be reconstructed with functional gracilis or latissimus dorsi muscle flaps. Defects involving bone can be reconstructed with free fibula flaps. Composite defects in forearm can be reconstructed with pedicled or free radial forearm flap.

Common regional flap options for chest wall reconstructions are pectoralis major muscle or myocutaneous flaps of size upto 15 by 23 cm. These flaps cannot be used if internal mammary artery on that side has been used for Coronary artery revascularization.

The abdominal wall can be divided into 4 zones i.e. epigastric, periumbilical, hypogastric and lateral. The lateral

abdominal defects can be closed with latissimus dorsi muscle flap. Free tissue transfer is for periumbilical defects as these areas are out of reach of local flaps. Hypogastric defects are closed with pedicled thigh flaps.

Discussion

Mohs is particularly useful in adult patients in anatomically or cosmetic locations like eyes, ears, and nose. The NCCN 2011 guidelines for the management of DFSP recommend immediate closure in diagnosed cases. The reconstruction can be delayed in cases involving extensive undermining or flaps until negative surgical margins are assessed by a frozen section. Radiotherapy has been planned as adjuvant therapy after surgical excision.

Neoadjuvant radiotherapy may reduce the risk of local recurrence, particularly in cases where positive surgical margins were outlined in biopsies. Based on the NCCN guidelines wide local excision was the most common modality in a series of 69% recurrent DFSP in this study and 23% cases operated by Mohs surgery.¹ Pathologically the fibrosarcomatous variety is aggressive with higher rates of recurrence and needs close follow-up after excision.² Pathological and immunohistochemical examinations are thus currently the gold standard for diagnosing DFSP, with surgical resection (at least 2cm margin) remaining the main treatment option similar to our case operated for the same.³ DFSP has the tendency to expand from the central focus and invade the surrounding tissues. The authors reported a median age of 11 years in a series of DFSPs most common in the head and neck region citing patient cooperation as the most common challenge.¹⁰ Hao et al. have described a staging system for DFSP as shown in the Table 3 below.

Dermatofibrosarcoma protuberans are considered to have a high rate of local recurrence, with reported rates in some series greater than 50%. Surgical defects created after the excision of tumors were closed with a layered primary closure in 5 patients, and 2 patients underwent reconstruction with plastic surgery.⁴ Eleven cases of recurrent DFSP were treated with reconstruction techniques.¹¹ This tumor activates platelet-derived growth factors which leads to rampant growth of the tumor. The appearance of distant metastases is preceded by a local recurrence of the tumor and positive margins. Imatinib is a selective PDGFR tyrosine kinase inhibitor, showing partial and complete remissions of DFSP.⁶ Stivala et al. reported 46 out of the 59 cases in their study were treated with wide local excision and exhibited tumor-free margins.¹² Woo and others had predicted margins of 1.5–2 cm to exhibit a higher degree of reconstruction options in group 3 patients and predicted re excision or adjuvant radiation therapy as a treatment option in patients with positive margins.¹³ The Clinical Practice Guidelines in Oncology for Dermatofibrosarcoma Protuberans published by the National Comprehensive Cancer

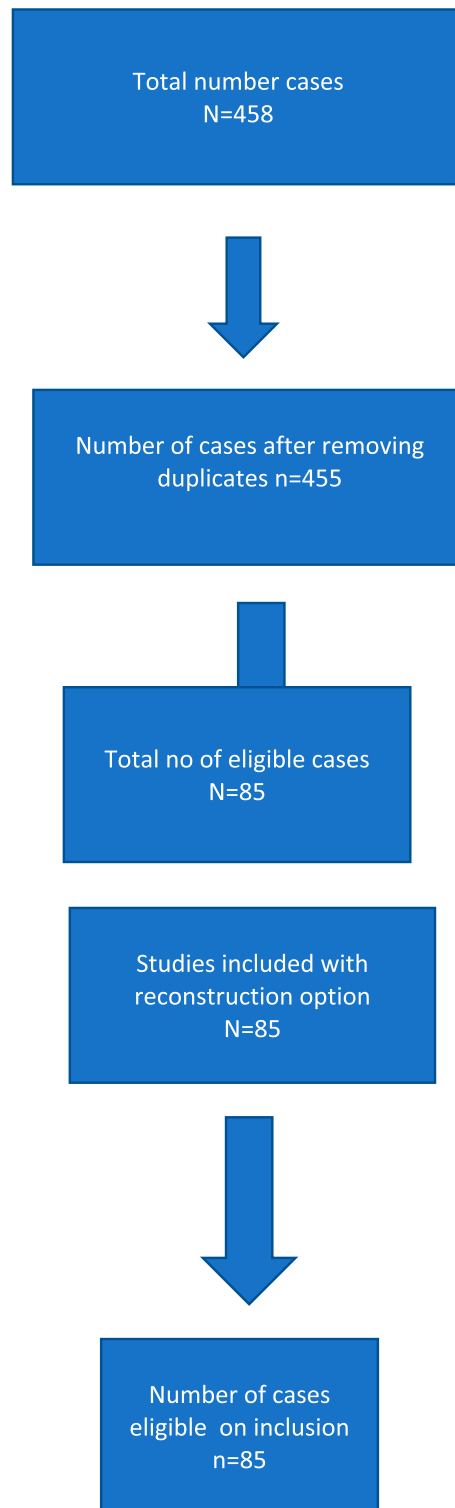
Table I. Prisma flow diagram.

Table 2. Screening of review articles based on reconstruction.

Review article	Tumor location	Margin	Flap	Recurrence
Guillen CS et al. (n = 222;2021)	Trunk	1.23	MMS	2(0.9%)
Popov (n = 40;2007)	Upper trunk	3.1	Flaps(23)	13(33%)
Goldberg et al. (n = 25,2015)	Trunk	2.5	Flaps	8(40%)
Durack et al. (n = 483,2021)	Trunk	3	WLE	82%
Verma et al. (n = ,2021)	Extremities	2	Flaps	2(6.5%)
Tsai (n = 13,2014)	Trunk	3	7 flaps	Nil

Table 3. Staging of DFSP.

Stage	Description
I	Non protuberant lesions
2A	Protuberant mass confined to skin
2B	Protuberant mass infiltrated through fascia
3	Lymph node metastasis
4	Distant metastatic lesion

Network recommends 2 cm to 4 cm margins to investing fascia if wide local excision is applied.

Archontaki and others have resected margins of at least 5 cm in cases where wide local excision was performed and subsequent radiotherapy was planned in this study.¹⁴ Follow-up of the cases should be extended beyond 5 years to look for late recurrences.

Mohs surgery involves a stepwise approach to the resection of a total tumor. The tumor is excised in quadrants with tangential margins done in real-time and in multiple stages. Indication for treatment is the plaque or scar lesions. It is also indicated in critical locations, such as the head or hands.^{15,16} A worse outcome is seen in patients with the following risk factors ie local recurrence, large tumors, deep invasiveness, myxoid type, and expression of cyclin D1. The meta-analysis of 8 observational studies showed excision with a margin <3 cm yielded a poor prognosis as compared to previous studies. There was an increased recurrence and positive margins.¹⁷

A systematic review revealed the risk of local recurrence, metastasis, and death from disease was higher in fibrosarcomatous variety as compared to the normal variant plus confounder s was not evaluated in this review.¹⁸ They found metastases in the lungs, bone, soft tissue, liver, kidney, gastrointestinal system, and lymph nodes.

Paradisi and others in their study showed the recurrence rates of Mohs surgery were significantly lower than those of wide local excision. This study was comparable to the studies done by Chen and Yao where the relative risk for recurrence between wide local excision versus Mohs surgery was 15.9.¹⁹

Free anterolateral thigh flaps were utilized in 5 cases of DFSP with good results and fewer chances of recurrence. A

giant keystone type 3 perforator flap was used in the posterior trunk for a wide skin defect avoiding skin grafting.²⁰ The authors have reported the use of a thoracoabdominal flap for reconstruction of superficial chest wall sarcomas consisting of dermatofibrosarcoma protuberans similar to our case based on the axial supply of the versatile flap and avoids microsurgical procedures.²¹ A supraclavicular artery island flap was used to reconstruct a defect in the parotid region with good results as this achieves microscopic free margins and good aesthetic results with better colour matches.²² Two patients underwent wide resection of forehead DFSP and reconstruction with a single rotational scalp flap based on the blood supply territories involving 3 major vessels namely occipital, superficial temporal, and posterior auricular artery. A free latissimus dorsi flap was done in stages to close an abdominal defect in this study with a 5cm margin and achieve a deeper resection for a disease-free result. The musculoaponeurotic plane was covered with a mesh followed by a free flap covering the superficial layer which was unfolded from the base to cover the defect.^{23,24}

For the other patient with the epigastric tumor, a synthetic mesh was placed, over the defect which was reconstructed with a reverse abdominoplasty flap and two thoraco-epigastric flaps with good results. A midline abdominal wall reconstruction with anterior component separation technique was done with bridge mesh plasty in a case of epigastric DFSP.^{25,26} 13 patients underwent wide excisions in this review.^{26,27} Post-excision reconstruction showed direct closure in three cases, skin grafting in three cases, and local or free flap reconstruction in seven cases.²⁸

Seventeen patients (10¹ male; mean age, 9.9 years) were managed during the study period. The median follow-up was 29 months. All patients had surgical excision. Three patients required further excision to achieve uninvolved final margins. There were no recurrences observed.²⁹ The choice of reconstruction, i.e., free, regional, or local flap was based on the size of the resultant defect.

A strict follow-up regimen is required in patients with DFSP as the recurrence rate is very high. The recurrence rates are higher in the head and neck region. The margins must be assessed by Mohs technique or 3D margin assessment. Lifelong surveillance is required to document the

progress of the disease. The examination of the primary site is done every 3–6 months to evaluate the progress of the disease. NCCN guidelines recommend are biopsy every 6–12 months. If disease-free survival is more than 3 years this surveillance is done every 6–12 months. European guidelines have recommended a clinical examination every 6 months for the first 5 years, and at yearly intervals thereafter for up to 10 years. Imaging should be preserved for recurrent DFSP or DFSP with sarcomatous change. Screening for metastasis is important in patients with sarcomatous change or patients with recurrence or margins that are positive.

All patients with reconstructions should undergo a strict screening protocol where the doctor can play crucial role by educating parents about the follow-up and reporting new lesions as early as possible. The health care workers caring for operated tumor patients can provide patients with cancer guidelines and educate patients on postop care after reconstruction with skin flaps or skin grafting. A multidisciplinary approach between the plastic surgeon, nurse, and radiation oncologist is needed for the effective management of these lesions.

Conclusions

In conclusion, DFSP is a rare cutaneous sarcoma with a high propensity for local recurrence. This however can be managed with wide surgical excision with clear resection margins which in most cases does not require any adjuvant therapy. However, since there is a 25% risk of recurrence within the next 5 years' patients should be kept in close follow up and need to be counselled regarding the consequences appropriately.

Patients can be made aware of the warning signs of skin malignancy. All operated tumor patients can be given safety tips and education on care after reconstruction with skin flaps or skin grafting. An algorithm must be selected to define surgical procedure based on tumour's location, size, stage and relationship with surrounding soft tissue and bone structures, A multidisciplinary approach between the plastic surgeon, nurse, and radiation oncologist is needed for the effective management of these lesions.

Choice of reconstruction can be based on site of the tumor i.e., abdomen where a local flap can be a viable option or free flaps like anterolateral thigh flap can be used to cover extensive defects over an on lay mesh. Recurrent lesions can be managed with local flaps or free flaps. Options for local flaps can be transposition or rotation flaps based on the location of the defect.

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