

# Oncoplastic approach in Dermatofibrosarcoma Protuberans: a Case Report

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June 1, 2020

## Abstract

We present the case of a 38-year-old woman with previous bilateral breast augmentation, who underwent wide local excision for Dermatofibrosarcoma protuberans, and reconstruction of the abdominal wall without shifting the inframammary fold.

## Introduction

Dermatofibrosarcoma protuberans (DFSP) is defined as a skin fibroblastic tumor which is locally aggressive, has a tendency to recur locally with rare metastasis.<sup>(1)</sup> The incidence rate is 4.1 persons per million person-years.<sup>(2)</sup> According to Lemm D. et al, the condition accounts for between 2% and 6% of all soft tissue sarcomas.<sup>1</sup> Contrary to more common skin cancers, the growth of which presents a concentric pattern, DFSP has unpredictable asymmetric growth with long fingerlike projections protruding between collagen bundles laterally, rendering its surgical management difficult.

With a rare distant metastasis (1% - 4%), complete removal is considered curative.<sup>2</sup> While fibrosarcomatous transformation is indicated by recent and rapid modification of the lesion, clinical suspicion must be confirmed by pathology before definitive surgery is performed. Definitive diagnosis of DFSP is made by incisional, or less frequently excisional, biopsy procedure.

When faced with tumors involving a cosmetically sensitive region, aggressive wide resection might result in some disfiguration to the region. This might require, to a certain extent, advanced reconstructive skills in order to limit disfiguration or to cover exposed defects, such as flaps or skin grafts.<sup>3</sup> For this reason a proper planning and a multi-disciplinary team approach, including a reconstructive surgeon, is part of best practice.

Our work presents a case of DFSP located at the superior part of the abdomen in a patient who had previously undergone breast augmentation with prosthesis. Some of the issues encountered in the aggressive oncological resection, while conserving an acceptable aesthetic result, are emphasized.

## Case Presentation

A previously healthy married 38-year old female patient, with a 6-year old bilateral implants breast augmentation, presented for wide local excision of an abdominal dermatofibrosarcoma protuberans (DFSP). History goes back to few weeks prior to presentation when the patient noticed, an indurated, erythematous skin lesion in the epigastric area, enlarging with time. She consulted her treating physician who recommended excisional biopsy. Excisional biopsy was done and the lesion was sent for definitive pathology. Pathology result showed skin with hyperplastic epidermis and underlying non-circumscribed dermal proliferation of spindle shaped cells arranged in a loose storiform pattern infiltrating deeply in the subcutaneous tissue entrapping fat cells. Mitotic activity was a low 1-2/10HPF. Immunohistochemistry showed tumor cells (positive for CD34; negative for SMA; Ki67 was low =2/100 cells), which was consistent with DFSP. Most importantly, the tumor was present at the surgical margin.

At two weeks post the excisional biopsy the patient was referred to the oncological surgeon who ordered a CT scan that showed only fibrosis at the previous surgical site, without any signs of extensive local invasion or distant metastasis. Wide local excision was done through an elliptical incision incorporating the old scar with an additional margin of 3 cm (8 x 6 cm). (**Fig.1**)

The skin, the subcutaneous fat, the linea alba at the center, and the anterior rectus muscle aponeurosis were excised en bloc and sent to frozen section that showed negative margins for tumor. Due to the proximity of the lesion to the Xyphoid area and the inframammary fold (IMF), the reconstructive surgeon made sure not to distort the IMF especially that the patient previously had a bilateral breast augmentation. In this regard, we proceeded with a more caudal undermining of the anterior rectus aponeurosis and rectus muscle than cranial (away from the IMF). Closure of the aponeurosis was done using PDS loop tension free. Vicryl 3-0 was used for subcutaneous closure and monosyn 4-0 for subcuticular closure. (**Fig.2**) . Post-operative course was uneventful.

Definitive pathology result showed residual DFS protuberans (1x0.5 cm), involving the dermis and subcutaneous adipose tissue and dermal fibrosis, traumatic fat necrosis, and foreign body giant cell reaction to suture material, compatible with previous surgery. All surgical margins were clear. The shortest distance between the residual neoplasm and the deep inked resection was 1 cm (linea alba).

Follow-up was recommended by self-physical exam, and a wound exam by the surgeon every 6 months for the first 5 years. The 6 months follow-up showed satisfactory result without clinical signs of recurrence. On a scale of 1 (very poor) to 10 (excellent), the cosmetic result was rated as 9 by the patient, which concurred with the same grade given by both involved attending surgeons. (**Fig.3**)

## Discussion

As reported in literature, DFSP is a very slow growth of flesh-colored or slightly yellow–brown skin tumor without epidermal invasion but with intracutaneous and subcutaneous spread. In most cases, the tumor appears mainly on the trunk but sometimes presents as a reddish, flat elevated, firm lesion with irregular borders or multinodular appearance on the extremities, head, and neck. The tumor's rare metastasis and the lesions benign appearance result in its late diagnosis and treatment. For diagnostic purposes, incisional or excisional biopsy is a rule of thumb. Diffuse infiltration of the dermis and subcutaneous fat by densely packed, cytological relatively uniform, spindle shaped, CD34 –positive tumor cells, arranged in a characteristic storiform shape are shown by haematoxylin and eosin stains. Tumor cells spread along the septae of the subcutaneous fatty tissue.<sup>4</sup>

Extensive work up is not routinely indicated except for patients with suspicion of metastases on clinical examination, for those with recurrent disease, and for DFSP with fibrosarcomatous transformation features.<sup>4</sup>

As in our case, in residual disease (after excisional biopsy), CT scan or MRI may be indicated before undergoing definitive curative debulking oncological resection. As a matter of fact, the gold standard treatment according to the NCCN Clinical Practice Guidelines in Oncology, is complete surgical excision with appropriate reconstruction. Surgical excision with a margin of at least 2 to 3 cm is recommended for treatment because the local recurrence rate is 20% to 50% in cases of incomplete resection. Unresectable lesion or resected lesion with a positive margin should be treated with adjuvant radiotherapy, which is known to be reliable in reducing the local recurrence rate and avoiding the mutilation and functional deficits caused by repeated surgery<sup>5</sup> . However, negative surgical margins are still considered the most significant prognostic factor because inadequate initial resection may result in uncontrolled local growth or metastases.<sup>6</sup> In our case, proper imaging allowed us to resect part of the anterior rectus muscle aponeurosis, and therefore to preserve most of the abdominal wall integrity while avoiding the need for mesh.

In addition to the correct identification of the extent of the disease, another challenge was faced due to the location of the lesion (epigastric area) and its proximity to the inframammary fold (IMF) when planning the excision with wide margin. For this reason we believe that proper prior planning with the plastic surgery team is part of the best standard of care. Having decided that flap surgery would not be necessary to

cover the defect, we opted for a full thickness advancement flap (cutaneous/subcutaneous/rectus fascia) of the lower part of the excised abdominal area (umbilical to xyphoid). This allowed us to keep the IMF attachment intact, and to preserve the previous harmonious and symmetrical breast augmentation result. Finally our results and limited follow-up showed a satisfactory cosmetic score, and most importantly the absence of clinical signs of recurrence.

## Conclusions

Dermatofibrosarcoma protuberans (DFSP), an aggressive, rarely metastatic tumor, can be cured with proper surgical margins. However, depending on its anatomical location, oncoplastic reconstruction should be part of the treatment. To restore the cosmetic and functional we strongly recommend a multidisciplinary approach (oncological and reconstructive surgeon) when facing DFSP.

## Author contribution

- **Conception and Design:** Wissam Dib and William Watfa
- **Acquisition of Data:** Ghassan Antoun and Antoine Challita
- **Drafting the Article:** Wissam Dib and Vincent Khoury
- **Revising It for Intellectual Content:** William Watfa
- **Final Approval of the Completed Article:** William Watfa

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## Legends

Figure 1. Gross specimen excised (8 x 6 cm) at the time of wide local excision before frozen pathology analysis.

Figure 2. Postoperative result after wide local excision and closure.

Figure 3. Anterior view of the patient at 6 months post-op showing acceptable cosmetic result.



