

# RECURRENT DERMATOFIBROSARCOMA PROTUBERANS OF THE CLAVICULAR REGION: RADICAL EXCISION AND RECONSTRUCTION WITH LATISSIMUS DORSI MYOCUTANEOUS FLAP

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## Summary

Dermatofibrosarcoma protuberans (DFSP) is a soft tissue tumor with high propensity for local recurrence. We present a case of a 53-year-old woman with a recurrent DFSP of the clavicular region. The patient underwent extensive local excision including the external cortical bone of the left clavicle. The resulting large defect was reconstructed using pedicled Latissimus Dorsi myocutaneous flap. The histopathological examination confirmed dermatofibrosarcoma protuberans with clear surgical margins and the patient was satisfied with the final aesthetic result.

**Key words:** LD flap, FALD flap, dermatofibrosarcoma protuberans, DFSP, reconstruction, pedicle flap, latissimus dorsi muscle

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## INTRODUCTION

The dermatofibrosarcoma protuberans (DFSP) is a rare soft tissue tumor that involves the dermis, with a strong tendency to infiltrate surrounding tissues. It is characterized by high propensity for local recurrence but low tendency to metastatic dissemination, being therefore recognized as an intermediate to low degree of malignancy tumor <sup>1</sup>. The typical clinical presentation is an irregular multiple mass or else a hard-indurated plaque affecting the skin. It is mostly seen on the trunk and extremities, but it may occur on the head and neck region also, such as the scalp, cheek, supra-clavicular region and the orbit <sup>2,3</sup>. The incidence is quite similar in women and men (4.4 vs 4.2% per million people per year) with a relative 5-year survival of 99.2% <sup>4</sup>. The most affected age group of DFSP is from 20 to 50 years <sup>5</sup>. The exact cause of the onset is not yet clearly understood. Studies have involved a chromosomal translocation, resulting in the fusion protein COL1A1-PDGFB, which promotes tumor growth through the overproduction of platelet-derived growth factor (PDGF). Confirmed diagnosis is obtained through skin biopsy <sup>6</sup>. Immunohistochemical analysis should be always utilized to support the diagnosis. Staining for Vimentin

and CD34 is commonly employed, and sensitivity has been reported between 84 and 100 percent <sup>7</sup>. The gold standard treatment is represented by surgery, since wide excision with at least 2 cm of safety margin should always be performed. The primary surgical goal is clean, tumor-free resection margins <sup>8</sup>. Unresectable DFSPs should be treated with radiation therapy and/or targeted therapy. The aim of this paper is to present a case report of a severe DFSP recurrence of the left supraclavicular region, treated with a wide and deep excision followed by reconstruction with a pedicled Latissimus Dorsi (LD) myocutaneous flap. This type of reconstruction allowed us to repair such a large loss of substance with nerve structures and large vessels exposition, and to fill the subcutaneous volume deficiency with the myo-adipose portion of the flap, that could also be enhanced with autologous fat tissue transfer <sup>9</sup>.

## CASE REPORT

We present a case of a 53-year-old female patient with a recurrent DFSP of the left supraclavicular region. The patient had already undergone several resection procedures of the tumor mass, which had recurred several times. At the time of the visit, the patient showed the result of the last surgery with a further 5.5 x 5.2 cm tumor excision and partial thickness skin graft reconstruction (Fig. 1). The histological examination showed a diagnosis of cicatricial fibrosis with chronic giant cell inflammation with hypercellular areas with morphological and immunophenotypic aspects (CD34+). The report was compatible with extended Dermatofibrosarcoma Protuberans, present at the deep margin of resection. The patient was scheduled for radicalization of the old excision and reconstruction using the pedicled LD myocutaneous flap. A 15 x 9 cm skin paddle over the LD muscle was drawn very low on the back (about 8 cm below the tip of the scapula) in order to ensure a proper transposition of the flap and to allow a tension-free reconstruction (Fig. 2). The patient was positioned on right lateral decubitus and an en-bloc excision was performed, including the muscular component, i.e. part of the clavicular portion of the pectoralis major muscle and clavicular fibers of the deltoid muscle, and the external clavicular cortical bone. We reached macroscopically disease-free margins, carefully identifying and preserving the external jugular vein and the branches of the brachial plexus (Fig. 3). Simultaneously, the LD flap was harvested from the back, basing it on pre-operative drawing. Once the skin paddle was isolated on the LD, the whole muscle was dissected from its iliac insertion to its tendinous origin at the humerus, which has been resected in order to ensure a tension-free transposition.



**Figure 1.** Pre-operative photograph in which the previous partial thickness skin graft is appreciated.



**Figure 2.** Pre-operative markings of the LD flap. It can be seen that the skin paddle was drawn very low to allow a tension-free transposition up to the defect in the clavicular area.

We left the thoracolumbar fascia on the back intact to avoid a potential lumbar hernia <sup>10</sup>. The neurovascular pedicle was identified from below and the nerve was not divided. The flap was then rotated through the axilla to the supraclavicular region and the donor area was closed, leaving one suction drain per surgical site. No postoperative complications occurred. Histopathological studies on the surgical specimen identified multiple foci of spindle cell neoplasia with morphological and immunophenotypic characters (CD34+) compatible with the diagnosis already known in the history of the patient of DFSP involving the skin, the subcutaneous tissue and the clavicular fibers of pectoralis major and



**Figure 3.** Intra-operative detail after wide excision including the external clavicular cortical bone, jugular vessels and cervical nerves.

deltoid muscles. The whole mass with overlying skin was excised with adequate safety margins.

## DISCUSSION

The DFSP is a rare mesenchymal tumor that accounts for 1% of soft tissue malignancies and 0, 1% of all other malignancies<sup>11</sup>. It has a strong local invasion and high recurrence rate (10-60%), due to the highly irregular shape and finger protrusions of the tumor. However, it rarely causes regional or distant metastases<sup>12,13</sup>. Although the etiology is unclear, some authors reported that it may develop from trauma, burn or surgery scar tissues<sup>14</sup>. Microscopically, it extends far beyond the assessed clinical margins, spreading in the dermis and subcutaneous tissue<sup>15</sup>. The high incidence of misdiagnosis of this cancer highlights the importance for pathological examination of early skin lesions to clarify any doubt<sup>16,17</sup>. The most common misdiagnoses are sebaceous cysts, keloid, scar, lipoma, dermatofibroma, morphea, neurofibroma, basal cell carcinoma, desmoid tumor, Kaposi sarcoma, nodular fasciitis, and sarcoidosis. Histopathological examination with immunohistochemistry remains the gold standard to

confirm an accurate diagnosis<sup>18,19</sup>. Treatment options include complete surgical excision, Mohs micrographic surgery, radiation, and imatinib mesylate<sup>20-22</sup>. Standard therapeutic approach is wide and deep tumor excision, achieving negative resection margins and simultaneously preserving the uninvolved tissue from resection. Series on outcomes of DFSP, demonstrated that wide local excision with adequate reconstruction can guarantee disease control in nearly 90% of the cases<sup>23</sup>. Other authors suggest the use of Mohs micrographic surgery<sup>24</sup>. Anyhow, reaching negative surgical margins is the primary endpoint of treatment<sup>25</sup>. There is not a consensus on the extension of safe surgical margins to obtain local control. The recurrence rate after surgical treatment with 2-3 cm surgical margins is 0-30%. When 5 cm of free surgical margins are obtained, recurrence rate is reported as 0-5%<sup>26,27</sup>. However, is strongly recommended to remove the tumor with the skin, subcutaneous tissue and the muscle fascia all together<sup>28</sup>. Since primary closure is not always practicable, reconstructive surgery may be required using either skin graft, local flaps or free flaps.

In our clinical case, skin, subcutaneous tissue, muscle and external cortical bone of the left clavicle were resected en-bloc. Negative surgical margins were obtained in the histopathological examinations and tissue defect was reconstructed with a LD myocutaneous flap, using the skin paddle over the muscle to replace the loss of skin substance. An MRI performed 6 months after surgery showed no signs of recurrence and the patient was satisfied with the final aesthetic result (Figs. 4-6). Similar cases are described in literature, using a wide range of reconstructions. Mishra GS et al. reported the case of a 30-year-old Hindu woman with recurrent DFSP of the clavicular region treated with complete excision and reconstruction with Pectoralis Major Myocutaneous flap<sup>29</sup>. The Pectoralis Major myocutaneous flap is a good reconstructive option, but the strong limitation is represented by the impossibility of guaranteeing a skin paddle as large as that of the LD flap. Moreover, the anterior scar result in a worse cosmetic result, particularly in women. LD myocutaneous flap is a valid option since it permits the harvesting of a large muscle flap with a reliable skin paddle<sup>30-32</sup>. It allows us to repair wide defects with exposure of nerve structures and vessels also, obtaining the replacement of the skin envelope and volume<sup>33</sup>. The use of radiotherapy in the treatment of DFSP has been investigated in many studies<sup>34</sup>. It is particularly encouraged if resection is inadequate but there is limited objective data to support its routine use currently<sup>35,36</sup>. However, successful application and recommendations have been reported in very few small series<sup>37</sup>. Imatinib mesylate was designed to treat Philadelphia chromosome positive leukemia (chronic





**Figure 4.** Post-operative result 9 months after surgical procedure: frontal view.



**Figure 6.** Post-operative result 9 months after surgical procedure: lateral view.



**Figure 5.** Post-operative result 9 months after surgical procedure: oblique view.

myelogenous leukemia). The application of imatinib for DFSP has been limited, in cases where surgery is not favorable or in locally advanced cases the use of systemic tyrosine kinas receptor blockers (imatinib mesilat) has hopeful results <sup>23</sup>, but its precise role in DFSP is currently under investigation in several clinical trials <sup>38</sup>.

## CONCLUSIONS

In making the differential diagnosis of the benign lesions of the skin, DFSP must be considered. This awareness will minimize the delay in the diagnosis process and will promote an adequate surgical approach from the outset, avoiding patient's incomplete resection surgery and subsequent re-operations. In our case, the use of pedicled LD myocutaneous flap allowed us to restore a wide loss of substance and to cover noble structures such as jugular vessels and cervical nerves at the same time, with a satisfactory aesthetic result.

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## CONFLICT OF INTEREST STATEMENT

The Authors declare no conflict of interest.

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## Abbreviations

A: conceived and designed the analysis  
D: collected the data  
DT: contributed data or analysis tool  
S: performed the analysis  
W: wrote the paper  
O: other contribution (specify contribution in more detail)

## ETHICAL CONSIDERATION

The research was conducted ethically, with all study procedures being performed in accordance with the requirements of the World Medical Association's Declaration of Helsinki.

Written informed consent was obtained from each participant/patient for study participation and data publication.

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