CASE REPORT

# Re-recurrent Dermatofibrosarcomaprotuberans of the chest wall

Niaz Hussain,[1] Mujtaba Zaki Naveed,[2] Ghulam Haider[3]

## Abstract

Dermatofibrosarcomaprotuberans is a rare, soft tissue tumour with high rate of recurrence. It is locally aggressive, with a low rate of metastasis. We describe the case of a 42 year old man who presented with a re-recurrent, large tumour situated on the anterior chest wall in the sternal region. We did a wide local excision and covered the resulting defect by using bilateral, pectoralis major myocutaneous flaps. Histopathology and immunohistochemical staining findings were consistent with the diagnosis of Dermatofibrosarcoma Protuberance. Post operatively the patient was treated with chemotherapy and radiotherapy.

**Keywords:** Dermatofibrosarcomaprotuberans, Chest wall, Soft tissue tumour.

## Introduction

Dermatofibrosarcomaprotuberans (DFSP) is an uncommon soft tissue tumour that tends to recur. It is a locally aggressive neoplasm with a high rate of recurrence despite rarely metastasising. It most commonly occurs on the trunk (42-72%), followed by the proximal extremities (16-30%) and rarely occurs above the neck (10-16%).[1]
DFSP comprises of 6% of all soft tissue sarcomas and less than 0.1% of all cutaneous malignancies.[2] The incidence rate is estimated to be at 0.8 cases per million persons per year.[2] We describe the case of a 42 year old male that presented with re-recurrent DFSP of the chest wall.

## Case Report

A 42 year old man came to the hospital with a painless mass in the sternal region since 4-5 years. He had a past history of a similar mass twice before, once in 2003 and then in 2008, both times the mass was excised in other centers. A histological biopsy taken in 2008 did not reveal DFSP. The patient had lost both previous surgery and biopsy records. During our physical exam we found the mass to belobulated, non tender, firm, mobile horizontally, 5.5 x 4cm in size, with well defined margins, a shiny surface and a previous surgery scar. There was no cervical or axillary lymphadenopathy and the rest of the physical exam was within normal limits. CT scan report revealed two closely applied oval shaped soft-tissue density enhancing masses located on the anterior aspect of the chest wall at the level of the manubrium. One lobule measured about 5.4 x 3.9cm and the other one measured about 5.3 x 4.5cm with no regional lymphadenopathy. Multiple hypodense areas in the center of these lesions suggestive of necrosis were noted.

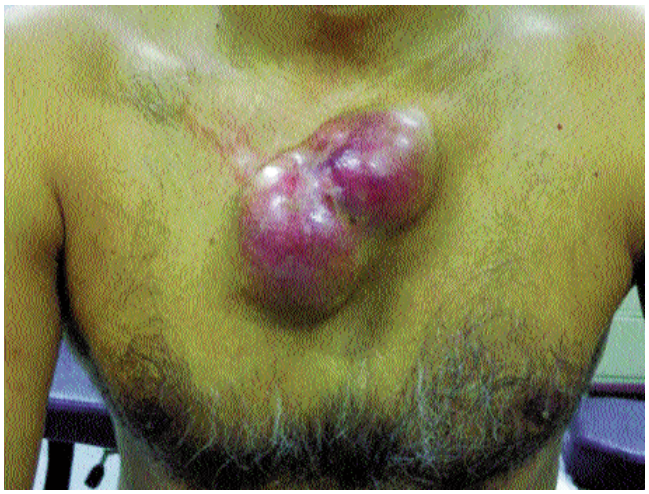

**Figure-1:** Dermatofibrosarcoma Protuberans of the anterior chest wall.

Wide local excision with a grossly 3cm tumour free margin all around was done. Bilateral, pectoralis majormyocutaneous flaps were used to cover the defect resulting from the excision. Biopsy revealed Dermatofibrosarcomaprotuberans. It also showed the lesion to be less than 1cm from the closest peripheral and 0.2cm from the deep margin of resection. Mitotic count ranges were from 6 to 8/10 HPF with few areas showing up to 10 mitosis/10 HPF. Mib-1 (Ki-67) index was approximately 15-20%. Immunohistochemical staining was diffusely positive for CD34 and negative for CD117. Post operatively the upper 4cm part of the wound became infected. This was treated by antibiotics and dressing. Medical and radiation oncologists were consulted, who after discussion in the tumour board decided to give post operative chemotherapy due to brisk mitotic activity and also radiation therapy because of the

[1]Ojha Institute of Chest Disease and Dow University Hospital, [2]Aga Khan University Hospital, [3]Jinnah Postgraduate Medical Center, Karachi, Pakistan.
**Correspondence:** Mujtaba Zaki Naveed. Email: zaki_mnv@hotmail.com

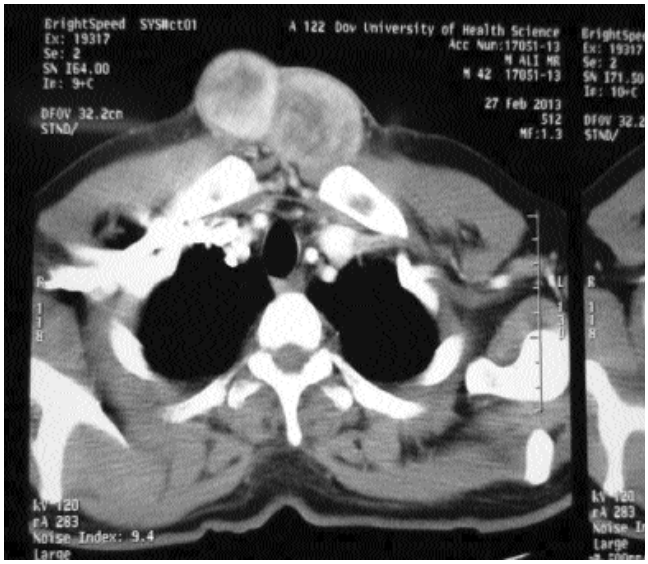


**Figure-2:** Preoperative CT scan of the chest showing the lobulated tumour in the sternal region.

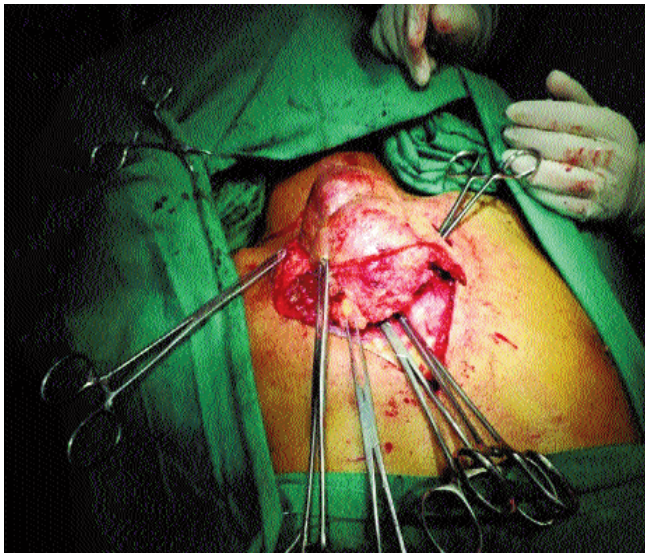

**Figure-3:** Wide local excision of the of the tumour.

close margin. The patient received six cycles of chemotherapy with Ifosfomide, mesna and Doxorubicin, all of which were tolerated well. After completion of chemotherapy, repeated staging workup did not show any evidence of disease at both local and distant sites. Later the patient completed 60 G of radiotherapy to the sternum uneventfully. Currently the patient is in complete remission for 21 months till date and he is in regular follow up every three months.

## Discussion

Chest wall tumours make up less than 5% of thoracic malignancies.[3] Among primary malignant chest wall tumours, soft tissue sarcomas constitute 50% of them. DFSP was initially reported in literature as early as 1890, but it was first described in 1924 by Darier and Ferrand as a distinct cutaneous disease entity called progressive and recurring dermatofibroma.[1] Hoffman termed it DFSP in 1925. The origin of DFSP are still unclear but a genetic defect is present in 95% of cases due to either a translocation of chromosomes 17 and 22, or the formation of a ring.[4] DFSP is slightly more common in males and is seen most often in adult life.[4]

Initially the tumour starts off as a small papule or non-indurated patch, that is often ignored. This progresses to a single, raised, red to blue, firm cutaneous nodule.[5] It further progresses to a painless, cutaneous, multilobulated mass, which is the most often encountered finding on presentation, also in our case. The initial lesion of DFSP can sometimes be mistaken for a keloid due to the similarity in appearance, therefore prompt diagnosis is vital for the treatment of this malignant tumour. Due to its local aggressivevess, it can grow into the surrounding fat, fascia, muscle and even bone. Rarely, distant metastasis is seen in upto 4-6% of patients, with the lungs being the most common site,[1,6] no metastasis was found in our patient. DFSP characteristically has a high rate of recurrence, varying in the literature from 10-80%.[7] Histologically it is identified by a pattern of monomorphous proliferation of cystological bland spindle cells with a visible storiform or whorled architecture.[7] CD34 antigen is a very common finding and its expression aids the diagnosis of DFSP.[8]

Among the various treatment modalities, surgery is the mainstay of treatment by wide excision with a safety margin of 3cm, including the underlying fascia since emphasis is on histological free margins for local control.[9] Conventional chemotherapy is not commonly used to treat DFSP. Radiation therapy (RT) may be recommended if adequate wide excision may result in major cosmetic defect or if the tumour margins are positive. Postoperative adjuvant RT may reduce the risk of recurrence when clear surgical margins are equivocal,[10] hence this was utilized in our case. Mohs surgery, a microscopic surgical technique, is widely gaining acceptance as the treatment of choice due its high oncological effectiveness and maximal tissue salvage. Imatinib, a tyrosine kinase inhibitorcan be used for the treatment of adult patients with unresectable, recurrent, and metastatic DFSP. Overall, the most significant prognostic factor in patients with DFSP has proved to be the extent of surgical resection, especially

the success of the initial surgical excision having a significant effect on the outcome.[8]

## Conclusion

As dermatofibrosarcoma is a locally aggressive neoplasm with high rate of recurrence, wide local excision with at least 3cms tumour free margin at the time of first surgery and oncologists consultation will prevent recurrence.

## References


1. Lemm D, Mugge LO, Mentzel T, Hoffken K. Current treatment options in dermatofibrosarcomaprotuberans. J Cancer Res Clin Oncol 2009; 135: 653-65.
2. Kshirsagar AY, Kanetkar SR, Nikam YP, Vasisth GOP. Recurrent Dermatofibrosarcoma Protuberans Over Anterior Abdominal Wall. J of Cutan Aesthet Surg 2010; 3: 167-9
3. David EA, Marshall MB. Review of Chest Wall Tumors: A Diagnostic, Therapeutic, and Reconstructive Challenge. Semin Plast Surg 2011; 25: 16-24
4. Camara ES, Kasse AN, Sane JC, Bousso A, Thiam B, Sy MH. Dermatofibrosarcoma. Internet J Orthop Surg 2013; 21: 1
5. Kara M, Saray A, Dikmen E, Atasoy P. An Uncommon Soft Tissue Tumour of the Chest Wall : Dermatofibrosarcoma Protuberans. Acta Chir belg 2006; 106: 116-8
6. Mendenhall WM, Zlotecki RA, Scarborough MT. Dermatofibrosarcomaprotuberans. Cancer 2004; 101: 2503-8
7. Brabant B, Revol M, Vergote T, Servant JM, Banzet P. Dermatofibrosarcomaprotuberans of the chest and the shoulder: wide and deep excisions with immediate reconstruction. Plast Reconstr Surg 1993; 92: 459-62.
8. Aiba S, Tabata N, Ishii H, Ootani H, Tagami H. Dermatofibrosarcomaprotuberans is a unique fibrohistiocytic tumour expressing CD34. Br J Dermal 1992; 127: 79-84.
9. Asuquo ME, Umoh MS, Ebughe G. Dermatofibrosarcoma protuberance: Case Reports. Ann Afr Med 2007; 6: 80-3
10. Stivala A, Lombardo GA, Pompili G, Tarico MS, Fraggetta F, Perrotta RE. Dermatofibrosarcoma protuberans: Our experience of 59 cases. Oncol Lett 2012; 4: 1047-55.