ORIGINAL RESEARCH PAPER
Volume - 14 | Issue - 06 | June - 2025 | PRINT ISSN No. 2277 - 8179 | DOI : 10.36106/ijsr
INTERNATIONAL JOURNAL OF SCIENTIFIC RESEARCH

TREATMENT JOURNEY OF A GIANT LUMBAR FIBROSARCOMATOUS DERMATOFIBROSARCOMA PROTUBERANS: A CASE REPORT

General Surgery

Amit Dashputra* Department Of General Of General Surgery, Grant Government And Medial College, Byculla, Mumbai- 400008 *Corresponding Author

Girish Bakhshi Department Of General Of General Surgery, Grant Government And Medial College, Byculla, Mumbai- 400008

Chandrakant Sabale Department Of General Of General Surgery, Grant Government And Medial College, Byculla, Mumbai- 400008

Urvashi Jain Department Of General Of General Surgery, Grant Government And Medial College, Byculla, Mumbai- 400008

Ranjitha Maiyya Department Of General Of General Surgery, Grant Government And Medial College, Byculla, Mumbai- 400008

Akshita Bhalerao Department Of General Of General Surgery, Grant Government And Medial College, Byculla, Mumbai- 400008

Harsh Gajjar Department Of General Of General Surgery, Grant Government And Medial College, Byculla, Mumbai- 400008

ABSTRACT

Dermatofibrosarcoma protuberans (DFSP) is a rare, low-grade soft tissue sarcoma and the fibrosarcomatous variant is known for its aggressive behaviour and local recurrence. Present case details the management of a 40-year-old male with a slow-growing nodular mass on the left lower back, evolving over 3 years, which presented with dull lower back pain, ulceration, and seropurulent discharge. Diagnostic evaluation included MRI showing multiple lobulations and cavitation, with no distant metastasis on PET-CT. Initial wide local excision with a 2 cm margin was performed, but margins were positive, necessitating re-resection. Final histopathology confirmed the fibrosarcomatous variant of DFSP. Subsequent interventions included flap reconstruction and planned radiotherapy. This case underscores the importance of comprehensive surgical therapy for achieving optimal outcomes in aggressive DFSP variants.

KEYWORDS

Dermatofibrosarcoma Protuberans, Fibrosarcomatous Variant, Wide Local Excision, Flap Reconstruction, Radiotherapy

INTRODUCTION

Dermatofibrosarcoma protuberans (DFSP) is a rare soft tissue sarcoma and local recurrence after incomplete resection is common, although distant metastases are rare [1,2]. The fibrosarcomatous variant is more aggressive, making management challenging. This case is unique due to the extensive surgical resection and flap reconstruction needed to address the large defect and achieve negative margins. Current literature emphasises the importance of complete surgical excision and adjuvant therapy, particularly for aggressive variants of DFSP. While acral DFSP are common, tumours over the trunk and abdomen are rare. Herein, we share a case of giant lumbar DFSP and its treatment challenges.

Case Report

The patient was a 40-year-old male who presented with a slow-growing mass on the left lower back. The mass had been progressively enlarging over the past 3 years. The patient reported persistent dull aching pain in the lower back for the past 3 months. Additionally, within the last month, the mass developed ulceration and began discharging seropurulent fluid.

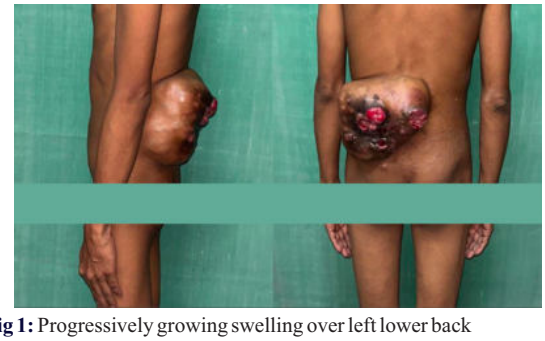

Fig 1: Progressively growing swelling over left lower back

The patient's medical history did not reveal any significant previous conditions or treatments. There was no notable family history of sarcoma or genetic disorders. Psychosocially, the patient expressed concern about the impact of the mass on his daily life and its potential implications for his future health. The patient had not received any prior interventions for this mass. Physical examination revealed a large, firm nodular mass with visible ulceration and seropurulent discharge on the left lower back (Fig. 1).

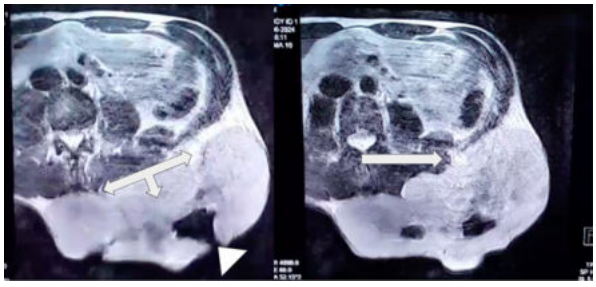

Fig 2: MRI showing (a) Multiple lobulations (arrow) with cavitation (arrowhead) (b) Suspicious loss of fat planes

Diagnostic methods included a four-quadrant biopsy, which revealed a spindle cell lesion of low grade. Imaging studies included MRI, which showed a subcutaneous tumor with lobulations and cavitation and a suspicious loss of fat planes involving adjacent muscle (Fig. 2).

PET-CT confirmed no distant metastasis. Bacterial cultures from the ulcerated area showed no organisms. The differential diagnosis included other spindle cell tumors such as malignant fibrous histiocytoma and leiomyosarcoma. However, the biopsy results and imaging findings were consistent with dermatofibrosarcoma protuberans, specifically the fibrosarcomatous variant. The prognosis was determined by a localized disease without distant metastasis, as confirmed by the PET-CT scan. The local aggressive nature of the fibrosarcomatous variant necessitated thorough surgical intervention and careful follow-up.

The wide local excision was performed with the initial goal of achieving a 2 cm margin (Fig. 3).

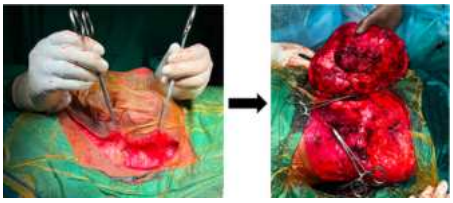

Fig 3: Excision of tumor with deep fascia and 2 cm margin

However due to positive margin at one place on histopathology, the intervention plan was adjusted to include re-resection to ensure negative margins (Fig. 4).

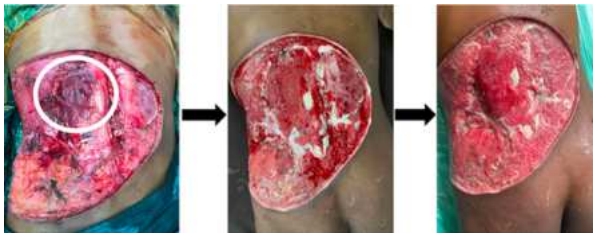

Fig 4: Area of suspicious loss of fat planes (circle) had positive margins. Subsequent re-resection and wound bed prior to flap cover.

The addition of flap reconstruction was necessary to address the large surgical defect created by the excision prior to radiotherapy (Fig. 5).

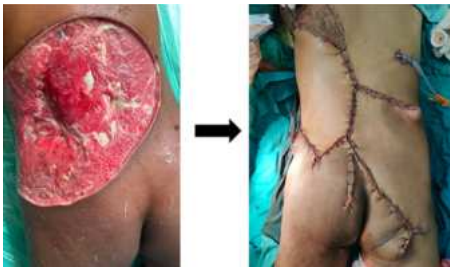

Fig 5: Local advancement and transposition flap coverage of the defect supplemented by split thickness grafting.

The patient's recovery was monitored, and flap coverage was successfully implemented.

The final histopathology confirmed the fibrosarcomatous variant of DFSP with negative margins after re-resection. Histopathology confirmed the diagnosis (Fig. 6). Immunohistochemistry showed positivity for CD34 and negativity for factor XIIIa, with negative stromelysin-3 and CD117

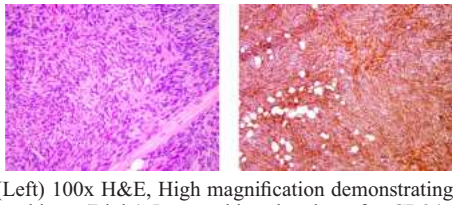

Fig 6: (Left) 100x H&E, High magnification demonstrating nuclear monomorphism. (Right) Immunohistochemistry for CD34 showing diffuse staining

Patient was advised adjuvant radiotherapy, however, he refused in spite of counselling. Hence the patient is kept under close surveillance. Follow-up of 6 months has shown him to be disease and symptom free .

DISCUSSION

Dermatofibrosarcoma Protuberans (DFSP) is a rare, locally aggressive mesenchymal tumour of the skin, representing less than 0.1% of all malignancies and approximately 1% of soft tissue sarcomas, with an annual incidence ranging from 0.8 to 4.5 cases per million people [2,3,4,5,6]. DFSP grows slowly but has a high recurrence rate due to its tendency to infiltrate subcutaneous tissue, fascia, and underlying muscle in a pseudopod-like manner [6,7].

Clinically, DFSP typically presents as an asymptomatic, multinodular, bluish or brownish erythematous plaque that develops over several years, often with a characteristic 'protuberant' appearance [1,6]. It can also appear as an atrophic plaque, sometimes mistaken for morphea [8,9]. The myxoid variant of DFSP was first described by Frierson and Cooper in 1983 [10], with subsequent cases like that of Hong et al. highlighting its rare presentation with prominent myxoid stroma [3]. Tantcheva-Poor et al. identified a vascular histological variant [11]. Most DFSPs occur on the trunk and extremities and are usually benign [3,5], but it is uncommon on the head and neck and extremely rare in the breast [5].

DFSP affects both men and women equally, although a slight male predominance has been noted [12]. It typically arises in individuals between the second and fifth decades of life [5]. There are cases where DFSP develops in previously traumatised areas or following multiple surgeries [13], but present patient had no such history.

DFSP is generally located in the dermis but can sometimes invade subcutaneous fat, displaying either a 'pastry' pattern (with neoplastic cell bands parallel to the epidermis) or a 'honeycomb' pattern (with adipocyte islands bordered by tumour tissue) in about 60% of cases [3,6]. It has a low metastatic potential, with less than 5% likelihood of regional or distant metastases, often limited to the lungs and less frequently to lymph nodes [5,14,15]. Differential diagnoses include recurrent dermatofibroma, hypertrophic scars, keloids, skin manifestations of myofibroblastoma, metaplastic carcinoma, fibromatosis, or other lesions with fusiform cells [5].

Histologically, DFSP is characterised by relatively uniform, densely packed spindle cells with elongated nuclei and minimal cytologic atypia, arranged in a storiform pattern [3]. Nodular lesions exhibit higher nuclear atypia compared to plaque forms. Fibrosarcomatous changes with a fish bone pattern may occasionally be present [5]. Immunohistochemistry typically shows positivity for CD34 (in 84-100% of cases) and vimentin, reflecting the fibroblastic nature of the tumour, with negativity for S-100, HMB45, desmin, and actin [1,6,16,17]. Present case exhibited the classic pattern of positivity for CD34 and negativity for factor XIIIa, with negative stromelysin-3 and Cd117.

Standard treatment involves wide local surgical resection with margins of 2-3 cm, including skin, subcutaneous tissue, and underlying fascia [5,14,18]. Larger surgical margins are associated with reduced local recurrence rates [5]. This procedure can lead to cosmetic and functional impairments. Role of frozen section to guide wide local excisions is controversial. Although the frozen section is preferred for intraoperative negative margins, final histopathology is confirmed after immunohistochemistry which is time consuming. High recurrence rates are linked to factors such as histological subtype, cellularity, tumour size, location on the head and neck, and a high mitotic rate [19,20]. Mohs micrographic surgery is advocated for limited tumors due to its tissue-sparing approach and lower recurrence rates especially in areas with functional loss such as face. [4,21,22,23].

DFSP frequently presents with translocations on chromosomes 17 and 22, leading to overexpression of platelet-derived growth factor, which stimulates tumour growth. Imatinib mesylate, a selective tyrosine kinase inhibitor, targets platelet-derived growth factor receptors and has shown positive effects in about 65% of patients with unresectable, recurrent, or metastatic lesions [1,4,5,24].

Radiation therapy serves as an adjuvant treatment for cases with inadequate surgical margins, aesthetic or functional defects, or positive margins post-resection. It is also used for inoperable macroscopic lesions [1,5]. Postoperative radiotherapy has been associated with cure rates exceeding 85% [25], though it carries risks such as acute or chronic radiodermatitis and potential development of secondary skin tumours [1]. Due to high local recurrence risk, re-resection for positive margins was considered in present case. However, in present case patient refused adjuvant radiotherapy and patient was kept on surveillance.

Due to the tumour's invasive nature, even wide excisions may leave residual tumour, leading to recurrence [1,26,27]. Local recurrences can appear up to 5 years post-treatment, emphasising the need for long-term monitoring and biannual follow-ups [1,5,16,28].

CONCLUSION

The strengths of the approach included comprehensive surgical management with meticulous margin assessment and the use of a

multidisciplinary team. Limitations included the need for re-resection due to initial positive margins, low sensitivity of frozen sections for sarcomas, which prolonged the treatment course.

A comprehensive management is essential for this variant is based on the need for thorough surgical excision. The aggressive nature of the fibrosarcomatous variant necessitates a robust approach to treatment.

The primary lesson is the necessity of a comprehensive treatment strategy for fibrosarcomatous DFSP, which includes wide excision, margin evaluation, flap reconstruction, and adjuvant radiotherapy to optimise outcomes and minimise recurrence.

Disclosure

The patient provided informed consent for the publication of case details and any associated photographic material.

REFERENCES

1. Bhambri S, Desai A, Del Rosso JQ, Mobini N: Dermatofibrosarcoma protuberans: a case report and review of the literature. J Clin Aesthet Dermatol 2008;1:34-6.
2. Gloster HM Jr: Dermatofibrosarcoma protuberans. J Am Acad Dermatol 1996;35(3 pt 1):355-74.
3. Hong YJ, Choi YW, Myung KB, Choi HY: A case of myxoid dermatofibrosarcoma protuberans. Ann Dermatol 2011;23:379-81.
4. Rutkowski P, Wozniak A, Switaj T: Advances in molecular characterization and targeted therapy in dermatofibrosarcoma protuberans. Sarcoma 2011;2011:959132.
5. Dragoumis DM, Katsohi LA, Amplianitis IK, Tsiftsoglou AP: Late local recurrence of dermatofibrosarcoma protuberans in the skin of female breast. World J Surg Oncol 2010;8:48.
6. Pérez OG, Righetti R, Woscoff A, Amante H: Case for diagnosis. Dermatofibrosarcoma protuberans. An Bras Dermatol 2010;85:245-7.
7. Pack GT, Tabah EJ: Dermato-fibrosarcoma protuberans: a report of 39 cases. AMA Arch Surg 1951;62:391-411.
8. Hanabusa M, Kamo R, Harada T, Ishii M: Dermatofibrosarcoma protuberans with atrophic appearance at early stage of the tumor. J Dermatol 2007;34:336-9.
9. Bakry O, Attia A: Atrophic dermatofibrosarcoma protuberans. J Dermatol Case Rep 2012;6:14-7.
10. Frierson HF, Cooper PH: Myxoid variant of dermatofibrosarcoma protuberans. Am J Surg Pathol 1983;7:445-50.
11. Tantcheva-Poor I, Marathovouniotis N, Kutzner H, Mentzel T: Vascular congenital dermatofibrosarcoma protuberans: a new histological variant of dermatofibrosarcoma protuberans. Am J Dermatopathol 2012;34:e46-e49.
12. Asquo ME, Umoh MS, Ebughe G: Dermatofibrosarcoma protuberance: case reports. Ann Afr Med 2007;6:80-3.
13. McKee PH, Calonje JE, Granter S, et al: Pathology of the Skin, with Clinical Correlations, vol 2. Philadelphia, Elsevier-Mosby, 2005.
14. Chang CK, Jacobs IA, Salti GI: Outcomes of surgery for dermatofibrosarcoma protuberans. Eur J Surg Oncol 2004;30:341-5.
15. Laskin WB: Dermatofibrosarcoma protuberans. CA Cancer J Clin 1992;42:116-25.
16. Labonte S, Hanna W, Bandarchi-Chamkhaleh B: A study of CD117 expression in dermatofibrosarcoma protuberans and cellular dermatofibroma. J Cutan Pathol 2007;34:857-60.
17. Adler N, Tsabari C, Sulkes J, Ad-El D, Feinmesser M: Cyclooxygenase-2 expression in dermatofibroma and dermatofibrosarcoma protuberans. J Cutan Pathol 2008;35:532-5.
18. Kimmel Z, Ratner D, Kim JY, Wayne JD, Rademaker AW, Alam M: Peripheral excision margins for dermatofibrosarcoma protuberans: a meta-analysis of spatial data. Ann Surg Oncol 2007;14:2113-20.
19. DuBay D, Cimmino V, Lowe L, Johnson TM, Sondak VK: Low recurrence rate after surgery for dermatofibrosarcoma protuberans: a multidisciplinary approach from a single institution. Cancer 2004;100:1008-16.
20. Goldblum JR, Tuthill RJ: CD34 and factor-XIIIa immunoreactivity in dermatofibrosarcoma protuberans and dermatofibroma. Am J Dermatopathol 1997;19:147-53.
21. Snow SN, Gordon EM, Larson PO, Bagheri MM, Bentz ML, Sable DB: Dermatofibrosarcoma protuberans: a report on 29 patients treated by Mohs micrographic surgery with long-term follow-up and review of the literature. Cancer 2004;101:28-38.
22. Xiang Q, Lu W, He X, Li H: Surgical treatment of dermatofibrosarcoma protuberans using wide local excision combined with Mohs micrographic surgery. Zhongguo Xiu Fu Chong Jian Wai Ke Za Zhi 2011;25:1350-3.
23. Galimberti G, Montaño AP, Kowalczuk A, Ferrario D, Galimberti R: Outcomes in 11 patients with dermatofibrosarcoma protuberans treated with Mohs micrographic surgery. Int J Dermatol 2012;51:89-93.
24. Jain S, Xu R, Prieto VG, Lee P: Molecular classification of soft tissue sarcomas and its clinical applications. Int J Clin Exp Pathol 2010;3:416-28.
25. Mendenhall WM, Zlotecki RA, Scarborough MT: Dermatofibrosarcoma protuberans. Cancer 2004;101:2503-08.
26. Ratner D, Thomas CO, Johnson TM, Sondak VK, Hamilton TA, Nelson BR, Swanson NA, Garcia C, Clark RE, Grande DJ: Mohs micrographic surgery for the treatment of dermatofibrosarcoma protuberans. Results of a multiinstitutional series with an analysis of the extent of microscopic spread. J Am Acad Dermatol 1997;37:600-13.
27. Haycox CL, Odland PB, Olbricht SM, Casey B: Dermatofibrosarcoma protuberans (DFSP): growth characteristics based on tumor modeling and a review of cases treated with Mohs micrographic surgery. Ann Plast Surg 1997;38:246-51.
28. Ruiz-Tovar J, Fernández Guarino M, Reguero Callejas ME, Aguilera Velardo A, Arano Bermejo J, Cabañas Navarro L: Dermatofibrosarcoma protuberans: review of 20-years experience. Clin Transl Oncol 2006;8:606-10.