SAS Journal of Surgery
Abbreviated Key Title: SAS J Surg
ISSN 2454-5104
Journal homepage: https://www.saspublishers.com

OPEN ACCESS

Dermatofibrosarcoma Protuberans of the Back – A Case Report

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DOI: 10.36347/sasjs.2021.v07i10.011 | **Received:** 07.09.2021 | **Accepted:** 12.10.2021 | **Published:** 13.10.2021

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Abstract **Case Report**

Dermatofibrosarcoma protuberans (DFSP) is a rare tumour of the soft tissue tumor involving the dermis, subcutaneous tissue, and in few cases, muscle and fascia. The tumour typically presents as a slowly growing, firm plaque or nodular on the trunk and extremities of young adults. The etiology of dermatofibrosarcoma protuberans has been attributed to a chromosomal translocation t (17;22)(q22;q13) that results in a fusion protein promoting tumour growth through the overproduction of platelet-derived growth factor (PDGF). Diagnosis is made via skin biopsy. Treatment options include complete surgical excision by performing conventional surgery with wide margins (>3 cm) or Mohs micrographic surgery. These lesions are locally aggressive with high rate of recurrence following surgery but the prognosis is excellent when it is treated effectively. These tumours have intermediate malignant potential.

Key words: Dermatofibrosarcoma protuberans, Rare, Clinical diagnosis, Surgery, Recurrence.

INTRODUCTION

This tumour was first described by Darier and Ferrand in 1924 but Hoffmann named it in 1925 [1, 2]. Dermatofibrosarcoma protuberans (DFSP) represents a low-grade soft tissue sarcoma that originates from the dermal layer of the skin. It is a rare tumour which accounts for about 5% of soft tissue sarcoma occurring in all age groups, especially in young adults and affecting both sexes equally. Simon *et al*. in 1997 identified that a chromosomal translocation between chromosome 17 and 22 resulting in the fusion protein COL1A1-PDGFB, which promotes tumour growth through the overproduction of platelet-derived growth factor (PDGF) was responsible for the development of DFSP [3]. The trunk and extremities are the most common sites of involvement, but has been described in other areas like the neck, head, breast, and vulva. It usually presents as a slowly growing superficial soft tissue mass having a good prognosis when treated properly. At times, these lesions become locally aggressive and invade the deep soft tissues. This is a clinical diagnosis, but CT or MRI can be done to assess the extent and depth of involvement [4-6]. The ideal treatment modality for dermatofibrosarcoma protuberans is Mohs micrographic surgery that allows complete margin assessment and tissue preservation. They can also be treated with wide local excision. The chemotherapeutic agent imatinib mesylate is currently used for adults with unresectable, recurrent, or metastatic tumours [7-11].

CASE REPORT

35 year old female presented to us with a large swelling on the back for the past 2 years. It was spontaneous in onset and gradually progressing to the present size. She gives history of occasional pain. There is no history of ulceration or bleeding from the swelling. There was no history of any co-morbid illnesses. On examination, a large swelling of size 15 x 9 x 3cm was located on the left upper back 2cm left of her spine. The swelling was pedunculated and lobulated with stretched and shiny skin on the surface with no

ulceration or bleeding. The swelling was not warm or tender. The underlying muscles were free. There was no regional lymphadenopathy. A clinical diagnosis of dermatofibromasarcoma protuberans was made. (Fig. 1) We planned for excision of the lesion and primary closure of the defect. Under general anaesthesia and prone position, the lesion was excised in layers with fascia with a 3cm margin all around. The muscle was free from tumour infiltration. Haemostasis was secured and the surrounding skin flaps were undermined and raised. The post excisional defect was closed primarily in layers with 2-0 polyglactin and 2-0 nylon sutures. (Fig. 2) Compression dressing was applied. Post-operative period was uneventful with sutures removed on the 14th post-operative day. Histopathology confirmed the lesion as a dermatofibrosarcoma protuberans and the patient is on regular follow-up.

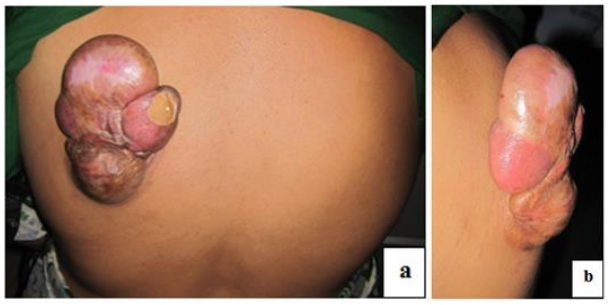

Fig-1: Clinical photograph of the lesion frontal (a) and lateral views (b)

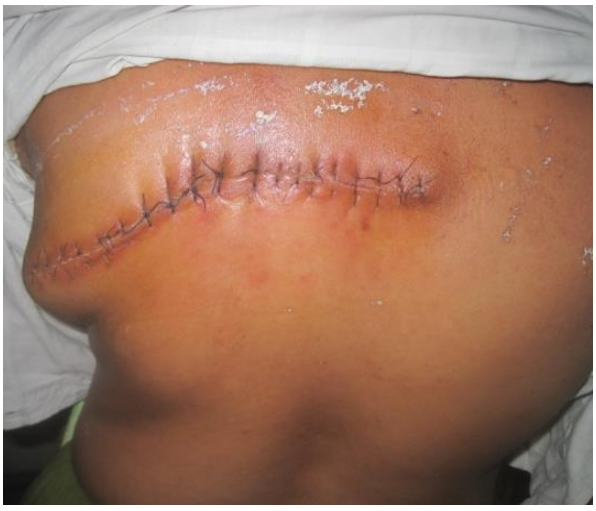

Fig-2: Immediate post-op photograph

DISCUSSION

Dermatofibrosarcoma protuberans usually develops in areas of previous trauma like surgical scars, trauma and burn scars, radiation injury and insect bites [12-16]. The most common site to be affected by DFSP in majority of the cases is the chest and trunk (42–72%). About 16 to 30% of cases are located on the proximal extremities (particularly on the legs) and up to 16% of cases affect the head and neck region [17, 18]. Clinically, they are a slow-growing asymptomatic plaque and presenting to the medical practitioner at a late stage [18]. Martin *et al.* identified three clinical forms of non-protruding DFSP: morphea-like, atrophoderma-like and angioma-like [19]. The most frequent presentation seen in adults is a large plaque presenting multiple nodules on its surface. DFSP is characterized by a low rate of metastasis and an eccentric growth rate, which may determine a high level of local invasion and was found that conventional surgery leads to local recurrence in up to 30% of cases [18]. Kim described seven histological subtypes' namely classic, giant cell fibroblastoma, Bednar tumour, sclerotic, myxoid, atrophic and fibrosarcomatous types, of which 90% of cases are of the classic type [20]. The first immunohistochemical marker identified for DFSP was the CD34 antigen. It is expressed in up to 90% of cases, differentiating DFSP from other fibrohistiocytic tumours [21, 22]. Molecular biology techniques, including reverse-transcriptase polymerase chain reaction and fluorescent *in situ* hybridization, have revealed that DFSP is characterized by supernumerary ring chromosomes or a reciprocal translocation between chromosome 17 and chromosome 22 t(17;22) (q22;q13) [3, 23]. This translocation involves the collagen type 1 α 1 (COL1α1) gene located on chromosome 17 and the PDGFβ gene located on chromosome 22. In DFSP, COL1α1 is highly expressed and acts as an inducer of gene transcription [24]. COL1α1-PDGFβ fusion leads to the transcription of a fully active PDGFβ protein, which triggers mitosis through the activation of the PDGFβ receptor (PDGFβR) via autocrine and paracrine stimulation of its functional ligand [25]. The PDGFβR is composed of three structural domains: an extracellular binding, a transmembrane and a cytoplasmic domain with tyrosine kinase activity. The tyrosine kinase activates an intracellular signaling cascade that affects physiological cell processes, including chemotaxis, proliferation and apoptosis [18]. The treatment of DFSP is primarily complete surgical excision of the lesion. It has been reported that standard surgical resection leads to a local recurrence rate of up to 60%, which is due to the occult spreading of the tentacle-like projections beneath the clinically normal-appearing skin margins [26]. The main challenge in DFSP surgery is to achieve satisfactory local control. To obtain the lowest recurrence rate, two surgical treatments may be performed: wide local excision and Mohs micrographic surgery (MMS). For recurrent and metastatic lesions, molecular targeted therapy with imatinib mesylate may be considered as a suitable alternative or additional treatment option. According to previous studies, it is recommended that surgical excision is performed at least 2–3 cm away from the gross margin. Furthermore, it is important to perform a three dimensional en bloc removal of the tumor, including skin, subcutaneous tissue and fascia. If the underlying bone structures are affected it is necessary to perform a wide resection of the periosteum and bone [17]. The prognosis for patients with completely excised lesions is good [27].

CONCLUSION

Dermatofibrosarcoma protuberans is rare superficial soft tissue tumour which does not require imaging before excision and histological diagnosis. The treatment is surgical removal and the key is to remove the entire lesion with clear margins. Local recurrences are very common when margins are positive. Dermatofibrosarcoma protuberans does not respond well to chemotherapy or radiation. All patients need long-term follow-up with imaging studies as recurrences are common.

REFERENCES

1. Darier, S., Ferrand, M. (1924). *Dermatofibrosarcoma: A progressive and recurring rigid fibrosarcoma of the skin. Ann Dermatol Venereol*, *5*; 545-62.
2. Hoffman, E. (1925). *About the dermatofibrosarcoma protuberans of the skin.* Acta Dermatol Venereol Suppl (Stockh), *43*; 1-28.
3. SIMMON, M. (1997). Deregulation of the platelet-derived growth factor B-chain gene via fusion with collagen gene COL1A1 in dermatofirosarcoma protuberans and giant-cell fibroblastoma. *Nature Genet*, *15*, 95-98.
4. Taylor, H.B., Helwig, E.B. (1962). *Dermatofibrosarcoma protuberans. A study of 115 cases.* Cancer, 15; 717-25.
5. Dimitropoulos, V.A. (2008). *Dermatofibrosarcoma protuberans. Dermatol Ther*, *21*; 428-32.
6. Torreggiani, W. C., Al-Ismail, K., Munk, P. L., Nicolaou, S., O'Connell, J. X., & Knowling, M. A. (2002). Dermatofibrosarcoma protuberans: MR imaging features. *American Journal of Roentgenology*, *178*(4), 989-993.
7. Neff, R., Collins, R., & Backes, F. (2019). Dermatofibrosarcoma protuberans: A rare and devastating tumor of the vulva. *Gynecologic oncology reports*, *28*, 9.
8. Du, K., Li, J., Tang, L., Lin, X., Kong, X., Liao, X., & Wu, J. (2019). Role of postoperative radiotherapy in dermatofibrosarcoma protuberans: a propensity score-matched analysis. *Radiation oncology*, *14*(1), 1-8.
9. Gladdy, R. A., & Wunder, J. S. (2019). Risk-stratified surveillance in dermatofibrosarcoma protuberans: Less is more.
10. Huis in 't Veld, E. A., van Coevorden, F., Grünhagen, D. J., Smith, M. J., van Akkooi, A. C., Wouters, M. W., ... & van Houdt, W. J. (2019). Outcome after surgical treatment of dermatofibrosarcoma protuberans: Is clinical follow-up always indicated?. *Cancer*, *125*(5), 735-741.
11. Parham, D. M. (2018). Fibroblastic and myofibroblastic tumors of children: new genetic entities and new ancillary testing. *F1000Research*, *7*.
12. Mbonde, M. P., Amir, H., & Kitinya, J. N. (1996). Dermatofibrosarcoma protuberans: a clinicopathological study in an African population. *East African medical journal*, *73*(6), 410-413.
13. Bashara, M. E., Jules, K. T., & Potter, G. K. (1992). Dermatofibrosarcoma protuberans: 4 years after local trauma. *The Journal of foot surgery*, *31*(2), 160-165.
14. Tanaka, A., Hatoko, M., Tada, H., Kuwahara, M., Iioka, H., & Niitsuma, K. (2004). Dermatofibrosarcoma protuberans arising from a burn scar of the axilla. *Annals of plastic surgery*, *52*(4), 423-425.
15. Argiris, A., Dardoufas, C., & Aroni, K. (1995). Radiotherapy induced soft tissue sarcoma: an unusual case of a dermatofibrosarcoma protuberans. *Clinical Oncology*, *7*(1), 59-61.
16. Lehmer, L. M., & Ragsdale, B. D. (2011). Digital dermatofibromas-common lesion, uncommon location: a series of 26 cases and review of the literature. *Dermatology online journal*, *17*(8).
17. Lemm, D., Mügge, L. O., Mentzel, T., & Höffken, K. (2009). Current treatment options in dermatofibrosarcoma protuberans. *Journal of cancer research and clinical oncology*, *135*(5), 653-665.
18. Sanmartín, O., Llombart, B., López-Guerrero, J.A. (2007). Dermatofibrosarcoma protuberans. *Actas Dermosifiliogr*, *98*; 77–87.
19. Martin, L., Piette, F., Blanc, P., Mortier, L., Avril, M. F., Delaunay, M. M., & French Group for Cutaneous Oncology. (2005). Clinical variants of the preprotuberant stage of dermatofibrosarcoma protuberans. *British Journal of Dermatology*, *153*(5), 932-936.
20. Kim, G. K. (2011). Status report on the management of dermatofibrosarcoma protuberans: is there a viable role for the use of imatinib mesylate? In which cases may it be therapeutically helpful and in which cases not?. *The Journal of clinical and aesthetic dermatology*, *4*(3), 17.
21. Aiba, S., Tabata, N., Ishii, H., Ootani, H., & Tagami, H. (1992). Dermatofibrosarcoma protuberans is a unique fibrohistiocytic tumour expressing CD34. *British journal of Dermatology*, *127*(2), 79-84.
22. Kutzner, H. (1993). Expression of the human progenitor cell antigen CD34 (HPCA-1) distinguishes dermatofibrosarcoma protuberans from fibrous histiocytoma in formalin-fixed, paraffin-embedded tissue. *Journal of the American Academy of Dermatology*, *28*(4), 613-617.
23. Pedeutour, F., Simon, M. P., Minoletti, F., Sozzi, G., Pierotti, M. A., Hecht, F., & Turc-Carel, C. (1995). Ring 22 chromosomes in dermatofibrosarcoma protuberans are low-level amplifiers of chromosome 17 and 22 sequences. *Cancer research*, *55*(11), 2400-2403.
24. Terrier-Lacombe, M. J., Guillou, L., Maire, G., Terrier, P., Vince, D. R., Somerhausen, N. D. S. A., ... & Coindre, J. M. (2003). Dermatofibrosarcoma

protuberans, giant cell fibroblastoma, and hybrid lesions in children: clinicopathologic comparative analysis of 28 cases with molecular data: a study from the French Federation of Cancer Centers Sarcoma Group. *The American journal of surgical pathology*, *27*(1), 27-39.

25. Shimizu, A., O'Brien, K. P., Sjöblom, T., Pietras, K., Buchdunger, E., Collins, V. P., & Östman, A. (1999). The dermatofibrosarcoma protuberans-associated collagen type Iα1/platelet-derived growth factor (PDGF) B-chain fusion gene generates a transforming protein that is processed to functional PDGF-BB. *Cancer research*, *59*(15), 3719-3723.
26. Chang, C. K., Jacobs, I. A., & Salti, G. I. (2004). Outcomes of surgery for dermatofibrosarcoma protuberans. *European Journal of Surgical Oncology (EJSO)*, *30*(3), 341-345.
27. Xiao, W., Que, Y., Peng, R., Ding, Y., Zhao, J., Wen, X., & Zhang, X. (2018). A favorable outcome of advanced dermatofibrosarcoma protuberans under treatment with sunitinib after imatinib failure. *Onco Targets and therapy*, *11*, 2439.