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DERMATOFIBROSARCOMA PROTUBERANCE; A CASE REPORT

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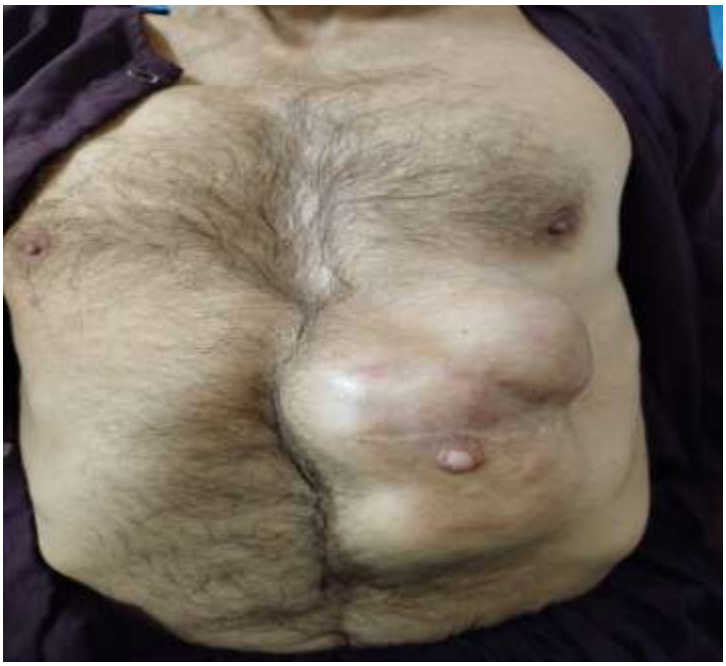
INTRODUCTION

Dermatofibrosarcoma protuberans (DFSP) is an uncommon, slow-progressing tumor originating from the skin's dermal layer (1). Initially identified by Darier and Ferrand in 1924, DFSP was more precisely characterized by Hoffman in 1925 (2). This neoplasm typically appears as a painless, skin-colored plaque that can exhibit dark red or blue tints(3). Over time, DFSP can grow larger, often becoming raised or ulcerated (2). While it is locally invasive (4) and can penetrate nearby tissues including the subcutaneous layer, muscles, tendons, and even bones (5), it rarely metastasizes (4).
DFSP primarily affects adults aged 20 to 50 (6), but it is also reported in children, constituting 6-20% of cases, and can occasionally be congenital (7). There is a slight male predominance. Children with adenosine deaminase-deficient severe combined immunodeficiency (ADA-SCID) are more frequently affected by DFSP (8). The tumor most commonly appears on the trunk (42-72%), followed by the proximal limbs (20-30%), and the head and neck regions (10-16%)(9).DFSP can arise in various sites, including areas with previous surgical scars, burns, trauma, radiation dermatitis, and even around central venous line punctures, vaccination sites, and insect bites (9)(10).

CASE REPORT

A 49 years old man was admitted to surgery ward of HMC Peshawar with a large protuberant mass located in the lower left side of anterior chest wall with extension in abdominal wall. The patient denied any pain, weight loss, fever, night sweats or chills (Figure 1).

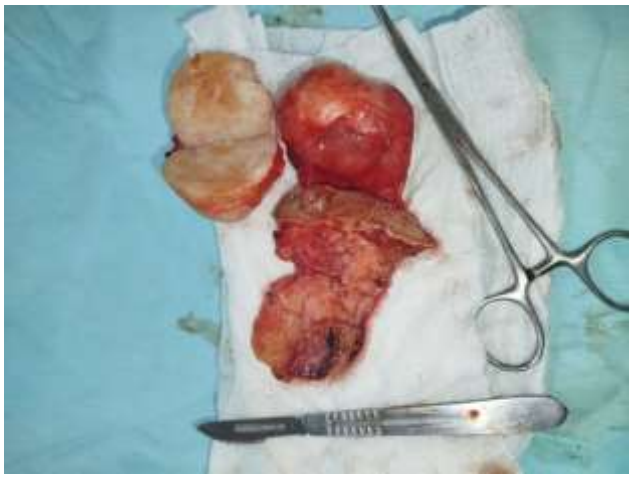


(Figure 1)

On physical examination , an about 8 * 4 cm , soft, painless mass was found with no sign of localized heat or tenderness. A soft tissue ultrasound was performed and a poorly defined heterozygous mass was described at left lower thoracic wall and anterior abdominal wall.

Computed Tomography of abdomen and pelvis (IV contrast and oral contrast given) demonstrated an about 8* 4 cm lobulated soft tissue mass lesion in the lower anterior wall of left sided chest with element of extension to the soft tissue of the upper anterior abdominal wall with no demonstrable evidence of calcification, areas of degeneration or erosion of the underlying ribs.

With the possible diagnosis of soft tissue sarcoma patient underwent wide excision of mass. Upon histological examination, a well circumscribed tumor measuring 9.5 cm* 7.5 cm * 4cm was described (Figure 2). Hematoxylin and eosin stained sections revealed a cellular neoplasm present in dermis. The tumor is arranged in storiform pattern and fascicles. The cells are spindled having hyperchromic nuclei and scant cytoplasm. 9 mitosis/10 HPFs are seen. No necrosis identified. On immunohistochemical stains, the spindle cells showed diffuse positivity for CD 34 antigens. Based on the histological and immunohistochemical findings diagnosis of DFSP was made. The patient's postoperative course was uneventful and was discharged on 3rd postoperative day.On follow up patient was having no complaints and stitches were removed on local anesthesia.



(Figure 2)

DISCUSSION

Dermatofibrosarcoma protuberance (DFSP) is an uncommon, slow-developing malignant tumor arising from fibroblastic mesenchymal cells in the skin. It represents less than 0.1% of all malignancies and about 1% of soft tissue sarcomas (11). DFSP typically presents as a violaceous, pink, or reddish-brown plaque, initially confined to the skin and growing gradually. Over time, it can form multiple nodules that may penetrate deeper into the subcutaneous tissue, fascia, muscles, and occasionally bone. In our observation, there was no invasion into the surrounding muscular or bony structures(9)(10).

In its early stages, DFSP needs to be distinguished from conditions such as lipomas, epidermal cysts, keloids, dermatofibromas, and nodular fasciitis. As the tumor progresses, the differential diagnosis should include pyogenic granuloma, Kaposi sarcoma, and other types of soft tissue sarcomas(12).

Ultrasound imaging of DFSP typically shows these tumors as predominantly hypoechoic or sometimes mixed hyperechoic masses, often with well-defined or irregular margins that can resemble pseudopodia(13). The vascularity of DFSP, which can indicate its malignancy, is variable(14). MRI scans are also not definitive for DFSP, as they may not always differentiate it from other soft tissue sarcomas(15). Consequently, a histological examination remains the gold standard for a conclusive diagnosis.

DFSP is histologically distinguished by its extensive infiltration into the dermis and subcutaneous tissue, while typically leaving the epidermis and skin appendages intact. It proliferates along the existing fibrous septa and penetrates fat lobules, creating a characteristic honeycomb-like appearance. Occasionally, DFSP may appear as an infiltrative subcutaneous lesion(8). The tumor cells show minimal atypia, and mitotic figures are uncommon. In smaller biopsy samples, the superficial portion of the tumor, which may be less cellular, can pose diagnostic challenges. Prominent vascular features and granular cell changes are seldom observed(8). It is crucial for the histopathology report to note the mitotic rate, presence of necrosis, and any fibro sarcomatous transformation, as these factors are associated with more aggressive behavior and reduced overall survival(16).

Immunohistochemically, tumor cells stain for vimentin, CD34, apolipoprotein D, nestin, and may be for EMA. Desmin, S100 protein, FXIIIa, stromelysin III, HMGA1&2, tenascin, D2-40, CD163, and keratins are negative. In myoid nodules, tumor cells stain for SMA. Fibrosarcomatous DFSP may show loss of CD34 positivity and increased expression of TP53. In our case tumour cells tested positive for CD34(8)(16).

The primary treatment for dermatofibrosarcoma protuberans (DFSP) is wide local excision, aiming for clear margins of 3 to 5 cm around the tumor, including the skin, subcutaneous tissue, and underlying fascia(17). If the tumor extends to the bone, the periosteum or a portion of the bone may need to be removed to ensure complete excision with negative margins(18). The likelihood of recurrence is closely linked to the extent of these margins. Studies have shown that using 5 cm margins results in recurrence rates of less than 5%(18).Post-surgical reconstruction may be necessary to address tissue loss, utilizing techniques such as local skin flaps, skin grafts, or myocutaneous flaps(1).

Currently, Mohs micrographic surgery is regarded as the preferred treatment for DFSP(19). This method involves progressive horizontal slicing (5-7 μm thick) of the tissue during resection, with each section immediately examined under a microscope until tumor-free margins are confirmed(9). This approach boasts high local cure rates, ranging from 93% to 100%(20).

For adjuvant treatment of dermatofibrosarcoma protuberans (DFSP), imatinib mesylate, a tyrosine kinase inhibitor, is utilized in managing cases that are unresectable, recurrent, or metastatic. Imatinib works by targeting the tyrosine kinase associated with PDGF and is particularly effective in DFSP patients exhibiting the t(17;22) translocation(3). Radiotherapy is another option, especially in situations where surgical margins are positive or insufficient, or if there is a recurrence. It is also considered when wide excision results in unacceptable functional or cosmetic outcomes(21).

DFSP is known for its high recurrence rate, with most local recurrences occurring within the first three years post-surgery, and approximately 50% appearing within the first year. However,

recurrences can also happen beyond five years post-operatively(22). Therefore, long-term follow-up is essential for these patients.

REFERENCES:

1. Wu, T. , Wu, H. , Tsai, Y. , Luo, T. and Lee, C. (2021) Dermatofibrosarcoma Protuberans of the Neck: A Case Report. *International Journal of Otolaryngology and Head & Neck Surgery*, **10**, 85-91. doi: 10.4236/ijohns.2021.102009.
2. Elafram R, Romdhane MB, Khessairi N, Sghaier M, Annabi H. Dermatofibrosarcoma protuberans of the hallux: A case report with review of the literature. Int J Surg Case Rep. 2022 Jul;96:107325. doi: 10.1016/j.ijscr.2022.107325. Epub 2022 Jun 22. PMID: 35810681; PMCID: PMC9284047.
3. Menon G, Brooks J, Ramsey ML. Dermatofibrosarcoma Protuberans. [Updated 2024 Apr 18]. In: StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; 2024 Jan-. Available from: https://www.ncbi.nlm.nih.gov/books/NBK513305/
4. AbuMoussa S, Roshan MP, Souza FF, Daley D, Rosenberg A, Pretell J, Fullerton N, Subhawong T. Soft Tissue Masses of the Hand: A Review of Clinical Presentation and Imaging Features. Curr Oncol. 2023 Feb 7;30(2):2032-2048. doi: 10.3390/curroncol30020158. PMID: 36826119; PMCID: PMC9955517.
5. Lyu A, Wang Q. Dermatofibrosarcoma protuberans: A clinical analysis. Oncol Lett. 2018 Aug;16(2):1855-1862. doi: 10.3892/ol.2018.8802. Epub 2018 May 24. PMID: 30008876; PMCID: PMC6036409.
6. Al Laham O, Atia F, Shaheen J, Alkhoury L, Ibrahim D, Alchikh Omar M. A unique case of Dermatofibrosarcoma Protuberans arising from an inguinal hernial repair scar in a Middle Eastern male - A Case Report. Int J Surg Case Rep. 2022 Jul;96:107334. doi: 10.1016/j.ijscr.2022.107334. Epub 2022 Jun 22. PMID: 35777334; PMCID: PMC9284050.
7. Paramythiotis D, Stavrou G, Panagiotou D, Petrakis G, Michalopoulos A. Dermatofibrosarcoma protuberans: a case report and review of the literature. Hippokratia. 2016 Jan-Mar;20(1):80-83. PMID: 27895450; PMCID: PMC5074405.
8. Kesserwan C, Sokolic R, Cowen EW, Garabedian E, Heselmeyer-Haddad K, Lee CC, Pittaluga S, Ortiz C, Baird K, Lopez-Terrada D, Bridge J, Wayne AS, Candotti F. Multicentric dermatofibrosarcoma protuberans in patients with adenosine deaminase-deficient severe combined immune deficiency. J Allergy Clin Immunol. 2012 Mar;129(3):762-769.e1. doi: 10.1016/j.jaci.2011.10.028. Epub 2011 Dec 6. PMID: 22153773; PMCID: PMC3294021.
9. Allen A, Ahn C, Sangüeza OP. Dermatofibrosarcoma Protuberans. Dermatol Clin. 2019 Oct;37(4):483-488
10. Thway K, Noujaim J, Jones RL, Fisher C. Dermatofibrosarcoma protuberans: pathology, genetics, and potential therapeutic strategies. Ann Diagn Pathol. 2016 Dec;25:64-71
11. Effiom OA, Olojede ACO, Akinde OR, Olawuyi AB, Amoo AT, Arotiba GT. Dermatofibrosarcoma protuberans: clinicopathologic presentation in Nigerians. Pan Afr Med J. 2018 Sep 12;31:25. doi: 10.11604/pamj.2018.31.25.13665. PMID: 30918552; PMCID: PMC6430858.
12. Pathak B, Maimoon S. Dermatofibrosarcoma Protuberans Over Right Lumbar Region: A Case Report. Cureus. 2023 Jan 1;15(1):e33208. doi: 10.7759/cureus.33208. PMID: 36733580; PMCID: PMC9887664.
13. Gong X, Li J, Ding A, Chen J, Tao X, Xiong P, Rao Y, Liu Y, Sun Q. Multimodal ultrasound for preoperative evaluation of dermatofibrosarcoma protuberans: a series of 40 cases. BMC Cancer. 2022 Nov 5;22(1):1137. doi: 10.1186/s12885-022-10211-4. PMID: 36335314; PMCID: PMC9637320.
14. Hao X, Billings SD, Wu F, Stultz TW, Procop GW, Mirkin G, Vidimos AT. Dermatofibrosarcoma Protuberans: Update on the Diagnosis and Treatment. J Clin Med. 2020 Jun 5;9(6):1752. doi: 10.3390/jcm9061752. PMID: 32516921; PMCID: PMC7355835.

15. Al Laham O, Atia F, Shaheen J, Alkhoury L, Ibrahim D, Alchikh Omar M. A unique case of Dermatofibrosarcoma Protuberans arising from an inguinal hernial repair scar in a Middle Eastern male - A Case Report. Int J Surg Case Rep. 2022 Jul;96:107334. doi: 10.1016/j.ijscr.2022.107334. Epub 2022 Jun 22. PMID: 35777334; PMCID: PMC9284050.
16. Silvia Stacchiotti, Annalisa Astolfi, Alessandro Gronchi, Andrea Fontana, Maria A. Pantaleo, Tiziana Negri, Monica Brenca, Marcella Tazzari, Milena Urbini, Valentina Indio, Chiara Colombo, Stefano Radaelli, Silvia Brich, Angelo P. Dei Tos, Paolo G. Casali, Chiara Castelli, Gian Paolo Dagrada, Silvana Pilotti, Roberta Maestro; Evolution of Dermatofibrosarcoma Protuberans to DFSP-Derived Fibrosarcoma: An Event Marked by Epithelial–Mesenchymal Transition–like Process and 22q Loss. Mol Cancer Res 1 September 2016; 14 (9): 820–829. https://doi.org/10.1158/1541-7786.MCR-16-0068
17. Hamid R, Hafeez A, Darzi AM, Zaroo I, Owais H, Akhter A. Dermatofibrosarcoma protuberans: Role of wide local excision. South Asian J Cancer. 2013 Oct;2(4):232-8. doi: 10.4103/2278-330X.119926. PMID: 24455646; PMCID: PMC3889049.
18. Sambri A, Caldari E, Fiore M, Zucchini R, Giannini C, Pirini MG, Spinnato P, Cappelli A, Donati DM, De Paolis M. Margin Assessment in Soft Tissue Sarcomas: Review of the Literature. Cancers (Basel). 2021 Apr 2;13(7):1687. doi: 10.3390/cancers13071687. PMID: 33918457; PMCID: PMC8038240.
19. Saporito F, Charalampoudis P, Kinney M, Knox S. Operative management of dermatofibrosarcoma protuberans of the breast. Proc (Bayl Univ Med Cent). 2017 Jan;30(1):128-129. doi: 10.1080/08998280.2017.11929558. PMID: 28154469; PMCID: PMC5287695.
20. Kong K, Rowlands CJ, Varma S, Perkins W, Leach IH, Koloydenko AA, Williams HC, Notingher I. Diagnosis of tumors during tissue-conserving surgery with integrated autofluorescence and Raman scattering microscopy. Proc Natl Acad Sci U S A. 2013 Sep 17;110(38):15189-94. doi: 10.1073/pnas.1311289110. Epub 2013 Sep 3. PMID: 24003124; PMCID: PMC3780864.
21. Muto P, Pastore F. Radiotherapy in the Adjuvant and Advanced Setting of CSCC. Dermatol Pract Concept. 2021 Oct 1;11(Suppl 2):e2021168S. doi: 10.5826/dpc.11S2a168S. PMID: 34877076; PMCID: PMC8609952.
22. Liang CA, Jambusaria-Pahlajani A, Karia PS, Elenitsas R, Zhang PD, Schmults CD. A systematic review of outcome data for dermatofibrosarcoma protuberans with and without fibrosarcomatous change. J Am Acad Dermatol. 2014;71:781–786.