Journal of Neonatal Surgery
ISSN(Online): 2226-0439
Vol.15 , Issue 1s (2026)
https://www.jneonatalsurg.com

OPEN ACCESS

From Diagnosis to Treatment: Dermatofibrosarcoma Protuberance – A Case Report

Samah Dehda[1], Samir Derouiche*[2,3], Ouahab Abdelmalek[1], Sahli Allel[1], Benazza Ali[1],Yahla Fatiha[1], Mohammed Kerkoubi[4]

[1]Department of General surgery, Faculty of medicine, university of Batna 2- Algeria

[2]Departement of Medicine, Faculty of medicine, university of El-Oued –Algeria

[3]Laboratory of Biodiversity and Application of Biotechnology in the Agricultural Field, Faculty of Natural Sciences and Life, University of El-Oued, El-Oued 39000, Algeria.

[4]Department of Chemistry, University Ouargla, Algeria

Corresponding Author:

Samir Derouiche

Cite this paper as: Samah Dehda, Samir Derouiche, Ouahab Abdelmalek, Sahli Allel, Benazza Ali,Yahla Fatiha, Mohammed Kerkoubi (2026) From Diagnosis to Treatment: Dermatofibrosarcoma Protuberance – A Case Report. Journal of Neonatal Surgery, 15, (1s) 162-165

ABSTRACT

Dermatofibrosarcoma protuberans (DFSP), also known as Darier–Ferrand dermatofibrosarcoma, is a rare soft tissue neoplasm and the most common type of cutaneous sarcoma. It is characterized by slow growth and a low metastatic potential, although it presents a high risk of local recurrence, mainly related to the quality of surgical excision. The standard treatment is wide local excision performed by an experienced multidisciplinary team. In locally advanced, unresectable, or metastatic forms, radiotherapy and targeted therapy with imatinib may be considered. We report the case of a 27-year-old man who underwent surgical treatment for dermatofibrosarcoma protuberans located in the left hypochondriac region

Keywords: Dermatofibrosarcoma protuberans, cutaneous sarcoma, wide local excision

INTRODUCTION

Dermatofibrosarcoma protuberans (DFSP), also known as Darier-Ferrand tumour, is a rare, locally aggressive soft tissue neoplasm of dermal origin, representing the most common subtype of cutaneous sarcoma [1]. It has an infiltrative growth pattern with slow growth and low metastatic potential, but high incidence of local recurrence, when incompletely excised [2]. DFSP is most common in young and middle-aged adults, usually involving the trunk and proximal extremities [3]. Clinically it manifests as an indurated plaque or multinodular mass that slowly enlarges and may ultimately become protuberant [4]. Histologically DFSP is characterised by a proliferation of monomorphic spindle-shaped cells infiltrating the dermis and subcutaneous tissue in a storiform or cartwheel pattern [5]. Immunohistochemical expression of CD34 is usually present in tumour cells and this aids in the differentiation of DFSP from other fibrohistiocytic tumours [6]. Cytogenetically, most of them are associated with the t(17;22)(q22;q13) translocation resulting in the COL1A1–PDGFB fusion gene that accounts for the sensitivity of advanced forms to the targeted therapy with imatinib [7].

Wide local excision or Mohs micrographic surgery with negative margins is the standard treatment for localised disease [8]. In cases of locally advanced unresectable recurrent or metastatic disease, radiotherapy and targeted therapy may be considered [9]. The delayed diagnosis and poor surgical management of DFSP, a low-grade sarcoma, can lead to multiple recurrences and significant morbidity, emphasising the importance of early recognition and appropriate treatment [10].

Case Report

We report a 27 year old man, with no significant medical history, who has 04 nodules in the left hypochondrium, over a period of 03 months, followed by a dermatologist. The physical examination revealed a patient is in good health, present in left hypochondriac region, 04 purplish pink, indurate, fixed nodules, two measured 02cm and two measured 01cm, painless. Patient was followed by a dermatologist who has prescribed a medical treatment; as result of no improvement and also nodules' size increase, a biopsy was performed, which revealed: a cutaneous fibro-histiocytic mesenchymal proliferation, and on immune histochemical stains revealed CD34 antigen positivity, the DFSP was confirmed. The patient was referred to our surgical department, performed a wide resection with safety margins of 03 cm and wound closure without tension (figure 1). On the immediate postoperative, there were no side effects

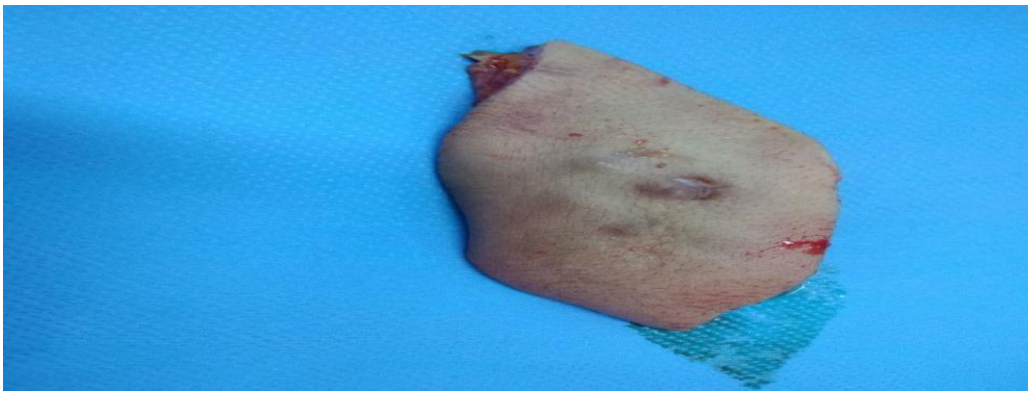

Figure 1. Surgical specimen of tumour with resection margins of 03 cm

On day 05 post-operative; the patient presented with an 8 cm subcutaneous haematoma; treated by its pancture and medical treatment with antibiotic and NSAIDs (figure 2).

Figure 2. The wound condition on the twentieth day after surgery

The histopathology report; confirmed the DFSP diagnostic; the dermis and the hypodermis were infiltrated, also revealed that all the surgical margins are free. Every six months for at least five years; it is planned that the patient is being clinically evaluated.

DISCUSSION

Le Dermatofibrosarcome is a tumor of soft tissues of the dermis, fibroblastic origin, rare, 0,8 to 5 cases/millions inhabitants per year [11]. DFSP is the most common type of cutaneous sarcoma , about 0,1% of malignant cutaneous tumor. Characterized by slow evolution, and it is a locally aggressive and rarely metastatic occurs around 3 %, it is an intermediate malignancy tumor [12, 13]; recurrence rate is high about 20 à 40 %. DFSP is diagnosed between 30 and 50 years old, a slight male predominance is reported. Less than 6% of DFSP are rare congenital and pediatric forms [14,15]. Clinically, DFSP are presented as cutaneous and subcutaneous lesions, and it can spread further deep down, as a mobile, fixed, an indurated, nodules or plaques red, violaceous, bluish, regular or not with a different sizes [16]. The predilection Sites of DFSPs include the trunk (40 à 50 % des cas), the extremities (30 à 40 %), head and neck (10– 15 %) [11, 14]. Fibrosarcomatous is the multi nodular form due to the late evolution [16], and it presents a higher rate of metastasis. the positive diagnosis of DFPS based on histopathology and immunohistochimical marker CD34 antigen that expressed by the tumor. A reciprocal translocation results in a fusion of the *COL1A1* and *PDGF*-β genes in 90% DFSP [17]. The MRI imaging is consider as the choice exam for the evolution of DFSP, also MRI assessed the depth and local extend. The main DFSP treatment is surgery, resection tumor with free margin from 01cm to 05cm [18,19], Mohs micrographic surgical technique; which consists of an excision with immediate histological examination of margins , limiting the parietal sacrifice [20].DFSP is sensitive to radiation and chemoresistant tumor. The radiotherapie may be considered after R1 resection without second surgery, also recurrence or inoperable tumor [18]. Imatinib is a targeted therapy for dermatofibrosarcoma protuberans (DFSP) that inhibits PDGFb-R and shows a response in approximately 50%. [19]. Imatinib indicated for patients with inoperable, locally advanced recurrent or

metastatic DFSP [21]. DFSPs characterized by high local recurrence rates, for that, it is recommended that every six months for at least five years, patients are clinically evaluated for the primary tumor, and every three months for two years, there after every six months for at least three years by clinical examination and imaging studies for lymph node metastases [22]. DFSP is tumor with good carcinological prognosis , overall survive about 92 % at 05 years, which is related tumor size,its local characteristics, evolution, et the quality of management [23].

CONCLUSION

DFSP is rare tumor, locally aggressive, its management is mainly surgical, Requires rigorous surveillance due to high recurrence rate. DFSP is rarely metastasizing tumor for that it is considered as good prognosis.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given his consent for his images and other clinical information to be reported in the journal. The patient understands that the name and initials will not be published and due efforts will be made to conceal the identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest

REFERENCES

1. Menon G, Brooks J, Ramsey ML. Dermatofibrosarcoma Protuberans. [Updated 2024 Apr 18]. In: StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; 2026 Jan-. Available from: https://www.ncbi.nlm.nih.gov/books/NBK513305/
2. Deffner F, Magradze G, Melzer K, et al. Impact of histological and clinical parameters on resection status and recurrence probability in head and neck basal cell carcinomas. Front Oncol. 2025;15:1679804.
3. Oikonomidi C, Lagogianni E, Filippou D, Chrysikos D. Dermatofibrosarcoma Protuberans: A Rare Anatomical Location of Dermal Sarcoma. Cureus. 2025;17(10):e93809.
4. Eason CR, Stovall C, Corley SB. Ulcerated indurated plaques on the upper extremities. JAAD Case Rep. 2019;5(9):749-751.
5. Nimodia D, Dudhe S, Parihar PH, Patil R, Kotla R, Choudhary A. From subtle to striking: A rare case of dermatofibroma protuberans and its clinical journey. Radiol Case Rep. 2025;20(5):2557-2561. Published 2025 Mar 8. doi:10.1016/j.radcr.2025.02.045
6. Petrovic V, Petrovic A, Nikolic IR, et al. Immunohistochemical Pattern of CD34 Distribution in Different Types of Basal Cell Carcinoma and in Peritumoral Skin. Medicina (Kaunas). 2026;62(1):158. Published 2026 Jan 13. doi:10.3390/medicina62010158
7. Das S, Ahlawat S. Beyond COL1A1::PDGFB: Rare fusions and their clinical implications in dermatofibrosarcoma protuberans. World J Clin Cases. 2025;13(33):113167. doi:10.12998/wjcc.v13.i33.113167
8. Carrasquillo OY, Cancel-Artau KJ, Ramos-Rodriguez AJ, Cruzval-O'Reilly E, Merritt BG. Mohs Micrographic Surgery Versus Wide Local Excision in the Treatment of Merkel Cell Carcinoma: A Systematic Review. Dermatol Surg. 2022 Feb 1;48(2):176-180. doi: 10.1097/DSS.0000000000003331.
9. Wang X, Meng J, Zhang X, et al. The Role of Radiotherapy for Patients with Unresectable Locally Advanced Breast Cancer following Neoadjuvant Systemic Therapy. J Oncol. 2023;2023:5101078. Published 2023 Feb 17. doi:10.1155/2023/5101078
10. Jozwik M, Bednarczuk K, Osierda Z. Dermatofibrosarcoma Protuberans: An Updated Review of the Literature. *Cancers (Basel)*. 2024;16(18):3124. Published 2024 Sep 11. doi:10.3390/cancers16183124
11. 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.
12. Llombart B, Serra C, Requena C, Alsina M, Morgado-Carrasco D, Través V, Sanmartín O. Guidelines for Diagnosis and Treatment of Cutaneous Sarcomas: Dermatofibrosarcoma Protuberans. Actas Dermosifiliogr (Engl Ed). 2018 Dec;109(10):868-877. English, Spanish. doi: 10.1016/j.ad.2018.05.006. Epub 2018 Jul 4. PMID: 30539729.

13. Bilger G, Nicolaï M, Perez M, Simon, E.. Cas clinique: reconstruction par abdominoplastie inversée et transposition ombilicale d'un dermatofibrosarcome épigastrique. In Annales de Chirurgie Plastique Esthétique. Elsevier Masson. 2021 ; 66(3) : 261-267.
14. Penel N, El Bedoui S, Robin Y. M, Decanter G. Dermatofibrosarcome: prise en charge. Bulletin du Cancer. (2018 ; 105(11) : 1094-1101.
15. Grizzetti L, Gaide O, Kuonen F.. Dermatofibrosarcoma protuberans: aspects clinico-pathologiques et prise en charge [Dermatofibrosarcoma protuberans: clinico-pathological aspects and management]. 2020.
16. Osché M, Mitcov M, Lipsker D, Cribier B. Étude anatomoclinique et immunohistochimique de 8 cas de dermatomyofibromes [Anatomoclinical and immunohistochemical study of 8 cases of dermatomyofibroma]. Ann Dermatol Venereol. 2020 Nov;147(11):721-728.
17. Marcoval J, Moreno-Vílchez C, Torrecilla-Vall-Llosera C, Muntaner-Virgili C, Pérez Sidelnikova D, Sanjuán X, Penín RM. Dermatofibrosarcoma Protuberans: A Study of 148 Patients. Dermatology. 2024;240(3):487-493. doi: 10.1159/000536172. Epub 2024 Jan 16. PMID: 38228098; PMCID: PMC11168446.
18. Ugurel S, Kortmann RD, Mohr P, Mentzel T, Garbe C, Breuninger H, Bauer S, Grabbe S. S1 guidelines for dermatofibrosarcoma protuberans (DFSP) - update 2018. J Dtsch Dermatol Ges. 2019 Jun;17(6):663-668. doi: 10.1111/ddg.13849. Epub 2019 May 22. PMID: 31115967.
19. Diagnosis and treatment of dermatofibrosarcoma protuberans. European consensus-based interdisciplinary guideline.
20. Granier G, Agrici V, Habib F, Meunier L, Moustey C, Marty-Double C. La chirurgie micrographique de Mohs dans la prise en charge des carcinomes basocellulaires: Application au CHU de Nîmes. In Annales de Pathologie. Elsevier Masson. 2007; 27(2) : 74-79).
21. Navarrete-Dechent C, Mori S, Barker CA, Dickson MA, Nehal KS. Imatinib Treatment for Locally Advanced or Metastatic Dermatofibrosarcoma Protuberans: A Systematic Review. JAMA Dermatol. 2019 Mar 1;155(3):361-369. doi: 10.1001/jamadermatol.2018.4940.
22. Fields RC, Hameed M, Qin LX, et al. Dermatofibrosarcoma protuberans (DFSP): predictors of recurrence and the use of systemic therapy. Ann Surg Oncol. 2011;18(2):328-336. doi:10.1245/s10434-010-1316-
23. Remiszewski P, Pisklak A, Filipek K, et al. Dermatofibrosarcoma Protuberans (DFSP): Current Treatments and Clinical Trials. Curr Treat Options Oncol. 2025;26(11):967-989. doi:10.1007/s11864-025-01348-y.