Dermatofibrosarcoma Protuberans of the Scalp: Case Report

Dermatofibrossarcoma protuberante no couro cabeludo: Relato de caso

Jefferson Lessa Soares de Macedo[1] Jovita Maria Rafael Batista de Albuquerque Espíndola[1] Thamara de Oliveira Vasconcelos[1] Luna D'Angelis Barbosa de Albuquerque[1] Sarah Pereira Lima Xavier da Silva[1] Luis Felipe Rosa de Macedo[2] Cecilia Rosa de Macedo[3] Simone Correa Rosa[1]

[1] Hospital Regional da Asa Norte, Plastic Surgery Unit, Brasília, DF, Brazil
[2] Universidade Católica de Brasília, Undergraduate Course in Medicine, Brasília, DF, Brazil
[3] Centro Universitário de Brasília (UniCEUB), Undergraduate Course in Medicine, Brasília, DF, Brazil

Address for correspondence Jefferson Lessa Soares de Macedo, Hospital Regional da Asa Norte, Unidade de Cirurgia Plástica, Brasília, DF, 70710-100, Brazil (e-mail: jlsmacedo@yahoo.com.br; coremehran@gmail.com).

Rev Bras Cir Plást 2025;40:s00451807279.

Abstract

Dermatofibrosarcoma protuberans is a rare cutaneous tumor of low malignant grade, with slow, infiltrative growth and a strong tendency towards local recurrence after surgical excision. The most frequently affected anatomic site is the thorax (65%), and the tumor rarely affects the scalp (5%). Its high recurrence rate correlates with poor surgical management, as lesions may be confused with dermatofibroma or keloid. We herein report a case of scalp dermatofibrosarcoma protuberans posing particular challenges due to the proximity to the periosteum, calvaria, dura mater, and brain; the resulting defects often require extensive reconstructive procedures. This case confirms the importance of the accurate diagnosis of the primary lesion and the need for aggressive surgical treatment (excision of 2–3 cm of the lateral margins) to lower the incidence of local relapse.

Keywords

- dermatofibrosarcoma
- head and neck neoplasms
- scalp
- surgery
- plastic surgery
- tissue expansion devices

Resumo

Dermatofibrossarcoma protuberante é um tumor cutâneo raro, de baixo grau de malignidade, caracterizado por um padrão de crescimento lento e infiltrativo e grande tendência de recorrência local após excisão cirúrgica. A localização anatômica mais comum é o tórax (65%), e o tumor raramente localiza-se no couro cabeludo (5%). Sua alta taxa de recorrência está associada ao manejo cirúrgico inadequado pelo fato de as lesões serem confundidas com dermatofibroma ou queloides. Relatamos aqui um caso de dermatofibrossarcoma protuberante do couro cabeludo que representa um desafio particular devido à proximidade do tumor com o periósteo, a calota craniana, a dura-máter e o cébebro. Os defeitos resultantes geralmente necessitam de procedimentos reconstrutivos extensos. Este caso confirma a importância do diagnóstico primário da lesão e a necessidade do tratamento cirúrgico agressivo (excisão de 2–3 cm de margens laterais) para diminuir a incidência de recorrência local.

Palavras-chave

- cirurgia plástica
- couro cabeludo
- dermatofibrossarcoma
- dispositivos para expansão de tecidos
- neoplasias de cabeça e pescoço

received
September 21, 2024
accepted
February 6, 2025

DOI https://doi.org/10.1055/s-0045-1807279.
ISSN 2177-1235.

© 2025. The Author(s).
This is an open access article published by Thieme under the terms of the Creative Commons Attribution 4.0 International License, permitting copying and reproduction so long as the original work is given appropriate credit (https://creativecommons.org/licenses/by/4.0/)

Thieme Revinter Publicações Ltda., Rua Rego Freitas, 175, loja 1, República, São Paulo, SP, CEP 01220-010, Brazil

Introduction

Dermatofibrosarcoma protuberans (DFSP) is an uncommon, locally invasive cutaneous sarcoma with an indolent clinical course and an annual incidence of 4.1 cases per 1 million people in the United States.[1] It typically occurs in middle-aged adults, with a slight predominance in men, and accounts for 1% of soft tissue sarcomas. Its preferred locations are the trunk (40–50%) and the extremities (30–40%). The incidence in the head and neck ranges from 10 to 15%, and 5% of the cases occur in the scalp. The main characteristic of this tumor type is its high local recurrence rate after surgical excision, especially for head and neck neoplasms.[2]

We herein report a case of DFSP whose diagnosis was confused with dermatofibroma despite tumor recurrence after two resections. With the evolution in cutaneous oncology, surgeons have been increasingly required to diagnose and provide therapeutic guidance for less frequent tumors.

The Research Ethics Committee of Fundação de Ensino e Pesquisa em Ciências da Saúde/Secretaria de Estado de Saúde do Distrito Federal (Fepecs/SES-DF) approved the current study under CAAE 48838921.2.0000.5553/opinion number 5.784.230. The study followed all ethical aspects from Resolution 466/2012 of the Brazilian National Health Council (Conselho Nacional de Saúde, CNS, in Portuguese).

Case Report

A 25-year-old female, white, single patient from the town of Paracatu, state of Minas Gerais, went to the Plastic Surgery Outpatient Clinic of our hospital with a scalp tumor that had been present for 2 years. The neoplasm had undergone two excisions but recurred. On both occasions, the histopathological diagnosis was of dermatofibroma. After the last procedure 6 months ago, the lesion had grown rapidly and reached 12 cm in diameter. The patient was in good general condition, and palpation did not reveal lymphadenomegaly or visceromegaly. A computed tomography scan of the head showed the scalp tumor did not infiltrate the underlying skull. Magnetic resonance imaging demonstrated the soft tissue tumor had well-defined borders. Chest radiography was negative for the disease. Finally, the patient underwent bone scintigraphy with technetium-99m (^{99m}Tc), which ruled out bone involvement. We performed an incisional biopsy under local anesthesia, and the immunohistochemical analysis diagnosed dermatofibrosarcoma protuberans. The patient underwent wide local excision in the subgaleal plane under general anesthesia, with a 3-cm macroscopic free margin. We covered the defect with a partial skin graft from the external aspect of the left thigh. Subsequently, the patient underwent two surgeries for skin expander placement to cover the alopecic area (►**Fig. 1A–E**). Since then, she has remained free of disease recurrence for 12 years.

Discussion

Dermatofibrosarcoma protuberans (DFSP) is a low-grade superficial sarcoma of dermal origin. Its histological margins are usually well beyond the macroscopic margins, as the tumor spreads horizontally. Even apparently small tumors may present distant projections interspersed in fat lobules, explaining the high local recurrence rate. Ten to 20% of the cases present a history of trauma as a triggering factor. However, there are several reports of DFSP developing in surgical scars, burns, or vaccination (bacillus Calmette-Guérin, BCG) scars, as well as rapid growth during pregnancy, as the tumor has progesterone receptors. The most common skin sarcoma, approximately 5% of cases of DFSP affect the scalp.[1,2]

Microscopically, the tumor consists of atypical spindle cells in a dermal fibrotic stroma involving the subcutaneous tissue and organized in irregular, intertwined fascicles,

Fig. 1 (**A**) Dermatofibrosarcoma protuberans measuring 12 cm in diameter on the scalp. (**B**) Four months after dermatofibrosarcoma excision and partial skin grafting over the intact periosteum. (**C**) Rectangular expanders implanted for scalp expansion. (**D**) Postoperative aspect 1 year after coverage of the alopecic area (anterior view). (**E**) Postoperative aspect 1 year after coverage of the alopecic area (lateral view).

resulting in a storiform or cartwheel pattern (►**Fig. 2**). Mitotic activity is mild, and nuclear pleomorphism is minimal, explaining the low-grade behavior of the tumor. Metastases are rare, occurring in fewer than 4% of patients. Multiple recurrences and long-standing tumors usually precede metastasis. Although the lung is the main site of metastatic involvement, there have been reports of brain and bone lesions.[3,4]

Accurate diagnosis is crucial, as DFSP is occasionally confused with dermatofibromas or keloid lesions. It initially appears as a pink nodule with approximately 1 cm in diameter attached to the subcutaneous plane. The nodule tends to grow slowly under the normal epidermis. Recent immunohistochemical studies showed that the probable tumoral origin is perineural. The fusion of chromosome 17 of the *collagen type-1 alpha-1* (*COL1A1*) gene with chromosome 22 of the *platelet-derived growth factor-beta* (*PDGF-β*) gene as a linear translocation is the key molecular abnormality of the tumor,[3,4] resulting in an increased activation of PDGF-β. Therefore, neoadjuvant and adjuvant therapies may be helpful in association with the surgical treatment of recurrent tumors, using medications to block the receptors for these molecules, such as imatinib mesylate.[5,6]

On immunohistochemistry, DFSP tumor cells are positive for CD34 and vimentin, but not for CD44, S-100 protein, and factor XIIIa. In contrast, dermatofibromas react strongly to CD44 and slightly to CD34 and vimentin. In addition to CD34 and vimentin, stromelysin 3 (ST3) is a characteristic immunohistochemical marker for DFSP.[5]

The treatment of choice is surgical resection with wide margins of 2 cm to 3 cm or Mohs micrographic surgery. Surgical margins should be free in the depth and periphery of the primary tumor resection and undergo adequate assessment. It is possible to use postoperative radiotherapy in lesions with unreachable surgical margins, such as in facial tumors. A selective tyrosine kinase inhibitor is being studied, and it has shown some activity in patients with unresectable or metastatic DFSP. Imatinib inhibits the PDGF receptor by blocking its autocrine stimulation.[6]

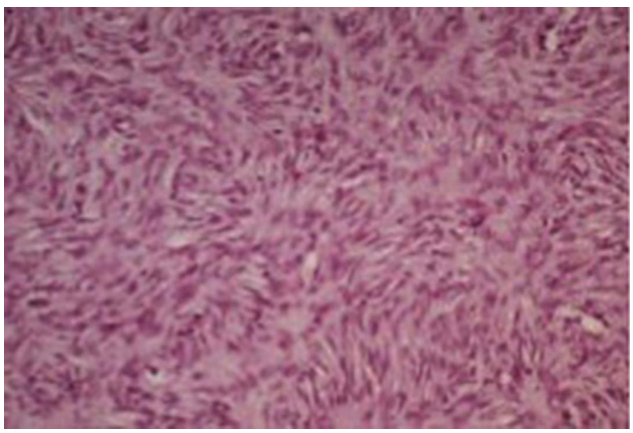

Fig. 2 Photomicrograph showing the high cellularity of the tumor, characterized by the proliferation of monomorphic spindle cells arranged in an irregular pattern (hematoxylin and eosin, 400x).

Dermatofibrosarcoma protuberans of the scalp represents approximately 5% of all cases of this tumor type. Its peculiarities include the probability of periosteal, skullcap, or dural infiltration. Furthermore, the resection extent can generate vast defects, with difficult reconstruction of the region due to the low distensibility of the scalp.

A multicenter review[5] of cases of dermatofibrosarcoma of the scalp totaled 53 cases in the world literature, mainly primary tumors.

Our patient presented a primary tumor with no periosteal invasion. We performed the resection in the subgaleal plane, and we covered the defect with a partial skin graft to wait for the histopathological report and define the free peripheral and deep margins. Definitive reconstruction used rectangular tissue expanders to expand the remaining scalp. The patient has been disease-free for 12 years.

Reconstruction using expanders after dermatofibrosarcoma resection is a good option, and it can start preoperatively in selected cases.[7,8] Likewise, a dermal matrix with negative pressure therapy is a good option to manage complex scalp defects after malignant tumor excision.[9]

The fibrosarcomatous form of DFSP has a worse prognosis. It differs from the classic type due to the presence of more spindle cells, higher number of nuclei, more frequent mitoses, and decreased expression of CD34, explaining the more aggressive nature of the tumor. This subtype of DFSP has a high recurrence rate and a greater chance of distant metastases.[10]

The most reported predisposing factors for recurrence are age over 50 years, resection close to a positive microscopic margin, fibrosarcomatous variant, high mitosis rate, increased cellularity, and a disease-free surgical margin lower than 2 cm.[10]

Ultimately, the only way to reduce the recurrence incidence is initial wide tumor excision. However, the margins are often smaller in neoplasms in the head and neck region to spare significant structures. In these cases, the treatment of choice is Mohs microsurgery.[11] Alternatively, many authors[12] suggest postoperative radiotherapy of 50 cGy to 60 cGy when the resection margin is inadequate, or the histological examination reveals the fibrosarcomatous variant of DFSP.

In conclusion, DFSP is a locally-infiltrative malignant tumor with a high recurrence rate but a low metastatic potential. The main differential diagnosis is dermatofibroma. One must always consider DFSP in cases of lesion recurrence with the histopathological diagnosis of "fibroma." The histopathological diagnosis of DFSP can be difficult due to the absence of nuclear atypia and low mitotic index, requiring immunohistochemical analysis for confirmation and great attention to the peripheral and deep margins free of the tumor. The treatment of choice is wide excision and regular and prolonged postoperative follow-up.[13]

Authors' Contributions

JLSM: data analysis and/or interpretation and final manuscript approval; JMRBAE: data collection and methodology; TOV: conceptualization, methodology, and performance of surgeries and/or experiments; LDABA: data analysis and/or interpretation, methodology, and writing – review & editing;

SPLXS: project management, investigation, methodology, software, and validation; LFRM: data collection and conceptualization; CRM: writing – original draft and software; and SCR: investigation and writing – review & editing.

Financial Support
The authors declare that they did not receive financial support from agencies in the public, private, or non-profit sectors to conduct the present study.

Clinical Trials
None.

Conflict of Interests
The authors have no conflict of interests to declare.

References

1 Kreicher KL, Kurlander DE, Gittleman HR, Barnholtz-Sloan JS, Bordeaux JS. Incidence and survival of primary dermatofibrosarcoma protuberans in the United States. Dermatol Surg 2016;42 (Suppl 1):S24–S31. Doi: 10.1097/DSS.0000000000000300
2 Reha J, Katz SC. Dermatofibrosarcoma Protuberans. Surg Clin North Am 2016;96(05):1031–1046. Doi: 10.1016/j.suc.2016.05.006
3 Thway K, Noujaim J, Jones RL, Fisher C. Dermatofibrosarcoma protuberans: pathology, genetics, and potential therapeutic strategies. Ann Diagn Pathol 2016;25:64–71. Doi: 10.1016/j.anndiagpath.2016.09.013
4 Denadai R, Raposo-Amaral CA, Raposo-Amaral CE, Stelini RF, Cintra ML, Araújo KM. Scalp dermatofibrosarcoma protuberans: a potential diagnostic pitfall. J Craniofac Surg 2016;27(02): 530–532. Doi: 10.1097/SCS.0000000000002327
5 Kuhlmann C, Ehrl D, Taha S, Wachtel N, Schmid A, Bronsert P, et al. Dermatofibrosarcoma protuberans of the scalp: Surgical management in a multicentric series of 11 cases and systematic review of the literature. Surgery 2023;173(06):1463–1475. Doi: 10.1016/j.surg.2023.02.026
6 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;155(03):361–369. Doi: 10.1001/jamadermatol.2018.4940
7 Portugal EH, Alves JCRR, Silva Filho AF, Fonseca RPLd, Pereira NA, Almeida ACd, Andrade Filho JdS. Expansão prévia no tratamento de dermatofibrossarcoma protuberante envolvendo o osso frontal. Rev Bras Cir Plást 2017;32(04):599–602. Doi: 10.5935/2177-1235.2017RBCP0097
8 Rodrigues KKT, Ono MCC, Novais JR, Oliveira IC, Menegazzo HC, Freitas RS. Avaliação do uso do expansor de tecido em um serviço universitário. Rev Bras Cir Plást 2020;35(03):309–315. Doi: 10.5935/2177-1235.2020RBCP0055
9 Otsuka ACVG, Santos Filho IDAO, Campagnari M, Lobo MdM, Bertolli E, Neiva RO, et al. Manejo de defeitos complexos de couro cabeludo após excisão de tumores malignos. Rev Bras Cir Plást 2022;37(04): 494–497. Doi: 10.5935/2177-1235.2022RBCP.627-en
10 Macedo JLS, Rosa SC. Dermatofibrossarcoma protuberante. Rev Bras Cir Plást 2015;30(03):506–509. Doi: 10.5935/2177-1235.2015RBCP0187
11 González A, Etchichury D, Rivero JM, Adamo L. Treatment of dermatofibrosarcoma of the head and neck with Mohs surgery with paraffin sections. J Plast Reconstr Aesthet Surg 2021;74(05): 1061–1070. Doi: 10.1016/j.bjps.2020.10.062
12 Castle KO, Guadagnolo BA, Tsai CJ, Feig BW, Zagars GK. Dermatofibrosarcoma protuberans: long-term outcomes of 53 patients treated with conservative surgery and radiation therapy. Int J Radiat Oncol Biol Phys 2013;86(03):585–590. Doi: 10.1016/j.ijrobp.2013.02.024
13 National Comprehensive Cancer Network. NCCN Clinical Practice Guidelines in Oncology: dermatofibrosarcoma protuberans, version 2, 2022. https://www.nccn.org/professionals/physician_gls/pdf/dfsp.pdf. Accessed 13 September 2023

 © 2025. The Author(s).