

## Dermatofibrosarcoma Protuberans: A Case Report

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### Key Words

Dermatofibrosarcoma protuberans · Neoplasia · Skin tumor · Malignancy

### Abstract

We present a typical case of dermatofibrosarcoma protuberans with local recurrence 2 months after surgery and, motivated by this patient, make a review of the most important aspects. This is a rare tumor and we call special attention to the fact that its recurrence is extremely frequent, so there is absolute need to observe these patients periodically after surgery.

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### Introduction

Dermatofibrosarcoma protuberans (DFSP) is a rare, slow-growing fibrohistiocytic neoplasm considered of low to medium malignancy. It mainly affects young and middle-aged adults. It emerges as an asymptomatic bluish or brownish erythematous multinodular plate, and the involved areas are most frequently the trunk, proximal extremities, head and neck. It affects adults between the second and fifth decade of life. Local recurrence after incomplete resection is common, although distant metastases are rare [1, 2]. In histopathology, the tumor is located in the dermis and consists of uniform fusiform cells, densely grouped, with an elongated nucleus, showing strong positive reaction to CD34 and negative to S-100 and desmin [3]. This DFSP case is reported for its rarity.

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## Case Report

A 41-year-old black man, born in the State of Paraíba, Northeastern Brazil, living in Rio de Janeiro, working as an electrician, came to the hospital with a lesion on his lower dorsum. He reported that a similar tumor present for the past 2 years on that same location had been removed 2 months prior to this consultation. No histopathological examination had been performed at that time. There was local pain. On examination, an exophytic, lobulated tumor, brownish in the periphery and lighter in the center, was observed. Presence of telangiectasia was noted in the lower right paravertebral region (fig. 1). Local and distant metastases were dismissed by clinical examination and imaging. Histopathology showed monomorphic fusiform cell neoplasm, in a 'swirled' focal arrangement, infiltrative of dermal tissue, compatible with DFSP (fig. 2). Immunohistochemistry showed a positive reaction to CD34 (fig. 3) and CD99 (fig. 4) and negativity to factor XIIIa. The patient was referred to surgical resection and will be accompanied at the ward following the operation.

## Discussion

DFSP is a rare and locally aggressive dermal mesenchymal neoplasm. It consists of a sarcoma of cutaneous origin corresponding to less than 0.1% of all malignancies and to approximately 1% of all soft tissue sarcomas, with an annual incidence of 0.8–4.5 cases per million individuals [2–6]. Its growth is indolent, with a high recurrence rate due to its strong capacity to infiltrate subcutaneous tissue, fascia and underlying muscle (infiltration in the form of pseudopods) [6, 7].

Asymptomatic multinodular bluish or brownish erythematous plate, developing over years, with its typical 'protuberant' aspect, is the most frequently observed clinical aspect [1, 6]. It can also present as an atrophic plaque, resembling morphea, and is often misdiagnosed as such [8, 9]. Frierson and Cooper [10] reported the first case of the rare DFSP myxoid variant in 1983, and Hong et al. [3] showed another case of this clinical form rarely documented in the literature, with prominent myxoid stroma alterations. Tantcheva-Poor et al. [11] described a vascular histological variant of DFSP. The most common DFSP sites are the trunk and extremities, with a benign aspect [3, 5]. It is unusual above the neck and extremely rare in the breast region [5].

The proportion of involvement between men and women is 1:1; however, Asquo et al. [12] reported a slight predominance in the male gender. DFSP mainly occurs between the second and fifth decade of life [5]. There are reports of lesions in areas that suffered previous trauma or in patients who underwent several previous surgeries [13]. Our patient had no history of trauma or surgery, but only a similar lesion in the same area, excised 2 months before presentation.

In general, the tumor is located in the dermis, but it can sometimes present infiltrative growth in the subcutaneous fatty tissue, forming a pastry pattern (60% of cases; neoplastic cell bands parallel to the epidermis) or a honeycomb pattern (delimitation of adipocyte islets between the tumoral tissue) [3, 6]. Its low potential for metastasis is typical, with less than 5% probability for regional or distant metastases, with these often being restricted to the lungs and less frequently to the lymph nodes [5, 14, 15]. The differential diagnoses are recurrent dermatofibroma, hypertrophic scars, keloid, skin manifestations of myofibroblastoma, metaplastic carcinoma, fibromatosis or other underlying breast lesion with fusiform cells [5].

Histopathology reveals relatively uniform densely grouped fusiform cells, with elongated nuclei without significant cytologic atypia or pleomorphism in characteristic storiform arrangement [3]. The degree of nuclear atypia is higher in nodular lesions than in plates. Fibrosarcomatous focal alterations with a characteristic fish bone pattern are occasionally observed in DFSP [5]. Immunohistochemical findings are positivity to CD34 in 84–100% and to vimentin (which would relate to the fibroblastic nature of the tumor), and negativity to other markers such as S-100, HMB45, desmin and actin. The characteristic pattern is positive for CD34 and negative for factor XIIIa. Our patient had this pattern. Stromelysin-3 is negative (dermatofibroma marker), as is CD117 [1, 6, 16, 17].

The standard treatment of the localized disease consists of wide local surgical resection with recommended surgical margins of 2–3 cm and three-dimensional resection including skin, subcutaneous tissue and underlying fascia [5, 14, 18]. The local recurrence rate tends to decrease with the increase of surgical margins [5]. This procedure can generate cosmetic deformities and even functional damage. The factors associated with high rates of recurrence are histological subtype, cellularity, size, location on the head and neck, and high mitotic rate [19, 20]. There are current reports praising the use of Mohs micrographic surgery as a first-line therapeutic measure in cases of limited tumors for tissue preservation and reduction of recurrence rate [4, 21–23].

Over 90% of DFSP cases present a translocation in chromosomes 17 and 22, which leads to changes in the quantity of B platelet-derived chain growth factor, resulting in the activation of platelet-derived growth factor receptor and consequent stimulus for tumor growth. Imatinib mesylate is a potent selective inhibitor of tyrosine kinases, which include platelet-derived growth factor receptor. This medicine inhibits the growth of tumor cells and the transformation of fibroblasts, both *in vivo* and *in vitro*. Some clinical studies have shown a positive response in 65% of patients who used the drug, thus it constitutes an alternative for adults with non-resectable, recurrent or metastatic lesions [1, 4, 5, 24].

Radiation therapy is an adjuvant therapy in cases where adequate surgical margins are not easily reached or result in aesthetic/functional defect, or in cases of positive margins, even after maximum resection. It is also indicated for patients with inoperable macroscopic lesions [1, 5]. Postoperative radiotherapy has been associated with a cure rate of more than 85% [25]; however, it presents risks such as acute or chronic radiodermatitis and subsequent development of other skin tumors [1].

Since the tumor projects in multiple directions, reaching deep structures, not even wide excision can prevent residual tumor in single or multiple focus, and this would explain lesion recurrence [1, 26, 27]. Local recurrence may occur late in 24–90% of cases [4]. Most appear within 3 years, with half of them occurring in the first year after resection. There are reports of new lesions appearing 5 years after treatment, which justifies the need for biannual clinical and patient monitoring for an extended period after surgery [1, 5, 16, 28].

## Conclusion

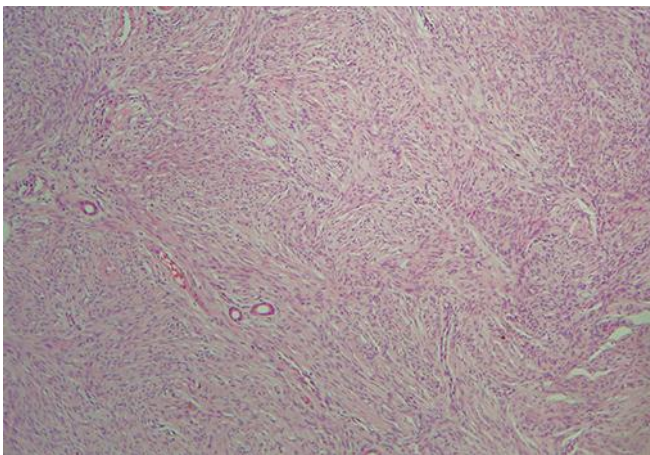
This case motivated us to review the most important aspects of this rare tumor, besides presenting an interesting and typical case of DFSP with local recurrence 2 months after surgery. All dermatologists and surgeons must know about the frequent recurrence of this tumor, sometimes even when excised with wide margins. For this reason these patients must be observed periodically after surgery for a long time.

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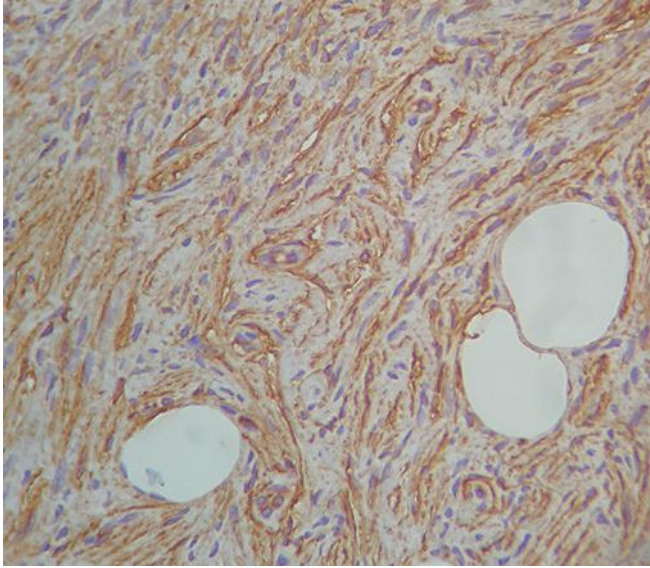


**Fig. 1.** Tumor in the lower right paravertebral region, compatible with DFSP.

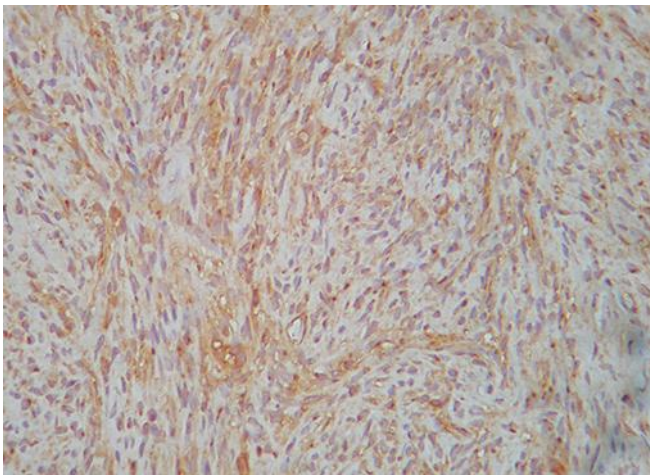


**Fig. 2.** Panoramic view at low magnification shows proliferations of monomorphic fusiform cells (HE, obj. 20).





**Fig. 3.** Immunohistochemistry: positivity for CD34 in tumoral cells (obj. 100).



**Fig. 4.** Immunohistochemistry positivity for CD99 in tumoral cells (obj. 100).