



## Dermatofibrosarcoma in glabella: An uncommon location

### *Dermatofibrossarcoma na glabela: Uma localização incomum*

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#### ■ ABSTRACT

Dermatofibrosarcoma is a rare cancer with a slow growth pattern and aggressive tissue invasion. The treatment for this condition involves surgical intervention to achieve clear margins. In this particular case, we have a 60-year-old man who had a mass in the glabella area that was excised twice under local anesthesia at a different facility. The results of both biopsies indicated dermatofibroma. However, the patient experienced a new recurrence, and during the immunohistochemistry analysis of the initial excision, it was revealed to be dermatofibrosarcoma. As a result, a broad excision was performed until negative margins were obtained based on frozen sections. Additionally, a frontal flap was employed to reconstruct the surgical defect. The outcome of the treatment was deemed successful, without complications.

**Keywords:** Dermatofibrosarcoma; Skin neoplasms; Mohs surgery; Head and neck neoplasms; Reconstructive surgical procedures.

#### ■ RESUMO

O dermatofibrossarcoma é um câncer raro que apresenta padrão de crescimento lento e invasão tecidual agressiva. O tratamento para esta condição envolve intervenção cirúrgica com o objetivo de obter margens livres. Neste caso particular, temos um homem de 60 anos que apresentava uma massa na região da glabella que foi extirpada duas vezes sob anestesia local em outro serviço. Os resultados de ambas as biópsias indicaram dermatofibroma. No entanto, o paciente apresentou nova recidiva, que durante a análise imuno-histoquímica da excisão inicial revelou tratar-se de dermatofibrossarcoma. Como resultado, uma ampla excisão foi realizada até que margens negativas fossem obtidas ao exame de congelação. Além disso, um retalho frontal foi empregado no procedimento para reconstrução do defeito cirúrgico. O resultado do tratamento foi considerado bem-sucedido, sem complicações.

**Descritores:** Dermatofibrossarcoma; Neoplasias cutâneas; Cirurgia de Mohs; Neoplasias de cabeça e pescoço; Procedimentos cirúrgicos reconstrutivos.

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Article received: November 27, 2022.  
Article accepted: August 20, 2023.

Conflicts of interest: none.

DOI: 10.5935/2177-1235.2023RBCP0769-EN

### INTRODUCTION

Dermatofibrosarcoma is a rare cancer, representing 1% of all skin sarcomas and less than 0.1% of all tumors<sup>1</sup>. This disease was initially reported by Darier and Fernand in 1924 as an unfamiliar condition and only later identified by Hoffman as dermatofibrosarcoma<sup>2</sup>.

It is considered an uncommon soft tissue tumor, affecting approximately 0.8 to 5 people per million<sup>3</sup>. The proportion of affected individuals is similar between

men and women. Although it can occur at any age, it is most commonly observed from the second to the fifth decade of life<sup>2</sup>.

The tumor exhibits characteristics such as slow growth, aggressive tissue invasion, and a high risk of recurrence. However, metastasis is rarely observed<sup>3</sup>. Initially, it was believed to originate from fibroblasts, but immunohistochemistry analysis suggested its origin from dendritic cells of the skin<sup>4</sup>.

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Dermatofibrosarcoma is most frequently found on the chest and extremities of the body, with approximately 10% to 15% of cases occurring on the head and neck. It initially presents as a painless, flat lesion with the same color as healthy skin or rose-colored. As a result, it can be mistaken for some benign conditions, leading to delayed diagnosis, sometimes even up to a few years<sup>3</sup>.

The recommended treatment for dermatofibrosarcoma is surgical excision of the lesion with wide margins and primary reconstruction. An excisional biopsy alone is insufficient, as at least 2 cm of free margins or Mohs micrographic surgery is necessary<sup>5,6</sup>. Currently, Mohs micrographic surgery is considered the gold standard for treatment, regardless of the tumor's location. Case series analyses and retrospective studies have demonstrated that this technique has lower recurrence rates and preserves more healthy tissue compared to the traditional technique with wide margins<sup>3</sup>.

### CASE REPORT

This study was accepted by the ethics committee – #24859519.0.0000.5509.

A sixty-year-old man, social drinker, and smoker for 27 years (from 18 to 45) complained of a forehead skin lesion next to the right orbit in the glabella for 6 years. He underwent surgical resection under local anesthesia in 2013 and 2017, with a pathological diagnosis of dermatofibroma spreading to muscle with the recommendation for immunohistochemistry evaluation; however, it had not been performed. A mobile subcutaneous nodular lesion measuring 2.5 cm located on the right glabella was detected (Figure 1). There were no palpable lymph nodes. The ultrasonography showed a highly vascularized nodule measuring 19 x 17 x 16 mm with a hypoechoic halo. The immunohistochemical analysis of both previous biopsies indicated dermatofibrosarcoma protuberans with a proliferation index (Ki67) of 10%. MRI scan revealed an expansive lesion with 25 x 18 x 21 mm in contiguity with the bone, however, without erosion. Surgical resection with intraoperative frozen sections was performed to determine free margins (Figure 2). After two margin augmentations next to the nasogenian groove, they were tumor-free. The reconstruction was performed with a midfrontal flap based on the left trochlear artery (Figures 3 and 4).

Histopathological examination showed low graded cell spindle tumor with discreet nuclear atypia, low mitotic index, and no necrosis, measuring up to 32 mm in diameter, spreading into dermis,



Figure 1. Lesion before excision



Figure 2. Lesion excision, including a portion of the nasal bone.

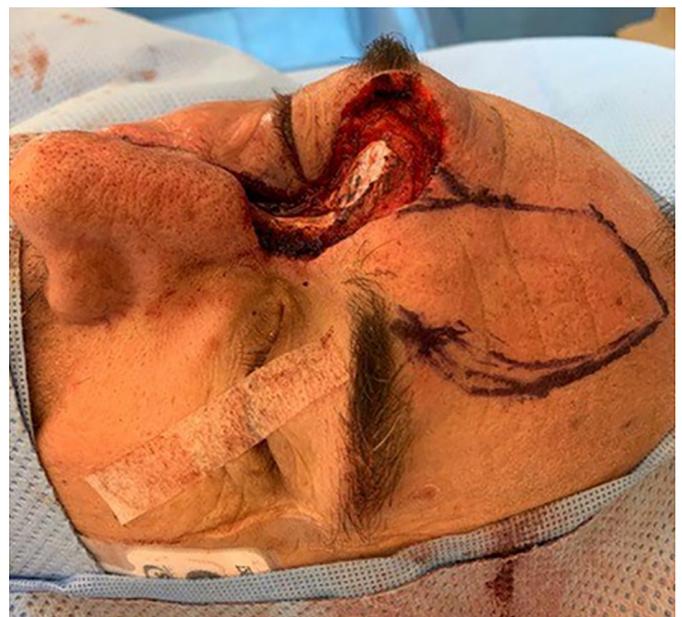
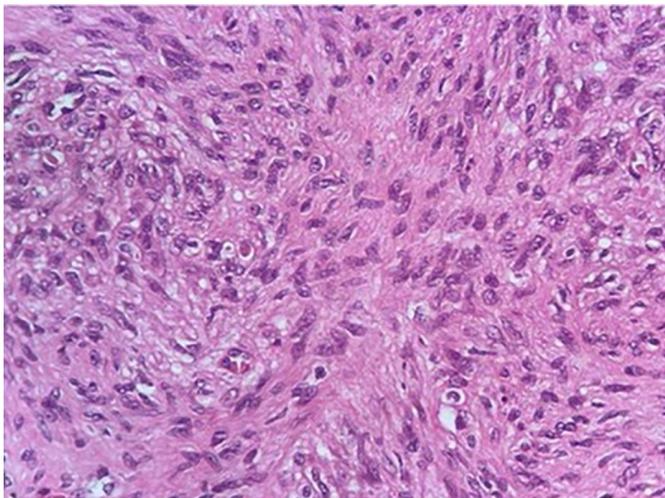


Figure 3. Demarcation of flap for reconstruction after margins clearance.



**Figure 4.** Patient's face after the reconstruction with a flap based on the left trochlear artery.

hypodermis, and striated muscle tissue with free margins (Figure 5). Thus, the final diagnosis was dermatofibrosarcoma protuberans. The patient evolved uneventfully.



**Figure 5.** Histopathological examination.

## DISCUSSION

Seldom does dermatofibrosarcoma affect the glabella, resulting in few reported cases in this location. It typically exhibits a slow growth pattern, local aggressive behavior, and negligible metastatic potential, with high rates of local recurrence<sup>1</sup>.

It commonly presents as a violaceous red-blue plaque with gradual growth over the years, eventually developing multiple nodules within the plaque<sup>1</sup>. The tumor demonstrates aggressive local invasion and a

high recurrence rate after excision, although metastasis is rare<sup>2</sup>. The diagnosis of dermatofibrosarcoma is often delayed due to its insidious onset and appearance that is relatively nonspecific<sup>7</sup>, and these lesions can be clinically misdiagnosed as benign entities, as was the case in our patient<sup>3</sup>.

Dermatofibrosarcoma has been reported to occur on various body surfaces, primarily the trunk, followed by the extremities, and less commonly on the head and neck<sup>4</sup>. Initially, it superficially adheres to the overlying skin, leading to mobility upon palpation. However, it may infiltrate adipose tissue in later stages and become fixed to deeper structures such as fascia and muscle. Delayed diagnosis and misdiagnosis are common, often due to the absence of symptoms or confusion with benign dermal fibrohistiocytic lesions<sup>4</sup>. The initial lesion was misdiagnosed as dermatofibroma in our case due to the lack of further immunohistochemical evaluation before the patient was referred to our facility. Other possible differential diagnoses in the early stages include lipomas, epidermal cysts, nodular fasciitis, and even keloids. As the lesion becomes more protuberant in the late stages, other soft tissue sarcomas should be considered in the differential diagnosis<sup>4</sup>.

MRI scan findings are not specific, typically showing a well-delineated tumor that appears isointense or mildly hyperintense on T1-weighted images and hyperintense on T2-weighted images<sup>1</sup>.

Dermatofibrosarcoma protuberans occupies a central position within a spectrum of tumors known as fibrous histiocytomas. These tumors can exhibit relatively benign behavior, similar to fibrous histiocytomas, or demonstrate more aggressive features akin to malignant histiocytomas. Histologically, dermatofibrosarcoma protuberans is highly cellular and composed of monomorphic, fusiform cells with elongated nuclei that show little or no pleomorphism or hyperchromasia. Mitotic activity is usually moderate. Immunohistochemical evaluation is necessary for accurate diagnosis<sup>5</sup>. The diagnosis is confirmed by demonstrating strong immunoreactivity for CD 34<sup>4</sup>. This patient underwent two surgeries at another facility, and, most likely, due to the omission of immunohistochemistry evaluation, they were misdiagnosed as dermatofibroma.

Due to its histological characteristics of poor circumscription and frequent extension into the dermis, subcutis, and muscles, resection should include wider and deeper margins than those appreciated clinically or on MRI studies<sup>1</sup>. The tumor often extends beyond the clinically determined margins during surgical resection<sup>3</sup>.

Successful treatment requires deep and wide excision and extensive reconstruction involving flaps or grafts<sup>5</sup>. Micrographic Mohs surgery is considered the optimal treatment for this disease in all anatomical locations<sup>3,6</sup>. Recommended excision margins are 2 cm or even 3 cm for block excision<sup>4,5</sup>. The recurrence rate depends on the adequacy of the excision margins<sup>4</sup>. In our case, intraoperative frozen sections confirmed disease-free margins, subsequently confirmed by the final histopathologic examination using paraffin-embedded tissue.

Dermatofibrosarcoma protuberans is uncommon in the glabella region. Given its proximity to the orbits, its occurrence in this location presents a significant challenge due to the requirement for radical excision and the potential impact on both functional and cosmetic outcomes.

### COLLABORATIONS

**CEB** Conception and design study, Conceptualization, Final manuscript approval, Methodology, Project Administration, Writing - Original Draft Preparation, Writing - Review & Editing.

**MAC** Analysis and/or data interpretation, Final manuscript approval, Realization of operations and/or trials, Writing - Review & Editing.

**RAD** Conception and design study, Conceptualization, Final manuscript approval, Methodology, Project Administration, Realization of operations and/or trials, Supervision, Writing - Review & Editing.

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