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Case Report

Dermatofibrosarcoma Protuberans in Face, A Challenging Case

Carlos José Gaspar Junior[*1]

[1] Plastic Surgery and Burns Service of Santa Casa de Misericórdia de São José do Rio Preto - Rua Fritz Jacobs, 1236. Good view - São José do Rio Preto, Brazil.

Introduction

Dermatofibrosarcoma Protuberans (DFSP) is a rare type of cancer. A soft tissue sarcoma that develops in the deep layers of skin. DFSP is often misdiagnosed as a benign skin disease and thus, mistreated [1-3]. Therefore, patients usually come to their plastic surgeons and dermatologists in advanced stages. Doctors around the globe must always keep in mind DFSP when facing a suspicious lesion [4]. After the correct diagnose, the treatment for DFSP requires a large margin (2 to 4 cm) and the design of local flaps. In this case, we present a 45-year-old male, with a facial DFSP [5-8]. The lesion spread underneath the skin, affecting a large part of the face (Figure 1 & 2). The local recurrence rate for DFSP in studies ranges from 0% to 60%, whereas the rate of development of regional or distant metastatic disease is only 1% and 4% to 5%, respectively. Differentiation of DFSP from dermatofibroma can sometimes be difficult. In these instances, immunostaining with CD34, factor XIIIa, metallothioneins, tenascin, and/or stromelysin-3 may be useful. The NCCN Clinical Practice Guidelines in Oncology, recommends that appropriate and confirmatory immunostaining be performed in all cases of suspected DFSP [9]. Because of its proclivity for irregular and frequently deep subclinical extensions, every effort should be made to completely remove this tumor at initial therapy.

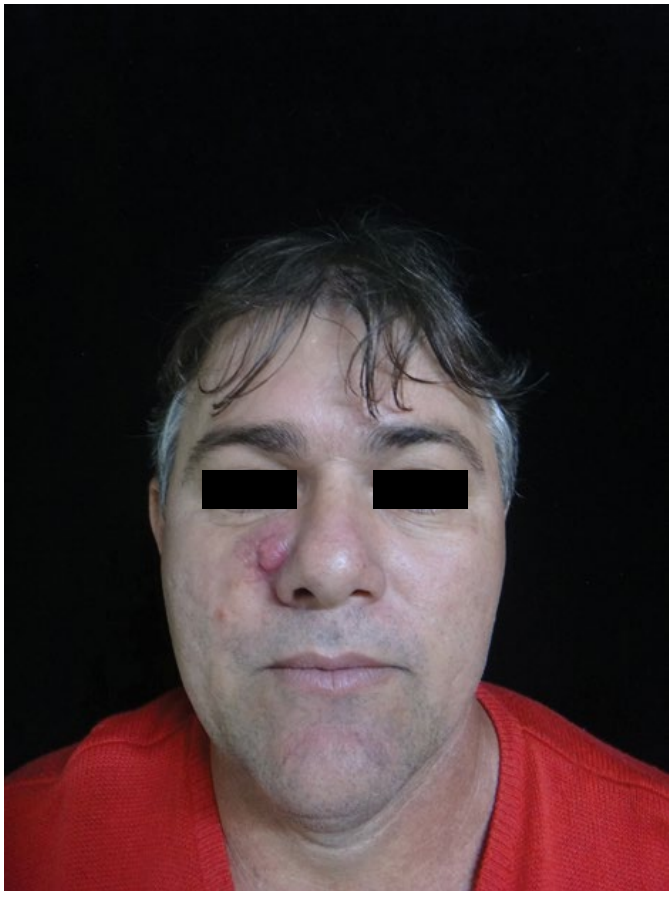

Figure 1: shows skin lesion in face. Note the facial units are being affected – right malar and zygomatic.

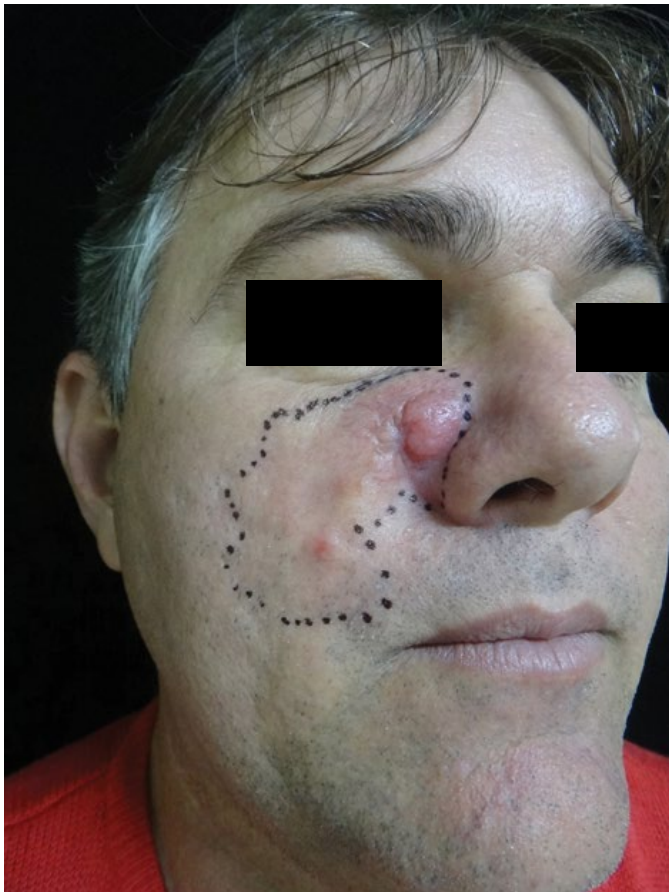

Figure 2: Shows the underskin affection and the design of margin (2 cm in lower right eyelid and 3 cm to the rest).

Corresponding author: Carlos José Gaspar Junior, Plastic Surgery and Burns Service of Santa Casa de Misericórdia de São José do Rio Preto - Rua Fritz Jacobs, 1236. Good view - São José do Rio Preto, Brazil, Tel: ; Fax: ; E-mail: dr.gaspar.jr@gmail.com

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Objective

The purpose of this work is to show how important is the early diagnose and treatment for this problem. DFSP requires a margin-free of 2 to 4 cm. We must put all the efforts towards the early diagnose.

Materials and Methods

45-year-old, Male presented at our service complaining about a face injury that was taking a long time to heal. The patient had visited two other physicians before, and had been treating the lesion as a Facial Acne. We proceeded with a biopsy [10]. The result was Dermato-Fibrosarcoma-Protuberans. The patient had no other comorbidities. As the DFSP spread under the skin to a wide part of the face – Right Malar and Zygomatic. By palpation we underlined the entire under-skin area affected, and designed a Mid-Forehead Flap to cover the most Malar-Nasal wing part of the defect and a Skin graft (full length graft) for the zygomatic part of the defect [11-15]. We performed the resection with a 2 to 3 cm margin and went through the deep layer resecting the muscle fascia. Frozen section biopsy was performed, and after confirmation of margin free, we performed the reconstruction. As shown in Figure 3 & 4.

Results

After 1 year of DFSP resection, the patient had no disease recurrence. (Figure 5). We recommend the 2 cm margin for DFSP in the Face. More studies are required for conclusion that DFSP can be treated with a smaller margin for Facial Lesions [16-19]. Until then, surgeons must keep all efforts for the resection with a 2 to 4 cm margin. Since DFSP has a high level of recurrence.

Discussion

DFSP is characterized by a translocation between chromosomes 17 and 22 (t(17:22)) resulting in the overexpression of platelet-derived growth factor receptor (PDGFR) β [20]. 26–28 These findings suggest that targeting PDGFRs may lead to the development of new therapeutic options for DFSP. In recently published results, imatinib mesylate, a protein tyrosine kinase inhibitor, has shown clinical activity against localized and metastatic DFSP tumors containing t(17:22).29–33 Imatinib mesylate has recently been approved by the FDA for the treatment of unresectable, recurrent, and/or metastatic DFSP in adult patients.34 Because tumors lacking the t(17;22) translocation may not respond to imatinib molecular, analysis with cytogenetics may be useful before initiating imatinib therapy [21-24].

DFSP continues to challenge surgeons. We must be prepared to receive patients in advanced stages of the disease. DFSP has high level of local recurrence in one year. The disease is on local aggressive [25,26]. There are almost no case-reports of distant spreading

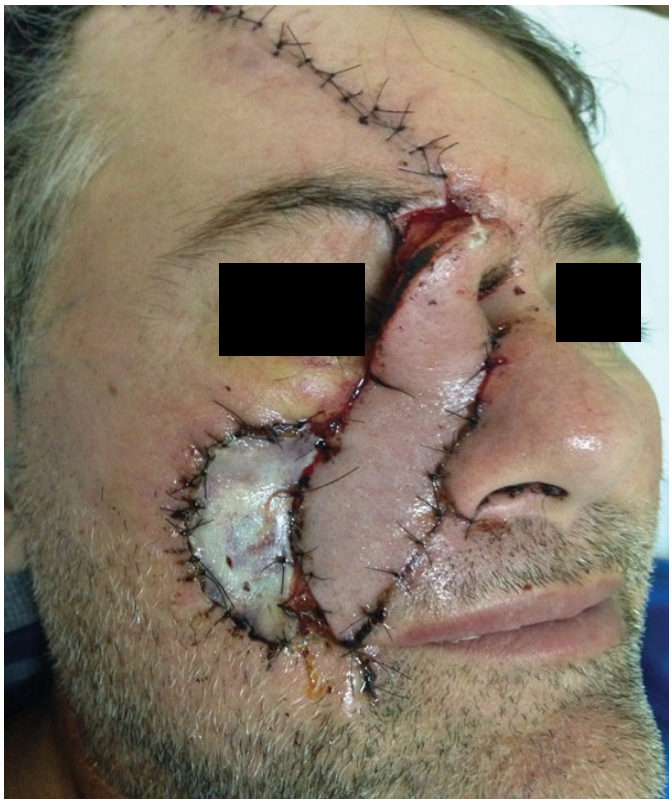

Figure 3: shows 7 days after reconstruction. Mid-Forehead Flap for the most medial defect and a Full-Length Skin graft for the most lateral defect.

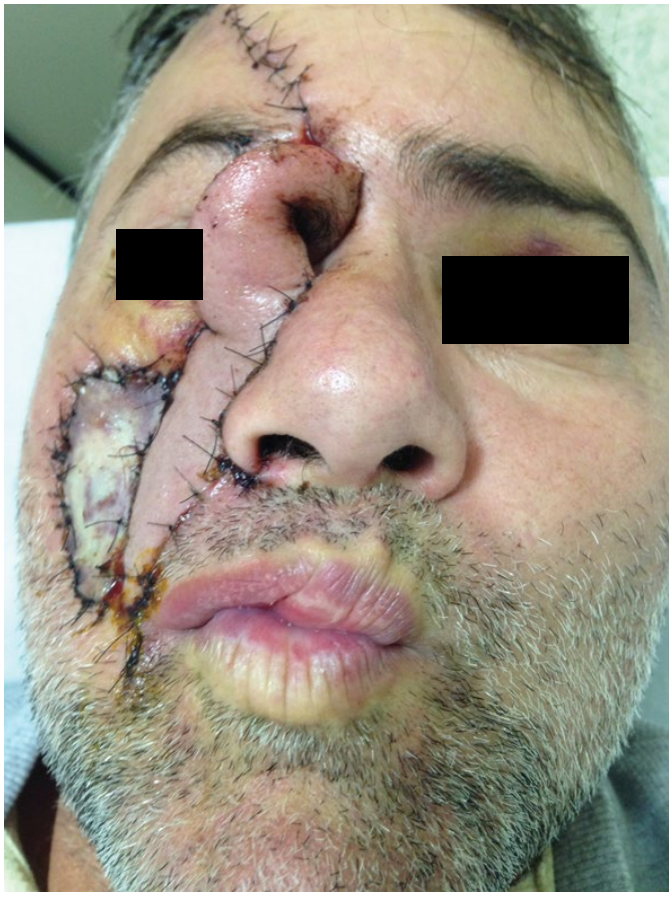

Figure 4: shows the paresthesia of the buccal branch – Facial Nerve.

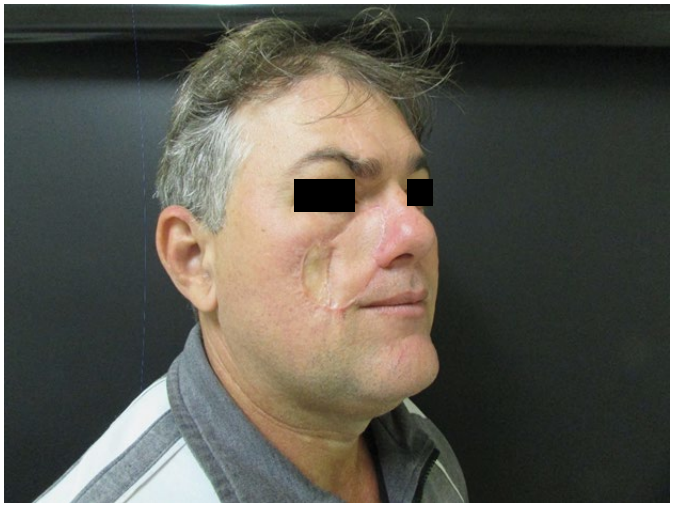

Figure 5: shows 1-year post operation. No recurrence detected and recover from Facial Nerve paresthesia.

(metastasis) [27-29]. At our service we never had a metastatic DFSP. Our suggestion is that the three to five margin-free must be observed, and the frozen section biopsy is mandatory [30]. When available, Mohs technique can also be used, and should be encouraged.

In a recent series of 244 patients with DFSP, tumor depth was the only factor associated with disease-free survival in the primary setting, underscoring the importance to excise the deep fascia to remove any infiltrating tumor cells [31-34].

In another retrospective review of 48 patients, positive margins were more frequent with wide excision than with Mohs, but the local recurrence rates were statistically similar (3.6% vs. 0%, respectively; P = 1.0) [35].

Conclusion

With our case report we concluded that in some facial lesions the margin levels could be smaller. But studies should be performed to confirm this affirmation. We hope to contribute somehow to the medical community by showing our case. We believe that once we see what DFSP looks like, diagnose can come to mind when facing patients with similar lesions.

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