Reconstructive strategies for dermatofibrosarcomas of the face: Role of regenerative dermal templates

Leonardo Sartore, MD,1 Erica Dalla Venezia, MD,1 Alessandro Della Puppa, MD,2 Alberto Bedogni, MD,3 Luca Campana, MD,4 Giorgio Giatsidis, MD,1 *

1Clinic of Plastic Surgery, Department of Surgery, Padua University Hospital, University of Padua, Padova, Italy, 2Department of Neurosurgery, Padua University Hospital, University of Padua, Padova, Italy, 3Unit of Maxillofacial Surgery, Verona University Hospital, University of Verona, Verona, Italy, 4Sarcoma and Melanoma Unit, Veneto Institute of Oncology (IOV-IRCCS), Padova, Italy.

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ABSTRACT: Background. Dermatofibrosarcomas protuberans is a challenging cutaneous tumor from an oncologic and reconstructive surgical point of view. Involvement of functionally and aesthetically sensitive areas, such as facial units, in young patients accounts for more demanding cases. An updated evaluation of most beneficial excisional/reconstructive strategies in these cases is still lacking.

Methods. We investigated the potential of regenerative dermal templates in staged postoncologic reconstructive management of a young woman affected by a dermatofibrosarcomas protuberans of the forehead involving the frontal bone.

Results. Final result was optimal in terms of cosmetic and functional recovery, obtaining a pliability, softness, and color similar to surrounding healthy skin.

Conclusion. In facial dermatofibrosarcomas protuberans, staged reconstruction with regenerative dermal templates provides a conservative yet safe and effective management, achieving optimal aesthetic outcomes. We suggest its adoption as first-line treatment in facial dermatofibrosarcomas protuberans that cannot be repaired by direct suture and in equivalent benign yet challenging cases. © 2014 Wiley Periodicals, Inc. Head Neck 37: E8–E11, 2015

KEY WORDS: dermatofibrosarcoma protuberans, regenerative dermal template, facial tumors, conservative treatment, reconstructive strategies

INTRODUCTION

Malignant cutaneous tumors of the face represent a considerable surgical challenge because of the need to achieve an oncologic radical excision while preserving functional and aesthetic features of specific facial units. Despite that these two needs are closely related, they are often not equally addressed when investigating the most adequate therapeutic strategies, which focus on excision procedures and leave aside the evaluation of most beneficial reconstructive techniques. This issue has long been debated for malignant tumors in which oncologic safety is an undeniable priority, but it has been recently emerging as a critical aspect in the management of benign tumors characterized by a locally invasive behavior, in particular, in younger patients.1–6 Among these, dermatofibrosarcoma protuberans is an uncommon low/intermediate-grade fibrohistiocytic sarcoma often affecting young adults and showing uncontrolled infiltrative local growth.1–6 Despite the malignant potential and metastases are rare, for tumors affecting the head (10% to 15% of cases), invasion of surrounding tissues may provide mutilating or even a life-threatening consequences.2–20 In the last decade, numerous studies have provided evidence that a wide surgical excision (<2.5 cm margins) or a more conservative Mohs micrographic surgery can significantly affect the rate of recurrences: nonetheless, literature has scarcely investigated how to best profit from novel reconstructive strategies.21–28 Regenerative dermal templates have been successfully adopted in the management of malignant cutaneous tumors of the scalp for which their use is now well established.29–35 Even so, they have never been described in the management of facial dermatofibrosarcoma protuberans. The purpose of this study was for us to report the case of a young woman affected by a dermatofibrosarcoma protuberans of the forehead and we discuss therapeutic, functional, and aesthetic benefits of the procedure.

CASE REPORT

In April 2011, a 35-year-old woman was admitted to our Clinic of Plastic Surgery for a single voluminous subcutaneous nodule in the median region of the forehead. The patient had first noticed the neoplasm 5 years before, observing a slow progressive growth in subsequent months. Ultrasounds initially misdiagnosed the lesion as a sebaceous cyst. No history of trauma or other relevant systemic disorders could be recorded; however, she reported to have undergone several intravenous injections of Fe2+ in the central vein of the forehead, when
newborn, causing frequent extravasation injuries. A subsequent mild subcutaneous atrophy of the forehead developed in the following years and it was treated during adolescence (age 16 years) with autologous adipose tissue injection. At medical examination, the nodule appeared nontender, firm, fixed on overlying skin, but movable over bony structures; the surrounding skin was otherwise healthy. The patient reported no symptoms and absence of pain. X-rays of the skull showed a superficial involvement of the underlying bone. Given the growth and the clinical progression of the neoplasm, in agreement with the patient and after obtaining written informed consent, we decided to proceed with a multistep strategy. First, we performed a small incisional biopsy, which confirmed the diagnosis of dermatofibrosarcomas protuberans. Hence, an en bloc surgical excision of the nodule was performed preserving 2-cm margins from macroscopically affected surrounding skin and extending to the galea; because of evidence of erosion of the outer table of the frontal bone (eventually because of multiple injections received when a newborn), we combined a bone excision (Figure 1). Because intraoperative histological analysis of the margins was negative for the presence of disease, dura mater was repaired by a temporal fascia graft, whereas final closure was obtained by implant of a double-layer regenerative dermal template covered by a protective silicone sheet (Integra dermal regeneration template; Integra LifeSciences Corporation, Plainsboro Township, NJ). The patient was discharged on the following day and monitored at our outpatient clinic twice a week without any adjuvant treatment. After 3 weeks, histological analysis confirmed absence of disease at resection margins, hence, providing the chance for definitive surgical reconstruction. The dermal layer appeared well-vascularized and vital, but the underlying fascia graft only partially survived (Figure 2). Hence, we decided to perform a regenerative dermal template overgrafting in order to obtain an adequate dermal thickness that could match surrounding tissues and, 3 weeks later, an autologous split-thickness skin graft from the left thigh over the well-vascularized dermal template. No postsurgical complications were observed at follow-up; in the following months, the skin graft gradually gained a similar color, smoothness, and appearance of the surrounding skin with a remarkable aesthetic outcome (Figure 3). No signs of recurrences were noticed at a 26-month follow-up.

**DISCUSSION**

The dermatofibrosarcomas protuberans, so named by Hoffmann in 1925 after a first description by Darier and Ferrand in 1924, is an uncommon benign spindle cells neoplasm; it represents only 0.1% of all cancers and 1% to 5% of all soft tissue sarcomas affecting adult individuals.1-6 Localization to the head and neck region (10% to 15% of cases) is even more rare because dermatofibrosarcomas protuberans usually affects the trunk (50% to 60% of cases) or extremities (20% to 30%).1-6 Unfortunately, the highest incidence affects young patients (male:female ratio is almost 1.5-4:1) in their second to fourth decade of life.1-6 Despite being considered a benign tumor,
Dermatofibrosarcomas protuberans is burdened by a relevant tendency to local growth with invasion of surrounding tissues and structures. In addition, recurrence rates exceed, on average, 50% to 75% of cases affecting the head and neck regions. Recurrence potential is likely related to extension of resection and to the need for a more conservative surgical excision in sensitive areas in which preservation of functional and aesthetic properties is a priority. However, because recurrence may be associated with further local malignancy, intracranial invasion (rare), distant metastasis, or presence of isolated areas of fibrosarcoma (or malignant fibrous histiocytoma), recent literature has advocated the need for a more radical primary surgical approach. Adoption of wide (2.5–3 cm) margins of excisions with resection of the underlying (temporalis) muscle, the deep fascia, and part of the periosteum is today considered the standard of treatment and may decrease recurrence rates to 18%. Beside oncologic benefit, this strategy poses substantial reconstructive challenges in a very peculiar anatomic region, such as the face. The advent of Mohs micrographic surgery has offered the opportunity to maximize preservation of tissues with even lower recurrence rates (2% to 5%). However, as a first-line treatment, Mohs micrographic surgery is laborious, not available in every center, and not suitable for large tumors; hence, wide excision is still frequently adopted. The debate between radical surgery or Mohs micrographic surgery has been the main focus of recent literature, whereas the role of reconstructive strategies for these disfiguring defects has been marginally considered and mostly reduced to old-fashioned procedures, each presenting intrinsic limits. Standard reconstructive procedures reported in literature include full-thickness or split-thickness skin grafts, fascia lata grafts, various allografts, local or pedicled flaps, distant free flaps (radial forearm flap or groin flap), staged reconstruction with skin expanders, or even healing by second intention. None of these options is free from complications (up to 25% of cases), which include necrosis of grafts or flaps, infection, dehiscence. In addition, some of these treatments may be inadequate for larger defects (local flaps) or full-thickness excisions (allografts), they may require more invasive surgeries, longer hospitalization, and longer postoperative recovery (pedicled/distant flaps, skin expanders), or they may not be adequate for a temporary coverage (decellularized allogeneic dermis), while margins are assessed for final histological results. Significantly, most of these techniques can provide only scarce to moderate aesthetic results. Soon after their introduction in clinical practice, regenerative dermal templates have proved to be a reliable and valuable resource for the reconstructive management of many postsurgical defects. The template allows a prompt and effective reconstruction by means of a low invasive surgery and without any sacrifice of major autologous donor areas. In the last 10 years, surgeons have applied these features in the challenging management of excision of severe malignant tumors of head and neck regions, in particular in the scalp. The characteristics of dermal regenerative templates, in our opinion, make them an ideal candidate as a first-line reconstructive treatment also in dermatofibrosarcomas protuberans of the face. As in our report, the template alone allows the immediate coverage of large defects with a conservative strategy preserving surrounding tissues (no need of local flaps) as well as of other distant donor areas. This issue is of primary importance if considering the high rate of recurrences and the potential need to preserve as many surgical options as possible for a second reconstructive surgery. Recurrence occurs, on average, after 18 to 22 months, but adequate follow-up should be prolonged for at least 5 years; our patients showed no signs of recurrence after 24 months. These surgeries may be well tolerated also by patients affected by multiple comorbidities and require short hospitalization periods. The artificial graft may be shaped and adapted accordingly to specific needs and in order to provide the same dermal thickness; it may be overgrafted with low invasive staged surgeries (3 weeks are required to induce revascularization of the graft) that are usually minimally burdened by postoperative complications. In addition, the adoption of a staged strategy offers the opportunity for an effective temporary coverage without immediate definitive reconstruction, which may represent a significant benefit if final histological examination requires a new excision of positive margins in the following weeks. Dermatofibrosarcomas protuberans is considered resistant to conventional chemotherapy and radiation therapy. Metastases have been treated with novel targeted molecular therapies, such as selective tyrosine kinase inhibitors (imatinib mesylate) for adjuvant treatment. Even so, some authors have addressed the role of radiation therapy for adjuvant treatment in locally invasive cases. In these patients, temporary coverage could be achieved by means of a regenerative dermal template during radiation therapy, postponing final reconstruction at the end of the therapy to not affect viability of reconstructed tissues. Finally, given the incidence of the tumor in young adults, attention should be placed on overall aesthetic outcome of reconstructive procedures and quality of life; regenerative dermal templates provide an ideal recovery of the dermal layer allowing a positive healing of overgrafted partial-thickness skin grafts. As for our young female patient, the final result was considered optimal in terms of cosmetic and functional recovery, obtaining a pliability, softness, and color similar to the surrounding healthy skin.

**CONCLUSIONS**

In conclusion, dermatofibrosarcomas protuberans is a tumor in which the aggressive behavior makes a radical oncologic excision mandatory. In cases involving facial units, in particular in young patients, reconstructive options should be adequately considered to preserve and optimize functional-aesthetic outcomes. In this light, staged reconstruction with regenerative dermal templates provides a unique opportunity for a conservative yet safe and effective management (lower rate of complications, adequacy for larger defects or full-thickness excisions, low invasive surgery, short hospitalization, and postoperative recovery, and adequacy for temporary coverage while margins are assessed for final histological results or during radiation therapy for adjuvant treatment), achieving optimal aesthetic outcomes. For all these reasons, this reliable approach should be considered as a first-line treatment in all dermatofibrosarcomas protuberans of the
face that cannot be repaired by direct suture but also in a broader range of benign cases involving large or challenging facial defects.

REFERENCES