Journal of Surgery
El Danaf AA, et al. J Surg 9: 11129
www.doi.org/10.29011/2575-9760.11129
www.gavinpublishers.com
Case Report

Combined Wide Excision, Mohs Micrography, and Reconstruction of Recurrent Dermatofibrosarcoma Protuberans

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Citation: El Danaf AA, Rabie AA, Eldanaf HA, Mohsen MA (2024) Combined Wide Excision, Mohs Micrography, and Reconstruction of Recurrent Dermatofibrosarcoma Protuberans. J Surg 9: 11129 DOI: 10.29011/2575-9760.11129

Received Date: 23 August 2024; **Accepted Date:** 29 August 2024; **Published Date:** 31 August 2024

Abstract

Both the wide deep local excision and Mohs micrographic surgery are standard treatment for Dermatofibrosarcoma (DFSP). This report aims to evaluate the association of these two surgical policies with immediate reconstruction of the defect by a distant flap for treating large repeatedly recurrent DFSP. An adult patient presented with a 10x12 cm DFSP on the neck anterior surface and another with an 11x25 cm DFSP on the chest right side. They underwent excision of the tumor entire lesion with 2-4 cm safety margins including the apparently healthy underlaying tissue layer associated with frozen sections testing of the defect periphery and floor. The regularly advisable reconstruction by split-thickness skin grafting for easy detection of recurrence could not be performed because it would have end with cervical skin shortening contractures in one patient and would not have survived on bare clavicular bone in the other. An island latissimus dorsi musculocutaneous flap ensured satisfactory functional and aesthetic results. Hematoxylin and eosin-stained tissues reported peripheral mass free from residual tumor. Follow-up of 18 months in the first case and 12 months in the second case did not show recurrence.

Keywords: Dermatofibrosarcoma; Latissimus Dorsi Flap; Mohs Surgery; Recurrence

Introduction

Dermatofibrosarcoma Protuberans (DFSP) is a rare slowly growing locally aggressive dermal malignancy that rarely metastasizes and accounts for less than 0.1% of all malignancies [1-3]. Its mean size ranges from 2 to 3.5 cm [4]. DFSP is known for its high recurrence rate following excision. The standard therapy has been wide local excision with 2-5 cm safety margins, or Mohs controlled surgery [4,5]. Defect coverage by a split-thickness skin graft is recommended for easy detection of potential recurrence. However, in this article, a reconstructive flap was used to avoid graft shrinkage in the front neck region in one case and to cover the clavicle bone denuded from periosteum in the other. The presentation aims to encourage therapy by simultaneous wide

deep tumor excision, Mohs micrographic defect examination, and durable reconstructing flap for presented cases with giant repeatedly recurrent DFSP.

Case Presentations

Two adult male patients presented with DFSP stage IIB according to Hao et al. [4] staging system, one seated the front aspect of the neck and the other occupied the right-side upper chest and shoulder. Lesions were painless and history revealed repeated recurrences after simple excisions. There were no palpable regional lymphadenopathies. Investigations included local histopathologic assessments and systemic search for secondaries though chest x-ray, abdominal ultrasonography, and bone scintigraphy; magnetic resonance imaging was not available. Both patients underwent wide and deep in-bloc excision with 2-4 cm safety margins and concomitant Mohs micrographic periphery and floor verification. The resulting large defect was reconstructed by Latissimus Dorsi Musculo-cutaneous (LD) island flap. The muscle insertion to the humerus bicipital groove was sectioned to increase its arc of rotation. A couple of centimeters thick muscular sleeve around the long thoracodorsal artery and its venae concomitants, the main blood supply, was preserved to protect the transfer against potential excessive tractions or torsions. Fixing stitches between the skin paddle periphery and the muscle were used to avoid eventual vascular compromise by accidental enter-layer sheering during translocation. Flap inset in the recipient area was secured by sutures in two layers. The flap donor area was closed over suction-drainage. Histopathological results of hematoxylin and eosin-stained tissues did not show any peripheral mass residual tumor characters. Wounds healed within two to three weeks without problems.

Case 1: A 32-year-old patient presented with a 30-mm-diameter red ball-like nodule, pedunculated and partially ulcerated on a 10x12 cm scarred area of skin in the sub-hyoid neck region (Figure 1a). The mass had started a few years before and the last conventional excision had been one year prior to presentation. A punch biopsy confirmed the presence of spindle fibroblasts arranged in storiform clusters, characteristic of DFSP. The entire lesion along with the surrounding scarred skin was excised under general anesthesia (GA) without intraoperative histological control and the resulting lack of skin was addressed with two opposing Limberg apparently heathy skin and platysma flaps (Figure 1b). Unfortunately, postoperative paraffin sections of the excised tissue detected DFSP cells. Within ten days, the affected skin area including the local flaps had to be excised again and a temporary split skin graft was used to close the wound when the arrangements for frozen-section testing were disrupted (Figure 1c). Three weeks later, a wider and deeper excision was under GA performed involving healthy tissues, namely 2 cm upper, 2 cm left and right, and 4 cm lower margins, the deep investing fascia of the neck, and the front layer of the pre-tracheal and lower sternocleidomastoid muscles. Frozen sections of the defect edges and floor did not show residual tumor cells and the front aspect of the neck was reconstructed with a LD flap, tunneled through an infraclavicular skin bridge. Over 18 months of follow-up, no signs of recurrence were observed (Figure 1d).

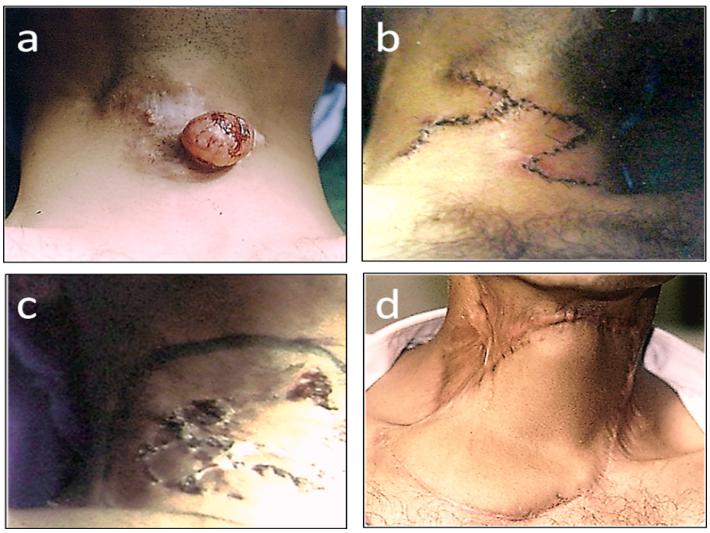

Figure 1: A cervical recurrent DFSP a: Preoperative image showing ulcerated 3 cm nodule on top of an old unhealthy skin graft in the central area of the neck, diagnosed by biopsy as DFSP; b: Wide excision without frozen section study followed by reconstruction with local tissue double opposing flaps; c: Recurrence again after repetition of wide excision and defect reconstruction by skin graft without frozen tissue assessment; d: Eighteen-months postoperative photograph of reconstruction by LD flap following a wide deep excision combined with Mohs verifications

Case 2: A 35-year-old patient presented with a recurrent ulcerating nodular skin lesion on his right upper chest (Figure 2a), initially misdiagnosed elsewhere as a spontaneous keloid. An excisional biopsy revealed DFSP. The tumor measured 11x25 cm. A plain x-ray of the right clavicle showed no signs of infiltration. A chest computed tomography scan revealed no lung metastases. Under GA, a wide and deep excision involving the apparently normal medial and lateral couple of centimeters, superior and inferior 3 cm skin boundaries, the healthy underlying superficial fibers of the deltoid, pectoralis, and trapezius muscles, and the intact clavicular periosteum. An island homolateral LD flap was used to reconstruct the thoracic and shoulder large defect area. Follow-up over a year showed no relevant issues (Figure 2b).

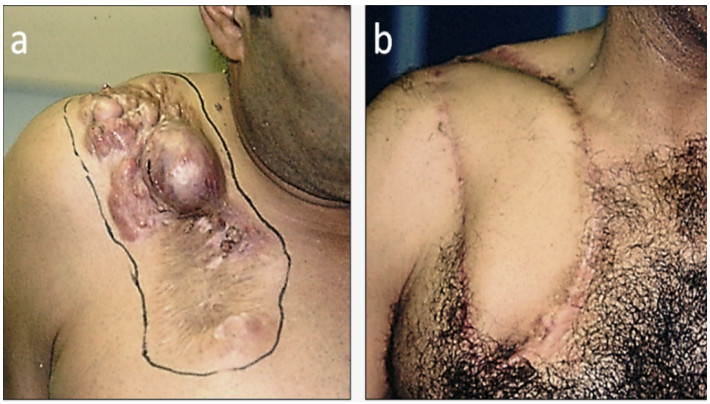

Figure 2: A giant recurrent DFSP on the right shoulder and upper chest region a: Preoperative image; and b: One-year postoperative image.

Discussion

DFSP was in 1924 labeled as "dermatofibroma precursor of sarcoma" by Darier and Ferrand [6] and a year later, the name was updated by Hoffman [7]. DFSP is a low-grade sarcoma of the dermis. It usually begins as an indurated bluish-red, fibrotic extended plaque base on which multiple slowly growing nodules subsequently arise and may ulcerate. It is usually painless and in early stages it may be quite difficult to distinguish it from a keloid or histiocytomas. DFSP occurs at any age, with its incidence being so rare in children [5] and increasing between the 3rd and 4th decades of age [4,8]. Differential diagnoses include fibroma, schwannoma, neurofibroma, lipoma, and melanoma. A dermal histopathologic uniform spindle-like fibroblasts arranged in a storiform pattern represent the specific character. Patients usually seek treatment first at dermatology clinics. The earlier the patient reaches the surgeon and oncologist the less morbidity and easier excision and reconstruction are expected. Nodal and systemic metastases are rare and may regress with chemo and radio therapy. DFSP has a 50% recurrence rate after conventional surgical excision [7]. It has always been recommended to include the apparently normal underlying tissue layer with the excised lesion [8, 9,10]. Mohs micrographic surgical discipline reduces recurrence rates by almost 10 times [11,12]. Reconstruction with split-thickness skin grafts has been advised to aid in early detection and diagnosis of eventual recurrence, despite the high risk of inducing contractures in flexion areas. Flaps yield durable cosmetic contours and are necessary to cover bones, cartilages, and tendons without periosteum, perichondrium, and paratenon, respectively. Reconstructive flaps have been favored after wide excision of large DFSP [8,13-16] and in 64 out of 103 immediate reconstructions after excision of sarcomas in extremities [17]. In conclusion, the ideal wide local excision of recurrent extensive DFSP should include 2 to 4 centimeters of safety margins and a deeper layer of apparently normal tissue. The borders and floor of its remaining defect should be checked by frozen sections authentication before reconstruction with a flap.

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