
Three-dimensional histology for dermatofibrosarcoma protuberans: Case series and surgical technique

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Background: Dermatofibrosarcoma protuberans (DFSP) is a low-grade malignant skin tumor that may also infiltrate dermis and subcutaneous tissue. Although the mainstay of treatment has been wide local excision, during the last decade three-dimensional (3D) histology surgery has proven very effective for the treatment of this tumor.

Objective: We sought to evaluate the effectiveness of 3D histology surgery for the treatment of DFSP.

Methods: We retrospectively reviewed charts of patients with DFSP treated in our unit with 3D histology surgery between April 2000 and May 2011. Age at onset, gender, duration of tumor, previous treatment, lesion site, number of surgical stages, postsurgical defect size, closure technique, and follow-up were registered.

Results: A total of 29 patients were included. Mean patient age was 40.7 years. Fifteen lesions were located on the trunk, 11 on the extremities, and 3 in the head and neck region. Twelve patients had primary tumors and 17 were referred to us after incomplete excision. The average number of 3D histology surgical stages required for tumor clearance was 1.4. Mean postsurgical defect size was 26 cm². All lateral and deep borders excised were tumor-free. Mean follow-up period was 68 months (range 12-142 months) with a 0% recurrence rate.

Limitations: This was a retrospective review.

Conclusion: Three-dimensional histology surgery with paraffin sections is effective for the treatment of DFSP and feasible in an outpatient setting. The low recurrence rates obtained confirm the oncologic efficacy of the procedure. Furthermore, designing closure on the basis of focally affected margins improves functional and aesthetic outcomes without compromising oncological effectiveness. (J Am Acad Dermatol 2012;67:991-6.)

Key words: Burow triangles; dermatofibrosarcoma protuberans; Mohs micrographic surgery; paraffin sections; recurrence; three-dimensional histology surgery.

Dermatofibrosarcoma protuberans (DFSP) is an uncommon, mesenchymal tumor of the skin, which originates in the dermis and extends deep into the subcutaneous tissue.¹ It has an estimated incidence of 0.8 cases per million per year and accounts for less than 2% of soft-tissue sarcomas.^{2,3} The clinical appearance may vary. Initially, it may present as a slowly growing indurated plaque that can eventually transform into a violaceous to red-brown painless nodule, most commonly on the

Abbreviations used:

DFSP: dermatofibrosarcoma protuberans
MMS: Mohs micrographic surgery
3D: three-dimensional

trunk (50%-60%) or upper limbs (25%) and less often on the head and neck.³ Clinical differential diagnosis includes keloids, large dermatofibroma, epidermal

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cyst, anetoderma, and morphea. DFSP diffusely infiltrates the dermis and subcutaneous tissue with peripheric tentacle-like projections; furthermore invasion of fascia and muscle can also occur.⁴⁻⁷ Even though DFSP are low-grade malignant tumors, after multiple recurrences, they may degenerate into fibrosarcomatous DFSP, a more malignant form of fibrosarcoma with the ability to metastasize regionally and distantly.⁸⁻¹¹

As for management, DFSP has been initially treated by wide local surgical excision, with the recommended standard treatment being en bloc resection of the tumor with a 3-cm margin of surrounding nonaffected skin, including the underlying fascia.^{5,12}

One of the major disadvantages of this approach is that such margins are often impossible to obtain, especially in cases in which the tumor is located near critical facial structures. Currently the best surgical treatment of DFSP is that which allows complete analysis of the piece, sparing surrounding tissue, while still achieving clear margins.¹²⁻¹⁵ The concept of histologic study of all margins of any given tumor was first introduced in 1941 by the American surgeon Frederic Edward Mohs, who originally used chemosurgery.¹⁴ After its original description, several schools modified both the name and the technique itself. Recently, Moehrle et al¹⁶ recommended replacing all these appellations by the expression “three-dimensional histology,” which is a generic and universal name for a surgical concept, and a term that, in our opinion, all those performing this type of surgery should use.

The following work presents our experience in treating DFSP with 3D histology surgery with paraffin sections.

METHODS

The medical records of all patients with DFSP treated in our unit between April 2000 and May 2011 were reviewed. Diagnosis of DFSP was confirmed in all patients by biopsy. Patient demographics, along with clinical and surgical data, are summarized in [Table I](#).

Surgical method

Oral and written informed consent was obtained before surgery. All patients were treated by a single surgeon (P. R.). Depending on tumor and patient characteristics, the procedure was performed on an inpatient or outpatient basis, most frequently under

local anesthesia. First the lesion was delimited as a geometric figure with a 1- to 1.5-cm margin, depending on the local anatomy and tumor characteristics ([Fig 1](#)). Then perpendicular incision (at a 90-degree angle) following the marked borders was performed including fascia and muscle fibers based on the prior biopsy. After careful hemostasis, the wound was

covered with antibiotic ointment, petrolatum gauzes, and a compressive dressing that was left in place until the second stage. To orient the excised piece for histologic analysis, a suture was placed in both the specimen and the patient.

The horizontal blocks (ie, deep margins) were sectioned horizontally from the excised tumor (as in conventional Mohs micrographic surgery [MMS]) and the vertical blocks (ie, lateral margins) were sectioned with

margin strips of 3 to 4 mm, and placed in pieces in cassettes used for routine procedures, pressed with their outer margin facing down ([Fig 1](#)). After rush fixation for 2 hours in 60°C hot formalin, all specimens were embedded in paraffin and sectioned using routine procedures. Twenty hours later, the hematoxylin–eosin–stained slides were reviewed by the dermatopathologist together with the dermatologist, for identification of possible positive margins.

The next day further excision or repair was performed. Whenever deep margins were affected, excision of precisely localized positive areas with additional 10- to 15-mm margins including fascia or muscle was performed ([Fig 1](#)). Conversely, when the excised borders were focally and superficially affected, when feasible and without compromising the oncological resection, they were resected within the Burrow triangles, generally used for closure either directly or with local flaps. Thus, reconstruction was conditioned by the affected margins, which avoided further surgery that delayed final closure ([Fig 1](#)).

Follow-up

Postoperative revisions were scheduled every 6 months for the first 5 years and from then on, once a year. At each revision thorough clinical examination was performed and if any sign of recurrence was suspected, biopsy and imaging tests (ultrasonography or magnetic resonance imaging) were performed.

CAPSULE SUMMARY

- Dermatofibrosarcoma protuberans is an uncommon infiltrative dermal and subcutaneous tumor.
- Three-dimensional histology surgery with paraffin sections is an effective and reproducible treatment in an outpatient setting and does not require a steep learning curve.
- Designing the closure on the basis of the focally affected margins improves functional and aesthetic outcomes.

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