Dermatofibrosarcoma protubercans (DFSP) is a low grade fibrosarcoma with a tendency for local recurrence and limited potential for metastasis. If inadequately treated, it can be very destructive. Tumors with a significant sarcomatous component histologically are at higher risk for developing metastases. There have been varying degrees of success in the management of this growth, therefore, DFSP can represent a challenge for dermatologists. The most recent research suggests that wide excision or a modified Mohs technique represent the best interventions with lowest recurrence rates.

**Diagnosis and Management**

Current management guidelines from the National Comprehensive Cancer Network stress the importance of careful follow-up whenever there is a high index of suspicion for DFSP. The guidelines note that delayed diagnosis or initial misdiagnosis and subsequently large tumor size at time of diagnosis are common, prompting the authors to recommend appropriate and confirmatory immunostaining in all cases of suspected DFSP.

Light microscopy may be sufficient to diagnose DFSP. If samples are non-representative or if analysis of the specimen does not confirm the diagnosis, immunostaining for CD34 and factor XIIIa is indicated. DFSP is CD34 positive and non-reactive to FXIIIa. Even though immunostains using CD34 are helpful, they may be negative in transitional zones and in the fibrosarcomatous component of a DFSP.

Once the diagnosis is confirmed, surgical excision is indicated with the goal of removing the entire malignancy in the initial surgery. The size and location of the tumor may determine the ideal approach—either local excision with wide margins or modified Mohs technique. Tumor extensions can be deep and difficult to appreciate clinically.

Margin recommendations for local excision range from 1-3cm, though the most current data support margins at the upper end of the range at 3cm.

Retrospective analysis of records of 24 DFSP patients treated with wide resection (one patient with fibrosarcoma had adjuvant radiation) found a 100 percent local recurrence-free survival at median follow-up of 54 months. The 24 patients in the analysis included 11 who received definitive wide resection after diagnostic excisions elsewhere and 13 who were treated for recurrent tumors following surgical treatment elsewhere. Upon presentation, DFSPs in patients in the recurrent group were larger and deeper than in the primary group. Eighty percent of cases with bone involvement were in the recurrent group.

Retrospective review of another set of patients treated for DFSP through local excision with margins of 3cm or more confirmed that wide local excision decreases local regional recurrences. Disease-free survival rates were 86 percent at five years and 76 percent at 10 years. Overall recurrence rate was 16.7 percent. Thirty percent of these recurrences were after five years. Biopsy-confirmed negative margins are essential prior to reconstruction.

Mohs micrographic surgery (MMS) is a tissue-sparing alternative to wide excision that allows the surgeon to immediately confirm negative mar-

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**Diagnostic Considerations**

A recent survey has provided some insight into a reported clinical variant of DFSP known as pre-protuberant DFSP. Of 143 patient cases reviewed, DFSP was described as protuberant ab initio in 81 cases and initially non-protuberant (npDFSP) in 62 cases. Among the findings regarding npDFSP:

- Initial misdiagnosis was common
- Age at onset was similar to that for DFSP
- Mean duration of non-protuberant stage was 7.6 years
- 29 percent of npDFSP were described as morphea-like
- 19 percent of npDFSP were described as atrophoderma-like
- 42 percent of npDFSP were described as angioma-like

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**Identifying Optimal Surgical Approaches for Dermatofibrosarcoma Protuberans**

Recent data confirm the role of wide excision or Mohs micrographic surgery for the management of this soft-tissue tumor.

By Jonathan Wolfe, MD
gins. This approach may be preferable to wide local excision in cosmetically sensitive areas or in cases of recurrence. One review reported the average recurrence rate of DFSP after MMS was 0.6 percent with a total recurrence rate of 1.6 percent. A recent study further supports the role MMS in management of DFSP. Evaluation of clinical outcomes of modified Mohs surgery in 22 patients with histologically confirmed DFSP revealed no local recurrence at a mean follow-up of 54 months. The authors recommend paraffin sectioning and three-dimensional histological evaluation as “an accurate additional tool for treatment optimization.”

Follow-Up
Depending on patient history and anatomic location of the tumor, wide local excision or modified Mohs micrographic surgery are clearly surgical interventions of choice for DFSP. Though adjuvant radiation therapy has been shown beneficial in management of DFSP, it is not indicated for the majority of patients. Instead, current recommendations suggest radiation when positive margins are present following resection or when further surgery would result in cosmetic or functional impairment. Radiation therapy may be of benefit for those tumors with a significant fibrosarcomatous component.

Careful follow-up is crucial for all patients with DFSP. Though data support the efficacy of wide excision or MMS to eradicate tumors with low risk of recurrence, DFSP nonetheless has long been associated with a high rate of recurrence and thus we suggest follow-up at six- to 12-month intervals. Additionally, patients must learn the proper approach to skin self-exams and conduct them regularly.

New In Your Practice

Melanoma Update. Women who have had any kind of skin cancer may be at greater risk for developing melanoma. A study (Cancer, Feb. 1, 2006) of more than 67,000 white postmenopausal women found that those with a history of non-melanoma skin cancer were 70 percent more likely than other women to develop melanoma during the study period, suggesting genetic influences make some people vulnerable to skin cancers of all kinds.

Ultimate Protection. Finding the right product to recommend to patients can be difficult. Fallene Ltd. has released its Solar Protection Formula, a new professional brand of products for dispensing that protect skin with physical UVB and UVA sunblocks. The line includes Tinted/Concealer SPF65, which comes in three shades; non-comedogenic Clear SPF65; water- and sweat-resistant Chemical Sunscreen Free SPF58; and lanolin-free LipTECT SPF45.