Atrophic Dermatofibrosarcoma Can be Tough Call

BY BRUCE K. DIXON
Chicago Bureau

CHICAGO — Atrophic dermatofibrosarcoma protuberans is an underrecognized variant that must be treated with wide local excision, Dr. Shari Clarke said at the annual meeting of the American Society for Dermatologic Surgery.

Although both wide local excision and Mohs micrographic surgery are considered treatments of choice, the latter quickly is becoming the favored method because of its lower rate of recurrence, she said.

In presenting three cases of the atrophic variant of dermatofibrosarcoma protuberans (DFSP), she explained that these lesions are slow-growing, locally aggressive fibrohistiocytic tumors that rarely metastasize but have a marked tendency toward local recurrence.

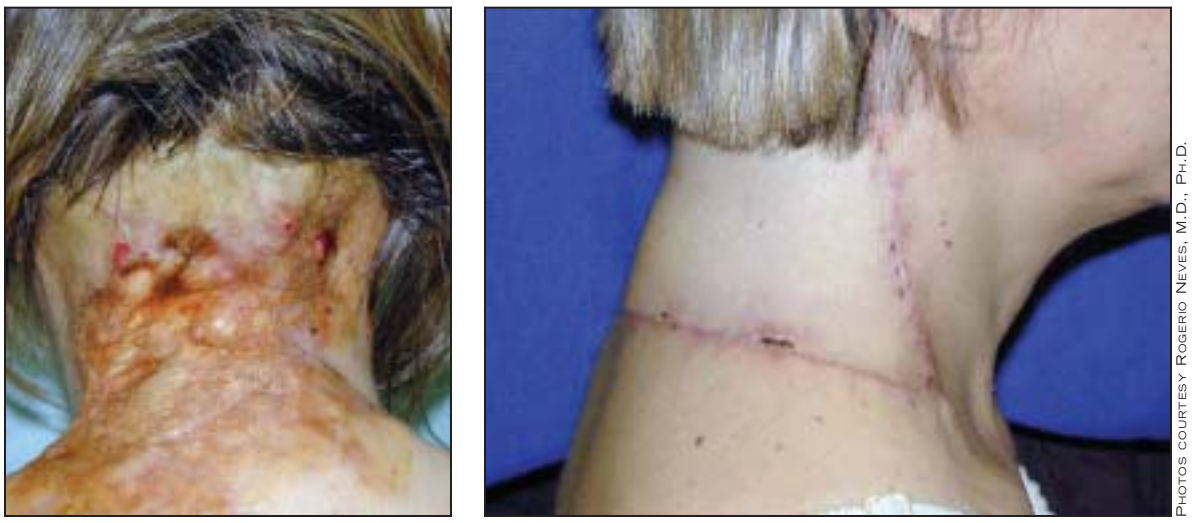
"Clinically, DFSP presents as indurated violaceous plaques which later develop nodules, and they're most commonly located on the trunk," explained Dr. Clarke of the department of dermatology at the Milton S. Hershey Medical Center, Pennsylvania State University, Hershey.

Histologically, DFSPs are characterized by monomorphous spindle cells in a storiform pattern that infiltrate between adipocytes. Later, the tumor can involve the upper dermis, deeper subcutaneous fat, or striated muscle, which correlates with the development of nodules and, as "we've demonstrated in our cases, the CD34 staining is very useful in distinguishing DFSPs from other fibrohistiocytic tumors," Dr. Clarke explained.

A total of five distinct early clinical variants of DFSP have been described in the literature, including confluent nodules forming sclerotic plaques, keloidlike sclerotic plaques, tumor, angiomalike, and atrophic DFSP. As exemplified in three cases described by Dr. Clarke, atrophic DFSP may or may not develop nodules in the later stages. In the first two cases, wide excision was used because the procedures were performed by a plastic surgeon.

The third case, involving the crural fold, was performed by a Mohs surgeon.

The first patient, a 43-year-old woman with a 13-year history of asymptomatic nodules that began as an atrophic plaque on the back of her neck, was clinically diagnosed as having multiple neurofibromas. She presented for a second opinion as the nodules continued to enlarge.



Photos courtesy Rogerio Neves, M.D., Ph.D.

A patient with DFSP is shown before undergoing wide local excision (left) and 3-months post excision (right).

A punch biopsy showed monomorphous spindle cells in a fibrous stroma infiltrating the septa between the adipose. CD34 stains were positive and she was given a diagnosis of DFSP, but since her original clinical exam showed atrophic plaque, the diagnosis was changed to atrophic DFSP, Dr. Clarke explained.

A wide excision with 2-cm margins was performed, producing a gross examination specimen measuring 20 by 15 cm. Intraoperative frozen sections with CD34 showed clear 2-cm margins and the wound was reconstructed with a transverse rectus abdominal muscle cutaneous flap utilizing the inferior epigastric artery and vein for blood supply.

The second patient was a 14-year-old girl who presented with a 7-year history of an enlarging, asymptomatic plaque on her right thigh. She was clinically diagnosed as having a keloid and treated without improvement. Surveillance was stopped for a number of years because the patient thought it was "just a scar," Dr. Clarke said, but the nodules continued to enlarge.

On examination, there was an atrophic plaque with scattered nodules that was found on biopsy to be a DFSP.

As with the first patient, the diagnosis was changed to atrophic DFSP.

"She had an excision with 2-cm margins, and the excised specimen was about 30 by 15 cm. Resection was performed down to the quadriceps complex with the 2-cm margins and intraoperative frozen sections showed clear margins," Dr. Clarke said. Reconstruction consisted of a split thickness skin graft from the uninvolved leg.

The third patient was a 40-year-old woman with a 4-by-2.5 cm violaceous atrophic plaque of the right crural fold, said Dr. Clarke, explaining that the patient initially was biopsied and misdiagnosed as having a dermatofibroma.

A repeat biopsy and tumor appearance confirmed atrophic DFSP, which was cleared in two stages using Mohs surgery. The final defect measured 11 by 6 cm. The reconstruction used an advancement flap. Histology showed spindle cells, and a CD34 stain was grossly positive. There has been no evidence of recurrence in the intervening 8 years, said Dr. Clarke, who had no relevant conflicts to disclose.

"Atrophic DFSP, otherwise known as DFSP nonprotuberans or morpheaform DFSP, has been reported infrequently in the dermatologic literature, and like typical DFSP, the atrophic variant has an insidious and aggressive local growth pattern, a similar age of onset, and predominant truncal location," she said, noting that there appears to be a slight female predominance.

Although the atrophic variety is clinically distinct, the treatment and histology are the same as they are for typical DFSP. Adjuvant treatment with imatinib looks promising for very difficult cases.

"Exciting advances in the molecular genetics behind DFSPs have recently been elucidated and have important implications for treatment," she explained, adding that 90% of DFSPs overexpress platelet-derived growth factor β.

"This is important because imatinib mesylate [Gleevec] currently approved for treating chronic myeloid leukemia, acts as a selective tyrosine kinase inhibitor of the platelet-derived growth factor receptor β and has been used successfully in isolated case reports for the presurgical treatment of locally advanced DFSPs and inoperable recurrent or metastatic disease," she said.

Dr. Clarke suggested using 400 mg of imatinib mesylate, the dose used for gastrointestinal stromal tumors, either daily or twice daily. "The side effects are typically mild, although severe edema and liver toxicity have been reported in the elderly," she said. ■

MAL-PDT Reveals Cosmetic Edge Over Excision in BCC

BY NANCY WALSH
New York Bureau

VIENNA — Treatment with methyl aminolevulinate-photodynamic therapy was as effective as surgery for superficial basal cell carcinoma, Dr. Rolf-Markus Szeimies reported in a poster session at the 16th Congress of the European Academy of Dermatology and Venereology.

Among the methods for removal of basal cell carcinomas are simple excision, Mohs surgery, radiotherapy, curettage/electrodessication, and cryosurgery, with the choice of treatment depending on type, size, depth, and location of the lesion.

Methyl aminolevulinate-photodynamic therapy (MAL-PDT) has previously been shown to be as effective as cryotherapy for removal of these lesions and to have superior cosmetic results. Now, in the first multicenter randomized trial comparing MAL-PDT with simple excision, similar findings have been found, according to Dr. Szeimies of the department of dermatology, Regensburg (Germany) University Hospital.

A total of 196 patients whose mean age was 63.8 years were included in the study. The mean number of lesions per patient was 1.4, and the mean diameter of the lesions was 12.4 mm.

Patients randomized to MAL-PDT underwent two treatment sessions 7 days apart, with the option of repeat treatment at 3 months if clinical response was incomplete. Those randomized to surgery underwent simple elliptical excision with 3-mm margins from the estimated edge of the lesion.

The lesion complete response rate was 87% with MAL-PDT and 89% with excision, confirming the noninferiority of MAL-PDT to surgery, wrote Dr. Szeimies.

Results were similar in the two groups for lesions on the trunk and neck, with MAL-PDT and excision having complete response rates of 85% and 89%, respectively. For lesions on the face and scalp, MAL-PDT and excision showed complete response rates of 95% and 67%.

Complete response was not related to size of the lesion.

Investigator-rated cosmetic outcome favored MAL-PDT, with 87% of lesions having good to excellent outcome, compared with 58% of those in the surgery group.

Patients also preferred the cosmetic outcome with MAL-PDT, with 93% rating the outcome as good to excellent, compared with 81% of the patients in the excision group.

The study was sponsored by Galderma, which makes the MAL-PDT used in the study.

In a recent review of experience with MAL-PDT for basal cell carcinoma, Dr. Szeimies noted that, while surgery remains the preferred method of treatment, some patients—such as those with large lesions, poor vasculature, and concomitant use of anticoagulants or immunosuppressives—may be poor candidates for surgery.

Moreover, postsurgical keloid or dystrophic scarring is common, particularly on the trunk. "Because of the relatively low-risk nature of superficial [basal cell carcinoma], scarring problems should be taken into consideration when choosing a suitable therapy. Therefore, PDT may offer significant advantages over surgical or other destructive techniques" he wrote (Dermatol. Clin. 2007;25:89-94).

Dr. Szeimies disclosed no conflicts of interest. ■