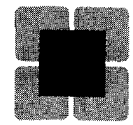

KENNETH J. TOMECKI, MD, SECTION EDITOR

Dermatofibrosarcoma protuberans of the hand

Report of a case treated with Mohs micrographic surgery

EDMUND R. HOBBS, MD; JOHN LOUIS RATZ, MD, FACP

■ A 35-year-old man had a dermatofibrosarcoma protuberans of the thumb. Instead of the wide excision conventionally used to manage this frequently recurrent tumor, Mohs micrographic surgery was performed. This conservative, tissue-sparing technique provides total margin control and yields cure rates that are superior to standard surgical methods.

□ INDEX TERMS: HAND NEOPLASMS; HAND, SURGERY □ CLEVE CLIN J MED 1988; 55:252–256

DERMATOFIBROSARCOMA protuberans (DFSP) is a relatively uncommon, malignant soft-tissue neoplasm that rarely presents on the hand. The tumor is locally aggressive and recurs frequently, even after wide excision. We report a case of DFSP occurring on the thumb and discuss its treatment by Mohs micrographic surgery (fresh-tissue technique).

CASE REPORT

A 35-year-old white man came to The Cleveland Clinic Foundation with a 6-mm firm dermal nodule in a surgical scar overlying the radial aspect of the metacarpophalangeal joint of the left thumb (*Fig. 1*). Two months earlier, his local physician had excised a tumor there and closed the resulting defect primarily. Histologic analysis disclosed a dermis replaced by spindle-shaped cells in a storiform pattern created by cartwheel-like arrays about collagenous centers (*Fig. 2*). The interlacing bands and whorls of fibroblasts extended to the deep margin of the resection. A diagnosis of DFSP was rendered, and the patient was referred for additional treatment.

With local anesthesia, the tumor was excised with the Mohs micrographic surgery (fresh-tissue technique), a procedure that permits the surgeon to view the lateral and deep margins of the excisional specimen in their entirety. The first excisional specimen was bisected, and Mohs frozen sections revealed tumor involving the deep margin of both segments (*Fig. 3*). Because the lateral margins were free of tumor, the original wound did not need to be extended. A second layer was therefore excised horizontally below the level of the first excision and processed with the Mohs technique. The specimen showed all margins to be free of tumor. The resulting 1.9 x 2.2-cm defect (*Fig. 4*) was allowed to heal by second intention. On follow-up at 3, 6, 12, and 25 months postoperatively, healing was complete and the patient had full range of motion and no functional limitation (*Fig. 5*). There has been no recurrence.

DISCUSSION

DFSP arises in the dermis, invades locally, and rarely metastasizes. Its incidence has been estimated at 0.8 cases per million persons per year.[1] It accounts for only about 0.1% of all malignancies[2,3] and is most often detected in

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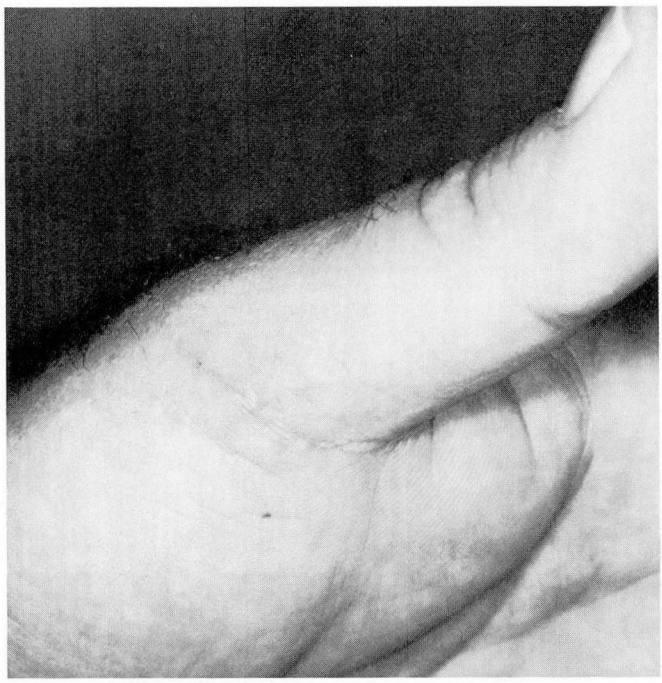

FIG. 1. Dermatofibrosarcoma protuberans is located in the surgical scar overlying the metacarpophalangeal joint of the left thumb.

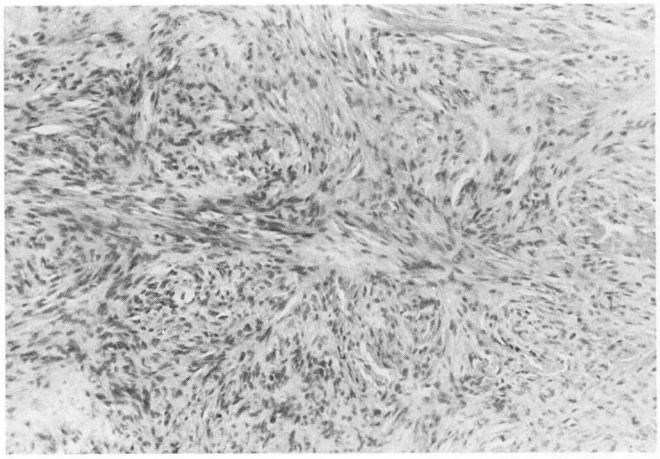

FIG. 2. Histopathologic specimen reveals the interlacing bands and whorls of fibroblasts that form the storiform pattern typical of dermatofibrosarcoma protuberans (hematoxylin-eosin, original magnification x10).

the second through fifth decades of life.[2,4] Men are affected slightly more often than women.[1–3] Evidence suggests that the fibroblast is the cell of origin.[5]

While DFSP can occur in skin in any anatomic location, most cases involve the trunk and proximal extremities.[6] In the seven largest series reported to date,[2–4,6–9] representing a total of 483 cases, only one case involved the hand.[6] In 1977, Schvarcz[10] reported a case thought to represent congenital DFSP of the hand, and in 1981, Wirman et al[11] also reported a case. DFSP has not been reported to arise on the palm.

The lesion usually presents as a dusky reddish-blue to reddish-yellow plaque. Its indolent behavior and nonspecific clinical features often cause it to be dismissed by patient and physician alike. In time, it can assume a large multinodular configuration, may become tender or painful, and may ulcerate. It is fixed to skin, but usually mobile over deep structures. In advanced cases, the tumor invades fascia and muscle.

As the tumor grows, its main body gives off multiple, pseudopod-like extensions that can only be examined microscopically. These projections infiltrate both laterally and deeply between normal collagen bundles, rendering the border of the tumor impossible to appreciate macroscopically. Additionally, the periphery of the

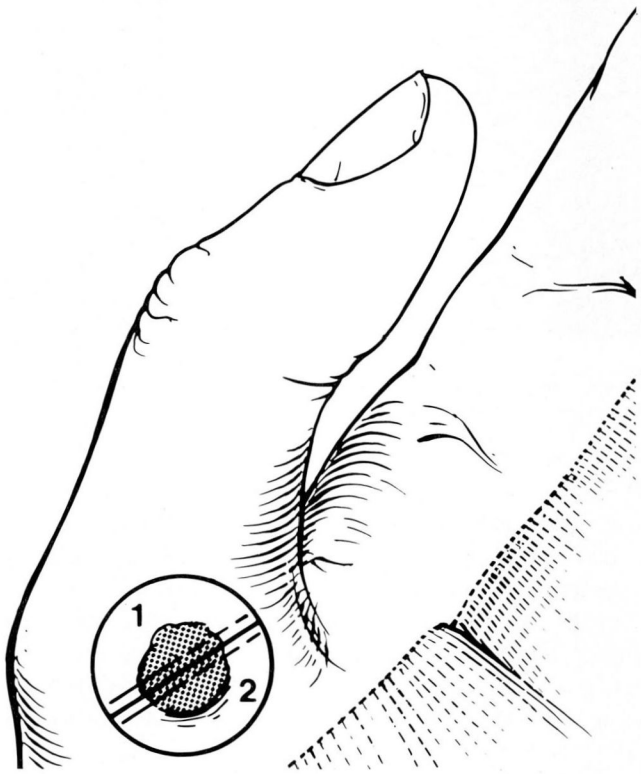

FIG. 3. A sketch map is used to facilitate orientation. The initial Mohs sections contained tumor at the deep margins (shaded areas), but the lateral edges were not involved. Tissue edges were marked with dyes that are visible histologically and are coded on the map as bars (for red) and dashes (for blue).

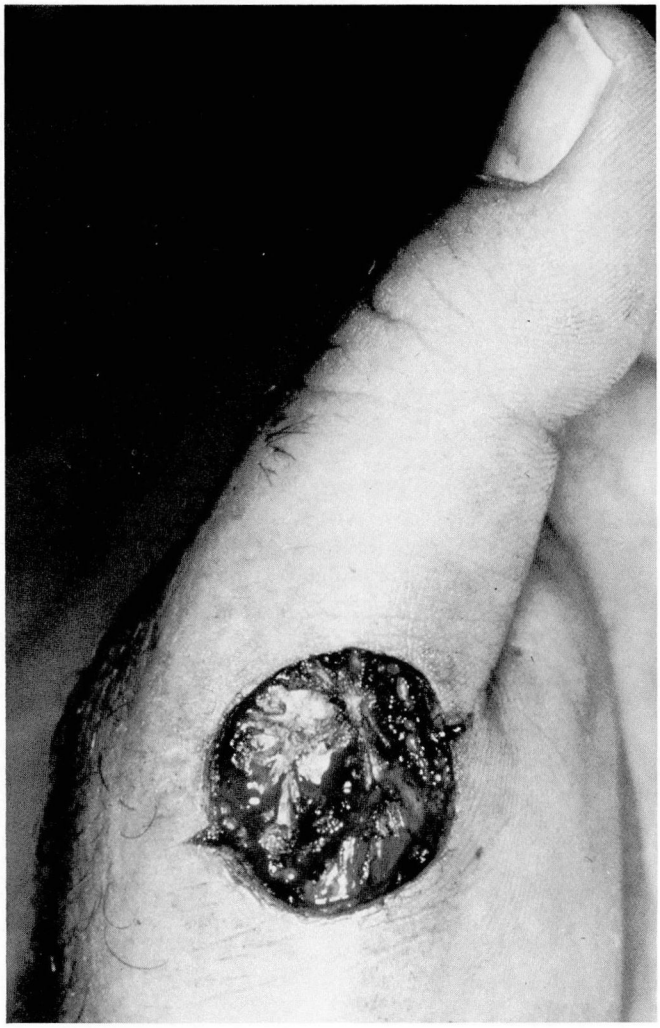

FIG. 4. The operative defect, after the second and final Mohs layer, measured 1.9 x 2.2 cm.

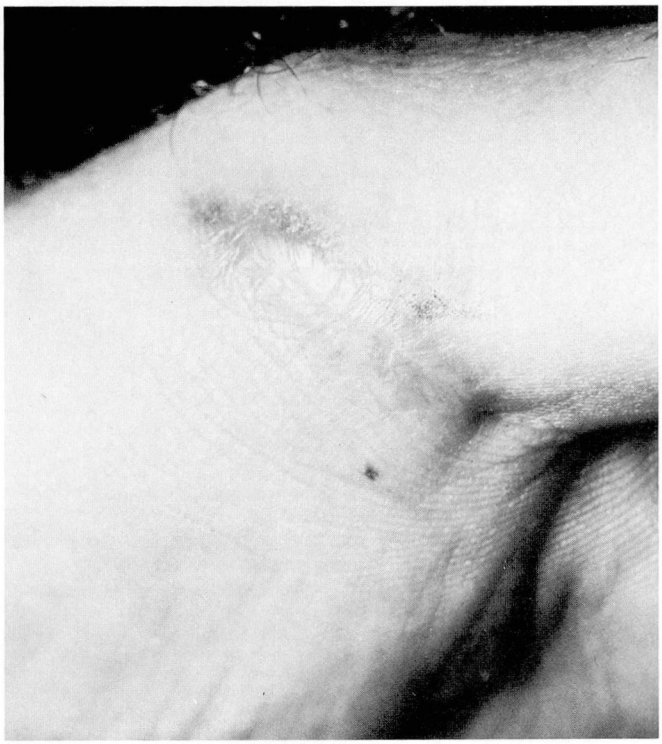

FIG. 5. The wound's appearance six months postoperatively, with healing by second intention. There is no functional deficit, and the patient has remained free of tumor after 25 months.

DFSP is more fibrous than the center,[6] often conveying an erroneous impression of encapsulation. These features of DFSP frequently result in incomplete excision or persistence of growth, even after wide excision.[2]

In 98 cases with follow-up reported by Taylor and Helwig,[6] the recurrence rate was 49%, with nearly 40% recurring in the first year after surgery. Of Hajdu's 119 cases,[7] 54% recurred after wide excision, mostly within one year. One fourth of those treated by Burkhardt et al[4] recurred, while 24% of those treated by Pack and Tabah[2] recurred. Such statistics have resulted in the current recommendation that DFSP be treated by wide excision with a 3.0-cm[1,3,12] or more[13] margin of clinically uninvolved skin down to and including fascia. However, even when managed in this fashion, recurrences are common. McPeak et al[3] reported an 11% recurrence rate in 11 patients so treated. Roses et al[12] had two recurrences in 10 patients in whom 3-cm margins were used, while Bendix-Hansen et al[1] took 3-cm margins in seven cases and had no recurrences. While such generous margins might appear prohibitive when applied to such a functionally important area as the hand, failure to remove the tumor completely inevitably results in more mutilating surgery.

A more rational and potentially more effective means of treating DFSP is Mohs micrographic surgery. With this technique, the clinically apparent tumor is excised with a conservative margin. The tissue is divided into smaller segments, carefully mapped, and marked with color-coded dyes so that the precise anatomic position of each piece can always be determined. Tangential frozen sections are cut to establish a continuous plane (*Fig.* 6) lateral and deep to the tumor. The pliable nature of fresh tissue allows it to be stretched or compressed as necessary to establish the needed component planes. The Mohs surgeon inspects each stained segment histologically and marks on the map the precise location of any tumor involving the margin. The surgeon then removes addi-

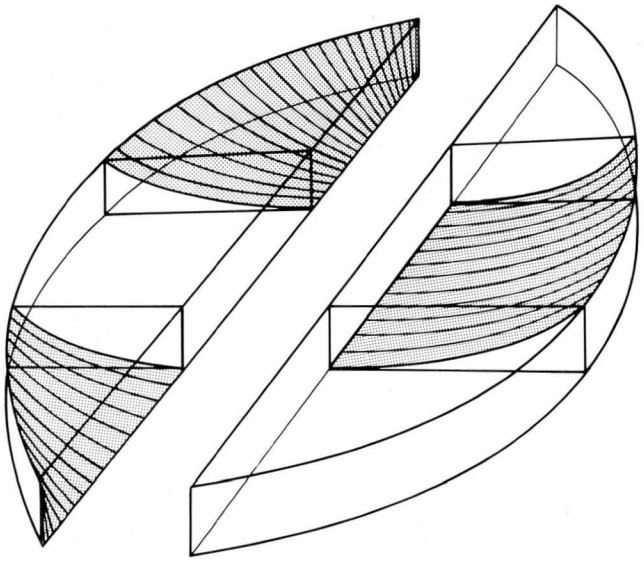

FIG 6. During processing, sections are cut to establish a continuous plane around the tumor. The tissue is stretched or compressed as needed to facilitate planes of section that would not be possible were the tissue rigid.

tional tissue from those exact sites.

Mohs sections permit the surgeon to view the lateral and deep margins of the excisional specimen simultaneously and to view them in their entirety. This total margin control, with its meticulous mapping procedure, differentiates Mohs micrographic surgery from standard margin control techniques and yields significantly better results. Just as the microscopic extensions of tumor cannot be appreciated grossly, so too can they be missed by any technique (including vertical step sectioning) that does not examine the margins completely. Mohs micrographic surgery is an ideal treatment for neoplasms that invade locally and seldom metastasize, such as DFSP. To date, no recurrences of DFSP following Mohs surgery have been reported. Mohs,[14] Robinson,[5] and Mikhail and Lynn[15] excised seven, four, and two lesions of DFSP, respectively, using Mohs' technique and did not observe any recurrences more than five years later. Peters et al[16] excised a facial DFSP with Mohs micrographic surgery and observed no recurrence within 3 $^1/_2$ years; Hess et al[17] used the technique for a myxoid DFSP and noted no recurrence after 18 months. A total of 10 cases of DFSP have been managed with Mohs micrographic surgery at the Cleveland Clinic,[18] with no recurrences in an average of 43 months of follow-up. These successes lead to the conclusion that Mohs micrographic surgery is the treatment of choice for DFSP.[17,19]

Mohs micrographic surgery is especially advantageous when DFSP involves the hand. The procedure completely eradicates the tumor, yet spares the maximum possible amount of normal tissue. Wide excision with arbitrarily specified margins invariably necessitates split-thickness skin grafts,[3] sometimes impairing appearance and function. The confidence generated by Mohs margin control allows immediate repair by the complete range of reconstruction methods, including second-intention healing where appropriate, as in the small defect that resulted in the case reported here. The Mohs procedure is often suited to the outpatient surgical setting and the use of local anesthesia. But, most importantly, Mohs micrographic surgery appears to provide a cure rate superior to that of wide excision.

While most recurrences of DFSP are noted within the first three years following surgery,[6,12] there are sporadic reports of late recurrences as much as 20 years later.[6] For this reason, the case reported here warrants continued close follow-up. We have treated a tumor that is rare on the hand, using the technique most likely to cure and least likely to cause disfigurement and functional impairment.

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REFERENCES

1. Bendix-Hansen K, Myhre-Jensen O, Kaae S. Dermatofibrosarcoma protuberans: a clinico-pathological study of nineteen cases and review of world literature. Scand J Plast Reconstr Surg 1983; 17:247–252.
2. Pack GT, Tabah EJ. Dermatofibrosarcoma protuberans: a report of thirty-nine cases. Arch Surg 1951; 62:391–394.
3. McPeak CJ, Cruz T, Nicastri AD. Dermatofibrosarcoma protuberans: an analysis of 86 cases—five with metastasis. Ann Surg 1967; 166:803–816.
4. Burkhardt BR, Soule EH, Winkelmann RK, Ivins JC. Dermatofibrosarcoma protuberans: study of fifty-six cases. Am J Surg 1966; 111:638–644.
5. Robinson JK. Dermatofibrosarcoma protuberans resected by Mohs' surgery (chemosurgery): a 5-year prospective study. J Am Acad Dermatol 1985; 12:1093–1098.
6. Taylor HH, Helwig EB. Dermatofibrosarcoma protuberans: a study of 115 cases. Cancer 1962; 15:717–725.
7. Hajdu SI. Pathology of Soft Tissue Tumors. Philadelphia, Lea & Febiger, 1979, pp 60, 83.
8. Gutierrez G, Ospina JE, deBaez NE, deEscorcia EK, Gutierrez R. Dermatofibrosarcoma protuberans. Int J Dermatol 1984; 23:396–401.
9. Gentele H. Malignant fibroblastic tumor of skin: clinical and pathological-anatomical studies of 129 cases of malignant, fibroblastic tumors from cutaneous and subcutaneous layers observed at Radiumhemmet

during period 1927–1947. Acta Derm Venerol (Stockh) 1951; suppl **27**:1–20.
10. Schvarcz LW. Congenital dermatofibrosarcoma protuberans of the hand. Hand 1977; **9**:182–186.
11. Wirman JA, Sherman S, Sullivan MR. Dermatofibrosarcoma protuberans arising on the hand. Hand 1981; **13**:187–191.
12. Roses DF, Valensi Q, LaTrenta G, Harris MN. Surgical treatment of dermatofibrosarcoma protuberans. Surg Gynecol Obstet 1986; **162**:449–452.
13. Martin RG. Neoplasms of the Skin and Malignant Melanoma. A Collection of Papers Presented at the Twentieth Annual Clinical Conference on Cancer, 1975, at the University of Texas System Cancer Center, M.D. Anderson Hospital and Tumor Institute, Houston, Texas. Chicago, Year Book, 1976, pp 243–250.
14. Mohs FE. Chemosurgery: Microscopically Controlled Surgery for Skin Cancer. Springfield, IL, Charles C Thomas, 1978, p 251.
15. Mikhail GR, Lynn BH Dermatofibrosarcoma protuberans. J Dermatol Surg Oncol 1978; **4**:81–84.
16. Peters CW, Hanke CW, Pasarell HA, Bennett JE. Chemosurgical reports: dermatofibrosarcoma protuberans of the face. J Dermatol Surg Oncol 1982; **8**:823–825.
17. Hess KA, Hanke CW, Estes NC, Shideler SJ. Chemosurgical reports: myxoid dermatofibrosarcoma protuberans. J Dermatol Surg Oncol 1985; **11**:268–271.
18. Hobbs ER, Wheeland RG, Bailin PL, Ratz JL, Yetman RJ, Zins JE. Treatment of dermatofibrosarcoma protuberans with Mohs micrographic surgery (in press).
19. Sagi A, Ben-Yakar Y, Mahler D. A ten-year-old boy with dermatofibrosarcoma protuberans of the face. J Dermatol Surg Oncol 1987; **13**:82–83.