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Distant Metastasis of Dermatofibrosarcoma Protuberans of the Scalp

—Case Report—

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Abstract

A 49-year-old man presented with a rare dermatofibrosarcoma protuberans (DFSP) of the scalp associated with local recurrence and distant metastasis to the lung and abdomen. An elastic-hard small mass on the right occipital scalp was initially treated by simple resection in another clinic. Ten years later, recurrent tumor was associated with infiltration to the calvarium, and resection was performed again also in another clinic. Approximately 1.5 years later, the patient was transferred to our clinic because of recurrence with intracranial involvement. Repeated relapses and metastasis to the lung were recognized despite surgery, chemotherapy, and local radiation. Eventually, the patient died of distant metastasis to the abdomen 17 years after the initial diagnosis. Scalp DFSP is very uncommon but is an aggressive scalp tumor, so initial wide local resection and local radiation therapy after surgery are important to prevent local recurrence and distant metastasis.

Key words: dermatofibrosarcoma protuberans, distant metastasis, recurrence, scalp tumor

Introduction

Dermatofibrosarcoma protuberans (DFSP), first described in 1924 as ''progressive and recurring dermatofibroma,''[5] is a nodular cutaneous tumor characterized by a prominent storiform pattern.[10] DFSP has since been considered as a fibrohistiocytic tumor of intermediate malignancy characterized by local recurrence.[2,7] DFSP has an incidence of only 0.8 cases per million persons per year,[13] and typically presents during early- or mid-adult life and predominantly in males. The location is most frequently on the trunk and proximal extremities, but may occur at almost any site.[2,7] Sites on the head and neck occur in up to 15% of cases.[2,7] The scalp is the most common site, accounting for about 30% of such cases.[2]

Here we describe a rare case of recurrent scalp DFSP followed by local infiltration and distant metastasis to the lung and abdomen.

Case Report

A 49-year-old man found an elastic-hard small mass on the right occipital scalp in 1981 and consulted a neighborhood orthopedician for resection of the growing mass in 1983. He received two further resections of the recurrent tumor in another clinic in February and March 1992. The histological diagnosis of the mass was DFSP (Fig. 1). Subsequently he was transferred to our clinic because of a recurrence

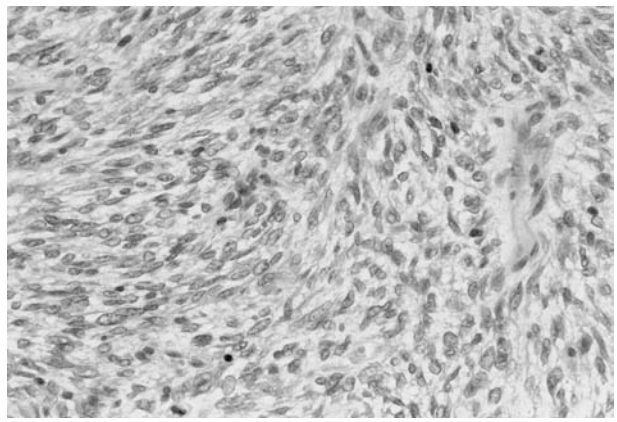

Fig. 1 Photomicrograph showing spindle cells arranged in the storiform pattern. Hematoxylin-eosin stain, ×360.

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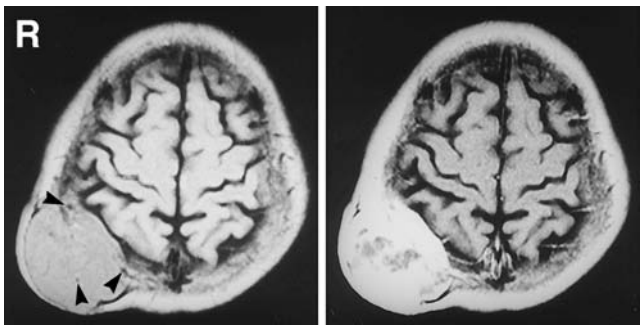

Fig. 2 T_1-weighted magnetic resonance images without (*left*) and with gadolinium (*right*) showing an isointense mass with flow void signs (*arrowheads*) and partly heterogeneous enhancement.

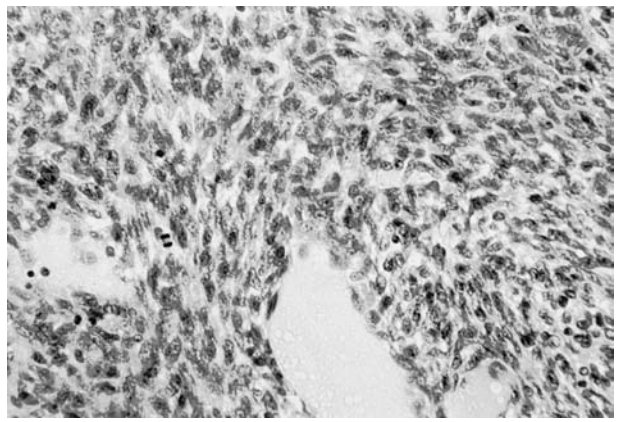

Fig. 3 Histological appearance. Part of the fibrosarcoma shows the fascicular pattern. Hematoxylin-eosin stain, ×350.

with bone infiltration in July 1993.

On admission, examination found a nodular scalp tumor on the right occipital region, which appeared highly vascularized and elastic-hard. Computed tomography (CT) revealed an enhanced mass in the subcutaneous and epidural space, associated with bone destruction. Magnetic resonance imaging also showed an enhanced mass with partial ring enhancement (Fig. 2). Cerebral angiography showed a highly vascular tumor fed by branches of the right external carotid artery.

Simple resection of the tumor was performed without difficulty in dividing the tumor from the epidermis and the dura mater. Huge multinodular scalp masses appeared in December 1993. Wide resection was recommended but rejected by the patient in favor of simple resection. Nine operations and three gamma knife procedures were performed for local recurrences, including sinus involvement and intradural masses for about 4 years. Gamma knife surgery only achieved mild growth inhibition of the tumor. A course of chemotherapy of middle dose of methotrexate (500 mg/m^2 body surface area) was also administered based on the chemosensitivity test using 3-(4,5-dimethylthiazol-2-yl)-2,5-diphenyltetrazolium bromide (MTT) assay. However, the disease remained refractory to treatment.

Metastatic involvement of the lung was also treated surgically twice during this period. The histology was same as that of the intracranial involvement. Skin necrosis and liquorrhea appeared in June 1997. He underwent wide resection and free latissimus dorsi myocutaneous flap transplantation. However, the histology of the tumor had become more malignant with sarcomatous transformation in some areas (Fig. 3). Moreover, he developed right facial nerve paresis and hearing disturbance in July 1998.

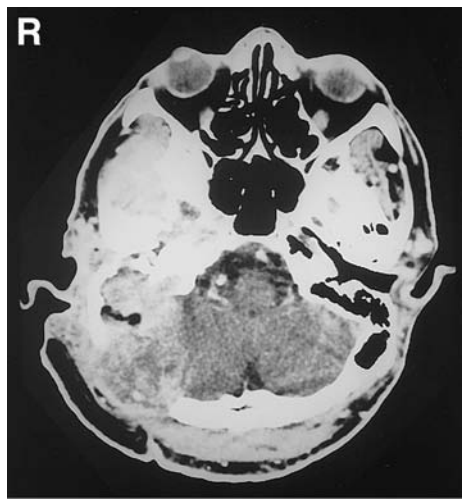

Fig. 4 Computed tomography scan showing intradural recurrences infiltrating into the petrosal bone and tympanic cavity.

Multiple intradural recurrences were recognized infiltrating into the petrosal bone and tympanic cavity (Fig. 4). He developed dysphagia and pneumonia in September 1998, as well as abdominal pain. Abdominal CT revealed a huge mass attached to the liver (Fig. 5). Sepsis developed despite antibiotic administration, resulting in death on November 2, 1998.

Discussion

The initial clinical manifestation of DFSP is usually the development of a firm, plaque-like lesion of the skin, often with a surrounding red to blue discolora-

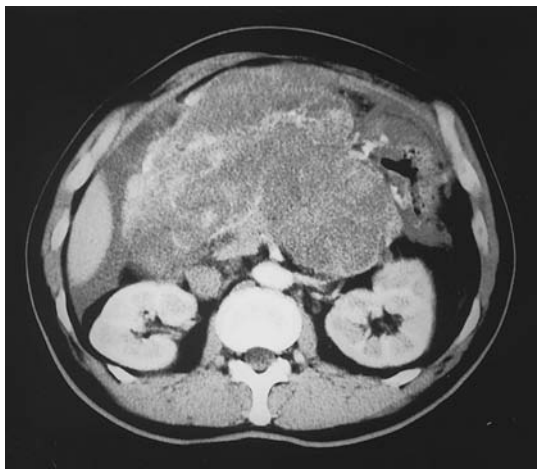

Fig. 5 Computed tomography scan showing an isodense huge mass with heterogeneous enhancement in the abdomen.

tion.[19] Then, one or more nodular lesions with the typical ''protuberant'' appearance develop. The mean size at surgery is approximately 5 cm and the skin overlying the tumor is taut or even ulcerated. Hemorrhage and cystic change are sometimes seen, but necrosis is rare.[7,19] The tumor on the trunk and extremities is fixed to the overlying skin but moves freely over the deeper tissues. However, the paucity of deeper tissue on the scalp means the tumor mass is fixed to the underlying periosteum and does not move freely early in its course.[17]

DFSP is locally aggressive and recurs in 50% to 75% of all patients.[2,14,19] Local recurrence usually develops within 3 years of the initial surgery. Recurrence after several years as seen in this case has also been reported.[14] The recurring and nonrecurring tumors show no differences in location, duration, size, or histology.

Metastasis is infrequent despite the locally aggressive behavior, and should be clearly discriminated from conventional sarcomas. Metastasis is seen in no more than 6% of cases.[2,14,16,21] The most common metastatic sites are the lung, bone, and lymphnodes.[7,14,16,21] In this case, lung and abdominal metastases developed 11 and 17 years after the initial surgery, respectively. Thus, long-term follow up may indicate higher metastatic rates than previously reported. Scalp DFSP, like this case, deeply infiltrates and destroys the bone and dura mater, and then invades into the subdural space and dural sinus. If invasion into the dural sinus develops, distant metastasis may occur more frequently.

DFSP must be differentiated from other fibrohistiocytic tumors, neurofibroma, myxoid liposarcoma, keloid, and hypertrophic scars.[8] DFSP has more uniformity and smaller cells displaying a more distinct storiform pattern than benign or malignant fibrous histiocytoma. In addition, there are fewer secondary changes such as giant cells and inflammatory cells. CD34 immunohistochemistry is very useful for the differentiation from a benign fibrous histiocytoma, since CD34 is almost consistently expressed in DFSP and rarely in benign fibrous histiocytoma.[20] Neurofibroma usually contains tacoid structures or other features of neural differentiation, and lacks high cellularity with mitotic figures. S-100 protein is present in virtually all neurofibromas and absent in DFSP.[8] Highly myxoid DFSP resembles myxoid liposarcoma in the prominent vasculature and bland stellate or fusiform cells. However, lipoblasts are completely absent in DFSP. DFSP contains fibrosarcomatous areas appearing as a fascicular pattern and usually plumper cells with more atypia as shown in this case.[6,22] Sarcomatous areas have lower immunoreactivity for CD34 than DFSP.[9] Sarcomatous areas are more commonly encountered in recurrent lesions and are associated with more local recurrence.[4] However, the cases with sarcomas arising in DFSP do not have an increased risk of distant metastasis, provided wide local resection with negative margins is performed.[9]

The optimum therapy for DFSP is initial wide local resection. Wide resection of more than 5 cm from the margin of the tumor[1] and Mohs micrographic surgery should be performed.[2,15] The risk of local recurrence correlates well with the extent of the wide excision.[9,15] A margin of excision of 3 cm or more results in a recurrence rate of 20%, compared with 2 cm or less leading to 41% recurrence.[18] In addition, skin transplantation such as latissimus dorsi myocutaneous flap transplantation must be considered as well.[11,13] Obviously a conservative approach as seen in this case cannot be justified if local recurrences of an incompletely excised lesion would seriously jeopardize a vital structure. The involvement of the dural sinus should be considered in cases of scalp lesion. Control of the bleeding is extremely important as DFSP is highly vascularized[17] and hemorrhagic.

There is little support for the value of radiotherapy[3,12,19] or chemotherapy. However, radiotherapy was effective in 10 cases, and three cases showed complete regression after only radiotherapy.[12] The present tumor was recurrent and more resistant to radiotherapy, so gamma knife surgery was mildly effective for the inhibition of tumor growth. Gamma knife radiotherapy may be more effective than conventional radiation against the primary lesion.

Chemotherapy using medium doses of

methotrexate was tried but had no apparent efficacy based on the MTT assay. Although the MTT assay is generally useful for the elimination of ineffective drugs, the results are not always predictors for the determination of effective agents.

Neurosurgeons should be aware of DFSP as an aggressive scalp tumor and the importance of initial wide local resection, and we recommend local radiation therapy after surgery.

References

1) Arnaud EJ, Perrault M, Revol M, Servant JM, Banzet P: Surgical treatment of dermatofibrosarcoma protuberans. *Plast Reconstr Surg* 100: 884–895, 1997
2) Barnes L, Coleman JA, Johnson JT: Dermatofibrosarcoma protuberans of the head and neck. *Arch Otolaryngol* 110: 398–404, 1984
3) Burkhardt BR, Soule EH, Winkelmann RK, Ivins JC: Dermatofibrosarcoma protuberans. Study of fifty-six cases. *Am J Surg* 111: 638–644, 1966
4) Connelly JH, Evans HL: Dermatofibrosarcoma protuberans: A clinicopathologic review with emphasis on fibrosarcomatous areas. *Am J Surg Pathol* 16: 921–925, 1992
5) Darier J, Ferrand M: Dermatofibromas progressifs et recidivants ou fibrosarcomes de la peau. *Ann Dermatol Syph* 5: 545–562, 1924 (Fre)
6) Ding JA, Hashimoto H, Enjoji M: Dermatofibrosarcoma protuberans with fibrosarcomatous areas. Clinicopathological study of nine cases and a comparison with allied tumors. *Cancer* 64: 721–729, 1989
7) Enzinger FM, Weiss SW: Chapter 14: Dermatofibrosarcoma protuberans. Fibrohistiocytic tumors of intermediate malignancy, *in*: *Soft Tissue Tumors, ed 3*. St Louis, Mosby, 1995, pp 325–349
8) Flethcer CDM, Evans BJ, Maccartney JC, Smith N, Wilson JE, McKee PH: Dermatofibrosarcoma protuberans: A clinicopathological and immunohistochemical studies with a review of the literature. *Histopathology* 9: 921–938, 1985
9) Goldblum JR, Reith JD, Weiss SW: Sarcoma arising in dermatofibrosarcoma protuberans: a reappraisal of biological behavior in eighteen cases treated by wide local excision with extended clinical follow up. *Am J Surg Pathol* 24: 1125–1130, 2000
10) Hoffmann E: Uber das knollentreibende fibrosarkom (Dermatofibrosarkoma protuberans). *Dermat Ztschr* 43: 1–28, 1925 (Ger)
11) Lakshman D, Shabnam BG, Karam C, Leelawati D: Intracranial extension of a dermatofibrosarcoma protuberans of the scalp: A case report with brief review of literature. *Surg Neurol* 54: 452–454, 2000
12) Marks LB, Suit HD, Rosenberg AE, Wood WC: Dermatofibrosarcoma protuberans treated with radiation therapy. *Int J Radiat Oncol Biol Phys* 17: 379–384, 1989
13) McLoughlin PM, Girach M, Wood GA: Dermatofibrosarcoma protuberans of the scalp. *Br J Oral Maxillofac Surg* 30: 401–403, 1992
14) McPeak CJ, Cruz T, Nicastri AD: Dermatofibrosarcoma protuberans. An analysis of 86 cases — Five with metastasis. *Ann Surg* 166: 803–816, 1967
15) Parker TL, Zitelli JA: Surgical margins for excision of dermatofibrosarcoma protuberans. *J Am Acad Dermatol* 32: 233–236, 1995
16) Petoin DS, Verola O, Banzet P, Dufourmentel C, Servant JM: Dermatofibrosarcome de Darier et Ferrand: Etude de 96 cas sur 15 ans. *Chirurgie* 111: 132–138, 1985 (Fre)
17) Rockley PF, Robinson JK, Magid M, Goldblatt D: Dermatofibrosarcoma protuberans of the scalp: A series of cases. *J Am Acad Dermatol* 21: 278–283, 1989
18) Roses DF, Valensi Q, Latrenta G, Harris MN: Surgical treatment of dermatofibrosarcoma protuberans. *Surg Gynecol Obstet* 162: 449–452, 1986
19) Taylor HB, Helwig EB: Dermatofibrosarcoma protuberans. A study of 115 cases. *Cancer* 15: 717–725, 1962
20) Weiss SW, Nickoloff BJ: CD34 is expressed by a distinctive cell population in peripheral nerve, nerve sheath tumors, and related lesions. *Am J Surg Pathol* 17: 1039–1045, 1993
21) Wooldridge WE: Dermatofibrosarcoma protuberans: A tumor too lightly considered. *Arch Dermatol* 75: 132–134, 1957
22) Wrotnowski U, Cooper PH, Shmookler BM: Fibrosarcomatous change in dermatofibrosarcoma protuberans. *Am J Surg Pathol* 12: 287–293, 1988

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