

A Case of Recurrent Dermatofibrosarcoma of the Scalp

Tae-Yeon Jo, M.D., Sang-Dae Kim, M.D.,
Se-Hoon Kim, M.D., Jung-Yul Park, M.D.

Department of Neurosurgery, Korea University Medical Center, Ansan Hospital, Ansan, Korea

We report a case of recurrent dermatofibrosarcoma in a 30-years-old woman who had undergone operations three times during 60 months and had received post-operative radiotherapy. On neurological examination, no neurological deficits were noticed. In brain magnetic resonance image(MRI), there was right parieto-occipital scalp mass with high signal in T2-weighted image, low signal in T1-weighted image with homogeneous enhancement. The removal was done including about 2cm uninvolved margins and pathologic examination of the lesion revealed dermatofibrosarcoma protuberans(DFSP). The prognostic factors of local recurrence may be related to surgical margins for resection; the length from the grossly intact margins, and the microscopically controlled excision in margins.

KEY WORDS : Dermatofibrosarcoma protuberans · Prognostic factor · Resection · Recurrence.

Introduction

Dermatofibrosarcoma protuberans(DFSP) is a rare, locally aggressive, cutaneous neoplasm of low-grade malignancy^{2,5,7,8,10,12}. The most common site of origin is the skin of the trunk (50% to 60%), proximal extremities (20% to 30%) and the head and neck (10% to 15%)¹¹. In spite of its slowly infiltrative growth and tendency to local recurrence, metastasis is very unusual^{4,5,8}.

The mainstay of treatment for DFSP is surgical resection. Resection with conservative margins has led to unacceptable local tumor control, with published local recurrence rates varying from 26% to 60%. To improve local control after excision, many authors have recommended wide excision with more than 2cm grossly uninvolved margin of skin and underlying deep fascia. This treatment approach has reduced local recurrence rates to a mean of about 20%¹¹. Surgery followed by post-operative radiation therapy(RT) may effectively reduce the local recurrent rate and preserve more tissue than radical surgery^{1,12}.

We report a case of recurrent dermatofibrosarcoma of the scalp after three operations followed by radiation therapy.

- Received : June 8, 2004 • Accepted : September 24, 2004
- Address for reprints : Jung-Yul Park, M.D., Department of Neurosurgery, Korea University Medical Center, Ansan Hospital, 516 Gojan-Dong, Danwon-gu, Ansan 425-707, Korea
Tel : 031) 412-5053, Fax : 031) 412-5054
E-mail : jypark@kumc.co.kr

Case Report

A 30-year-old woman was referred to our institution because of recurrent scalp mass on parieto-occipital area. She was initially operated on at other hospital with a presumed benign lesion. But tumor recurrence occurred one year later. Despite of salvage excision at second hospital, the second recurrence appeared again two years later. And the third operation was given and she received radiotherapy following operation. Finally she visited our clinic with a large recurrent scalp mass after 18 months from the latest operation.

On neurological examination, there were no abnormal findings. In Magnetic resonance image(MRI) showed a 6×4×6cm soft tissue mass on right parieto-occipital scalp area. In T1-weighted image, the mass was demarcated, with hypointensity. In T2-weighted image, it consisted of hyperintense mass. In gadolinium enhancement image, the mass represented relatively homogeneous enhancement (Fig. 1). There was a defect from invasion in outer table of skull but inner table was intact. In 6-vessel angiography, distal branch of superficial temporal artery was supplying the soft tissue mass. After angiographic embolization was done, there was diminished blood flow from distal branch of superficial temporal artery to soft tissue mass following embolization.

The gross total tumor removal with wide resection more than 2cm of margin and the pathologic findings were compatible with dermatofibrosarcoma protuberans. Pathologically, the specimen showed pinkish gray in external

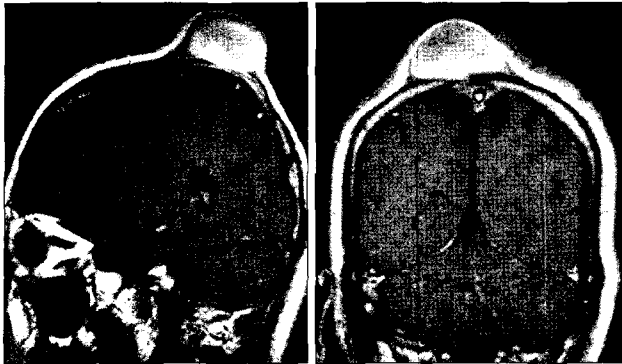


Fig. 1. In the right parieto-occipital scalp area, there is a 6 x 4 x 6 cm soft tissue mass which represents low signal with relatively homogeneous enhancement in T1-weighted magnetic resonance image.

surface, yellowish tan in resection margin. necrotic tissue and focal atypical cells, with high cellularity, irregular nucleus and prominent nucleoli in H-E stain (Fig. 2A).

Immunohistochemistry of this showed Vimentin (+), CD34 (+), Ki-67 index positive in 30~40% and S-100, SMA, C-Kit negative (Fig. 2B).

Postoperative course was uneventful and there is no evidence of tumor recurrence two years postoperatively.

Discussion

Dermatofibrosarcoma protuberans (DFSP), a low-grade spindle cell neoplasm involving both dermis and subcutis thus freely mobile on the fascia, is an unusual, locally aggressive, cutaneous neoplasm of low-grade malignancy^{5,8}.

It affects mainly the trunk and proximal extremities of young and middle-aged adults. Males are affected four times as often as females. It typically appears during the 3rd or 4th decade of life². Approximately 5% of DFSP occur in the pediatric population. In adult patients, 40~50% of these tumors arise from the trunk. Involvement of the head and neck is observed in about 10% of patients and in spite of its slowly infiltrative growth and tendency to local recurrence, metastasis is very rare^{4,8}.

The presentation for DFSP of the head and neck region is that of a slow-growing, painless, firm, solitary subcutaneous nodule¹¹. Its surface is characterized by irregular protuberant swellings, and a hard, indurated plaque of irregular outline forms the base².

The tumor can invade surrounding tissues with local, regional and distant metastases (mainly to the lungs), and its resistance to chemotherapy makes surgery the treatment of choice. In fact, early and radical surgery remains the first choice treatment to improve prognosis^{1,10}.

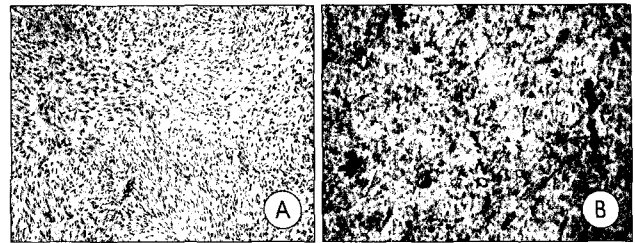


Fig. 2. A : There are necrotic tissue and focal atypical cells, with high cellularity, irregular nucleus and prominent nucleoli. It consists of fascicles of fibroblast-like spindle cells, 'cartwheel' or 'storiform' appearance (H&E stain, x100). B : The result of staining for CD34 is positive which had been considered for the past few years to be the most important immunohistochemical marker for the diagnosis of DFSP (CD34 stain, x200).

Recent cytogenetic studies have shown that a reciprocal translocation, (17;22)(q22;q13), and a supernumerary ring chromosome derived from the translocation r(17;22) are highly characteristic of DFSP. These chromosomal rearrangements fuse the collagen type I $\alpha 1$ (COL1A1) and the platelet-derived growth factor B-chain (PDGFB) genes⁹.

DFSP is confirmed only after microscopic evaluation. The multiformity of this tumor in conjunction with its paucity of symptoms contributes to the difficulty in correct diagnosis on the basis of clinical findings alone. The seemingly benign appearance of this tumor can be misleading to patients and physicians. Preoperative diagnosis is usually difficult on account of the tumor's variable presentation. Microscopically, it consists of fascicles of fibroblast-like spindle cells, usually forming quite dense bands that interweave or radiate like spokes of a wheel; this is the so-called 'cartwheel' or 'storiform' appearance. Positive anti-CD34 immunostaining is also helpful in the diagnosis¹⁰. The tumor diffusely infiltrates the dermis and subcutis and spreads along the connective tissue².

The choice of treatment for DFSP is surgical resection. In fact, early and radical surgery remains the first choice treatment to improve prognosis¹⁰. Prognostic factors for distant disease failure included fibrosarcomatous variant of DFSP, positive microscopic margin, increased cellularity, high mitotic rate, age more than 50 years, and multiple prior recurrences.

The most significant factor that predicts outcome for DFSP is extent of resection. Deep lesions were more likely to have positive microscopic margin resections than superficial lesions¹¹. Resection with conservative margins has led to unacceptable local tumor control, with published local recurrence rates varying from 26% to 60%.

To improve local control after excision, many authors have recommended wide excision with more than 2cm grossly

uninvolved margin of skin and underlying deep fascia. This treatment approach has reduced local recurrence rates to a mean of about 20% lesion. The high local recurrence rate after resection has been attributed to failure to excise these clinically inapparent projections of tumor. In this point, some authors recommend Mohs Surgery the treatment of choice for DFSP because of its high cure rate and maximal conservation of tissue⁶. Moh's micrographic surgery is a method for removal of non-melanoma skin cancer in thin layers, allowing frozen-section examination of all peripheral and deep margins. Subsequent tissue layers are removed as dictated by microscopic examination, allowing for maximal sparing of normal tissue. This method offers cure rates significantly higher than excision or other modalities. Moh's micrographic surgery is the method of choice for removal of large, recurrent or incompletely excised skin cancers or for tumors located in regions of high recurrence.

This may be reasons for our patient who had three operations elsewhere prior to admission. Moreover, any micro-pathological sectioning technique that fails to examine all lateral margins and the deep aspect of the tumor inevitably will overlook these neoplastic extensions. We also have done six frozen examinations from all around the resected margins to make sure there were no tumor extension.

Surgery followed by post-operative radiation therapy may effectively reduce the local recurrent rate and preserve more tissue than radical surgery¹². In our case, radiation therapy was not given because of previous radiation therapy, and there is no signs of recurrence for one year.

Conclusion

DFSP can be managed with appropriate surgical treatment and wide excision. However, it can recur despite total resection followed by radiotherapy as in our case. In this case, the recurrence of this patient maybe result

from insufficient surgical resection margins in past operation. Thus, it is recommended to be aggressive to resect sufficient margins of more than 2cm grossly uninvolved margin initially to prevent recurrence.

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