MR Imaging Findings of Recurred Dermatofibrosarcoma Protuberans of the Scalp: A Case Report

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A 48-year-old man presented with a dermatofibrosarcoma protuberans (DFSP) of the scalp associated with local recurrence. Axial T1- and T2-weighted images demonstrated a well-circumscribed hypointense and intermediate hyperintense mass in the skin and subcutaneous layer of the scalp, respectively. Contrast-enhanced T1-weighted images showed the strongly enhanced mass invasion to the skin, subcutaneous layer and adjacent galeal layer. Scalp DFSP is very uncommon but is an aggressive tumor, so MR imaging diagnosis of the extent of the lesion to underlying structures, and initial wide local resection is important to prevent recurrence.

Index words : Dermatofibrosarcoma Protuberans
Brain MRI, Scalp tumor
Recurrence

Introduction

Dermatofibrosarcoma protuberans (DFSP) is a relatively rare cutaneous spindle cell neoplasm that is characteristically diffuse, poorly circumscribed, and has a high propensity for local recurrence after excision (1). Without accurate diagnosis and surgical treatment, DFSP tends to recur and destruct structures around. The tumor originates within the dermal layer of the skin, and it is believed that the cell of DFSP is a dermal stem cell or an undifferentiated mesenchymal cell with fibroblastic, muscular, and neurologic features (2). Accurate preoperative assessment of the tumor extent is mandatory for appropriate therapeutic planning. There have been articles regarding recurrent or metastatic DFSP of the scalp (3, 4). However, as far as we know, there is no report regarding the MR imaging findings of recurred DFSP of the scalp. We present a rare case of recurred DFSP of the scalp, and MR findings and pathologic confirmation were thought to be useful for the proper surgical management.

Case Report

A 48-year-old man visited our hospital with recurrent scalp mass and dull headache. Personal medical history included essential hypertension and congestive heart

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failure. Three years before, he had a history of surgical resection of the scalp tumor that presented as a very slow growing and skin colored asymptomatic mass. Physical examination revealed about 3 cm sized bulging mass on the right occipital area. Top of the mass was alopecic and colored in pink. It adhered around the grossly normal scalp, and was slightly movable on the skull. The previous operation scar laid on the edge without evidence of infection. The mass was well circumscribed, nodular and moderate tender, firm, and non-pulsatile. Cervical lymph nodes were neither tender nor hypertrophied. On neurologic examinations, the patient showed no pathologic finding. Laboratory evaluation showed normal range. Blood pressure was stable after antihypertensive medication. On cardiac examination, 2D M-mode cardiac Doppler showed the increased left ventricular chamber size and decreased systolic function to 37% of the normal ejection fraction.

Plain skull radiograph showed bulging soft tissue mass in the occipital scalp. On bone scintigrams, there was no increased accumulation of radiopharmaceutical. Cranial MR images demonstrated 4.6x1.9x4.3 cm sized soft tissue mass in right occipital scalp and intact bony calvarium. Axial T1- and T2-weighted images demonstrated a well-circumscribed hypointense and

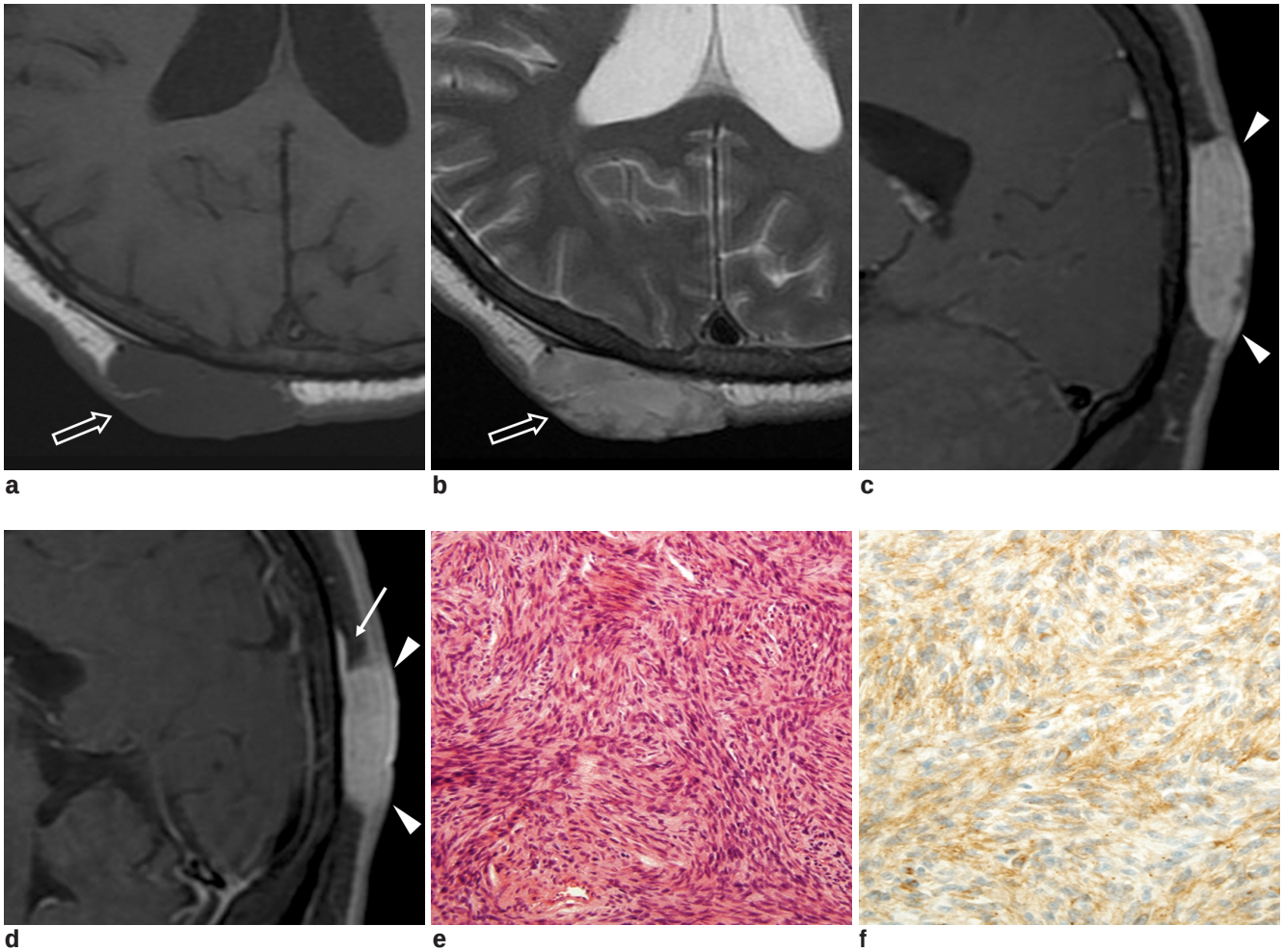

Fig. 1. A 48-year-old man with recurred scalp DFSP.
a, b. Axial T1-weighted image (**a**), and T2-weighted image (**b**) demonstrates a well-circumscribed hypointense and intermediate hyperintense scalp mass (arrow) in the skin and subcutaneous layer. There is no evidence of invasion into the skull. **c, d.** Contrast enhanced sagittal fat-suppression T1-weighted images demonstrate homogeneously enhancing the scalp mass (arrowheads) involving skin and subcutaneous layers. Peritumoral galeal enhancement (arrow) suggests perilesional extension. **e.** Photomicrograph shows spindle cells arranged in the storiform pattern (H & E stain, ×200). **f.** CD34 immunohistochemical stain shows diffusely positive in the tumor cells (CD34, ×400).

intermediate hyperintense mass in the skin and subcutaneous layer of the scalp, respectively (Fig. 1a, b). Contrast-enhanced fat-suppressed T1-weighted images showed the strongly enhanced mass invasion to the skin, subcutaneous layer and adjacent galeal layer (Fig. 1c, d).

On the next day, under the general anesthesia, bilateral scalp incision for wide resection, rotation graft and scalp reconstruction was performed. On operation field, the extracranial tumor was partially adhered to the scalp without gross evidence of bone infiltration. En bloc excision including the tumor and skin was carried out. The extracted specimen involved subcutis and skin of the scalp, and very closed to the periosteum. The cut surface of the tumor showed the tan colored and well demarcated mass without hemorrhage or necrosis. Microscopic examination demonstrated that the tumor diffusely infiltrated the dermis and subcutis of the scalp and was composed of uniform, monotonous spindle-shaped cells with a distinct storiform pattern in Hematoxylin and Eosin stain (Fig. 1E). Neither lymphovascular nor perineural invasion was found. On immunohistochemical stain, CD34 is diffusely positive in the tumor cells (Fig. 1f). Ki-67 was less than 1%. Negative staining for S-100 was observed.

Discussion

DFSP is a relatively rare and locally aggressive soft tissue tumor. Whereas DFSP of the scalp accounts for less than 5% of all DFSP cases, local recurrence rates for all cases of DFSP of the head and neck are extremely high, ranging from 50 to 75% (5). Local recurrence usually develops within 3 years of the initial surgery (4). The recurring and nonrecurring tumors show no differences in location, duration, size or histology (4). It is possible that any manipulation in the form of inadequate excision, which cuts through the tumor and simultaneously opens vascular channels, may be essential for vascular or lymphatic spread (6). Metastasis is infrequent despite the locally aggressive behavior, and should be clearly discriminated from conventional sarcoma. Metastasis is seen in no more than 6% of cases. The most common metastatic sites are the lung, bone, and lymphnodes (4).

DFSP tumors were well-defined lesions on MR imaging with low signal on T1-weighted images and higher than fat or intermediated signal intense on T2-weighted sequences (7, 8). On intravenous contrast enhancement MR images, there was uniform and patch central enhancement (8). In our case, postcontrast fat-suppression T1-weighted image depicted strong enhancement of the mass with adjacent galeal extension. MR images are well suited to show the skin and subcutaneous adipose tissue layer, and the relation of the mass to underlying structures (3, 7). Tumor invasion in to subcutaneous fat and skull was clearly visible on T1-weighted images. T2-weighted images and contrast enhanced T1-weighted images were sufficient to visualize the whole extent of DFSP of the scalp. Especially, postcontrast T1-weighted imaging with the fat saturation technique was also able to reveal tumor invasion into the subcutaneous fat and galea aponeurotica, which was not seen on superficial inspection. These MR imaging findings were useful for therapeutic planning for the patients with DFSP of the scalp. Arteriograms will show mild to moderate hypervascularity, and bone scintigrams will show increased accumulation of radiopharmaceutical (7). In our case, however, in spite of strong enhancement of the tumor on postcontrast enhanced T1-weighted image, there was no increased uptake on bone scintigrams.

Although the storiform arrangement of spindle cells in DFSP is relatively characteristic, histologic patterns simulating other benign as well as malignant neoplasm such as dermatofibroma, neurofibroma, malignant fibrous histiocytoma, and atypical fibroxanthoma have been described (1). Spindle cells in DFSP stain diffusely positive for CD34 but usually stain negatively for S-100 protein presented in virtually all neurofibromas (1). However, CD34-positive dendritic cells have been identified within the endoneurinum of normal nerve, neurofibromas, neurilemmomas, and dermatofibrosarcomas, as well as in blood vessels (1, 5). Therefore, it is important for clinicians and histopathologists to be aware that features of benign neural differentiation may be found within DFSP and may result in underdiagnosis as a benign neural neoplasm and inadequate treatment (1).

The goals of effectively managing DFSP of the scalp are to ensure complete surgical excision and to restore anatomic integrity with a functional and cosmetically

acceptable result. Mohs micrographic surgery may be the treatment of choice for facial and scalp dermatofibrosarcoma protuberans since this technique enhances reconstruction options and decreases the chance of recurrence of this uncommon tumor (2). Because of the subclinical and infiltrative nature of DFSP, wide excision with defined margins has been recommended and the periosteum should also be removed and analyzed for residual tumor (5). To cure DSFP, the wide and complete surgical tumor excision with inter-department cooperation should be planned. Although DFSP is not radiosensitive, postoperative RT may reduce the risk of local recurrence in patients with DFSP who have a high risk of residual disease after surgery (9). Gamma knife radiotherapy was mildly effective for the inhibition of tumor growth, and may be more effective than conventional radiation against the primary lesion (4). Chemotheraphy using medium doses of methotrexate was tried but had no apparent efficacy base on the 3-[4.5-dimethylthiazol-2-yl]-2, 5-diphenyltetrazolium bromide essay (4). Close follow-up every 3 months for the first year and then every 6 months for 5 years is also advocated (5).

In conclusion, we describe a 48-year-old man with recurred scalp mass of DFSP. DFSP tumor was well-defined lesion with low signal intense on T1-weighted images and intermediated signal intense on T2-weighted sequences. On intravenous contrast enhancement MR images, there was uniform and patch central enhancement. Although local recurrence or distant metastasis of scalp DFSP is not common, it is an aggressive scalp tumor, so MR imaging diagnosis of the lesional extent to the underlying structures, and initial wide local resection is important to prevent recurrence.

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두피에서 재발한 융기성 피부섬유육종의 MR영상: 증례 보고

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두피의 융기성 피부섬유육종이 국소 재발한 48세 남자의 예를 보고하고자 한다. MR영상 소견상 두피의 피부와 피하층에 T1 강조영상에서 저신호강도를 T2 강조영상에서 중등도 고신호강도를 지닌 종괴가 있었고, 조영증강 영상에서 강하게 조영증강되는 종괴는 피부와 피하 및 모상층(galea)으로 파급되는 소견을 보였다. 두피의 융기성 피부섬유육종은 매우 드문 병변이나 침습적인 종양으로서, MR 영상에서 주변 조직으로의 병변 파급을 진단하고 초기에 광범위 국소절제술을 시행하는 것이 재발을 방지하는 데 중요하다.

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