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Intracranial Fibromatosis
– A Case Report –

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Fibromatosis can occur at various sites, but intracranial fibromatosis is exceptionally rare. Here, we report a case of intracranial fibromatosis arising in the suprasellar area of a 52-year-old woman who had undergone a surgery at that site. A computed tomography scan revealed a heavily calcified, highly enhancing, poorly demarcated mass in the left sellar area that extended into the left suprasellar, parasellar areas, and orbital apex and completely encased the left distal inferior cerebral artery. Histologic and immunohistochemical features were compatible with those of fibromatosis, although the cellularity was focally higher than usual. The etiology of extra-abdominal fibromatosis is unknown, but physical injuries such as trauma and irradiation have been reported to be associated with its occurrence. Although fibromatosis is rare in the intracranial area, it should be considered as a differential diagnosis when an intracranial mass occurs at a previously injured site.

Key Words: Desmoid disease; Fibromatosis, aggressive; Sella turcica; Central nervous system

Fibromatosis encompasses a broad group of benign fibroblastic proliferations of similar microscopic appearance whose biologic behavior is intermediate between that of benign fibroblastic lesion and fibrosarcoma. Like fibrosarcoma, fibromatosis is characterized by infiltrative growth and frequent recurrences. But, unlike fibrosarcoma, fibromatosis never metastasizes. Fibromatosis can occur in various organs but intracranial fibromatosis is very rare. Only 1.9% of all extra-abdominal fibromatoses have been reported to occur in this location. Here, we report a case of intracranial fibromatosis arising at the suprasellar and parasellar area where trans-sphenoidal resection of pituitary adenoma had been performed.

CASE REPORT

A 52-year-old woman presented with 1-year history of ptosis of the left eyelid and headache. The patient also complained of dysarthria and impaired memory and orientation. Four years ago, the patient had undergone a resection of non-functioning pituitary adenoma through a trans-sphenoidal approach. The patient had also been diagnosed with a papillary thyroid carcinoma by fine needle aspiration in the previous year. The patient was chronically ill-looking and showed a decreased visual acuity on neurological examination.

Computed tomography (CT) and magnetic resonance imaging scans revealed a heavily calcified, highly enhanced, poorly demarcated mass in the left sellar area that extended into the left suprasellar, parasellar areas, and the orbital apex, and which completely encased the left distal inferior cerebral artery (Fig. 1). The deep gray matter of the left frontal lobe and the left anterior temporal lobe was compressed by the mass. Compressed normal pituitary gland was deviated to the right side along with the pituitary stalk. The patient underwent a debulking of the lesion through a craniotomy. At operation, a grayish pink tumor involving the sella and left cavernous sinus was identified. The tumor was compressing the left basal ganglia, optic chiasm, and temporal lobe. Several pieces of the tumor tissue measuring about 5 mL in aggregates were obtained and all were subjected to microscopic examination.

Histologically, the tumor was poorly demarcated and showed a variable cellularity ranging from low to moderate degree (Fig.

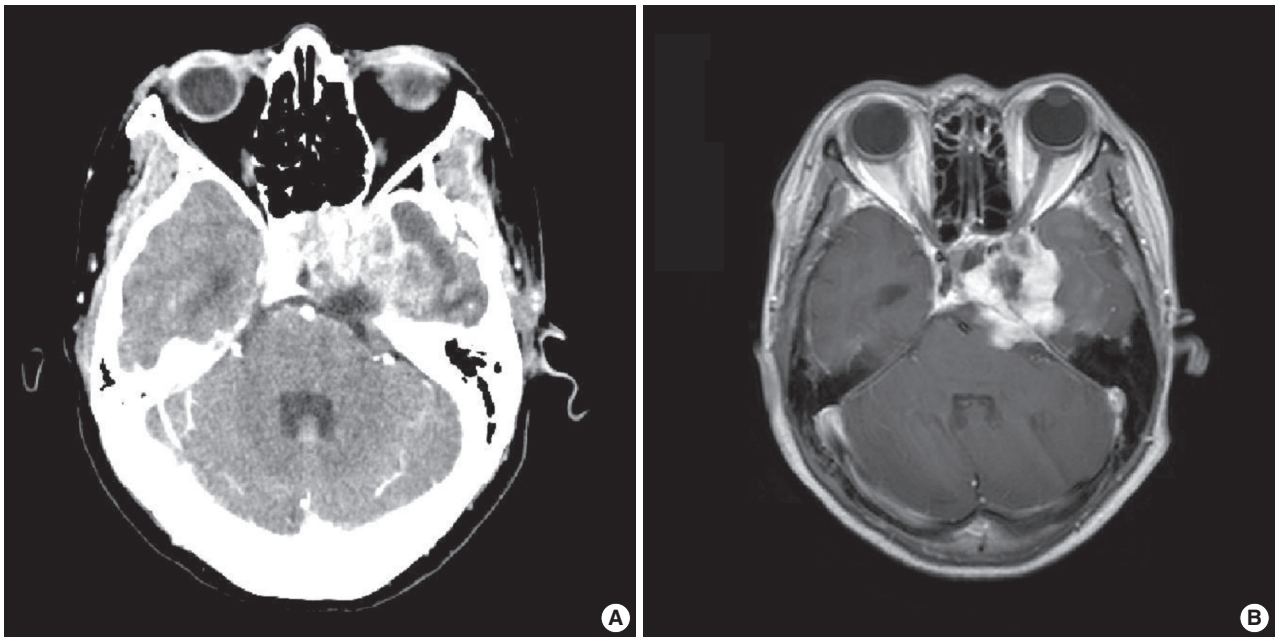

Fig. 1. Computed tomography scan (A) and T1-weighted magnetic resonance imaging scan (B) of the tumor. These axial plane views show a heavily calcified, poorly demarcated mass in the left parasellar, sellar area. The mass is compressing the left temporal lobe and is destroying the basal skull bone.

2A, B). The tumor consisted of bland-looking spindle cells with focal mild nuclear atypia and inconspicuous nucleoli (Fig. 2C). Between tumor cells was abundant collagen. Mitosis was rarely observed (less than one per 10 high power fields) and necrosis was absent. Although heavy calcification was noted on radiographic imaging, no calcification was identified on microscopic examination. The tumor cells were diffusely reactive for smooth muscle actin (Fig. 2D) but were negative for S-100 protein, desmin, and CD34. The Ki-67 labeling index was less than 1%.

There was no immediate post-operative complication. However, a follow-up CT scan showed a residual nodular enhancing lesion at the site of surgery and hydrocephalus. Six months after the surgical procedure, an episode of seizure occurred. Consideration will be given to either radiation therapy or chemotherapy if there is evidence of further tumor growth on follow-up radiological examination.

DISCUSSION

Extra-abdominal fibromatosis occurs most frequently in the musculature of the shoulder, followed by the chest wall, back, thigh, head, and neck. However, its occurrence within the central nervous system is extremely rare, accounting only for 1.9% of extra-abdominal fibromatosis. Our case is the first report in Korea and is particularly interesting because the occurrence of this tumor is associated with the previous surgical manipulation.

Differential diagnoses include fibrosarcoma, low grade osteosarcoma, and pseudosarcomatous myofibroblastic proliferation. In contrast to fibromatosis, fibosarcoma usually show uniformly high cellularity and a peculiar herring-bone arrangement. In addition, the tumor cell nuclei of fibrosarcoma are usually hyperchromatic and have prominent nucleoli. However, in the present case, the cellularity was only mild to moderate and there was no distinct herring-bone growth pattern. Moreover, the tumor cells of the present case were bland-looking. Finally, mitotic rate of fibrosarcoma in general is also higher than that of the present case. Because intratumoral calcification was observed in the CT scan, the possibility of low grade osteosarcoma should also be taken into account. However, that possibility was considered to be less likely because no osteoid was observed in the submitted tissue specimen. In addition, the advanced age of the patient and the absence of precipitating factors of secondary osteosarcoma were not compatible with osteosarcoma. Pseudosarcomatous myofibroblastic proliferation can also exhibit similar histological features as the present case but differs from this case by its well-defined border, extravasated red blood cells, frequent secondary changes and tissue culture-type fibroblasts. Furthermore, immunonegativity for S-100 protein, desmin, and CD34 excluded the possibility of neurogenic tumor, smooth muscle

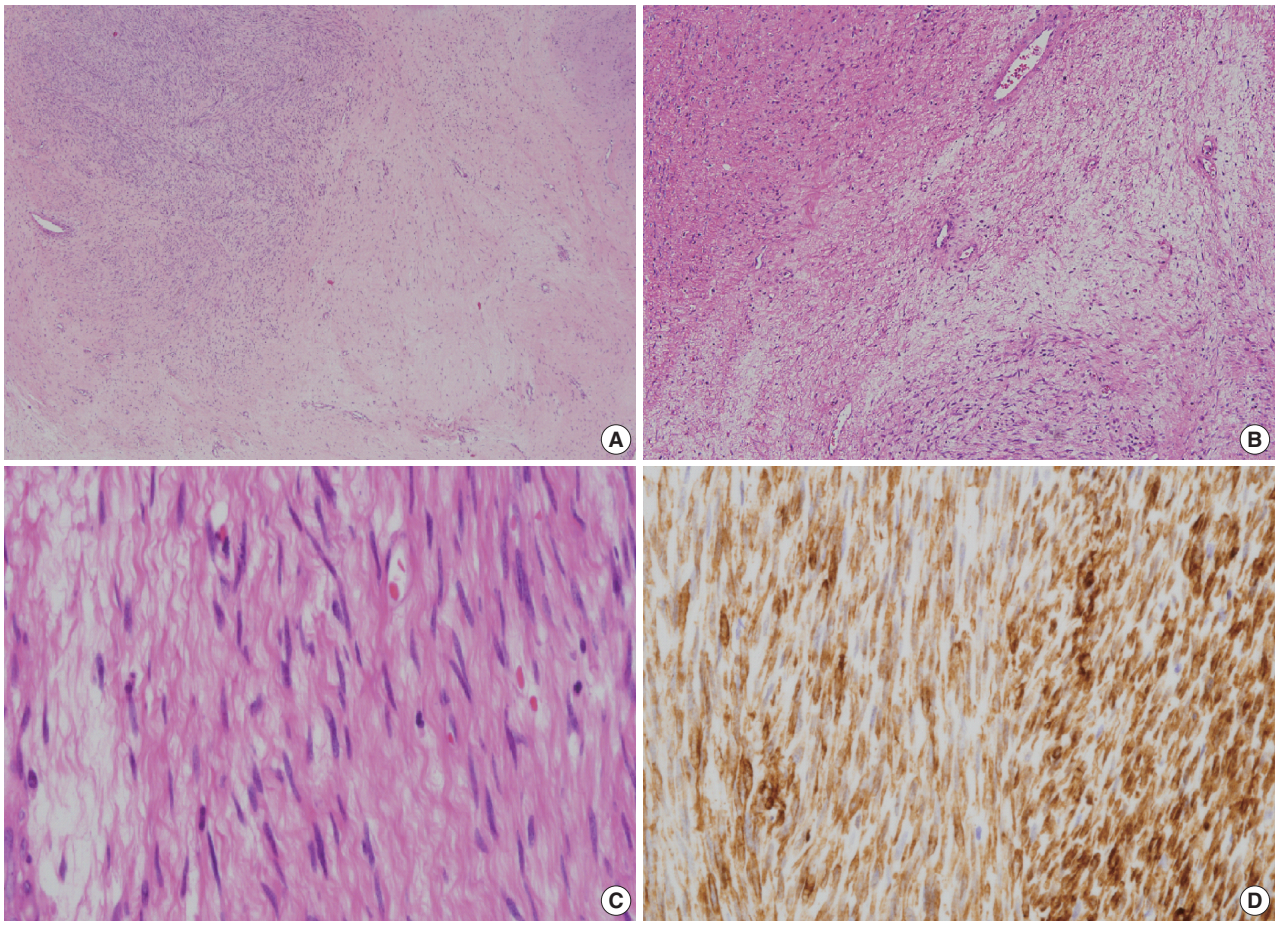

Fig. 2. View of the central portion of the tumor. (A) A gradual transition to the less cellular peripheral fibrous tissue is evident, suggesting an infiltrative tumor border. (B) The cellularity varies from place to place. (C) The tumor cells are bland-looking spindle cells arranged in a fascicular growth pattern and abundant collagen fibers are noted among the tumor cells. (D) The tumor cells are diffusely positive for smooth muscle actin.

tumor, or solitary fibrous tumor.

Several etiologies have been proposed for extra-abdominal fibromatosis. Hereditary syndromes (e.g., Gardner syndrome),[1] trauma or surgical incisions,[2] or hormonal effects[3] have all been attributed to the occurrence of extra-abdominal fibromatosis. Among them, trauma has been reported to be associated with deep fibromatoses most frequently. In a series of 30 extra-abdominal fibromatoses, 19 (63%) patients had a history of trauma at the site of tumor occurrence.[2]

Four cases of intracranial fibromatosis occurring at the site of previous surgery have been reported to date[4-7] and they are summarized in Table 1. The time interval between previous surgical manipulation and occurrence of fibromatosis ranged from 9 to 25 months and two cases also had a history of radiation therapy at the site of surgery. Histological features of those reported cases were similar to those of the present case and one case of them showed focal myxoid stroma. Together with those previous reports, our case further supports the notion that a previous surgical manipulation could be a risk factor for fibromatosis.

Table 1. Intracranial fibromatoses occurring at the previous surgery site (reported cases and present case)

Case No.	Age (yr)	Sex	Location	Previously history	Reference
1	7	M	Frontal lobe	Subependymal giant cell astrocytoma	Scougall *et al.*[4]
2	17	F	Left occiput, cerebellum	Arteriovenous malformation	Mitchell *et al.*[5]
3	30	F	Subocciput	Juvenile pilocytic astrocytoma	McCall *et al.*[6]
4	65	F	Cervical spine	Meningioma	Kenning *et al.*[7]
Present case	52	F	Sella, suprasella	Pituitary adenoma	

M, male; F, female.

In summary, our case was a rare intracranial fibromatosis associated with a previous surgical manipulation. Mechanistically, the unique occurrence of fibromatoses at the surgically manipu-

lated site is considered most likely to be linked to physical trauma. Although fibromatoses are rare in the intracranial area, they should be considered as a differential diagnosis especially when they occur at the sites with a history of previous surgery or trauma.

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