CASE REPORT

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Intracranial Solitary Fibrous Tumor

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Solitary fibrous tumor is a spindle cell neoplasm that can arise in any place of the body. Intracranial solitary fibrous tumors are rare. To our knowledge, only 57 cases with intracranial lesion have been reported. In Korea three cases have been reported. Our case was a 23-year-old woman who presented with morning headache. MRI showed a large intra-axial mass involving falx with typically isointense and heterogeneous strong enhancement on T1 weighted image in the right parieto-occipital region. Histologically the tumor showed spindle shaped cells within matrix with thick collagen deposition, hypercellularity, focal necrosis, and pleomorphism. Immunohistochemical study demonstrated diffuse positivity for CD34, Vimentin, Reticulin. In case of the intracranial tumors involving the meninges, we also should consider the solitary fibrous tumor with immunohistochemical staining for accurate diagnosis.

KEY WORDS : Solitary fibrous tumor · CD34 · Intracranial.

Introduction

Solitary fibrous tumors(SFTs) are rare fibrous proliferating neoplasms consist of the spindle cells. SFTs usually occur in the pleura-based intrathoracic region[8], although these tumors have also been diagnosed in every organ such as urinary bladder[7,13], lung parenchyma, upper respiratory tract, thyroid gland, and the meninges in central nervous system. The histopathological findings of SFTs in the brain are similar in SFTs in the pleura. The SFTs are usually benign but often have the malignant feature clinically or histologically. Although the precise prognosis of SFTs is unkown, complete removal rather than histological appearance seems to be the most important prognostic factor of SFTs[13]. Therefore we should use immunohistochemical study for accurate diagnosis and differentiate the SFTs from other tumors of mesenchymal origin which have poor prognosis.

In this report, we describe a case of intracranial SFTs and review the literatures.

Case Report

This 23 year-old female patient presented with a gradual increasing morning headache for 12 months. She had IgA nephropathy about 5 years ago.

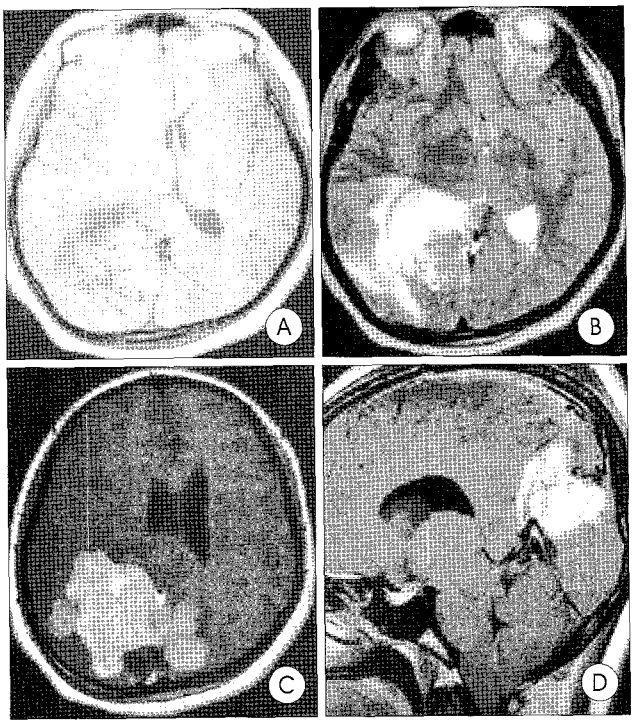

Fig. 1. Preoperative magnetic resonance image. T1 weighted axial (A), T2 weighted axial (B), Gd-enhanced T1 weighted axial (C) and Gd-enhanced T1 weighted sagittal (D) images show a lobulated solid mass in the right parietooccipital lobe, extending to the posterior falx and left occipital lobe. Note the dense enhancement and intratumoral hypervascularity.

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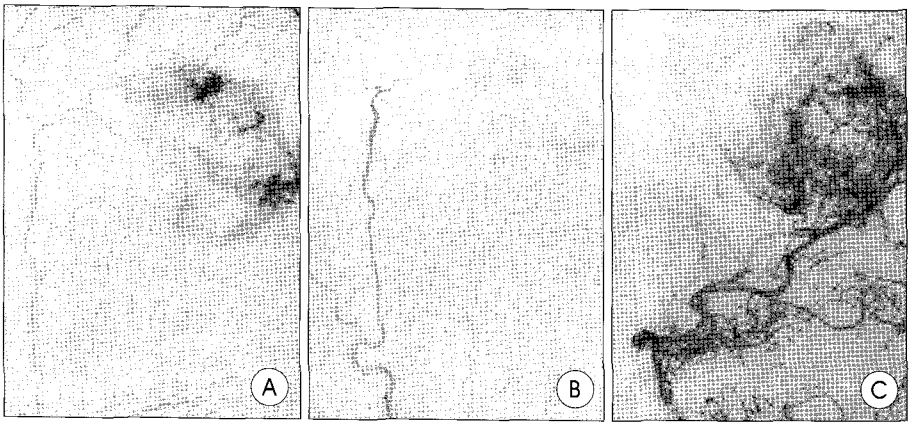

Fig. 2. Right external carotid angiography before embolization (A). Right vertebral (C) and external carotid angiography (B) after embolization of the feeding arterial branches from right external carotid artery. The angiography demonstrates a large hypervascular mass in the occipital region fed by branches from the distal posterior cerebral artery.

Preoperative brain MRI showed a irregular, round and well-demarcated tumor mass(6×5×5cm) which was based on posterior falx involved with the vein of Galen, straight sinus, extending from right parieto-occipital lobe to left occipital lobe. The mass was isointense with gray matter on T1-weighted image and showed the heterogeneous strong enhancement on Gd-enhanced T1 weighted image. On T2 weighted image the mass showed the hyperintense edema around the hypointense core of mass(Fig. 1). Angiographically the vascular supply of the tumor was very rich and mainly originated from right posterior cerebral artery and meningeal branches of right occipital artery and right STA. After the embolization of right occipital artery and Rt. STA, the vascular supply to the mass from right external carotid artery diminished remarkably. But the most part of mass was still stained via posterior cerebral artery(Fig. 2). At the first operation, there was copious bleeding from the tumor mass despite of embolization. So only a partial removal was achieved. Brain CT scan in 7 months after the first operation showed the bulging of tumor out of the craniectomy site(Fig. 3). At the second operation, a near total resection was achieved except a little tumor remnant near the vein of Galen and straight sinus(Fig. 4). Histologically the tumor showed hypercellular spindle cell proliferation with area of collagenization, prominent dilated vessels and pleomorphism. Mitoses were less than 4/10 HPF and cellular atypism was not definite. The immunohistochemical stain revealed positive staining for vimentin and CD34(Fig. 5), but negative staining for epithelial membrane antigen(EMA). Silver staining shows diffuse reticulin network in the tumor. The patient was free from recurrence, distance metastasis. Recently the patient was treated with the gamma-knife for the residual tumor, followed up at 9 months.

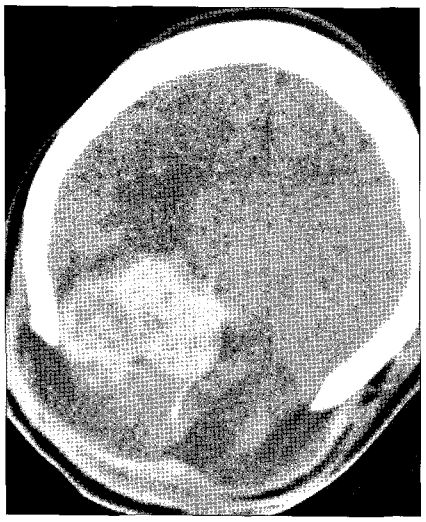

Fig. 3. Follow-up computed tomography(CT) scan in 7 months after the first operation. The postcontrast CT scan shows a large enhancing tumor mass in the operation site of the right occipital lobe. Note the bulging out of tumor mass through the craniectomy.

Discussion

The solitary fibrous tumors arise in any place of the body such as the pleura, peritoneum, me-

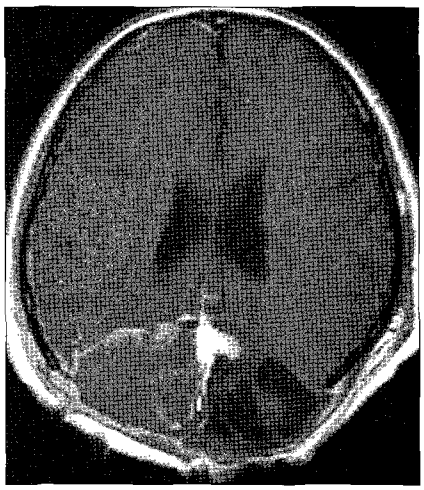

Fig. 4. Follow-up magnetic resonance image after the second operation. The Gd-enhanced T1 weighted axial sagittal image demonstrating the part of small residual tumor near the vein of Galen and straight sinus.

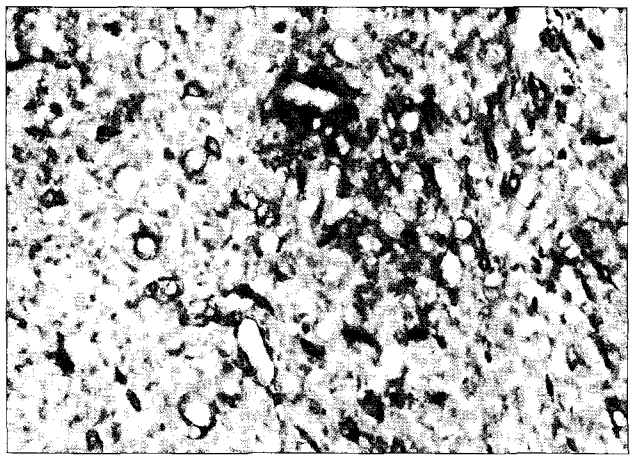

Fig. 5. The photograph showing the positivity for CD34 immunostaining as well as vimentin. (Immunohistochemistry by Anti-CD34, X 200).

ninges, even the organ without the serosa[2,13,15].

The histopathogenesis of SFTs has been a matter of debate. At the beginning of studies the mesothelial origin had been proposed, but recent studies confirmed the mesenchymal origin of SFTs[1] and revealed the various histological features[4]. The immunohistochemical, electron microscopic, and tissue culture studies show that the tumor originates in the mesenchymal fibroblast. To our knowledge, only 77 solitary fibrous tumors in central nervous system have been described in the literature[2]. Among them the 57 cases were intracranial SFTs and the 20 cases spinal SFTs[2]. The number of cases have been increasing continuously. The 57 cases of intracranial SFTs are summarized in Table 1. Caroli et al.[2] summarized 45 cases and reported another 12 cases recently. The age range is 11~73 years (mean : 50.9) and sex ratio is approximately 1 : 1. Clinically 15~20% of intrathoracic SFTs showed local invasion, intrathoracic spread, or distance metastasis[5]. But distance metastasis of the intracrnial SFTs seemed to be rare[2,13].

Table 1. The 57 cases of intracranial solitary fibrous tumors

Age/Sex	Tumor location	Treatment	Recurrence	Follow up
51/F	Posterior fossa	TR	No	240 months
47/M	Cerebellopontine angle	TR + RT	No	120 months
73/M	Frontal lobe	TR	No	8 months
62/F	Tentorium	TR	No	7 months
63/F	Cerebellopontine angle	TR	No	9 months
3 cases	Not state	Not state	Not state	Not state
43/M	Frontal lobe	TR	Not state	Not state
73/F	Optic nerve	TR	Not state	Not state
42/M	Tentorium	Not state	No	12 months
18/M	Craniospinal	TR	No	12 months
11/M	Occipital	TR	Not state	Not state
45/M	The 4th ventricle	Unknown	No	180 months
30/M	Frontal	Unknown + RT	No	9 months
64/F	Falcine	TR	Recurred	Died
44/M	Parietal parasagittal	STR	Not state	Not state
14/F	Parietal convexity	TR	Not state	Not state
58/F	Posterior fossa	TR	No	Not state
55/F	Posterior fossa	TR + RT	Recurred, Metastasis (lung, neck)	120 months
60/F	Frontal	Not state	No	18 months
51/F	Transverse sinus	STR	Recurred	84 months
54/F	Parieto-occipital	TR	No	36 months
61/F	Deep frontal	NTR+RT	No	10 months
46/F	Posterior fossa	TR	No	36 months
43/M	Cerebellopontine angle	NTR	No	24 months
72/F	Middle fossa	STR	No	12 months
71/M	Frontoparietal	IR	Not state	Died
58/M	Temporal lobe	TR	No	12 months
25/F	Occipital	TR	No	36 months
12 cases	9 supratentorial, 3cerebellar	Not state	Not state	Not state
38/M	Gasser's ganglion	TR	No	15 months
29/F	Cerebellar	TR	No	36 months
34/M	Frontal	TR	No	66 months
56/M	Pituitary fossa[6]	IR	No	5 months
33/M	Fronto-parietal[10]	TR	No	4 months
44/F	Tentorium[16]	TR	Recurred, Metastasis (lung)	312 months
48/F	Falcotentorial[14]	Surgery+RT	Recurred, CSF Dissemination	180 months
54/F	Sphenoid, carvernous sinus[3]	IR	Not state	Not state
50/F	Tentorium[17]	TR	No	6 months
53/F	Cerebellopontine angle[20]	Not state	Recurred	9 months
63/M	The third ventricle[9]	NTR	No	42 months
50/M	Parasellar[12]	Unknown + RT	Recurred	2 months
30/F	Frontal[19]	TR	Recurred	360 months
60/F	Parietal[11]	TR	No	12 months
*23/F	Parieto-occipital	NTR	No	9 months

* : Our case, M : male, F : female, RT : radiotherapy, CSF : cerebrospinal fluid, TR : total resection, STR : subtotal resection, NTR : near-total resection, IR : incomplete resection

The malignant histological features described for the intracranial SFTs include high cellularity, nuclear pleomorphism, necrosis, hemorrhage, and mitoses more than 4/10 HPF[2,5,15]. Because the histological finding does not always match with its clinical course, it is difficult to predict the biological course of the case or malignant change such as local recurrence and distance metastasis of SFTs. Even though the benign histological finding, the tumor can grow invasively, and in spite of the malignant histological finding, the mass can show the benign growth[5,15]. Furthermore it has been reported that local recurrence and distance metastasis of intracranial SFTs developed newly at 10 years follow-up[5,15]. In that reason, to predict precisely the clinical prognosis of intracranial SFTs is difficult, we should follow up them for the longer period.

The immunohistochemical stain revealed positive staining for CD34 and vimentin, but negative staining

for S-100 protein, EMA and desmin. Recently immunohistochemical studies about the SFTs revealed the strong positive reaction to CD34, and the electromicroscope showed microstructures with the fibroblast. The CD34 antigen is a transmembrane cell surface glycoprotein, which was originally described on haematopoetic stem cells, endothelial cells, and fibroblasts. Therefore instead of the mesothelial origin, the mesenchymal origin of SFTs had been proposed and confirmed[1].

CD34 reactivity can be identified also in fibrous meningioma and hemangiopericytoma because they can include the fibroblast within tumor. So the SFTs must be included in the differential diagnosis with hemangiopericytoma and fibrous meningioma[18,21]. The fibrous meningioma is characterized by a rich cytoplasm, collagen matrix, psammoma bodies, whorl formation of tumor cells, amorphous calcification, only focal or no positivity to CD34, and positivity to EMA[18,21]. CD34, EMA reactivity helps the differential diagnosis with SFTs and fibrous meningiomas. The hemangiopericytoma is typically cellular tumors composed of small, oval to slightly spindle cell. It shows a lot of branching, thin-walled vessels of varying caliber, "staghorn" vascular pattern. CD34 reactivity of the hemangiopericytoma is pachy, weak[18,21]. Tihan et al.[22] have reported the hemangiopericytoma showed malignant clinical course in many cases. The recurrence(83%), distance metastasis(27%), and mortality(22%) of hemangiopericytoma are very high in contrast to general benign nature of SFTs. Neurofibroma, fibrosarcoma, schwannoma including the fibrous meningioma and the hamangiopericytoma must be included in the differential diagnosis with intracranial SFTs.

SFTs usually have benign feature, but malignant behavior of SFTs cannot be predicted reliably on histological grounds. Total resection of tumor, rather than histological findings is the most important prognostic factor[13].

Conclusion

We experienced one case of the intracranial SFT. The patient shows good prognosis without regrowth of the residual tumor.

For diagnosis and treatment of intracranial neoplasm involving meninges, it is very important to be aware of intracranial SFTs and consider the differential diagnosis. In future, study of the radiotherapy for residual SFTs and long term follow up will be needed.

References

1. Brunori A, Cerasoli S, Donati R, Giangaspero F, Chiappetta F : Solitary fibrous tumor of the meninges : two new cases and review of the literature. **Surg Neurol** 51 : 636-640, 1999
2. Caroli E, Salvati M, Orlando ER, Lenzi J, Santoro A, Giangaspero F : Solitary fibrous tumors of the meninges : report of four cases and literature review. **Neurosurg Rev** 27 : 246-251, 2004
3. Cassarino DS, Auerbach A, Rushing EJ : Widely invasive solitary fibrous tumor of the sphenoid sinus, cavernous sinus, and pituitary fossa. **Ann Diagn Pathol** 7 : 169-173, 2003
4. Clayton AC, Salomao DR, Keeney GL, Nascimento AG : Solitary fibrous tumor : a study of cytologic features of six cases diagnosed by fine-needle aspiration. **Diagn Cytopathol 25** : 172-176, 2001
5. England DM, Hochholzer L, McCarthy MJ : Localized benign and malignant fibrous tumors of the pleura. A clinicopathologic review of 223 cases. **Am J Surg Pathol 13** : 640-658, 1989
6. Eom DW, Khang SK, Kim CJ, Ro JY : Solitary fibrous tumor of meninges in pituitary fossa. A Case Report. **Korean J Pathol 37** : 137-140, 2003
7. Hasegawa T, Matsuno Y, Shimoda T, Hasegawa F, Sano T, Hirohashi S : Extrathoracic solitary fibrous tumors : their histological variability and potentially aggressive behavior. **Hum Pathol 30** : 1464-1473, 1999
8. Klemperer P, Rabin CB : Primary neoplasm of the pleura. A report of five cases. **Arch Pathol 11** : 385-412, 1931
9. Kocak A, Cayl SR, Sarac K, Aydn NE : Intraventricular solitary fibrous tumor : an unusual tumor with radiological, ultrastructural, and mmunohistochemical evaluation : Case Report. **Neurosurgery 54** : 213-217, 2004
10. Kong TS, Son HJ, Choi HY, Moon WS, Chung MJ : Solitary Fibrous Tumor of the Meninges. **J Korean Neurosurg Soc 30** : 1439-1442, 2001
11. Lee JM : Meningeal solitary fibrous tumor. **J Korean Neurosurg Soc 37** : 232-234, 2005
12. Lo CP, Chen CY, Lin CK, Chin SC, Juan CJ, Hsueh CJ : Parasellar solitary fibrous tumor of meninges : magnetic resonance imaging features with pathologic correlation. **J Neuroimaging 14** : 281-284, 2004
13. Martin AJ, Fisher C, Igbaseimokumo U, Jarosz JM, Dean AF : Solitary fibrous tumours of the meninges : case series and literature review. **J Neurooncol 54** : 57-69, 2001
14. Miyashita K, Hayashi Y, Fujisawa H, Hasegawa M, Yamashita J : Recurrent intracranial solitary fibrous tumor with cerebrospinal fluid dissemination. Case report. **J Neurosurg 101** : 1045-1048, 2004
15. Moon WS, Lee KM : Solitary fibrous tumor clinico-pathological analysis of 11 Cases. **J Korean Cancer Assoc 31** : 847-854, 1999
16. Ogawa K, Tada T, Takahashi S, Sugiyama N, Inaguma S, Takahashi SS, et al : Malignant solitary fibrous tumor of the meninges. **Virchows Arch 444** : 459-464, 2004
17. Perez-Nunez A, Rivas JJ, Ricoy JR, Miranda P, Arrese I, Lobato RD, et al : Solitary fibrous tumor of the tentorium cerebelli. Case report. **J Neurosurg Sci 48** : 59-62, 2004
18. Perry A, Scheithauer BW, Nascimento AG : The immunophenotypic spectrum of meningeal hemangiopericytoma : a comparison with fibrous meningioma and solitary fibrous tumor of meninges. **Am J Surg Pathol 21** : 1354-1360, 1997
19. Pizem J, Matos B, Popovic M : Malignant intracranial solitary fibrous tumour with four recurrences over a 30-year period. **Neuropathol Appl Neurobiol 30** : 696-701, 2004
20. Rodriguez F, Scheithauer BW, Ockner DM, Giannini C : Solitary fibrous tumor of the cerebellopontine angle with salivary gland heterotopia : a unique presentation. **Am J Surg Pathol 28** : 139-142, 2004
21. Suzuki SO, Fukui M, Nishio S, Iwaki T : Clinicopathological features of solitary fibrous tumor of the meninges : An immunohistochemical reappraisal of cases previously diagnosed to be fibrous meningioma or hemangiopericytoma. **Pathol Int 50** : 808-817, 2000
22. Tihan T, Viglione M, Rosenblum MK, Olivi A, Burger PC : Solitary fibrous tumors in the central nervous system. A clinicopathologic review of 18 cases and comparison to meningeal hemangiopericytomas. **Arch Pathol Lab Med 127** : 432-439, 2003