CASE REPORT

J Korean Neurosurg Soc 41 : 425-428, 2007

Recurrent Sellar and Suprasellar Hemangiopericytoma

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Hemangiopericytoma (HPC) is a rare tumor with uncommon location in the central nervous system. We report a rare case of sellar and suprasellar HPC mimicking pituitary adenoma in a patient initially seen with the symptoms of bitemporal hemianopsia, headache, and panhypopituitarism. Magnetic resonance imaging of the brain revealed a contrast-enhancing soft tissue mass arising from the pituitary fossa, with apparent compression of the chiasm and involving the cavernous sinus. Subtotal resection of the tumor was achieved via a pterional approach. Histopathological examination identified the tumor as a HPC. Nine years later, the tumor recurred. To our knowledge, this is the first reported case of sellar and suprasellar HPC in Korea.

KEY WORDS : Hemangiopericytoma · Sellar · Suprasellar · Recurrent.

Introduction

Hemangiopericytoma (HPC) is a potentially malignant tumor originating from Zimmermann's pericytes around capillaries and postcapillary venules, which is most common in soft tissues[16]. Intracranial HPCs are very rare, accounting for less than 1% of primary central nervous system (CNS) tumors[2,14,15], and constitute 2% to 4% of large series of meningiomas[17]. Their location is usually supratentorial, most commonly parasagittal or falcine[15,17] (mimicking meningioma)[15], but they may develop wherever capillary vessels are present. We report here an extremely rare case of recurrent sellar and suprasellar HPC mimicking a pituitary tumor and review the relevant literature.

Case Report

A 44-year-old male patient was admitted with a 2-month history of headache, and visual loss in the both eyes via the local hospital. On admission to our hospital, ophthalmological examination found a visual acuity of 0.15 in the both eyes. A visual field examination uncovered a bitemporal hemianopsia. The other cranial nerves were intact. Endocrinological studies showed hypopituitarism. He had no past medical history of hypertension, diabetes mellitus, pulmonary tuberculosis, or hepatitis.

The brain magnetic resonance (MR) images revealed an intrasellar and suprasellar mass with the compression of the optic chiasm. The mass margin was lobulated. The cavernous portion of the left internal carotid artery was encased by the mass, but there was no evidence of invasion at MR angiography. The mass was isosignal with lower signal foci on T1-weighted images, inhomogeneous signal intensity on T2-weighted images, and diffuse homogeneous enhancement on gadolinium-enhanced T1-weighted images. The lesion measured 30 × 25 × 40 mm and extended cephalad, almost touching the chiasm (Fig. 1A, B). A presumptive diagnosis of nonfunctional pituitary adenoma was made on the basis of neuroradiological studies.

The patient underwent staged operations using bilateral pterional approaches with the interval of a month. After craniotomy, a highly vascularized tumor was encountered. There was profuse bleeding during the surgical procedure. A piecemeal subtotal resection was performed and fragments were obtained for histological diagnosis. Pathological examination of a frozen section established that the lesion was a meningioma. Particular attention was paid to obtaining complete hemostasis. Fat graft was placed in the sella after most of the tumor had been resected. The hemangiopericytoma was confirmed pathologically (Fig. 2A). Postoperative recovery was uneventful and visual

• Received : March 6, 2007 • Accepted : May 17, 2007

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function was recovered partially. Postoperative MR images suggested a residual tumor (Fig. 1C). The patient received radiotherapy (52 Gy in 30 doses) to the pituitary fossa.

Nine years later, the patient was referred to our institution because of patchy visual loss. MR images of the brain showed a contrast-enhancing soft tissue mass arising from the pituitary fossa (Fig. 1D). Cerebral angiography indicated that a branch of the meningohypophyseal trunk of the right internal carotid artery was supplying the soft tissue mass. Partial tumor removal was done and the pathologic findings were compatible with hemangiopericytoma (Fig. 2B). Immunohistochemistry of the tumor demonstrated reticulin and CD34 positive, MIB index positive in less than 1% of cells, topoisomerase II alpha expression index positive in 3% of cells and S-100, epithelial membrane antigen, and glial fibrillary acidic protein negative (Fig. 2C, D, E).

Postoperative recovery was uneventful and the patient now plans to undergo stereotactic radiosurgery.

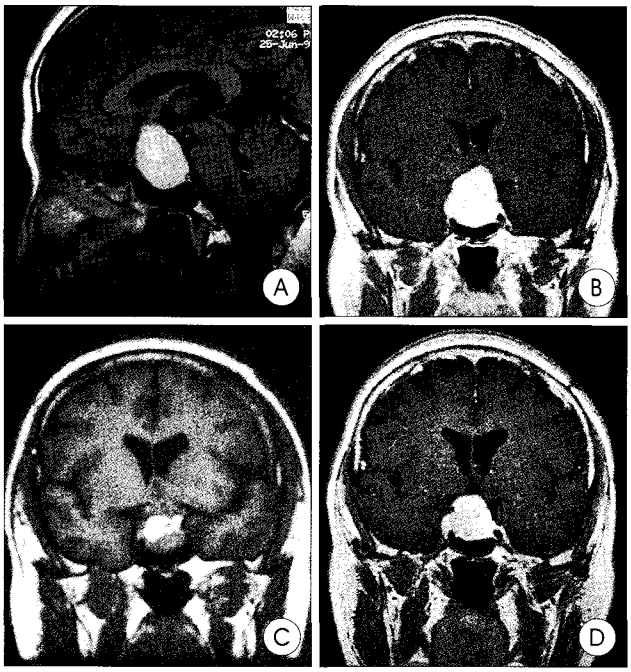

Fig. 1. Serial magnetic resonance images. A, B : Preoperative magnetic resonance images showing a sellar and suprasellar solid mass. C : Postoperative magnetic resonance image showing a remnant mass and packed fat tissue. D : Magnetic resonance image taken 9 years after operation and radiotherapy showing a recurrent tumor arising from the pituitary fossa.

Discussion

Intracranial HPC, a rare vascular tumor of the CNS, was accepted as a hemangiopericytic or angioblastic variant of meningioma[30]. HPCs are located similar to meningiomas, with 15% in the posterior fossa and 15% in the spine[4,15,28]. The majority of HPCs are supratentorial, commonly parasagittal or falcine[15]. Sellar or suprasellar HPC is very rare. Guthrie et al.[15] reported that only 2 out of 44 cases of HPC treated between 1938 and 1987 were located in the sellar or suprasellar area (Table 1). Jaaskelainen et al.[17] reported no cases of HPC in the sellar or suprasellar area out of 21 patients with primary intracranial meningeal HPC treated between 1954 and 1983. Very few cases of sellar or suprasellar HPC have been reported in the literature[15,25].

Male patients comprise 55% to 70% of patients with HPC[11,17], with the average age ranging from 38 to 42 years[17,26,28]. The average symptomatic interval for the supratentorial HPC was 8 months[15], while ordinary meningiomas are most often

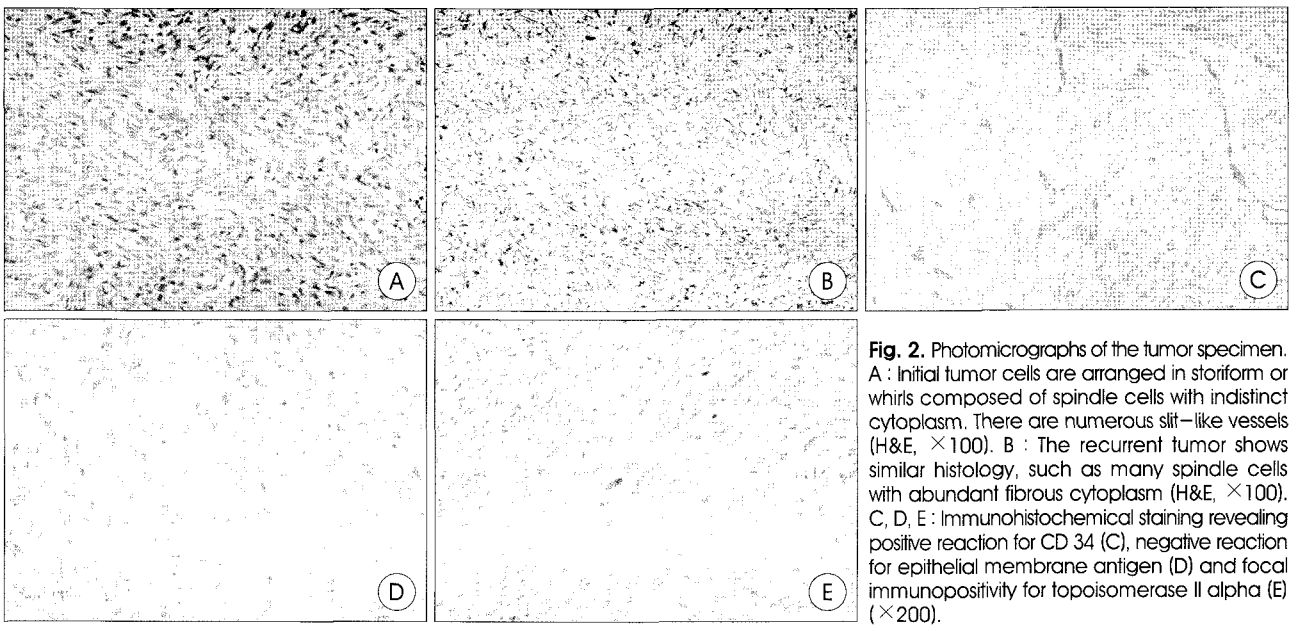

Fig. 2. Photomicrographs of the tumor specimen. A : Initial tumor cells are arranged in storiform or whirls composed of spindle cells with indistinct cytoplasm. There are numerous slit-like vessels (H&E, ×100). B : The recurrent tumor shows similar histology, such as many spindle cells with abundant fibrous cytoplasm (H&E, ×100). C, D, E : Immunohistochemical staining revealing positive reaction for CD 34 (C), negative reaction for epithelial membrane antigen (D) and focal immunopositivity for topoisomerase II alpha (E) (×200).

Table 1. Tumor Location

Location	Number of patients (%)
Supratentorial	27 (62)
Parasagittal/falx	14
Convexity	6
Middle fossa	3
Basifrontal/sphenoid	2
Suprasellar	1
Sellar	1
Posterior fossa	6 (13)
Tentorial	6 (13)
Torcular	2 (5)
Foramen magnum	2 (2)
Spine	2 (5)

symptomatic for 1 to 2 years[24], perhaps due to the more rapid growth rate of HPCs[15]. Presenting symptoms are related to tumor location[15,17,28].

Imaging features of intracranial HPCs are similar to those of meningiomas but, in HPCs, there is no calcification[15], and bone erosion is seen in more than 50% of them. They display prominent internal signal voids, suggesting feeding arteries and marked enhancement.

The definitive diagnosis of primary HPC of the CNS requires histological examination[15]. The tumors are very cellular, with round to oval cells. The cells are arranged around the thin-walled vascular space lined by a non-neoplastic endothelium (staghorn capillaries)[19]. Reticulin stains are useful in the diagnosis of HPC but the amount of reticulin fibers varies from case to case and even in different areas of the tumor[27]. Immunohistochemistry can differentiate HPC from meningioma : HPC stains positive for CD 34, factor VIII, and vimentin but negative for S-100 and epithelial membrane antigen (EMA)[12,21,22,30,33]. Immunohistochemical staining with PC10, a monoclonal antibody to proliferating cell nuclear antigen (PCNA), may be of particular value in the identification of patients at the greatest risk for rapid tumor metastasis and early death[36]. Tanaka et al.[34] found that topoisomerase II alpha expression was as well available for the marker of cell proliferative potential and one of the predicting factors for tumor reccurrence in meningioma and HPC as MIB-1 staining index. Chacko et al.[6] suggested that a topoisomerase II alpha proliferation index of 5% or greater is a reliable predictor of recurrence in intracranial HPC.

HPCs can recur locally or distantly in the neural axis or metastasize to extraneural sites[1-3,9,15,17,18,22,31,32]. Kim et al.[18] reported a 38.7% local recurrence rate, but other published rates of local recurrence for HPC are much higher, around 80%[2,3,10]. Many authors have reported that recurrence is late event[2,3,10,15,17]. Brunori et al.[3] reported that the mean interval to first recurrence was 84 months and a recent study by Kim et al.[18] determined the overall disease recurrence-free rates 5 and 10 years after initial surgery were 59.2% and 33.6%, respectively. Soyuer et al.[31] reported 5, 10, and 15-year overall survival rates of 85%, 68%, and 43%, respectively. Because recurrence is a late event in the natural history of HPC, we think that close follow-up for a long period after the first operation is needed.

Unlike most primary intracranial tumors, HPC metastasizes to extraneural sites, most commonly to the bone, liver, and lungs[8,15,17,18,23]. Koyama et al.[20] found that metastatic tumors appeared at a mean of 8 years, and as long as 16 years, after initial therapy.

Considering several current report, it seems that complete excision favorably affects recurrence and survival, as opposed to incomplete excision[2,9,15,17]. Kim et al.[18] reported that complete excision at the first operation significantly extended the average time before the first recurrence from 43 to 111 months, and the 5-year recurrence-free rates for the groups with complete excision and incomplete excision were 72.7% and 20.8%, respectively. In the Soyuer study, the 5-year local control rates for patients treated with gross total resection and subtotal resection were 84% and 38%, respectively[31].

Several retrospective studies have shown that radiotherapy after surgery is beneficial for patients with HPC[9,15,18]. Guthrie et al.[15] found that radiotherapy significantly prolonged the disease recurrence-free interval. Dufour et al.[9] reported that local disease recurrence rates for patients treated with surgery plus adjuvant radiotherapy and patients treated with surgery alone were 12% and 88%, respectively. Patients receiving more than 51 Gy irradiation experienced less local disease recurrence[1,9,10,15,23]. Some authors have also recommended preoperative radiotherapy in the management of case of HPC that involve considerable surgical risk[5,13,35].

Stereotactic radiosurgery is an effective treatment for recurrent HPCs[2,7-9,29]. Dufour et al.[9] suggested that radiosurgery is indicated for recurrent tumors measuring less than 30 mm in their greatest diameter. Sheehan et al.[29] recently reported a rate of 80% local control of recurrent HPC treated with radiosurgery. Generally, the early shrinkage of HPC after radiosurgery differs from the characteristic slow, modest response of meningioma treated using identical radiosurgical dose volume parameters[18]. This difference has been attributed to the rich blood supply of HPC[2].

In our case, local tumor recurrence seems to be attributable to incomplete excision, although the MIB index and topoisomerase II alpha proliferation index were both low and radiotherapy dose after surgery was sufficient.

Sellar and suprasellar HPC can mimic a pituitary tumor ophthalmologically, endocrinologically and radiologically. But, it carries a much graver prognosis. Histological confirmation

is essential for optimum management, even when clinical features appear to be diagnostic.

Conclusion

We report a case of recurrent sellar and suprasellar HPC mimicking a pituitary tumor without extraneural metastasis. It deserves a certain interest due to its rarity and can be also considered in the differential diagnosis of pituitary fossa tumors.

References

1. Alen JF, Lobato RD, Gomez PA, Boto GR, Lagares A, Ramos A, et al : Intracranial hemangiopericytoma : study of 12 cases. **Acta Neurochir (Wien) 143** : 575-589, 2001
2. Bastin KT, Mehta MP : Meningeal hemangiopericytoma : defining the role for radiation therapy. **J Neurooncol 14** : 277-287, 1992
3. Brunori A, Delitala A, Oddi G, Chiappetta F : Recent experience in the management of meningeal hemangiopericytomas. **Tumori 83** : 856-861, 1997
4. Cappabianca P, Mauri F, Pettinato G, Di Preisco B : Hemangiopericytoma of the spinal canal. **Surg Neurol 15** : 298-302, 1981
5. Carella RJ, Ransohoff J, Newall J : Role of radiation therapy in the mangement of meningioma. **Neurosurgery 10** : 332-339, 1982
6. Chacko G, Chacko AG, Rajshekhar V, Muliyil JP : Intracranial hemangiopericytomas : correlation of topoisomerase II alpha expression with biologic behavior. **Surg Neurol 65** : 11-17, 2006
7. Chang SD, Sakamoto GT : The role of radiosurgery for hemangiopericytomas. **Neurosurg Focus 14** : 1-5, 2003
8. Coffey RJ, Cascoino TL, Shaw EG : Radiosurgical treatment of recurrent Hemangiopericytomas of the meninges : preliminary results. **J Neurosurg 78** : 903-908, 1993
9. Dufour H, Metellus P, Fuentes S, Murracciole X, Regis J, Figarella-Branger D, et al : Meningeal hemangiopericytoma : a retrospective study of 21 patients with special review of postoperative external radiotherapy. **Neurosurgery 48** : 756-763, 2001
10. Dziuk TW, Woo S, Butler EB, Thomby J, Grossman R, Dennis WS, et al : Malignant meningioma : an indication for initial aggressive surgery and adjuvant radiotherapy. **J Neurooncol 37** : 177-188, 1998
11. Fabiani A, Favero M, Trebini F : On the primary meningeal tumors with special concern to the hemangiopericytoma pathology and biology. **Zentralbl Neurochir 41** : 273-284, 1980
12. Fountas FN, Kapsalaki E, Kassam M, Feltes CH, Dimopoulos VG, Robinson JS, et al : Management of intracranial meningeal hemangiopericytomas : outcome and experience. **Neurosurg Rev 29** : 145-153, 2006
13. Fukui M, Kitamura K, Ohgami S, Takaki T, Kinoshita K : Radiosensitivity of meningioma-analysis of five cases of highly vascular meningioma treated by preoperative irradiation. **Acta Neurochir 36** : 47-60, 1977
14. Goellner JR, Laws ER Jr, Soule EH, Okazaki H : Hemangiopericytoma of the meninges. Mayo Clinic experience. **Am J Clin Pathol 70** : 375-380, 1978
15. Guthrie BL, Ebersold MJ, Scheithauer BW, Shaw EG : Meningeal hemangiopericytoma : histopathological features, treatment, and long-term follow-up fo 44 cases. **Neurosurgery 25** : 512-522, 1989
16. Horten BC, Urich H, Rubinstein LJ, Montague SR : The angioblastic meningioma : a reappraisal of the nosological problem. Light-, electron-microscopic, tissue, and organ culture observation. **J Neurol Sci 31** : 387-410, 1977
17. Jaaskelainen J, Servo A, Haltia M, Wahlstrom T, Valtonen S : Intracranial hemangiopericytoma : radiology, surgery, radiotherapy, and outcome in 21 patients. **Surg Neurol 23** : 227-236, 1985
18. Kim JH, Jung HW, Kim YS, Kim CJ, Hwang SK, Paek SH, et al : Meningeal hemangiopericytomas : long-term outcome and biological behavior. **Surg Neurol 59** : 47-54, 2003
19. Kochanek S, Schroder R, Firsching R : Hemangiopericytoma of the meninges : Histopathological variability and differential diagnosis. **Zentralbl Neurochir 47** : 183-190, 1986
20. Koyama H, Harada A, Nakao A, Nonami T, Kurokawa T, Kaneko T, et al : Intracranial hemangiopericytoma with metastasis to the pancreas. Case report and literature review. **J Clin Gastroenterol 25** : 706-708, 1997
21. Kroh H, Figols J, Sobieraj A : Intracranial hemangiopericytoma. Histological and immunohistochemical study. **Folia Neruopathol 35** : 121-127, 1997
22. Mena H, Ribas JL, Pezeshkpour GH, Cowan DN, Parisi JE : Hemangiopericytoma of the Central Nervous System : A Review of 94 Cases. **Hum Pathol 22** : 84-91, 1991
23. Mira JG, Chu FC, Fortner JG : The role of radiotherapy in the management of malignant hemangiopericytoma. Report of eleven new cases and review of the literature. **Cancer 39** : 1254-1259, 1997
24. Mirimannoff RO, Dosoretz DE, L inggood RM, O Jemann RG, Martuza RL : Meningioma : Analysis of recurrence and progression following neurosurgical resection. **J Neruosurg 62** : 18-24, 1985
25. Morrison DA, Bibby K : Sellar and Suprasellar Hemangiopericytoma Mimicking Pituitary Adenoma. **Arch Ophthalmol 115** : 1201-1203, 1977
26. Pitkethly DT, Hardman JM, Kempe LG, Earle KM : Angioblastic meningiomas. Clinicopathological study of 81 cases. **J Neurosurg 32** : 539-544, 1970
27. Russell DS, Rubinstein LJ : **Pathology of Tumours of the Nervous System**, ed 5. Baltimore MD : Williams and Wilkins, 1989, pp473-479
28. Schroder R, Firsching R, Kochanek S : Hemangiopericytoma of the meninges. II.General and clinical data. **Zentralbl Neurochir 47** : 191-199, 1986
29. Sheehan J, Kondziolka D, Flickinger K, Lunsford LD : Radiosurgery for treatment of recurrent intracranial hemangiopericytomas. **Neurosurgery 51** : 905-911, 2002
30. Sim JE, Jeong JS, Choi SS, Choi YM, Kim KU : Cystic Hemangiopericytoma in the Third Ventricle. **J Korean Neurosurg Soc 40** : 467-470, 2006
31. Soyuer S, Chang EL, Selek U, McCutcheon IE, Maor MH : Intracranial Meningeal Hemangiopericytoma : The Role of Radiotherapy. Report of Cases and Review of the Literature. **Cancer 100** : 1491-1497, 2004
32. Stout AP, Murray MR : Hemangiopericytoma : Study of 25 new case. **Cancer 2** : 1027-1035, 1942
33. 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 flbrous meningioma or hemangiopericytoma. **Pathol Int 50** : 808-817, 2000
34. Tanaka M, Kubo O, Tajika Y, Takahashi T, Suzuki S, Takakura K : Immunohistochemical evaluation of intracranial recurrent meningiomas : correlation of topoisomerase II alpha expression and cell proliferative potential. **No To Shinkei 51** : 1033-1039, 1999
35. Uemura S, Kuratsu J, Hamada J, Yoshioka S, Kochi M, Ushio Y, et al : Effect of radiation therapy against intracranial hemangiopericytoma. **Neurol Med Chir (Tokyo) 32** : 328-332, 1990
36. Yu CC, Hall PA, Fletcher CD, Camplejohn RS, Waseem NH, Lane DP, et al : Hemangiopericytomas : the prognostic value of immunohistochemical staining with monoclonal antibody to proliferating cell nuclear antigen (PCNA). **Histopathology 19** : 29-33, 1991