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 Zimmerman's pericytes around capillaries and postcapillary venules, which is most common in soft tissues. Intracranial HPCs are very rare, accounting for less than 1% of primary central nervous system (CNS) tumors, and constitute 2% to 4% of large series of meningiomas. Their location is usually supratentorial, most commonly parasagittal or falciens (mimicking meningioma), 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 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...
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 meningohipophyseal 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 fibrillar acidic protein negative (Fig. 2C, D, E).

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

**Discussion**

Intracranial HPC, a rare vascular tumor of the CNS, was accepted as a hemangiopericytic or angioblastic variant of meningioma. HPCs are located similar to meningiomas, with 15% in the posterior fossa and 15% in the spine. The majority of HPCs are supratentorial, commonly parasagittal or falx. Sellar or suprasellar HPC is very rare. Guthrie et al. 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. 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.

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

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**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.

**Fig. 2.** Photomicrographs of the tumor specimen. A: Initial tumor cells are arranged in storiform or whorls 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 CD34 (C), negative reaction for epithelial membrane antigen (D) and focal immunopositivity for topoisomerase II alpha (E) (× 200).
Table 1. Tumor Location

<table>
<thead>
<tr>
<th>Location</th>
<th>Number of patients (%)</th>
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<tbody>
<tr>
<td>Supratentorial</td>
<td>27 (62)</td>
</tr>
<tr>
<td>Parasagittal/ Tempix</td>
<td>14</td>
</tr>
<tr>
<td>Cerebrally</td>
<td>6</td>
</tr>
<tr>
<td>Middle fossa</td>
<td>3</td>
</tr>
<tr>
<td>Basifrontal/sphenoid</td>
<td>2</td>
</tr>
<tr>
<td>Suprasellar</td>
<td>1</td>
</tr>
<tr>
<td>Suprasellar</td>
<td>1</td>
</tr>
<tr>
<td>Posterior fossa</td>
<td>6 (13)</td>
</tr>
<tr>
<td>Tentorial</td>
<td>6 (13)</td>
</tr>
<tr>
<td>Temporal</td>
<td>2 (5)</td>
</tr>
<tr>
<td>Foramen magnum</td>
<td>2 (2)</td>
</tr>
<tr>
<td>Spine</td>
<td>2 (5)</td>
</tr>
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</table>

Symptomatic for 1 to 2 years, perhaps due to the more rapid growth rate of HPCs. Presenting symptoms are related to tumor location. Imaging features of intracranial HPCs are similar to those of meningiomas but, in HPCs, there is no calcification, 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. 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 (tangential capillaries). 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. 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).

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. Tanaka et al. 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 recurrence in meningioma and HPC as MIB-1 staining index. Chacko et al. 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. Kim et al. reported a 38.7% local recurrence rate, but other published rates of local recurrence for HPC are much higher, around 80%. Many authors have reported that recurrence is late event. Bruno et al. reported that the mean interval to first recurrence was 84 months and a recent study by Kim et al. determined the overall disease recurrence-free rates 5 and 10 years after initial surgery were 59.2% and 33.6%, respectively. Suyer et al. 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 liver, bone, lung, and other organs. Koyama et al. found that metastatic tumors appeared at a mean of 8 years, and as long as 16 years, after initial therapy.

Considering several current reports, it seems that complete excision favorably affects recurrence and survival, as opposed to incomplete excision. Kim et al. 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 Suyer study, the 5-year local control rates for patients treated with gross total resection and subtotal resection were 84% and 38%, respectively.

Several retrospective studies have shown that radiotherapy after surgery is beneficial for patients with HPC. Guthrie et al. found that radiotherapy significantly prolonged the disease recurrence-free interval. Dufour et al. 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. Some authors have also recommended preoperative radiotherapy in the management of case of HPC that involve considerable surgical risk.

Stereotactic radiosurgery is an effective treatment for recurrent HPCs. Dufour et al. suggested that radiosurgery is indicated for recurrent tumors measuring less than 30 mm in their greatest diameter. Sheehan et al. 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. This difference has been attributed to the rich blood supply of HPC.

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