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# Meningeal Hemangiopericytoma : Study of 6 Cases and Review of the Literatures

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**Objective :** Hemangiopericytoma is known as a malignant tumor originating from pericytes and rarely occurs in the central nervous system. We present 6 cases of pathologically confirmed meningeal hemangiopericytoma.

**Methods :** Retrospective study was done based on patient's recordings including radiological studies. Each case of tumors was treated surgically and postoperative radiotherapy was done.

**Results :** There were 5 cases of intracranial and 1 case of spinal hemangiopericytomas. Three of 5 intracranial hemangiopericytomas were located at tentorial region. Total tumor removal was done in 4 cases and postoperative local recurrence (or regrowth) was noted in 3 cases despite of postoperative external radiation therapy, 2 of which had died.

**Conclusion :** Our cases show more frequent tentorial locations and poor clinical outcomes of hemangiopericytomas compared with meningiomas.

**KEY WORDS :** Meningeal hemangiopericytoma · Radiotherapy · Surgery.

## Introduction

Meningeal hemangiopericytomas have been reported as 2 to 4% of meningeal tumors and comprising less than 1% of all intracranial tumors[12]. Most intracranial hemangiopericytomas apparently arise from the meninges, and often misdiagnosed as meningiomas preoperatively. There have been reports about some imaging characteristics of hemangiopericytomas[2,5], but it seems yet to be documented. In addition, the prevalent locations of hemangiopericytomas have not been demonstrated. Total surgical removal is crucial in the treatment of hemangiopericytoma because of its tendency to recur and to metastasize[11]. But, profuse bleedings from the tumors and locations adjacent venous sinuses, such as tentorial region, often makes it difficult to achieve complete removal. Postoperative external radiation therapy and radiosurgery have shown significant effect on local control in cases of subtotal resection or recurrences[6,22]. We present our clinical experiences of hemangiopericytomas with review of the literatures.

## Materials and Methods

From 1994 to 2004, there were 6 cases of hemangiopericytomas which were surgically treated and pathologically diagnosed. It constituted 1.4% of all intracranial meningiomas during the same period in our institutes.

## Results

There were 4 women and 2 men and the age was ranged from 40 years to 72 years. Tumor locations were as follows : 4 cases were supratentorial, 1 was infratentorial and 1 was located at the spinal region, C6 to T1. 2 of the supratentorial and 1 of the infratentorial tumors were originated from the tentorium. Clinical symptoms at diagnosis were variable according to the locations of the tumors (Table 1). Most cases of tumors were dural-based and isointense on both T1 and T2 weighted image(WI) with heterogenous contrast enhancement in magnetic resonance imaging(MRI) (Fig. 1). No tumor-related calcification was noted. The preoperative diagnoses

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**Table 1.** Patients with meningeal hemangiopericytoma

| Patient's age / sex | Tumor location | Size (cm) | Symptoms | Rsection* | Recurrence (or regrowth) after surgery | Result after surgery |
|---|---|---|---|---|---|---|
| 1. 56 / F | Tentorium, left | 5x5x4 | Headache | II | 14 month | Follow up lost after 14 month |
| 2. 59 / F | Tentorium, right | 6x6x3 | Headache | IV | - | Follow up lost after 2 month |
| 3. 40 / M | Parietal convexity, left | 4x4x3 | Hemiparesis, disorientation | I | N.E.R.** | Not tumor related death (24month) |
| 4. 72 / M | Spinal (C6-T1) | 3x1 | Monoparesis | V | 19 month | Death(19month) |
| 5. 61 / F | Parietal convexity, right | 6x6x4 | Decreased visual acuity | II | 9 month | Death(25month) |
| 6. 66 / F | Tentorium, left | 4x4x3 | Dizziness | I | N.E.R. | Alive(8month) |

M : male, F : female, Resection* : Simpson grade I – V, N.E.R** : No evidence of recurrences, — : not evaluated

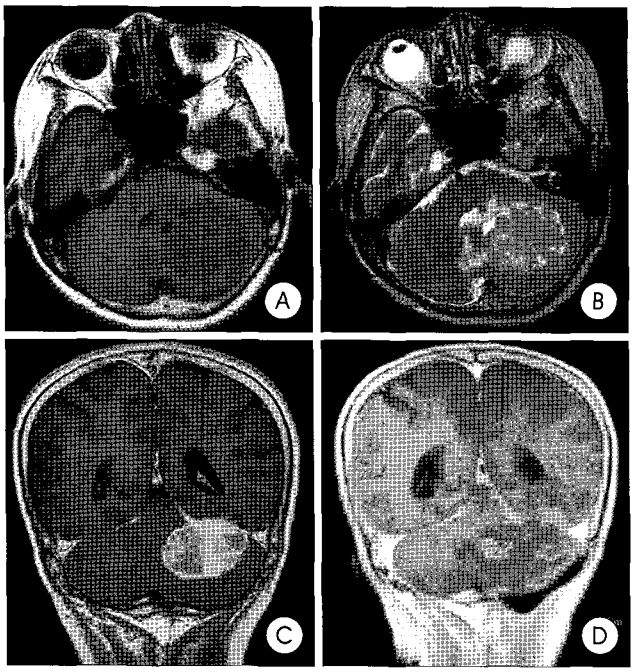


**Fig. 1.** Magnetic resonance(MR) imaging of hemangiopericytoma (Case 6) Axial T1-weighted (A) and T2-weighted (B) MR image shows a homogenous isointense mass on left cerebellar hemisphere compressing 4th ventricle. Coronal T1-weighted MR image after gadolinium injection (C) shows it is an extraaxial mass arises from tentorium cerebelli with heterogenous enhancement. Postoperative enhanced T1-weighted MR image shows no residual mass (D).

were meningiomas in all cases. Intraoperatively, the tumors showed soft, friable nature with moderate to heavy bleedings. Gross total tumor removal was done in 4 cases. Microscopically, all the tumors showed highly cellular and prominent vascular proliferation. Reticulin stains revealed dense reticular fibers around the individual tumor cells. Immunohistochemical stains showed focal positivity to CD34 and negative for epithelial membrane antigen(EMA). After surgical removal, all cases received external radiotherapy ranged from 6,120 to 7,200cGy. 3 patients have died during the follow-up period (1 case was not tumor-related death). Despite of the radiation therapy, local tumor recurrences were found in 3 cases, 2 of which were underwent re-operations but both of them died after several months (Table 1). Interestingly, tentorial locations were more frequently noted in our cases. 3 of the 5 intracranial hemangiopericytomas were originated from the tentorium.

## Discussion

Hemangiopericytoma is a rare, highly vascular and aggressive neoplasm occurs anywhere in the body[8]. Most common locations were reported as the extremities, the pelvis, and the head and neck[10]. It is thought to be arise from Zimmerman pericytes around capillaries and postcapillary venules[22]. Intracranial hemangiopericytomas are mostly dural-based and mimicking meningiomas. Once it has been considered as angioblastic subtype of meningioma but now it is classified as 'mesenchymal, non-meningothelial tumors' since 1993 when the World Health Organization separated it from meningioma. Ultrastructural findings demonstrate that intracranial hemangiopericytomas represent the counterpart to soft-tissue hemangiopericytomas[1]. Imaging studies have shown that the hemangiopericytoma usually hyperdense in computed tomography(CT) scan and variable enhancement following intravenous contrast injection. In the MRI studies it show heterogeneous, predominantly isointense on T1WI and T2WI, enhanced heterogeneously with internal vessel voids[5] and do not show calcification and hyperostosis[2]. In addition to lack of calcification, irregular or lobulated borders, mushrooming appearance, and more heterogeneous contrast enhancement have been proposed as suggestive findings of hemangiopericytomas rather than meningiomas[2]. But generally its radiological findings are so close to those of meningioma, pathological features are crucial in diagnosing this tumor. It is interesting that though the pathological findings is such important, the diagnosis of hemangiopericytoma is one of the most controversial among the vascular tumors[10]. For example, solitary fibrous tumor, which is clinically more benign than hemangiopericytomas, must be considered as differential diagnosis because it has such similar findings in immunohistochemical staining as well as in radiological imaging. Both tumors show prominent intercellular reticulin fibers, negative for S-100 protein and EMA, and diffuse positivity to CD34 though hemangiopericytomas shows more focal or patchy positivity[23]. On gross examination, hemangiopericytoma is well-circumscribed, brown lesion surrounded by a pseudo-capsule, while solitary fibrous tumor often shows gray color[10]. Electron microscopy may be helpful in differe-

**Table 2.** Tentorial locations of hemangiopericytomas in other literatures

| Authors | Total cases (intracranial) | Tentorial locations (%) |
|---|---|---|
| Jaaskelainen et al. (1985) | 21(21) | 3(15%) |
| Guthrie et al. (1989) | 44(42) | 6(14%) |
| Dufour et al. (2001) | 21(17) | 2(12%) |
| Kim et al. (2003) | 31(31) | 9(29%) |

ntiating the two tumors in that basement membrane substance may be found in hemangiopericytoma while absent in solitary fibrous tumors[9]. Some tumors may have both features of meningioma and hemangiopericytoma. Neurofibromatosis 2(NF2) gene analysis may be helpful in those situations that NF2 gene is frequently mutated in meningiomas[15].

Hemangiopericytomas are found mostly at supratentorial, less frequently at infratentorial and the spinal region[7,12]. Very rarely, intraventricular and even intraparenchymal locations have been reported[1,3,18,20]. This seems to be attributed to the fact that the origin of the tumor differs from the meningioma. Recent study suggested that hemangiopericytomas originate from meningeal pericytes, which are mesenchymal cells with contractile properties similar to smooth muscle located in the walls of capillaries, and not from arachnoid cap cells[13]. Chiechi et al.[5] reported that hemangiopericytomas occur in locations similar to meningiomas. Up to the present, the prevalent locations of hemangiopericytomas have not been described. Thus larger proportion of tentorial locations in our series, in contrast with the meningiomas whose tentorial locations are accounts for 3 to 6%[4], is rather unexpected. In our cases, 3 of 5 (60%) intracranial hemangiopericytomas were originated from the tentorium, though it is not statistically significant due to limited patient population. The incidence of hemangiopericytoma in tentorial location has not been described previously in detail. The large series of hemangiopericytomas, where tumor locations were described in detail, also showed higher incidence of tentorial locations (from 14 to 29%, Table 2). There have been reports about a tendency for the hemangiopericytoma to be located in the occipital or posterior fossa region[14,21], which may be associated with our findings. Thus one should consider hemangiopericytomas as differential diagnosis in case of meningeal tumors located in tentorium with such radiological findings as multilobulated, mushrooming appearance and lack of calcification.

The most distinguishing feature differentiating hemangiopericytomas from meningiomas may be its clinical aggressiveness. Large series of studies reported its tendency of metastasizing to other distant organs and high incidence of local recurrence after surgical removal[12,17]. Local recurrence rate was reported as ranges from 45 to 80%[6,12,19,22] and metastases ranges from 13 to 55%[11,12,14,16,22]. Complete tumor excision at the first operation extended the average time before first recurrence[17]. The mean time to local disease recurrence was 63 month to 104 month[17,22]. Postoperative radiotherapy is recommended by some authors to improve disease free survival rate[6,22]. Dufour et al.[6] suggested that postoperative radiotherapy provided benefit with regard to local disease recurrence. In our cases, 3 of 5 meningeal hemangiopericytomas (1 case was lost during follow-up period) have shown recurrence or regrowth within 19 month despite postoperative radiotherapy. Previous studies reported 5-and 10-year survival rates of 67 and 40%, respectively, with a mean survival of 7 years[12], while recent studies have shown more improved results, over 90% of 5-year survival rate[7,17]. However, our cases showed poor clinical outcomes despite of total surgical removal and postoperative radiotherapy, 2 of 5 have died within 25 month. It may be attributed to the high proliferative index of tumors but which were not studied except in the latest case (Ki labeling index about 5%).

## Conclusion

Our clinical experience of 6 cases of hemangiopericytoma shows frequent tentorial locations and poor clinical outcomes despite of total tumor removal and postoperative radiotherapy. Review of other literatures also showed prevalent tentorial locations of hemangiopericytomas than meningiomas. This preferred location may be helpful in preoperative diagnosis in addition to other radiological findings suggestive of hemangiopericytomas than meningiomas.

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## Commentary

I read with great interest the article titled 'Meningeal Hemangiopericytoma : study of 6 cases and review of the literature'. The authors reported their pathologically proven 5 intracranial hemangiopericytomas and one intraspinal case, stating tentorial location may be considered one of the differential point from meningioma in addition to other radiological differential features, such as lack of calcification, more heterogenous contrast enhancement, lobulated borders with mushrooming appearance, etc. They also attributed the poor clinical outcome of their patients than ordinary meningeal hemangiopericytomas(M-HPC) to high proliferative indices of the tumors, although only one case could be recorded to have high labeling index of 5%. As the authors reviewed the literature, these highly vascular and aggressive tumors occur most commonly in other parts of the body including extremities, pelvis, and retroperitoneum. Head and neck was also regarded not uncommon site. It is well established concept that M-HPCs are indistinguishable from those arising from other body parts and are no longer considered variants of meningiomas. M-HPCs are known to grow more often from or along the dural sinuses of falx and tentorium or skull base than conventional meningiomas having tendencies to recur and metastasize extracranially[3-5]. According to several large series of reports on locations of M-HPCs, parasagittal and falx tumors, skull base tumors were common than tentorial locations, in contrary to the authors' experience[1-3,5]. As for the 5-, 10- and 15-year survival rates, several large series reported 67~96%, 46~76%, and 23%, respectively[2,3,5]. As for the recurrence free 5-year survival rates, some 60~89% was reported mentioning that GTR and adjuvant radiotherapy or radiosurgery were the two most significant factors for recurrence free survival[1,3,5]. Salvage chemotherapy was reported not effective to prevent recurrence[2]. High grade M-HPCs recurred statistically significantly earlier than low-grade tumors[2], which might be the explanation of the poor prognosis of the authors. Guthrie et al. also demonstrated increasing extraneural metastases as time passes after the first treatments, reporting a 5-, 10-, and 15-year probability of developing metastasis of 13%, 33%, and 64%, respectively[3,5]. In conclusion, these highly vascular and aggressive tumors can be managed by radical surgery plus radiotherapy with a favourable long-term outcome. Tentorial predilection for tumor location and relatively poor clinical outcome associated with high labeling index of the tumors cannot be generalized due to the limited number of cases.

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