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# Meningeal Hemangiopericytoma Treated with Surgery and Radiation Therapy

## -Case Report-

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Meningeal hemangiopericytoma (HPC) is an uncommon dura-based tumor and can recur not only locally but also distantly in the neural axis or extraneural sites. We report our experience of radiation therapy, one preoperative and one elective postoperative, in two patients with meningeal HPC and reviewed the role of radiation therapy. A 41-year-old man (Case 1) presented with a 3-month history of headache and right hemiparesis. The mass was nearly unresectable at the first and second operation and diagnosed as meningeal HPC. Preoperative radiation therapy was given with a total dose of 55.8 Gy/31 fractions to the large residual mass of left frontoparietal area. Follow-up computerized tomography (CT) showed marked regression of tumor after radiation therapy. The third operation was performed to remove the residual tumor at 6 months after the radiation therapy and a 2×2 cm sized tumor was encountered. The mass was totally removed. The serial follow-up CT showed no evidence of recurrence and he is alive without distant metastasis for 4 years and 10 months after the first operation. A 45-year-old woman (Case 2) presented with suddenly developed headache and visual impairment. Tumor mass occupying right frontal lobe was removed with the preoperative diagnosis of meningioma. It was totally removed with attached sagittal sinus and diagnosed as meningeal HPC. Elective postoperative radiation therapy was performed to reduce local recurrence with a total dose of 54 Gy/30 fractions to the involved area of right frontal lobe. She is alive for 5 years maintaining normal activity without local recurrence and distant metastasis.

**Key Words**: Meningeal hemangiopericytoma, Surgery, Radiation therapy

Intracranial meningeal HPC is an uncommon dura-based tumor arising from Zimmerman pericytes around capillaries and postcapillary venules.[1] Most HPCs are located in the musculoskeletal system and the skin. Central nervous system HPCs are rare. Meningeal HPCs represent 2~4% in large series of meningeal tumors, thus accounting for less than 1% of all intracranial tumors. Meningeal HPCs resemble meningioma clinically and radiologically but behave more aggressively, with a tendency to metastasize. They can recur locally or distantly in the neural axis or extraneural sites.[1~10]

They are highly vascular tumors, which may bleed profusely at surgery. Therefore complete surgical resection is occasionally difficult. The currently preferred therapeutic modality is surgical resection and radical surgery is recommended when possible. However, the operative mortality in an early series was high, 14~20%. Preoperative tumor embolization with particles or glue appears to be less effective in the large meningeal tumors because of an arterial supply from multiple territories and has not widely been practiced. Therefore, in case of huge unresectable meningeal HPC, the preoperative radiation therapy could be performed and then removed successfully after radiation therapy.[11]

Radiation therapy is also necessary in operated case, postoperatively. Bastin and Mehta carried out a comprehensive review to define the role of radiation therapy and concluded that radiation therapy reduced the local recurrence rate and thereby prolonged disease-free and overall survival, particularly if the

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dose given was greater than 50 Gy.[12]

We review our experience of radiation therapy, one preoperative and one elective postoperative, in two patients with meningeal HPC and the role of radiation therapy.

## Case

### Case 1

A 41-year-old man presented in January 2001 with a 3-month history of headache and right hemiparesis (G4/ G4).

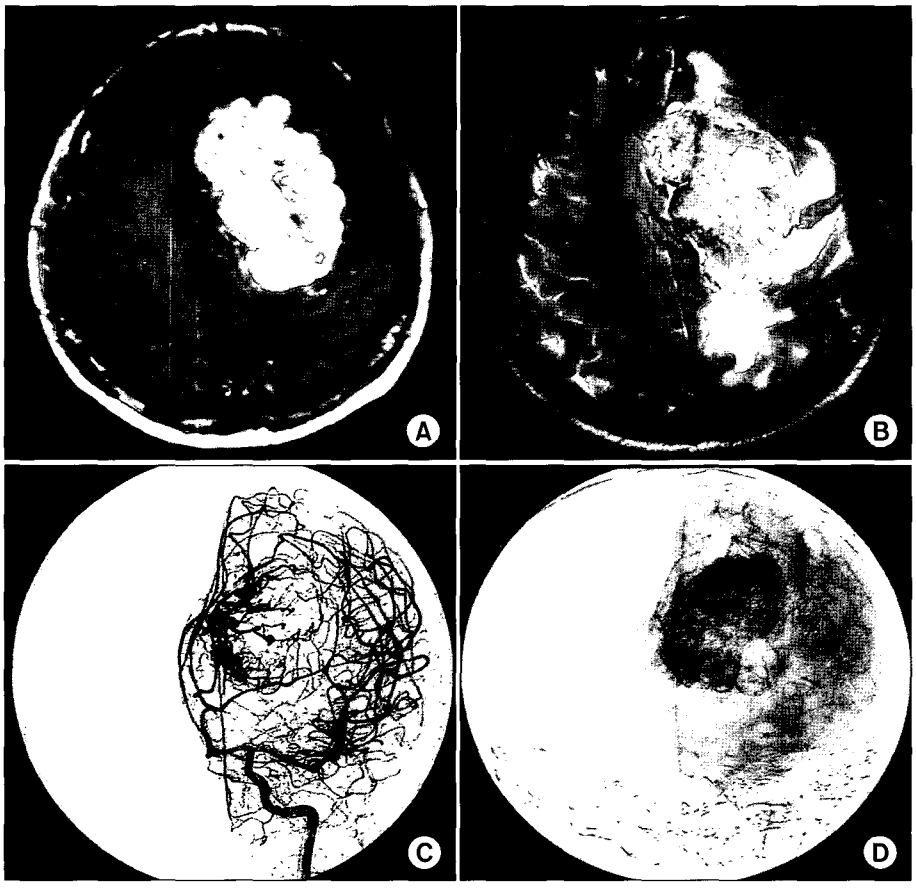


**Fig. 1.** MRI and angiogram before the first operation in Case 1, a 41-year-old man. (A) & (B) MRI showing a huge mass in the left frontoparietal lobe with surrounding edema and midline deviation. (C) & (D) Carotid angiogram showing corkscrew-like appearance of tumor staining.

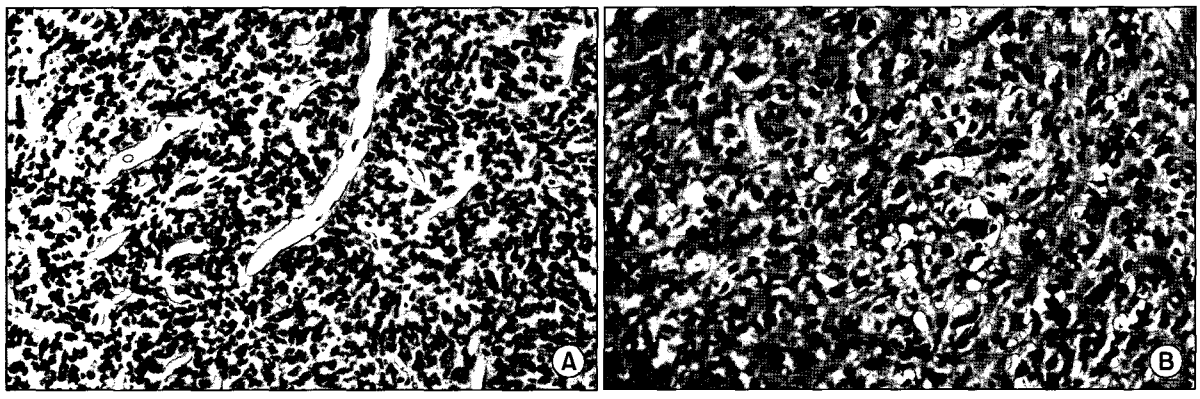


**Fig. 2.** (A) Before irradiation, the tumor is composed of a branching networks of capillaries surrounded by proliferation of polygonal and spindle cells. (B) After irradiation, the tumor shows bizarre, densely hyperchromatic nuclei and vacuolated changes. Hematoxylin and eosin stain, original magnification ×200.

Magnetic resonance imaging (MRI) showed a huge mass lesion on left frontoparieto-occipital area, suggestive of meningioma. Angiograms showed the corkscrew-like appearance of tumor staining (Fig. 1). At the first operation, the mass was not removed due to massive bleeding and only extraventricular drainage on right Kocher's point (frontal ventriculostomy) was performed to relieve brain swelling due to tumor mass. Preoperative embolization was performed on left external carotid artery for bleeding control at 2 weeks after the first operation. At the second operation, 1 week after the embolization, thetumor looked gray white, highly vascular, and relatively well marginated and surrounding parenchyme was edematous. The massive bleeding was seen again due to incomplete embolization, so the mass was not removed in fear of extensive blood loss. The pathologic diagnosis was meningeal HPC (Fig. 2A). Even after the second operation, the residual mass was very large and nearly the same as preoperative mass (Fig. 3). Preoperative radiation therapy was started 1 month later to change this unresectable tumor into resectable one. He was treated supine using anterior and two lateral wedged fields with 6 MV X-ray. A total dose of 55.8 Gy was given to left frontoparietal area in 31 fractions over a 9-week period. Treatment period was prolonged due to discharge from the scar edge after 30.6 Gy. The limping gait due to right hemiparesis was markedly improved after radiation therapy and follow-up CT showed marked regression of tumor after radiation therapy (Fig. 4). The third operation was performed to remove the residual tumor at 6 months after the radiation therapy and a 2×2 cm sized tumor was encountered and totally removed.

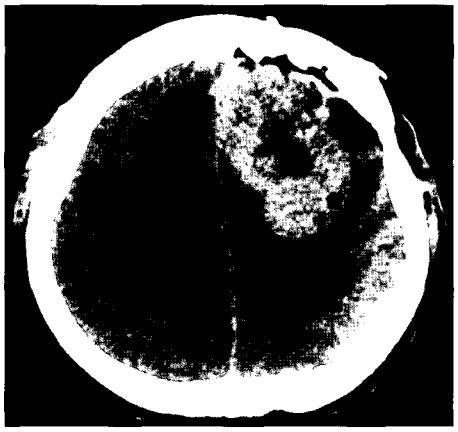

**Fig. 3.** CT after the second operation and before the radiation therapy in Case 1.

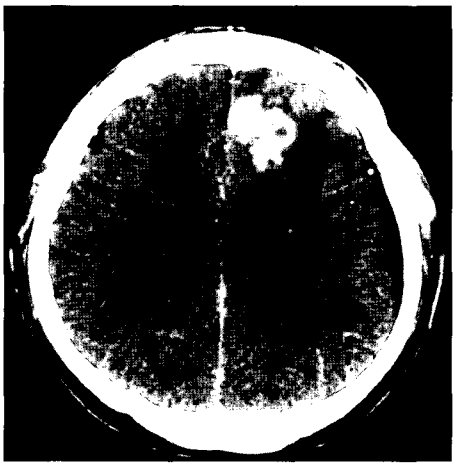

**Fig. 4.** CT at three months after the radiation therapy in Case 1. Nodular and irregularly marginated mass is seen in the left frontoparietal lobe, showing dense homogenous enhancement. The surrounding low density edema is also seen. The mass is markedly reduced after the radiation therapy.

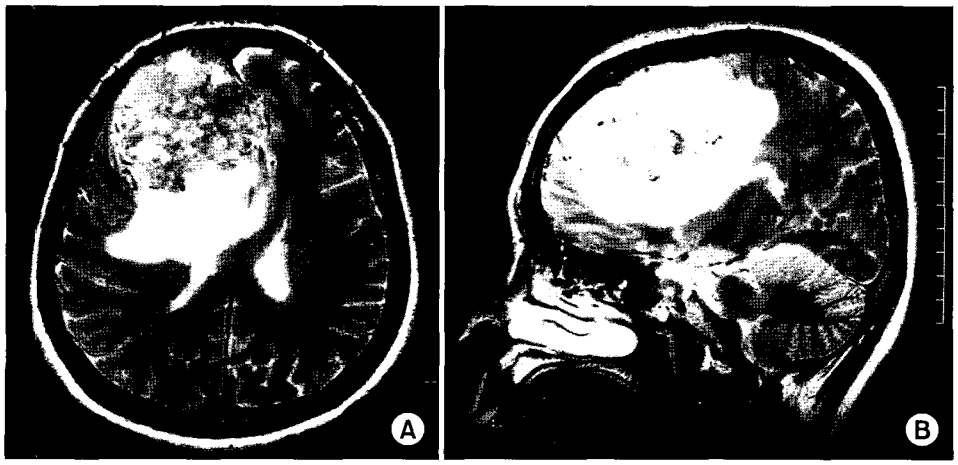


**Fig. 5.** Preoperative MRI of HPC in the right frontal lobe, Case 2, a 45-year-old woman. (A) & (B) Axial and sagittal MRI show a 3.5×4×5 cm sized mass in the right frontal lobe with surrounding edema.

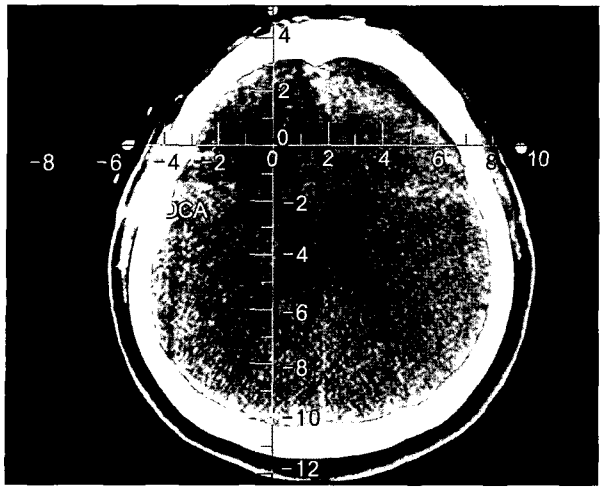


**Fig. 6.** Postoperative CT image shows complete removal of the tumor in the right frontal lobe.

It was hard, multi-lobe appearance, marble-like colored, and tightly attached to subgaleal layer. After irradiation the tumor showed bizarre, densely hyperchromatic nuclei and vacuolated changes (Fig. 2B). The follow-up MRI at 18 months after the completion of radiation therapy, showed no evidence of recurrence. The serial follow-up CT showed no evidence of recurrence and encephalomalacia with ipsilateral dilated ventricle in left frontoparietal area. He is alive for 4 years and 10 months after the first operation without distant metastasis.

**Case 2**

A 45-year-old woman presented with suddenly developed headache and visual impairment. MRI showed a well demarcated, 3.5×4×5 cm sized tumor mass in the right frontal lobe with attachment to sagittal sinus and well enhancement (Fig. 5). The preoperative diagnosis was meningioma. Tumor mass was totally removed with attached sagittal sinus and pathologic diagnosis was meningeal HPC. The headache was improved but visual impairment was not changed postoperatively. Elective postoperative radiation therapy was referred to reduce local recurrence even after total removal (Fig. 6).

The total dose of 54 Gy was delivered to the involved area of right frontal lobe in 30 fractions. The follow-up MRI was checked 12 months after operation and showed the absence of recurrence. She is alive for 5 years maintaining normal activity without distant metastasis after treatment.

## Discussion

Many authors have argued about the true origin of this tumor. The current World Health Organization classification of central nervous system tumors distinguishes HPC as an entity of its own, and classified it into the group of mesenchymal, nonmeningothelial tumors.[2] Meningeal HPCs usually occur during adulthood, particularly among men, and the average age of at diagnosis ranges from 32 to 41 years in different series. Approximately 10% of meningeal HPCs occur in children.[1]

HPCs have a relentless tendency for local recurrence and metastases outside the central nervous system which can appear even many years after diagnosis and adequate treatment of the primary tumor.[1~9] Unlike most primary intracranial tumors, meningeal HPC metastasizes to extraneural sites, most commonly to the bone, liver, and lung. Kim[9] reported that 5- and 10-year metastasis rates were 4% and 25%, respectively. Soyuer[1] reported that 55% developed extraneural metastases and 14% developed spinal metastasis. The average time to distant metastasis from the first surgery was 97~107 months in some series.[1,13,14] In meningeal HPCs, metastasis adversely affected survival, and was followed by death at an average of 24 months after its diagnosis.[13] In our both cases, distant metastasis has not been seen, yet.

Meningeal HPCs have a propensity for local recurrence. Therefore every attempt should be made to achieve complete removal of the tumor at the time of initial operation. The effect of the extent of surgery on local control is statistically significant. The 5-year local control rates for patients treated with complete removal and incomplete removal of tumor were 84% and 38% (p=0.0003) in the study of Soyuer et al., 73% and 21% (p=0.0060) in the study of Kim et al., respectively.[1,9] The effect of the extent of surgery on overall survival is statistically significant in the study of Guthrie et al.[13] (109 vs 65 months) and not statistically significant in the study of Soyuer et al.[1]

The role of radiation therapy could be divided into two parts, preoperative radiation therapy and postoperative radiation therapy.

Studies about preoperative radiation therapy is rare. Sometimes the size, location, and vascularity of the tumors make the surgical resection difficult. The first surgical attempts for

large tumors were sometimes discontinued due to profuse bleeding. If swelling occurs in HPC during surgery, tumor resection would be stopped immediately and intracranial pressure should be controlled. To change unresectable tumor into resectable one, the preoperative embolization and/or preoperative radiation therapy can be used according to the condition of tumor blood vessels. Preoperative tumor embolization with particles or glue appears to be less effective in the large meningeal tumors because of a complexity of the feeding vessels and has not widely been practiced.[11,15,16] In our Case 1, the embolization was not completely performed due to the communication between feeding vessel and ophthalmic artery.

Preoperative radiation therapy is useful in the treatment of HPC with considerable surgical risk.[11] The residual tumor can be easily removed without complications after preoperative radiation therapy. In the study of Uemura et al.,[11] seven cases of intracranial HPCs were retrospectively investigated about the efficacy of radiation therapy. Tumor response evaluated by CT and MRI was obviously seen after 20~30 Gy irradiation. The total reduction rate in Uemura's study was 80~90% and continued as long as 5~7 months after treatment. Five patients received preoperative radiation therapy before radical removal, and tumors were easily removed without massive hemorrhage. Histological inspection of specimens after irradiation showed a significant disappearance of tumor cells. Pyknosis frequently occurred in endothelial cells, and proliferating vessels with hyalinoid degeneration were also seen. Reticulin fibers between tumor cells were fewer, split, or absent. In our Case 1, the tumor was also totally removed at the third operation time after preoperative radiation therapy.

Studies about postoperative radiation therapy has been reported much more than preoperative radiation therapy in meningeal HPC. Recent data suggest that standard care would be to recommend postoperative radiation therapy for residual tumor and even in the setting of complete surgical resection.[1-3,9,12,14,17,18]

In the study of Bastin and Mehta,[12] data were gathered from their own tumor registry and compiled with an extensive analysis of published series and case reports. There was enough information about the original operation to allow grading of the tumor removal in 38 patients. Twenty-one cases were assigned grades 1 or 2 (no macroscopic residual), and 17 were assigned grades 3 to 5 (gross residual). That analysis revealed a 90% 9-year actuarial risk for local recurrence following surgical resection only. Interestingly, less than 33% of these recurrences were noted within the five years, which may account for false assumption that these tumors are highly curable with surgical resection only. Radiation therapy appeared to reduce this local recurrence rate (90% vs 48%), prolonging disease-free survival and overall survival (mean survival, 65 months vs 96 months). Radiation responses were dose-dependent, with over 50 Gy providing superior long-term disease-free survival.

The overall survival rate as well as the local control rates were much higher when patients received planned postoperative radiation therapy. Guthrie[13] reported that 9 of 17 irradiated patients relapsed within a median time of 58 months, but 13 of the 15 non-irradiated patients relapsed with a median of only 29 months. Radiation therapy after the first operation extended the average time before first recurrence from 34 to 75 months, and extended survival from 62 to 92 months. Dufour[3] also reported that surgical removal followed by external radiation therapy reduced the risk of local recurrence. The local recurrence rate was significantly lower in postoperative radiation therapy group, 12.5% in postoperative radiation therapy group and 88% in surgery only group.

Although the postoperative radiation therapy prolongs the recurrence-free survival as compared with surgery alone, radiation therapy alone does not show significant difference in recurrence-free survival. Lee[19] reported that the extent of tumor resection and presence of metastasis were the most important factors related to long-term outcome of patients with meningeal HPC. There was also statistically significant difference of median recurrence-free survival between the completely-resected group (Simpson grade 1 or 2) and partially-resected group (Simpson grade 3 or 4 or 5), 138 months compared to 47 months, respectively (p=0.009). The median recurrence-free period after subtotal resection of tumor and postoperative radiotherapy was 47 months compared to 117 months of the patients who underwent gross total resection of tumor and did not receive radiotherapy, but the difference was due to more patients of subtotal resection in postoperative radiation therapy group.

In summary, we report two cases of meningeal HPC, preoperative radiation therapy and postoperative radiation therapy each other. The role of radiation therapy could be divided into

two parts, preoperative and postoperative radiation therapy. Preoperative radiation therapy is useful in the patients with considerable surgical risk.[11] The large tumor can be easily removed without complications after preoperative radiation therapy. Postoperative radiation therapy is recommended in subtotal resection and also even in the setting of complete surgical resection.

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## 국문초록

# 수막 혈관주위세포종 환자에서의 수술과 방사선치료

**-증례보고-**

조선대학교 의과대학 방사선종양학교실

**장 지 영·오 윤 경**

수막에 발생하는 혈관주위세포종은 경질막에 기초한 드문 종양으로서 국소재발과 함께 신경축이나 신경외 장소로 원격전이를 일으킬 수도 있다. 저자들은 수막에 발생한 혈관주위세포종 환자 2예에서 수술 전 방사선치료와 수술 후 방사선치료를 각각 경험하였기에 방사선치료의 역할에 관한 문헌고찰과 함께 브고하고자 한다. 첫 번째 증례는 41세 남자로 3개월 동안 두통과 우측 반부전마비가 있었다. 1차 수술과 2차 수술 시 종양은 거의 제거할 수 없었고 수막 혈관주위세포종으로 진단되었다. 수술 전 방사선치료를 좌측 전두두정부의 큰 잔여종양에 일 회에 1.8 Gy씩 31회에 걸쳐 총 55.8 Gy를 조사하였다. 방사선치료 후 CT검사상 종양은 현저하게 크기가 줄었고 방사선치료 후 6개월에 시행한 3차 수술 시 잔여 종양의 크기는 2×2 cm로 완전히 제거되었다. 추적 CT검사상 국소재발의 소견은 없었고, 1차 수술 후 4년 10개월 동안 원격 전이 없이 생존하고 있다. 두 번째 증례는 45세 여자로 갑작스러운 두통과 시력 장애로 수막종이 의심되어 종양제거수술을 받았는데 조직 검사상 우측 전두엽에 발생한 혈관주위세포종으로 진단되었으며 시상동을 침습한 소견을 보였으나 완전히 제거되었다. 국소재발을 줄이기 위해 수술 후 방사선치료가 의뢰되었고 방사선치료는 우측 전두엽 부위에 일 회에 1.8 Gy씩 30회에 걸쳐 총 54 Gy를 조사하였다. 수술 후 5년 동안 국소 재발이나 원격 전이 없이 정상적인 활동을 유지하면서 생존하고 있다.

**핵심용어**: 수막 혈관주위세포종, 수술, 방사선치료