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Recurrence and Extraneural Metastasis in 31 Meningeal Hemangiopericytomas

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=Abstract=

31예 수막 혈관외피세포종에 있어서의 재발 및 신경계외 전이

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Purpose : Meningeal hemangiopericytoma(M - HPC), characterized by a high local recurrency and metastatic potential, is a rare neoplasm arising from perivascular pericytes. A retrospective study was performed to identify the recurrence and extraneural metastasis in M - HPC.

Materials and Methods : We reviewed the records of 31 M - HPC patients treated from 1982 through 1999 at our institution. The time to recurrence and the various parameters affecting recurrence were determined. Extreneural metastasis was also analyzed.

Results : The rate of local recurrency was 38.7%(12/31). The overall average recurrence - free period(RFP) before the first recurrence was 104 months, with overall recurrence - free rates(RFRs) at 5 and 10 years after first surgery of 59.2% and 33.6%, respectively. Of the 12 patients who experienced local recurrence, 4 had recurrences 5 years later after the first surgery. Complete excision at the first operation significantly extended the average time before first recurrence from 43 to 111 months. The 5 - year RFRs for the groups of complete excision and incomplete excision were 72.7% and 20.8%, respectively(p=0.0060). Although there was no statistical significance, complete excision followed by adjuvant radiotherapy of more than 50Gy extended the RFP. The 5 - year RFRs for the groups of complete excision and complete excision with adjuvant radiotherapy were 70.3% and 100%, respectively(p=0.3359). Four patients(12.9%) presented one or more extraneural metastases that were developed at an average of 107 months after the first operation with the 5 - and 10 - year metastasis rates of 4.4% and 24.9%, respectively.

Conclusions : M - HPC has a propensity to recur either locally or at distant sites after surgical resection. Complete excision is the most important factor to reduce recurrence. However, even with complete excision, adjuvant radiotherapy of more than 50Gy significantly reduces the risk of recurrence. Local and distant recurrences may occur after a prolonged disease - free interval, emphasizing the need for long - term follow - up.

KEY WORDS : Meningeal Hemangiopericytoma · Recurrence · Extraneural Metastasis · Complete Excision · Radiotherapy.

Introduction

Meningeal hemangiopericytoma(M-HPC) is a rare vascular tumor which is most commonly diagnosed in the early fifth decade of life[2)4)12)13)19)]. This tumor accounts for <1% of all central nervous system(CNS) tumors and has almost equal sex incidence[2)12)]. Although M-HPC initially

was believed to be a meningioma variant(angioblastic meningioma, hemangiopericytic type), it has been recognized as a distinct pathologic entity with different clinical behavior, immunohistochemical characteristics, and ultrastructural features as compared to meningioma[4)14)16)].

In contrast to ordinary meningioma, M-HPC is more aggressive with a high propensity for both local recurrence and extraneural metastasis in spite of aggressive treatment[4)8)11)13-15)19)26)]. According to a large study of M-HPC by Guthrie, et al.[13)], the average time before the first recurrence was 47 months, the average time to extraneural metastasis was 99 months, and the average survival period was 84 months. However, assessment of its current prognosis is difficult as the series described in the past literatures cover a long time span. We reviewed our personal experience to identify the current patient status, recurrence and extraneural metastasis in M-HPC.

Materials and Methods

During the years 1982 through 1999, 31 patients with primary M-HPC were treated at our institution. M-HPC constituted 2.4% of all intracranial meningiomas during the same period(31 M-HPCs : 1309 intracranial meningiomas). Data were collected from review of the clinical records and neuroradiological investigations, and histological slides were reexamined by an experienced neuropathologist. Information on the postoperative course was obtained from records of the outpatient clinic, phone contacts and whenever possible, new examinations. There existed a male predominance (22 males : 9 females), and the age at first operation ranged from 18 to 64 years with an average of 41 years. The mean duration of preoperative symptoms was 11 months, with the most frequent presenting symptom of headache. Nine cases were located in parasagittal, 7 in falx, 7 in tentorium, 3 in sphenoid ridge, 2 in falcotentorium, 3 in other sites. All patients received surgery : complete excision (Simpson grade Ⅰ+Ⅱ) was possible in 24 patients and remaining 7 patients underwent incomplete excision(Simpson grade Ⅲ+Ⅳ+Ⅴ). Among 24 patients who got complete excision, 5 patients received conventional external beam radiotherapy as a prophylactic measure after the first operation. We performed postoperative radiotherapy in 6 patients who underwent incomplete excision for a control of residual tumor after the first operation. Among these 6 patients, gamma knife radiosurgery(GKR) instead of conventional external beam radiotherapy was applied to 2 small residual tumors. None was treated with chemotherapy. Age, sex, size, location, extent of resection, and use of radiotherapy in the cases of complete excision were entered to test their impacts on recurrence. Survival and recurrence were calculated using Kaplan-Meier method, and the probability of recurrence rate of different groups was evaluated with log-rank test. The results were considered significant at $p<0.05$.

Results

Patient characteristics, including extent of resection, radiotherapy, recurrence, time to first recurrence as well as last follow-up status, are summarized in Table 1.

The average follow-up period was 77 months(follow-up range : 1 - 216 months). Six out of 31 patients died during the follow-up period(of the six patients, 2 died of unrelated disease), with the 5- and 10-year survival rates after fist surgery of 96.3% and 75.7%, respectively. The rate of local recurrency was 38.7%(12/31). The overall average recurrence-free period(RFP) before the first recurrence was 104 months, with overall recurrence-free rates(RFRs) at 5 and 10 years after first surgery of 59.2% and 33.6%, respectively(Fig. 1). Of the 12 patients who experienced local disease recurrence, 4 had recurrences 5 years later after the first surgery. Complete excision at the first operation significantly extended the average time before first recurrence from 43 to 111 months. The 5-year RFRs for the groups of complete excision and incomplete excision were 72.7% and 20.8%, respectively(p=0.0060)(Fig. 2). Although there was no statistically significant, complete excision followed by adjuvant radiotherapy of more than 50Gy extended the RFP. The 5-year RFRs for the groups of complete excision and complete excision with adjuvant radiotherapy were 70.3% and 100%, respectively(p=0.3359)(Fig. 3). One patient(case 2) underwent only Simpson grade V resection at first operation because of massive intraoperative bleeding, and received immediate postoperative conventional radiotherapy for the residual tumor. This case was responded dramatically to radiotherapy and tumor was markedly decreased in size after radiotherapy(Fig 4). When we performed second operation for the treatment of recurrence, tumor was easily removed without massive intraoperative bleeding. Six patients(19.4%) received stereotactic radiosurgery for the treatment of the residual or recurrent tumors. Of these 6

Table 1. Patient characteristics in 31 meningeal hemangiopericytomas

Case No.	Age/ Sex	Location	Size* (mm)	Resection**	RT	Recurrence	Time to first recurrence (mos.)	Survival	Last F/U (mos.)	Last F/U status	Comments
1	31/M	Tentorium	90	I	-	-		A	109	NED at primary site	
2	29/M	Sphenoid ridge	60	V	+	+	66	A	106	Local stable state at primary site	2nd operation(+), GKR (+) after recurrence
3	32/M	Falx	50	III	+	+	43	D	92	Expired due to extraneural metastasis	2nd operation(+), extra-neural metastasis to liver (at 92 mos.)
4	42/M	Parasagittal	55	II	-	+	111	D	116	Expired due to local extension	
5	44/M	Tentorium	25	III	-	+	41	A	175	NED at primary site	Reoperation(+) for 3 times, conventional RT(+) after recurrence
6	27/F	Falx	60	I	-	-		A	81	NED at primary site	
7	33/M	Parasagittal	30	IV	+	+	43	A	67	Local stable state at primary site	2nd operation(+), LINAC(+) after recurrence
8	23/F	Tentorium	30	II	+	-		A	129	NED at primary site	Extraneural metastasis to femur(at 112 mos.)
9	41/M	Parasagittal	80	I	-	+	104	A	194	Local stable state at primary site	2nd operation(+), conventional RT(+) after recurrence, extraneural metastasis to lung(at 192 mos.)
10	29/M	Falx	55	III	+***	-		A	23	Local stable state at priamry site	
11	30/M	Tentorium	90	I	-	+	182	A	216	NED at primary site	2nd operation(+), conventional RT(+) after recurrence
12	61/M	Parasagittal	50	II	+	-		A	32	NED at primary site	
13	58/M	Convexity	70	I	-	-		D	1	Expired due to postoperative hepatitis	
14	55/F	Falx	90	I	-	-		A	94	NED at primary site	
15	40/F	Tentorium	80	I	-	-		A	163	NED at primary site	
16	51/F	Falx	60	I	+	-		A	14	NED at primary site	
17	45/M	Falx	50	II	-	-		A	95	NED at primary site	
18	18/M	Parasagittal	80	II	-	-		A	32	NED at primary site	
19	33/F	Sphenoid ridge	30	I	-	+	38	A	82	Local extension state at primary site	2nd operation(+), conventional RT(+) after recurrence
20	50/F	Sphenoid ridge	30	II	-	+	31	D	158	Expired due to local extension	Reoperation(+) for 6 times, conventional RT and LIN-AC(+) after recurrence
21	55/M	Parasagittal	40	III	+	+	24	A	84	Local extension state at primary site	Reoperation(+) for 3 times, GKR(+) after recurrence
22	64/M	Falx	40	I	-	+	45	A	76	NED at primary site	2nd operation(+)
23	47/M	Parasagittal	70	II	-	-		A	70	NED at primary site	
24	37/M	Multiple	50	I	-	+	39	A	63	Local extension st-ate at primary site	2nd operation(+)
25	55/M	Parasagittal	40	II	-	-		D	2	Expired due to unrelated causes (acute renal failure)	

Table 1. Continued

Case No.	Age/ Sex	Location	Size* (mm)	Resection**	RT	Recurrence	Time to first recurrence (mos.)	Survival	Last F/U (mos.)	Last F/U status	Comments
26	45/M	Lateral ventricle	15	I	-	-		D	10	Expired due to leptomeningeal seeding	Conventional RT(+) after leptomeningeal seeding
27	55/F	Parasagittal	70	I	-	-		A	34	NED at primary site	
28	32/M	Tentorium	60	I	-	-		A	31	NED at primary site	Extraneural metastasis to right 8th rib and T10 dorsal mass(at 31 mos.)
29	44/M	Tentorium	60	III	+***	-		A	27	Local stable state at primary site	
30	27/M	Falcotentorium	30	I	+	-		A	7	NED at primary site	
31	36/F	Falcotentorium	60	I	+	-		A	7	NED at primary site	

RT : radiotherapy, GKR : gamma knife radiosurgery, LINAC : linear accelerator based radiosurgery, F/U : follow-up, mos. : months, NED : no evidence disease, A : alive, D : dead, Size(mm) : longest diameter of tumor ; Resection** : Simpson grade I-V, +*** : gamma knife radiosurgery instead of conventional beam radiotherpay after the first operation for the residual tumor

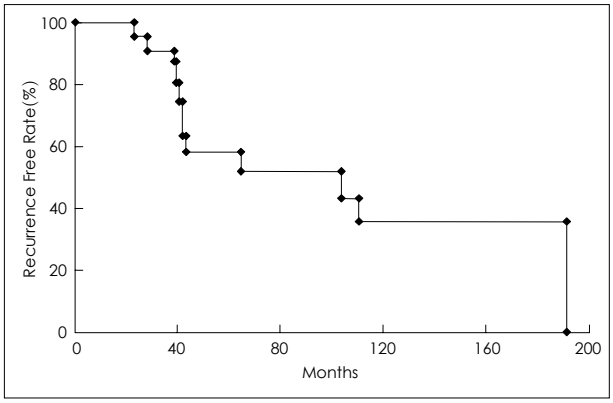

Fig. 1. Kaplan-Meier plot : overall recurrence free rates in 31 meningeal hemangiopericytoma patients

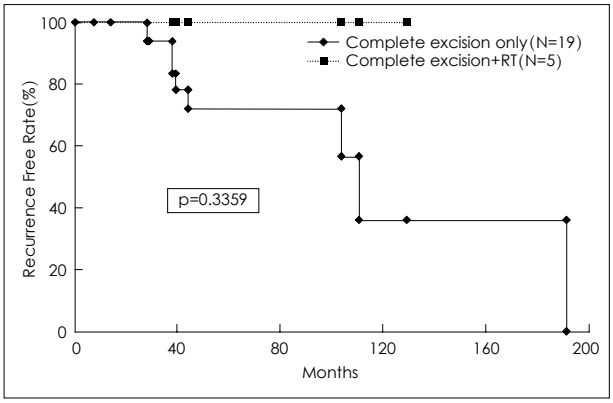

Fig. 3. Kaplan-Meier plot : recurrence free rates according to radiotherapy in complete excision group

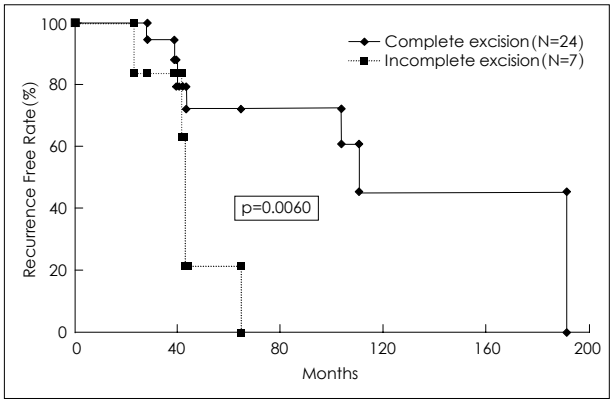

Fig. 2. Kaplan-Meier plot : recurrence free rates according to surgical resection grade

patients, 2 previously non-irradiated residual tumors which were treated with GKR remained stable. Among 4 patients who already were treated with conventional external beam radiotherapy and treated again with stereotactic radiosurgery (GKR : 2, linear accelerator based radiosurgery : 2) when tumor recurred, 2 patients achieved a stable response, but, 1 patient was in local extension state and 1 patient died of disease progression. Age, sex, size, and location of the primary tumor were not associated with recurrence in this population of patients with M-HPC. For the CNS recurrences, all except 1 case occurred at the primary tumor site. None had recurrences both at the initial and distant CNS sites. One patient(case 26) had diffuse leptomeningeal spread without recurrence at the initial CNS location. He received radiotherapy after leptomeningeal seeding(at 5 months later after fist operation), and died of disease progression. Four patients(12.9%) presented one or more extraneural metastases(lung, liver, femur, and rib/T10). Extraneural metastases were developed at an average of 107 months after the first operation with the 5- and 10-year metastasis rates of 4.4% and 24.9%, respectively(Fig. 5).

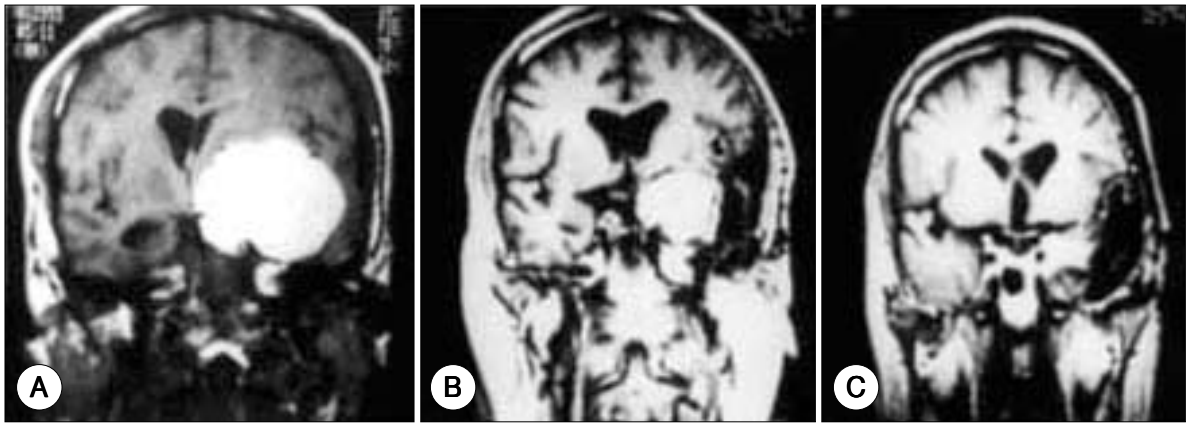

Fig. 4. Illustrative case(case 2) ; A : Initial T1-weighted coronal MRI scan with gadolinium-enhancement shows uniformly enhanced large tumor on left sphenoid ridge. This patient underwent only Simpson grade V resection at first operation and received immediate postoperative conventional radiotherapy for the residual tumor. B : Follow-up gadolinium-enhanced MRI obtained 4 months after radiotherapy demonstrates shrinkage of the tumor. C : Image obtained 24 months after radiotherapy shows a marked decrease in tumor size.

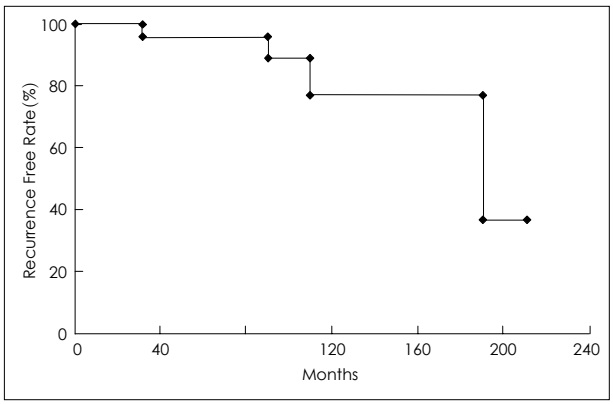

Fig. 5. Kaplan Meier plot : overall distant metastasis free rates in 31 meningeal hemangiopericytoma patients

Discussion

M-HPC was first described by Begg and Garret[3] who noted its similarity to angioblastic meningioma. Since that time, controversy has continued as to whether M-HPC should be classified as a variant of true meningioma or as a central form of peripheral HPC[16]. Histologic examinations, including ultrastructural studies, demonstrate the M-HPC tumor cells to be derived from a pericyte around the capillary wall. Further, recent immunohistochemical and genetic studies also have shown that M-HPC is distinct from meningioma and identical to peripheral HPC[4)14)16)].

Preoperative identification of M-HPC is important because of its aggressive nature that consistently has been correlated with decreased life expectancy and shortened recurrence-free survival comparing to meningioma[2)7-9)11-15)19)20)23)25)26)]. Several long-term studies have documented the propensity of M-HPC to recur either locally or regionally within the cranial or spinal meninges[2)4)7-9)11-14)19)22)]. This is also aggressive lesion that tends to metastasize extracranially, predominantly to bone, lung, kidney, and pancreas[1)2)4)6)7)9)11-19)21)].

As mentioned above, recurrent M-HPC represents a major problem[2)4)7-9)11-13)19)22)]. Dufour, et al.[9)], reported a 45% local recurrence rate, similar to 38.7% of our series. But, another published incidences of local recurrency for M-HPC were much higher and around 80%[2)4)12)22)]. Bastin, et al.[2)], showed a 90% risk for local recurrence by 9 years in a retrospective analysis of 19 patients with M-HPC treated surgically only. Goellner, et al.[12)], reported an 80% local recurrence rate for M-HPC, similar to 71% reported by Pitkethly, et al.[22)], These high local recurrence rates exceed those of meningioma, reported at between 20% for complete removal to 74% for subtotal removal[1)26)]. For the CNS recurrences, the majority occurs at the initial tumor site. Galanis, et al.[11)], noted that, of the 32 patients, 19(59%) occurred at the primary tumor site. Another 11(34%) had recurrences both at the initial and distant CNS sites, whereas the remaining 2(7%) had diffuse leptomeningeal spread without recurrence at the initial CNS location. In our series, all except 1 case(91.7%) occurred at the primary tumor site for the CNS recurrences. None had recurrences both at the initial and distant CNS sites, and 1 patient had diffuse leptomeningeal spread without recurrence at the initial CNS location. Many authors reported that recurrence is a late event[2)4)12)15)]. Bastin, et al.[2)], found that less than 33% of

their recurrent cases were noted within the first five years. Brunori, et al.[4], reported that the mean interval to first recurrence was 84 months, which matches well with the data of Jaaskelainen, et al.[15], Guthrie, et al.[13], showed the recurrence rate at 10 years after surgery was 76%, and Goellner, et al.[12], also noted that the recurrence rate at 15 years was 76%. In our series, the overall average RFP before the first recurrence was 104 months, with overall RFR at 10 years after first surgery of 33.6% Of the 12 patients who experienced local disease recurrence, 4 had recurrences 5 years later after the first surgery. We consider that, since recurrence is a late event in the natural history of M-HPC, close follow-up for longer period after the first operation must be needed.

Considering our review and the current literature, it seems that complete excision favorably affected recurrence and survival, as opposed to incomplete excision[2)9)13)14)15)18)]. Jaaskelainen, et al.[15], noted that 5 of the 18 postoperative survivors(28%) suffered recurrence at an average of 78 months. Among those surviving patients who had incomplete resections, an average time to recurrence was 68 months. Galanis, et al.[11], and Guthrie, et al.[13], emphasized the importance of complete removal at the first operation to prolong the time to recurrence and extend survival. They also insisted that aggressive surgical management is very important for the successful treatment of patients with even recurrent M-HPC. In our series, complete excision at the first operation significantly extended the average time before first recurrence from 43 to 111 months, and the 5-year RFRs for the groups of complete excision and incomplete excision were 72.7% and 20.8%, respectively. We agree that complete excision is a main stay of treatment in M-HPC.

Every effort should be made to eradicate the primary tumor, and many authors showed that prophylactic postoperative radiotherapy appears to reduce the local recurrence rate and prolong disease-free and overall survival[2)4)7)8)13)14)18-20)23)25)]. Guthrie, et al.[13], recommended postoperative radiotherapy before the first recurrence even after an apparently complete tumor resection. Nine of the 17 irradiated M-HPCs recurred in a median of 58 months while 13 of the 15 non-irradiated M-HPCs recurred in a median of 29 months. Radiotherapy after the first operation extended the average time before first recurrence from 34 to 75 months, and extended survival from 62 to 92 months in long-term follow-up of 44 cases study. Staples, et al.[23], showed that for all totally excised lesions, the 5-year disease-free survival was improved from 28% for surgery alone to 57% with adjuvant radiotherapy. Galanis, et al.[11], and Brunori, et al.[4], also reported that radiotherapy did not show any significant effect in the control of tumor regrowth after the second operation, and insisted that postoperative radiotherapy should be given even if removal is completed in the primary M-HPC at the first operation. Complete excision followed by adjuvant radiotherapy of more than 50Gy extended the RFP in our series. We think that because of the high rate of local recurrence after surgical excision, treatment strategies combining local excision of primary tumors with wide-field, high-dose radiotherapy are worthy of trial.

Some authors recommended preoperative radiotherapy in the management of M-HPC involving considerable surgical risk[5)10)25)]. Carella, et al.[5], noted that the " hemangiopericytic variant of angioblastic meningiomas " responded more favorably to radiation than common meningioma. Uemura, et al.[25], also showed that, in five M-HPC patients receiving radiotherapy before radical removal, the 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. We had the same experience in one patient of M-HPC who underwent only Simpson grade V resection at first operation because of massive intraoperative bleeding. He received immediate postoperative conventional radiotherapy for the residual tumor, and this case was responded dramatically to radiotherapy. When we performed second operation for the treatment of recurrence, tumor was easily removed without massive bleeding. We consider that preoperative radiotherapy is useful in the treatment of M-HPC involving considerable surgical risk.

Radiation response is dose-dependent, and radiotherapy, in doses of >50Gy should be incorporated as primary or adjuvant therapy for every M-HPC patients treated with curative intent[2)8)9)13)20)23)]. Dufour, et al.[9], and Bastin, et al.[2], showed that adjuvant radiotherapy of more than 50Gy significantly reduced the risk of recurrence and provided superior long-term disease-free survival in M-HPC. Guthrie, et al.[13], also proposed that a radiation dose-response relationship existed, having noted no local recurrences among patients receiving >51Gy. Of 8 patients receiving low-dose radiation(<45Gy), 7(88%) experienced local recurrence,

as opposed to 2 of 9(22%) in the high-dose group(>45 Gy). None of the 3 patients who received doses above 51Gy had experienced a recurrence.

Stereotactic radiosurgery may prove effective in treating some well-defined M-HPC[2)4)7)9)11)]. Dufour, et al.[9)], insisted that radiosurgery is indicated for recurrent tumors measuring less than 30mm in their greatest diameter. Bastin, et al.[2)], used stereotactic radiosurgery for recurrent M-HPC, and attained a dramatic tumor response following delivered in a single fraction. In Galanis, et al's., study[11)], radiosurgical techniques permitted the delivery of an additional high dose of focal irradiation to 17 tumors(in 7 patients) that had appeared, recurred, or persisted despite the previous administration of 50 - 60Gy of conventional fractionated radiotherapy. Despite the relatively large size of the treated lesions in their series(median, 32mm ; 35% of the lesions >40mm), all lesions responded to radiosurgical treatment, although the majority of responses lasted <1 year. Conversely, when steretactic radiosurgery was applied to small (<25mm) tumors in previously non-irradiated fields(3 patients), all patients achieved complete responses, and remained disease free at a median follow up time of 3 years from treatment. Generally, the early shrinkage of M-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 M-HPC[2)]. Six(19.4%) out of 31 M-HPC patients received stereotactic radiosurgery for the treatment of the residual or recurrent tumors in our series. Of these 6 patients, 2 previously non-irradiated residual tumors which were treated with GKR remained stable. Among 4 patients who already were treated with conventional radiotherapy and treated again with stereotactic radiosurgery(GKR : 2, linear accelerator based radiosurgery : 2) when tumor recurred, 2 patients achieved a stable response, but, 1 patient was in local extension state and 1 patient died of disease progression. We consider that, although prior data need further confirmation, radiosurgery seems an attractive option, especially in patients who have already undergone radiotherapy. But, the unpredictable appearance of new lesions distant from the original surgical site or radiosurgical treatment field has remained a vexing problem. Also, the tendency of M-HPC to recur just outside previous surgical or radiosurgical margins(sometimes after a delay of several years) has continued to limit the long-term effectiveness of any locally applied treatment modality for this neoplasm.

M-HPC has a propensity to recur at distant sites within the nervous system after surgical resection. Extraneural metastases also develop much more commonly in this lesion than in other types of histologically benign meningioma[1)2)4)6)7)9)11-19)21)]. Metastases may be multiple, to many organs and systems, including bone, lung, kidney, pancreas, adrenal gland, or liver, and rarely breast, thyroid, or lymph nodes. Galanis, et al.[11)], noted that bone and liver were the most common metastatic sites(82% and 41% of extraneural recurrences, respectively), and Brunori, et al.[14)], reported that the preferential sites were lung, bone, soft tissue and liver. Guthrie, et al.[13)], also showed that lung and bone were the most common metastatic sites. The published incidence of extraneural metastases for M-HPC ranges from 12% by Stout, et al.[24)],(similar to 12.9% of our series) to 57% by Adegbite, et al.[1)], One of the major factors accounting for this wide range is the varying time intervals from diagnosis when the given analysis is performed[4)13)18)19)]. The probability increases steadily with time, so long-term follow-up of this tumor is necessary[13)18)19)]. Guthrie, et al.[13)], demonstrated increasing metastases frequently from time of treatment, reporting a 5, 10, and 15 years probability of developing metastasis of 13%, 33%, and 64%, respectively (the average time to extraneural metastasis after the first operation : 99 months). Koyama, et al.[18)], found that metastatic tumors appear a mean of 8 years after initial therapy and have been discovered up to 16 years thereafter. Brunori, et al.[4)], also showed that, in one M-HPC patient who survived 15 years after the first operation, disseminated visceral metastases(lungs, kidney, liver, inguinal area) were detected at autopsy. In the present series, extraneural metastases were developed at an average of 107 months after the first operation with the 5- and 10-year metastasis rates of 4.4% and 24.9%, respectively. We consider that close long-term follow-up after the first operation is needed, and investigations should include complete physical examination, chest X-rays, abdominal ultrasound and total body CT, when indicated.

Conclusion

M-HPC is more aggressive, and tends to recur even after gross total resection. Complete excision followed by adjuvant radiotherapy of more than 50Gy significantly reduces the risk of recurrence. Stereotactic radiosurgery is of value

for treatment of CNS recurrences of smaller size(volume), even in previously irradiated fields or after multiple resections. Late extraneural metastases after a prolonged disease-free interval are common in the course of M-HPC, emphasizing the need for long-term follow-up. M-HPC still has a dismal prognosis, and more effective chemotherapy agents or combinations are clearly needed.

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31예 수막 혈관외피세포종에 있어서의 재발 및 신경계외 전이

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= 국문초록 =

목 적 : 높은 재발율과 전이 가능성의 특징을 갖고 있는 수막 혈관외세포종은 perivascular pericytes에서 기원하는 드문 종양으로, 저자들이 경험한 수막 혈관외세포종에 있어서 종양의 재발 및 신경계외 전이에 대하여 알아보고자 한다.

연구 대상 및 방법 : 1982년부터 1999년까지 수술을 시행하여 수막 혈관외세포종으로 진단받은 31명의 환자를 대상으로 후향적 연구를 시행하였다. 진단 후 재발까지의 기간, 재발에 영향을 미칠 수 있는 여러 가지 인자들, 그리고 신경계외 전이의 기간 및 부위 등을 분석하였다.

결 과 : 총 31명의 환자들 중 12명의 환자들에서 원발성 부위에 종양이 재발하였다(38.7%). 전체의 환자들에서 재발이 발생하지 않았던 평균 기간(recurrence - free period : RFP)은 104개월이었으며 처음 수술 후 5년, 그리고 10년의 재발이 발생하지 않았던 율(recurrence free period : RFP)은 각각 59.2%, 33.6% 이었다. 원발성 부위에 종양의 재발을 보였던 12명의 환자들 중 4명의 환자들에서 첫 수술 후 5년 이후에 재발을 관찰 할 수 있었다. 처음 수술시의 절제 정도가 재발에 가장 큰 영향을 미쳤다. 처음 수술시에 완전 절제한 군의 RFP는 111개월이었으며, 추적 관찰 5년째 RFR는 완전 절제한 경우 72.7%, 불완전 절제한 경우 20.8%로 그 차이는 통계적으로 유의하였다(p=0.0060). 통계적으로 유의성은 없었지만 완전 절제 후 부가적인 방사선 치료를 시행하였을 경우가 완전 절제만을 시행한 경우보다 RFR가 높아 추적 관찰 5년째의 RFR는 각각 100%, 그리고 70.3% 이었다(p=0.3359). 4명의 환자들(12.9%)에서 수막 혈관외세포종의 신경계외 전이가 발생하였으며, 이 경우 신경계외 전이의 평균 기간은 107개월, 그리고 추적 관찰 5년, 10년째의 신경계외 전이율은 각각 4.4%, 24.9% 이었다.

결 론 : 수막 혈관외세포종은 수술적 제거 후 원발성 부위에 또는 원발성 부위에서 멀리 떨어진 부위에 재발의 가능성이 무척 높은 종양이다. 이러한 재발의 가능성을 줄이는 가장 중요한 인자는 수술적 제거 정도로 처음 수술시에 가능한 완전 절제를 시도하여야 한다. 그리고 처음 수술시에 완전 절제가 가능하였다고 하여도 완전 절제 후 부가적인 방사선 치료를 시행하는 것이 재발의 위험성을 낮출 수 있을 것으로 생각한다. 원발성 부위의 재발 또는 신경계외 전이는 오랜 시간이 지난 후에도 가능할 수 있으므로 장기간의 추적 관찰이 필요하다고 생각한다.

중심 단어 : 수막 혈관외피세포종 · 재발 · 신경계외 전이 · 완전 절제 · 방사선 치료.