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Case Report

Intracavernous Hemangiopericytoma: Case Report and Review of the Literature

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Intracavernous hemangiopericytoma/solitary fibrous tumor is an extremely rare tumor, with only seven cases reported. We present a case of intracavernous hemangiopericytoma/solitary fibrous tumor and review all cases reported in the literature. A 67-year-old man experienced numbness over the left half of the face. Magnetic resonance imaging revealed a left intracavernous tumor extending into Meckel's cave and the posterior fossa. We performed gamma knife surgery (GKS) which a prescribed dose to the tumor of 12 Gy, but tumor recurred 43 months after GKS. We performed partial tumor resection via a subtemporal interdural approach. The pathological diagnosis was hemangiopericytoma. Postoperatively, we performed second GKS with a prescribed dose of 15 Gy. Diplopia and ptosis improved markedly and the tumor initially reduced in size, but tumor regrowth was seen again 29 months after second GKS. Third GKS was performed with a prescribed dose of 15 Gy. Recurrence was not seen at 18 months after third GKS, but was identified about 2 years after third GKS. We performed fourth GKS with a prescribed dose to the residual tumor of 16 Gy. We report a rare case of intracavernous hemangiopericytoma originating in the cavernous sinus, but distinguishing between hemangiopericytoma and schwannoma is difficult for round, intracavernous tumors showing homogeneous enhancement without flow voids. GKS might be one of the options for residual and recurrent intracavernous hemangiopericytomas.

Keywords: cavernous sinus, hemangiopericytoma, gamma knife surgery

Introduction

Hemangiopericytoma is a rare tumor, making up 1.6–4% of all dural-based tumors and <1% of all intracranial tumors.[1–12] Hemangiopericytomas can occur elsewhere intracranially, but mostly arise supratentorially.[2,10] We report a rare case of intracavernous hemangiopericytoma originating in the cavernous sinus.

Case Report

A 67-year-old man presented with an 11-month history of numbness over the left half of the face. Neurological examination showed hypalgia in the territories of all branches of the left trigeminal nerve. Magnetic resonance imaging (MRI) revealed a left intracavernous tumor extending into Meckel's cave and the posterior fossa (Fig. 1a). We diagnosed the tumor as trigeminal schwannoma and performed gamma knife surgery (GKS) with a prescribed dose to the tumor of 12 Gy. After GKS, symptoms improved and the tumor initially reduced in size. However, 42 months after GKS, he again presented with numbness over the left half of the face. After another month, drink spilled from the left corner of the mouth, and he presented with ptosis of the left eyelid and diplopia. Neurological examination revealed paralysis of the oculomotor, trigeminal, abducens and facial nerves on the left side. Imaging revealed tumor regrowth. The tumor showed slight hyperdensity on computed tomography (CT), and isointensity to slight hyperintensity on T_1- and T_2-weighted imaging. The lesion enhanced homogeneously on gadolinium-enhanced T_1-weighted imaging. Bone-density CT revealed erosion of the left petrous apex and CT angiography showed no apparent feeding vessels (Figs. 1b–1f). Tumor resection was performed via a left subtemporal interdural approach in December 2012, but the tumor bled easily, so the surgery resulted in partial resection of the tumor (Figs. 1g and 1h). Histological examination revealed the tumor cells had a round to spindle-shaped cytomorphology with stag-horn vessels (Fig. 2a). Some nuclei showed mitosis, but no apoptosis was apparent. Tumor cells expressed CD34 and were negative for S-100, glial fibrillary acidic protein and epithelial membrane antigen. STAT6 was expressed in the nuclei (Fig. 2b). We diagnosed the tumor as hemangiopericytoma. Mib-1 labeling index was 4.0%. Whole-body fluorodeoxyglucose-positron emission tomography after surgery revealed no other distant metastases. About 16 days postoperatively, we performed second GKS with a prescribed dose to the residual tumor of 15 Gy. About 8 months after second GKS, diplopia and ptosis were markedly improved, and the tumor reduced in size. About 23 months after second GKS, diplopia completely disappeared, and MRI showed no tumor recurrence (Figs. 3a and 3b). However, 29 months after second GKS, tumor regrowth was again identified. However, the patient showed only numbness over the left half of the face. We performed third GKS with a prescribed dose to the residual tumor of 15 Gy (Fig. 3c). After third GKS, numbness improved. About 15 months after third GKS, no recurrence was evident (Fig. 3d). However, about 2 years after third GKS, tumor regrowth was again identified. We performed fourth GKS, for which the prescribed

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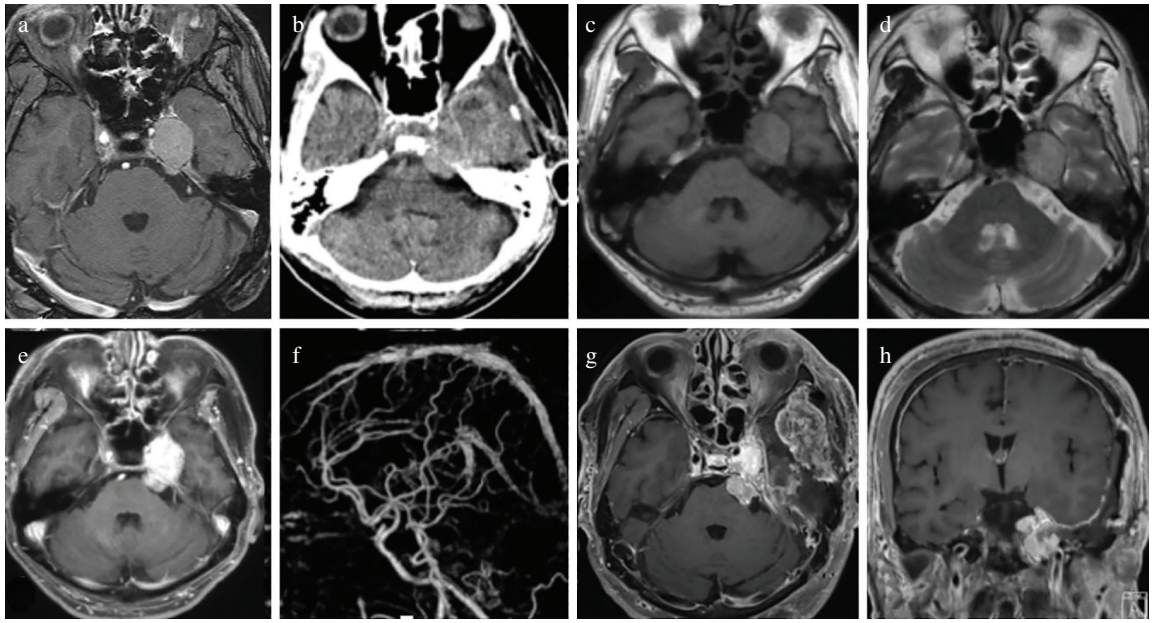

Fig. 1 (a) Gadolinium-enhanced T_1-weighted magnetic resonance imaging at the first GKS. The tumor located a left intracavernous tumor extending into the Meckel's cave and posterior fossa. The tumor volume was 6.9 cm^3. (b–f) Preoperative images. The tumor showed slightly high density on CT (b) and slightly high intensity on T_1 (c) and T_2-weighted image (d). The lesion enhanced homogenously on gadolinium-enhanced T_1-weighted image (e), and no apparent feeding vessels (f). The tumor volume was 12 cm^3 before the surgery. (g and h) Gadolinium-enhanced T_1-weighted magnetic resonance imaging after the surgery. The tumor was resected partially, and the volume was 8.5 cm^3. GKS, gamma knife surgery.

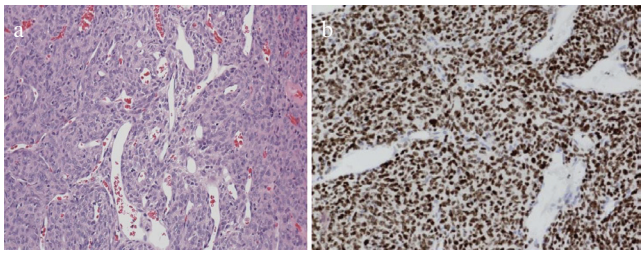

Fig. 2 Histological examination showed densely cellular neoplasm with stag-horn blood vessels (a). STAT6 was expressed in the nuclei (b).

dose to the residual tumor was 16 Gy (Fig. 3e). After fourth GKS, the patient reported no symptoms except numbness over the left half of the face.

Discussion

According to the latest WHO classification of tumors of the central nervous system, mesenchymal tumors of the fibroblastic type encompass a histological spectrum of tumors previously classified separately as hemangiopericytoma and meningeal solitary fibrous tumor.[13] Intracavernous hemangiopericytoma/solitary fibrous tumor seems extremely rare, with only seven cases reported in the literature, excluding the present case (Table 1).[1,2,14–17]

Features of intracavernous hemangiopericytoma/solitary fibrous tumor

From the previous seven cases and our case of intracavernous hemangiopericytoma/solitary fibrous tumor, some might show typical features of an intracranial hemangiopericytoma, such as multilobulate aspect and/or have prominent vessel voids.[1,2,14–17]

However, intracavernous round tumor enhancing homogeneously without flow voids, as in the present case, can be difficult to diagnose as hemangiopericytoma. Among the seven reported cases of intracavernous hemangiopericytoma/solitary fibrous tumor, four patients presented with facial numbness/pain or diplopia, and three of the six patients presented with headache or ptosis.

Surgical resection of cavernous sinus tumors

To resect around the cavernous sinus tumors, some authors have reported the usefulness of anterior transpetrosal approach,[14] endonasal endoscopic surgery[16] or radical removal with a high-flow bypass.[17] On the other hand, because complete resection of tumor in the cavernous sinus without deteriorating cranial nerve functions is difficult, some authors prefer to undergo less invasive treatment such as radiation therapy. Lee et al. reported the usefulness of stereotactic radiosurgery for cavernous sinus meningioma. They underwent primary radiosurgery for >50% of their patients with cavernous sinus tumors presumed to be meningiomas according to the neurodiagnostic criteria alone.[18]

Radiation treatment for hemangiopericytoma

Hemangiopericytoma tends to develop local recurrence as well as distant metastasis even after gross total resection (GTR).[3,4,6,7,9,11,19] Some reports have shown the utility of near-total resection and postoperative external beam

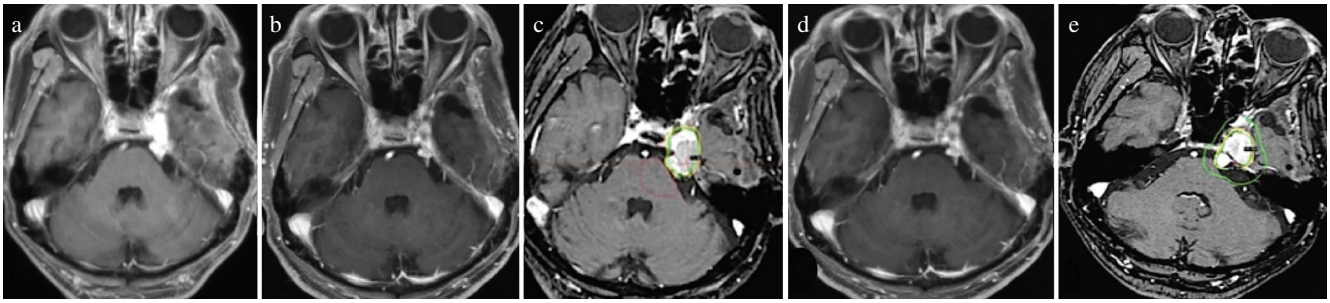

Fig. 3 Gadolinium-enhanced T_1-weighted magnetic resonance imaging after the surgery. 8 months (a) and 23 months (b) after second gamma knife surgery (GKS), the tumor reduced in size. 29 months after second GKS, we underwent third GKS which the prescription dose for the tumor was 15 Gy (c). 18 months after third GKS, the tumor reduced in size once (d). 24 months after third GKS, we underwent fourth GKS which the prescription dose for the tumor was 16 Gy (e).

Table 1 Literature review of eight cases of intracavernous hemangiopericytoma

Series	Age/Sex	Presenting symptoms	Imaging features	Operation type	Patient outcome
Bonde et al.[1]					
Case1	35/M	Numbness of face Difficulty in chewing Diplopia	Homogenous enhancement without flow voids	Extradural approach	Recurrence (10 years after surgery) →RT
Case2	57/F	Headache Numbness of face	Homogenous enhancement with flow voids	Basal temporal extradural approach	Recurrence (3 years after surgery) →→RT
Ganesan et al.[2]	56/F	Headache Loss of interest Recent memory loss	Multilobulated solid-cystic mass	Temporal craniotomy with near total resection	RT for residual tumor
Muto et al.[14]	51/M	Facial pain Diplopia, Ptosis	Homogenous enhancement with flow voids	Embolization before tumor resection via anterior petrosal approach	Sterotactic radiosurgery for residual tumor
Agarwal et al.[15]	11/M	Headache Diplopia, Ptosis Hemiparesis	Multilobulated mass with flow voids	Temporobasal craniotomy	Death due to septicemia (2 days after surgery)
Patrona et al.[16]	NA	Oculomotor, trocler nerve pulsy	NA	Endoscopic endonasal approach	Disappear symptom
Wanibuchi et al.[17]	44/M	NA	NA	Radical removal in combination with high-flow bypass	Maintained his activities of daily living at 21 months
Present case	67/M	Numbness of face Ptosis, Diplopia	Homogenous enhancement without flow voids	Subtemporal epidural approach	GKS for residual tumor

F: female, GKS: gamma knife surgery, M: male, NA: not available, RT: radiation treatment.

radiotherapy.[3,6,8–11,20] Sung et al.[21] also reported a lower recurrence rate in the GTR group than the STR group, though the difference was not statistically significant and GTR group had a significantly longer progression free survival rate compared with the STR group in the WHO grade II hemangiopericytoma/solitary fibrous tumor patients. Few reports have described the results of GKS for hemangiopericytoma and showed the usefulness of adjuvant GKS in the management of recurrent or residual hemangiopericytoma.[4,7] Sheehan et al.[4] concluded that GKS provided at least intermediate-term local tumor control and recommended a marginal dose exceeding 15 Gy. Kano et al.[7] also reported that the factors associated with improved progression-free survival are lower grade hemangiopericytomas and higher marginal dose >14 Gy. In the present case, the patient underwent GKS four times, with prescribed doses to the tumor of 12, 15, 15, and 16 Gy, respectively. The prescribed dose for initial GKS was 12 Gy (<14 Gy), which might thus represent a cause of tumor recurrence. We initially diagnosed this tumor as a cavernous sinus schwannoma and performed initially GKS without checking pathology. Snyder et al.[22] reviewed the literature of the patients who underwent stereotactic radiosurgery for trigeminal schwannoma and upfront stereotactic radiosurgery was performed in 52–81.2% of the patients. Tripathi et al.[23] also reviewed the results of

GKS for trigeminal schwannoma and reported that most of the studies succeeded GKS with a 12–14 Gy prescribed dose.

Treatment after recurrence

There is still a controversy regarding the optimal management of recurrence. Wang et al. recommended surgery at the first recurrence and radiotherapy should be administered if there is no history of radiation. They reported a longer survival rate with their patients who received surgery at their first recurrence.[24] Chemotherapy also might be useful at the recurrence time. Park et al.[25] reported that a combination therapy with temozolomide and bevacizumab is a beneficial regimen for hemangiopericytomas and solitary fibrous tumors. The usefulness of repeated GKS for progression after initial treatment with GKS has also been reported.[19,21,26] Sung et al.[21] concluded that a GTR and radiation therapy can increase the disease free time of patients, and also emphasized the fact that an active treatment such as reoperation, repeated radiation therapy or repeated GKS were meaningful even after recurrence.

However, adverse effects after GKS are not well known. Lee et al. reported that trigeminal nerve tends to be influenced by radiation. They analyzed 159 patients of cavernous sinus meningiomas, four patients remained permanent trigeminal neuralgia or keratitis and one patient showed transient facial paresthesias after GKS.[18] Roche et al.[27] also reported that half of their patients who underwent GKS for cavernous sinus meningioma with tumor related trigeminal neuralgia improved or recovered after GKS. On the other hand, Chang et al. reported the effects of stereotactic radiosurgery on secondary facial pain. They concluded that half of their patients showed recurrence of their facial pain during follow-up periods, though 86% of the patients achieved pain relief initially.[28] Adverse effects after repeated GKS are also not well known, further studies are needed.

In the present case, if the tumor was to increase in size again, surgery would be one of the alternative treatment, with a careful long term follow up.

Conclusion

We have reported a rare case of intracavernous hemangiopericytoma originating in the cavernous sinus. Although distinguishing an intracavernous hemangiopericytoma/solitary fibrous tumor from intracavernous schwannoma is difficult preoperatively, inclusion of hemangiopericytoma/solitary fibrous tumor in the differential diagnosis is important, even when the tumor is round, enhancing homogeneously without flow voids. GKS might be one of the options for residual and recurrent intracavernous hemangiopericytomas.

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Conflicts of Interest Disclosure

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