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Early Response to Radiation Therapy without Surgical Intervention in a Giant Cavernous Sinus Hemangioma with Hydrocephalus: A Case Report

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Abstract

Cavernous sinus hemangioma (CSH) is a rare vascular malformation, arising from the cavernous sinus. Because of its anatomically complex location, a large lesion can cause a variety of symptoms due to cranial nerve compression. A 69-year-old woman with an unsteady gait was admitted to our hospital, and magnetic resonance imaging revealed an extra-axial giant tumor in the cavernous sinus and enlarged ventricles. A radiographic diagnosis of CSH was made. As the risk of surgical removal was considered high, the patient underwent intensity-modulated radiation therapy of 50.4 Gy in 28 fractions. The size of the tumor decreased markedly over time, and the symptoms improved soon after treatment. A 61.8% reduction in tumor size was confirmed immediately after irradiation, and a 75.9% reduction was revealed at a follow-up visit one year later. We reported a case of a giant CSH with hydrocephalus, where tumor shrinkage was confirmed immediately after radiation therapy, and the symptoms of hydrocephalus improved without surgical intervention.

Keywords: cavernous sinus hemangioma, hydrocephalus, radiation therapy

Introduction

Cavernous sinus hemangioma (CSH) accounts for up to 3% of all cavernous sinus lesions.[1)] CSH is a benign extra-axial tumor but, as it grows, various symptoms occur because of its anatomic complexity. Patients may experience not only nonspecific symptoms, such as headache or dizziness, but also signs of cranial nerve palsy, such as diplopia, decreased visual acuity, ptosis, or facial hypoesthesia.[2-4)] Treatment strategies recommend radiation therapy, but may occasionally include surgical removal.[2-6)] Because the reported cases of CSH with hydrocephalus are limited, refinement of treatment strategies is even more difficult. We present a case of a giant CSH with hydrocephalus in which radiation therapy was performed without surgical intervention.

Case Report

A 69-year-old woman with a gait disturbance and urinary incontinence that had started several months earlier was referred to our hospital. Prior to the onset of symptoms, the patient was transferred to another hospital with transient loss of consciousness, and a computed tomography (CT) scan revealed a giant solid mass in the middle cranial fossa, with a density higher than that of the brain parenchyma but without calcification (Fig. 1A). Thereafter, the patient had no symptoms or signs of hydrocephalus and was monitored for a year, over which the lesion size gradually increased. Physical examination revealed mild

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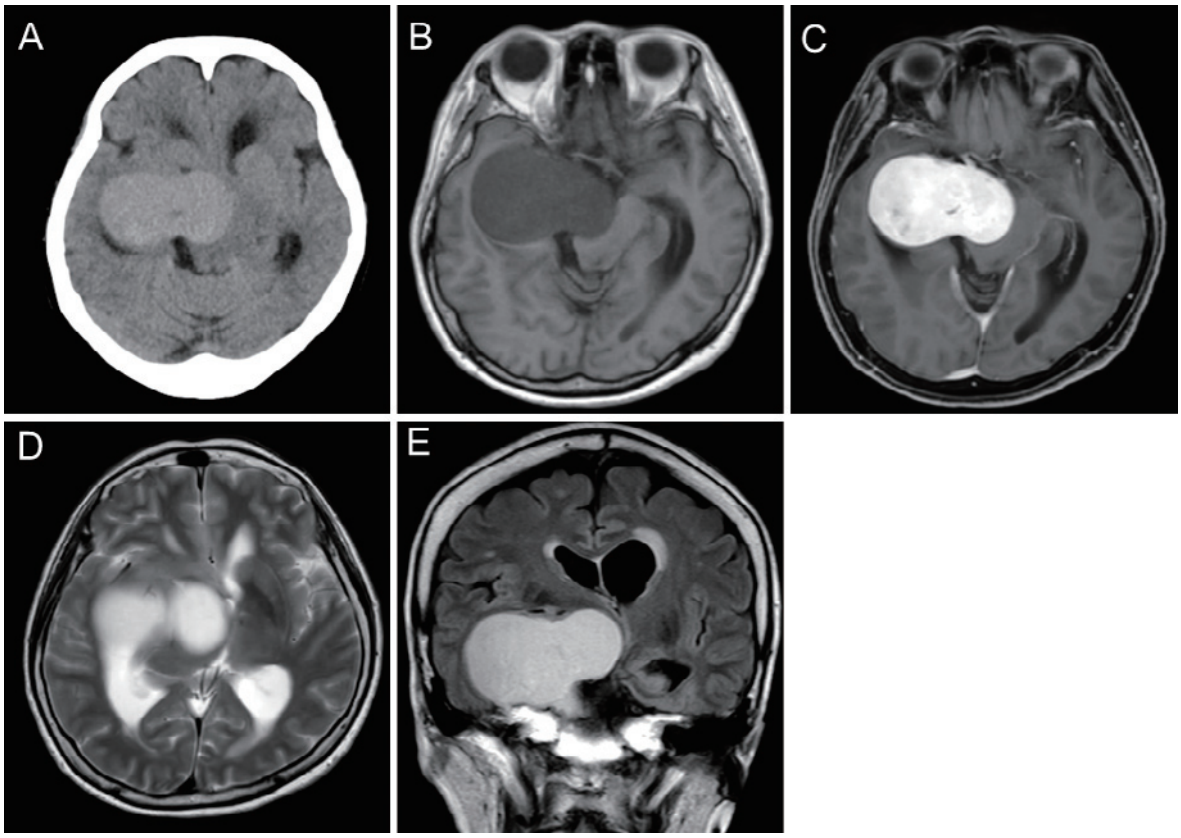

Fig. 1 CT scan and MRI on the presentation.
An extra-axial tumor, 64 × 40 × 48 mm in size, spread from the right cavernous sinus and sella turcica to the medial temporal fossa and markedly compressed the midbrain. The tumor density was slightly higher than that of brain parenchyma without calcification on CT (A). There was distinct hypointensity on T1WI (B), with a uniform enhancement on post-contrast T1WI (C), and prominent hyperintensity on T2WI (D). The foramen of Monro was obstructed (D), and the disproportional enlargement of ventricles were observed with slight periventricular hyperintensities on FLAIR (E). No signs of hemosiderin deposition were detected.

right hemiparesis, difficulty walking, urinary incontinence, and mild cognitive dysfunction (a score of 28 out of 30 points on the Mini-Mental State Examination). The Karnofsky Performance Scale (KPS) score was 80.

Magnetic resonance imaging (MRI) revealed an extra-axial tumor 64 × 40 × 48 mm in size, spreading from the right cavernous sinus and sella turcica to the medial temporal fossa, that was markedly compressing the midbrain. The tumor was hypointense on T1-weighed images (T1WI), hyperintense on T2-weighed images (T2WI), and uniformly enhanced on post-contrast T1WI (Fig. 1B-D). Because of the compression in the foramen of Monro, the ventricles were disproportionally enlarged with slight periventricular hyperintensities on fluid-attenuated inversion recovery images (Fig. 1D, E). On angiography, the right intracranial internal carotid artery was deviated medially, but flow to the distal end was not impaired. Small feeding arteries from the meningohypophyseal trunk (MHT) and the anterior branch of the middle meningeal artery were revealed; numerous branches from the MHT demonstrated gradual contrasting of the tumor from its inner part (Fig. 2). Based on these findings, a radiographic diagnosis of hemangioma of the cavernous sinus was made.

The patient underwent intensity-modulated radiation therapy (IMRT) of 50.4 Gy in 28 fractions, with emergency surgery as a backup. Regarding the target volume for IMRT, the gross tumor volume (GTV) was defined as an MRI-enhanced mass lesion, the clinical target volume (CTV) was defined as the GTV plus 5 mm margin, and the planned target volume was the CTV plus 2 mm margin. Trilogy (Varian Medical Systems, Palo Alto, California) and TrueBeam (Varian Medical Systems, Palo Alto, California) were used for treatment, with an X-ray energy of 10 MV. On MRI performed at the end of radiotherapy, the tumor size decreased from 85.9 to 32.8 cm^3 (a 61.8% tumor reduction), and the dilated ventricles decreased in size (Fig. 3A, B). A complete improvement in symptoms was soon confirmed, and the KPS score was 100 points. At one-year follow-up, there was an overall tumor reduction of 75.9%.

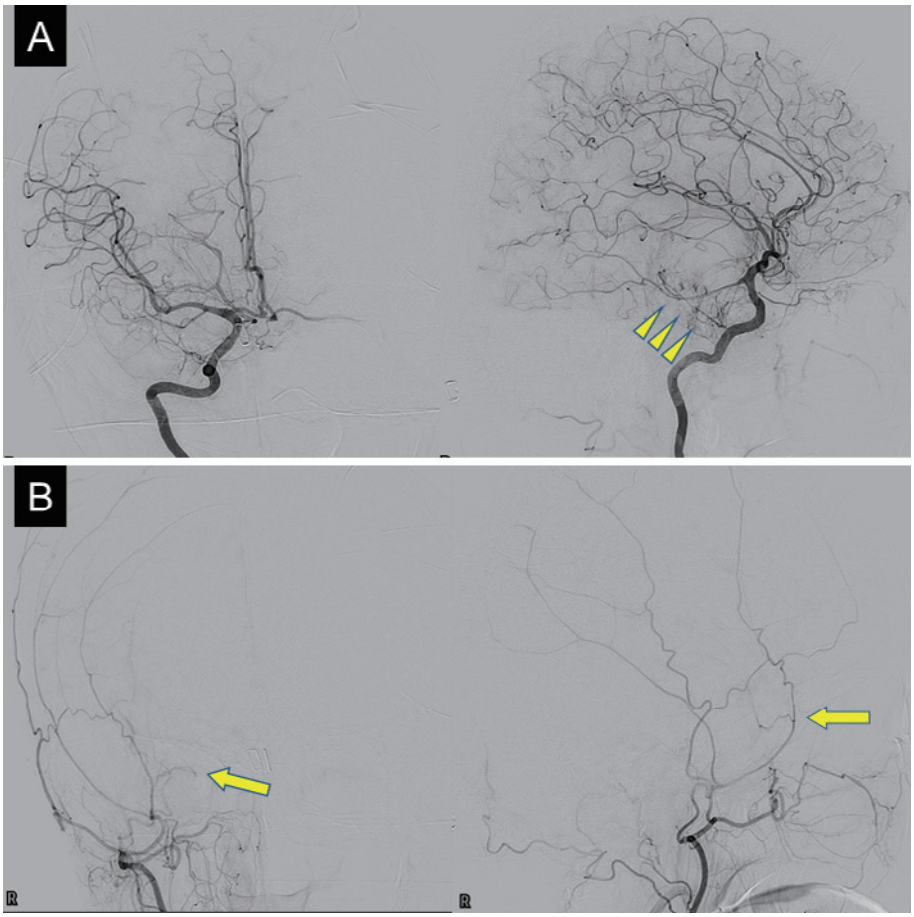

Fig. 2 Anteroposterior and lateral views of the internal carotid angiography (A) and the external carotid angiography (B). The right intracranial internal carotid artery deviated medially, and small feeding arteries from the meningohypophyseal trunk (MHT) (A: arrowheads) and the anterior branch of the middle meningeal artery (B: arrow) were observed. The MHT contrasted the tumor gradually, starting from the inner part.

Discussion

CSH is a cluster of thin-walled dilated vessels with fibrous connective tissue arising from the cavernous sinus. Compared with cavernous malformation (CM), CSH is neither in the brain parenchyma nor contains histological evidence of prior hemorrhage, thrombosis, or calcification.[1)] Radiographically, CSH shows low intensity on T1WI, markedly high intensity on T2WI, high density on CT without signs of hemosiderin deposition, and uniform enhancement on contrasted T1 images,[7-9)] whereas CM shows heterogenous intensity on T1WI and T2WI, with subtle or no gadolinium enhancement on T1, and is prominent on hemosiderin-sensitive sequences.[1,10)] CSH angiography may demonstrate a "blush" appearance due to contrast accumulation in small feeding arteries, as in the current case,[8,9)] whereas CM is angiographically occult.[1,10)] Differential diagnosis includes meningioma, especially microcystic meningioma, which shows patently low intensity on T1WI and high intensity on T2WI, which may be similar to those of the present case.[11)] However, a meningioma would be accompanied by peritumoral brain edema and demonstrate low-to-isodensity on CT scans; furthermore, a large-sized meningioma would show noticeable feeding arteries on angiography.[7,11,12)] Compared with microcystic meningioma, the current case shows high attenuation on noncontrast CT, suggesting blood congestion in dilated venous vessels, prominent homogeneity on contrasted T1WI, and angiographic images of the subtle feeding arteries.

Surgical removal is a CSH treatment strategy; however, it carries the risk of profuse bleeding and complications in the form of cranial nerve dysfunction due to the origin and location of the tumor, as well as the importance of the surrounding structures.[8,13)] Zong-Hao Li et al. performed surgical removal in 47 cases, resulting in 55.3% of gross total removal, with complications of newly developed or deteriorated cranial nerve injury in 76.6% of cases. They insisted that increased tumor size is a risk factor for an unfavorable postoperative KPS score, and sella turcica invasion was related to cranial nerve damage and unfavorable KPS scores.[4)] In our case, we observed a giant hemangioma invading the sella turcica; therefore, the risk of complica-

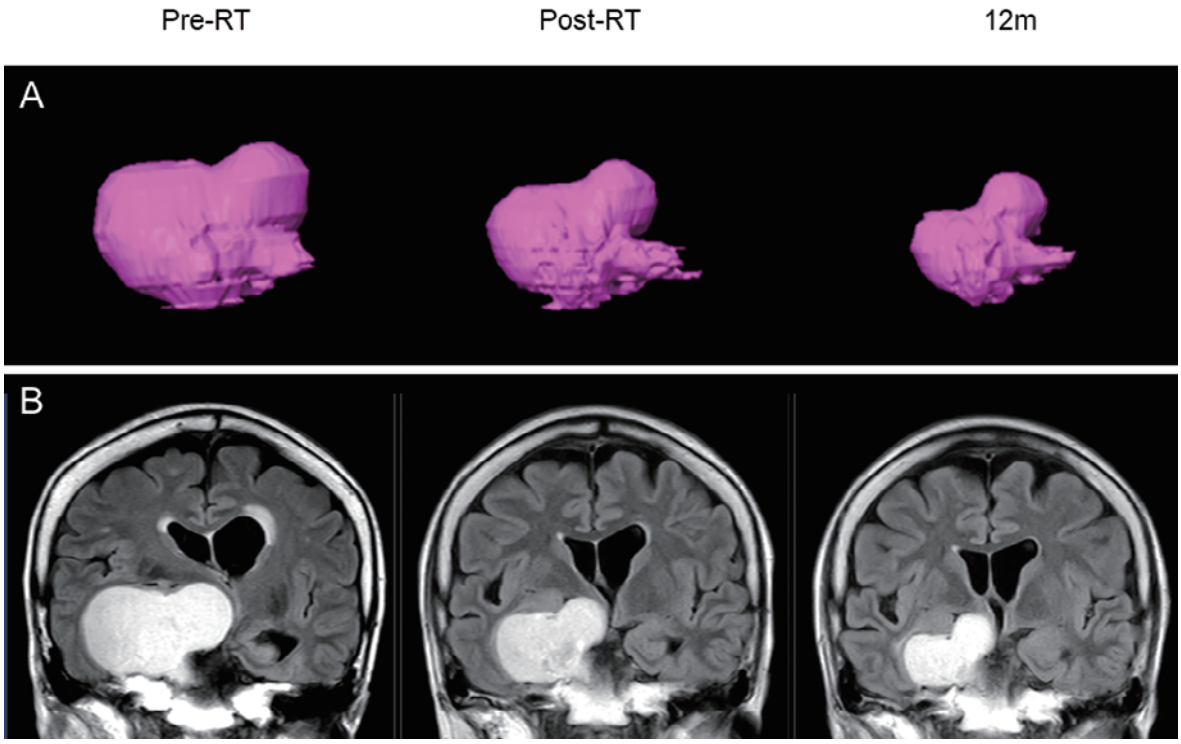

Fig. 3 Tumor size change over the follow-up period.
3D (A) and MRI on FLAIR (B) of the tumor before radiation therapy (RT), at the end of RT, and 12 months after RT. The tumor was gradually shrinking. Note that blockage of the foramen of Monro has already disappeared and the areas of periventricular hyperintensity are reduced at the end of RT.

Table 1 Summary of large studies of CSH radiation therapy

	Number of Cases	Age (year) [range]	TV (cm^3) [range]	Treatment Modality	MRD (Gy) [range]	Post-irradiation VR [range]	LFU (months) [range]	Permanent complication
Yamamoto M, et al. (2010) [14]	30	53 [19-78]	11.5 (mean) [1.5-51.4]	GK	13.8 [10.0-17.0]	>50% VR in 18 cases, slight VR in 11 cases	53 [12-138]	Hemifacial sensory disturbance in one case
Wang X, et al. (2012) [6]	14	56.9 [33-80]	50.2 (median) [22.6-96.3]	CK	21 in 3 Fr	Mean VR of 77%	15 [6-36]	None
Wang Y, et al. (2016) [15]	32	49 [21-73]	30.53 (mean) [2.5-78.6]	GK	14.22 [11-16]	Mean TV of 8.52 cm^3 [0-35.4 cm^3]	30.2 [12-67]	None
Wang X, et al. (2018) [3]	31	54 [22-80]	64.4 (mean) [40.9-145.3]	CK	21 in 3 Fr or 22 in 4 Fr	Median VR of 88.1% [62.3%-99.4%]	30 [6-78]	None
Xin Z, et al. (2020) [2] (*)	54	47.8 [22-64]	60.9 (mean) [20.2-230.5]	CRT	50 in 25 Fr	Mean VR of 79.7% [48.4%-98.5%]	35 [1-140]	None

TV: tumor volume, MRD: marginal radiation dose, VR: volume reduction, LFU: length of follow-up, GK: gamma knife, CK: cyber knife, CRT: conventional radiation therapy, Fr: fraction
*Tumor size decreased in all cases within 3 months after RT.

tions from surgical removal was considered high.

Several studies have shown the safety and efficacy of stereotactic radiosurgery for the treatment of CSH (Table 1).[2,3,6,14,15] In previous studies, small- and medium-sized tumors were usually treated by stereotactic radiosurgery because, in larger tumors, the risk of radiation-induced optic neuropathy increases since the lesions are adjacent to the optic apparatus.[2,3] This is why fractionated irradiation is recently favored. Three large studies have evaluated large-to-giant CSH treatment using a hypofractionated cyber

knife (CK) or conventional radiation therapy (CRT), where patients with large (tumor volume > 20 cm^3, 3-4 cm in diameter) and giant (tumor volume > 40 cm^3, >4 cm in diameter) tumors were treated, resulting in excellent tumor control without developing permanent neurological deficits.[2,3,6] According to a pooled analysis, patients who were managed with radiosurgery had a 100% decrease in the odds of post-treatment complications and significant improvement of the symptoms, compared with patients who were treated with surgical resection.[5]

Among several giant CSHs in the literature, there is a limited number of CSH with hydrocephalus cases in which patients underwent surgical removal due to signs of increased intracranial pressure, such as headache, nausea, or even a decreased level of consciousness. In these three reports, a diagnosis of CSH had not been made prior to surgeries, and two of the patients received radiation therapy after histopathological examinations.[7,16,17] As for our case, hydrocephalus symptoms developed gradually over a year of observation, and the diagnosis of CSH was made preoperatively. According to a previous study in which giant CSHs were treated with CRT, an average 34.3% peak reduction in tumor volume was observed at the end of radiotherapy.[2]

Therefore, based on the presumption that radiation therapy would become effective before the obstructive hydrocephalus became more severe, the patient was treated as described above. Fortunately, the tumor responded to the treatment so early that, by the end of radiotherapy, radiographic finings and hydrocephalus symptoms had improved. Compared with another previously published study, the percentage of tumor shrinkage in this case was significantly higher at the end of radiation therapy. Yamamoto et al. pointed out the efficacy of radiosurgical thrombolysis for a large CSH, describing one such case in their study. They suggested that irradiation of the lower part of the lesion could lead to a decrease in the blood supply to the tumor.[14] This hypothesis may explain the rapid response and disproportionate reduction of the inferior lateral portion of the lesion in our case (Fig. 3B).

However, the sudden worsening of hydrocephalus symptoms could have occurred by the time the tumor shrank. Although no cases of transient radiation-induced CSH enlargement have been reported, aggravated headache and vomiting have been reported during hypofractionated radiation therapy for giant CSHs, some of which required steroid administration.[3] These symptoms cannot explain an undetected transient tumor enlargement but could be confusing in a patient with a giant CSH with obstructive hydrocephalus. On the other hand, Xin Z et al. also treated large-to-giant CSHs, in which CRT was performed with a marginal dose of 50 Gy in 25 fractions with no such side effects. From this point of view, the treatment we performed was a more appropriate choice in the current case. Regardless of the choice of hypofractionated CK or CRT, in a giant CSH with obstructive hydrocephalus, it might be unnecessary to perform ventricular drainage in advance but ventricular drainage would be safer as a backup in case of sudden deterioration.

Conclusion

Instead of a high-risk surgery, we performed fractionated radiotherapy on a patient with a giant CSH, complicated by hydrocephalus but without urgent symptoms. The tumor size decreased rapidly after radiation, and the hydrocephalus improved without surgery. For the treatment of hemangioma with obstructive hydrocephalus, radiotherapy should be considered as a treatment option with emergent ventricular drainage as a backup.

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Conflicts of Interest Disclosure

The authors declare no conflicts of interest associated with this manuscript.

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