Capillary hemangioma of the cavernous sinus

Report of two cases

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Capillary hemangiomas are rare benign vascular tumors that tend to occur in children. Whereas the majority of hemangiomas may regress spontaneously, those associated with functional sequelae or severe symptoms may require treatment. Two patients with capillary hemangiomas of the cavernous sinus that caused neurological symptoms were treated with fractionated stereotactic radiation therapy. Both hemangiomas had shown a progressive increase in size during observation before radiation therapy; both tumors regressed after radiotherapy. Up to the time of the last follow-up evaluation both patients experienced symptomatic relief after radiation. One patient's tumor remains in complete remission and the second tumor continues to demonstrate minor residual contrast enhancement without progression.

The authors conclude that fractionated stereotactic radiation therapy is a useful treatment modality in the management of symptomatic capillary hemangiomas when these tumors arise in regions of the brain or skull base in which a complete resection cannot be accomplished.

KEY WORDS • hemangioma • cavernous sinus • stereotactic radiosurgery

Several types of angiomas are seen in the pediatric population. Coffin and Dehner reviewed vascular tumors found in 222 children and adolescents. Of these tumors, 89% were benign, 9% were borderline or indeterminate, and 2% were malignant. Fifty-eight percent of lesions were diagnosed when patients were in the 1st year of life and 57% were diagnosed when patients were in the first decade of life. Capillary hemangioma (32%) was the most common type.

Capillary hemangiomas in newborns are characterized by proliferative and involutinal phases. Although the majority of hemangiomas regress spontaneously, those associated with functional sequelae or significant symptoms may require treatment. Interventional modalities include surgery, embolization, steroid medications, interventional therapy, laser treatment, cryotherapy, and radiotherapy.

Two cases of capillary hemangioma in the CS are reported. Complete excision was not possible in either case because of tumor adherence to nerves and the risk of significant bleeding. Both patients achieved significant tumor regression (one had a complete response and the other had persistent minor residual contrast enhancement without progression) with stereotactic fractionated radiation therapy.

Case Reports

Case 1

This 15-year-old girl presented with right-sided sixth cranial nerve palsy; 2 months later she experienced right-sided third cranial nerve palsy. An MR image of her brain obtained at that time revealed a contrast-enhancing lesion of the right CS (Fig. 1). A right frontotemporal craniotomy was performed for biopsy sampling and partial removal of the lesion. Pathological studies revealed proliferation of capillary vessels, and the diagnosis of a capillary hemangioma of the right CS was made.

Serial imaging demonstrated that the lesion had grown from 1.2 (right/left) × 1.2 (superoinferior) × 3 cm (antero-posterior) to 1.5 (right/left) × 1.5 (superoinferior) × 3 cm.
over a 2-month period. During this time, the patient’s diplopia became progressively worse. She experienced increased proptosis and pain in her right eye.

Considering the clinically and radiographically confirmed progression of the tumor, the patient was offered stereotactic fractionated radiation therapy. To accomplish this, the patient was immobilized using a bite block frame. A 2-mm margin around the gross tumor volume to the prescribed isodose line (90%) was mapped using computerized tomography planning and MR image fusion. The 2-mm margin was used to account for potential day-to-day variation in setup and/or movement. A four-isocenter plan with 1-, 1.5-, 2.25-, and 3.5-cm cones was used. The patient was treated daily on a 6-MV linear accelerator by using a dynamic rotational technique. A dose of 4500 cGy given in 25 fractions prescribed to the 90% isodose line was delivered (Fig. 2). At 3600 cGy, the proptosis in the right eye was visibly reduced and the patient’s right globe pain improved.

A follow-up MR image obtained 1 month after radiotherapy demonstrated nearly complete resolution of the right-sided CS lesion with minor residual contrast enhancement in the right CS (Fig. 3). Five months after completion of radiotherapy, a repeated MR image demonstrated no regrowth. The patient underwent eye surgery shortly after this follow-up study to compensate for her persistent right-sided sixth cranial nerve palsy. Her latest MR image, which was obtained approximately 1.5 years after radiation therapy, demonstrated continued local tumor control with no change during the interval.

Case 2

This 19-year-old woman presented with double vision that occurred mainly when she looked to the left side. On physical examination she was found to have a left-sided sixth cranial nerve palsy. An MR image of the brain revealed a contrast-enhancing lesion of the left CS (Fig. 4 left). Biopsy sampling of this lesion was performed, and revealed a capillary hemangioma. A follow-up MR image obtained 6 months after the biopsy demonstrated a significant (three- to fourfold) increase in the size of the left-sided CS hemangioma (Fig. 4 right). This increase may have been due to hemorrhage. Despite this significant enlargement, the patient’s symptoms did not worsen and she continued to experience a complete sixth cranial nerve palsy. She was treated with fractionated stereotactic radiation therapy to a dose of 5000 cGy in 25 fractions prescribed to the 97.5% isodose line in four arcs (Fig. 5).

Four months after radiotherapy, a follow-up MR image demonstrated no residual enhancement (Fig. 6). The patient continues to exhibit a complete response on follow-up MR images obtained 1.75 years posttreatment. Her sixth cranial nerve palsy has persisted and she underwent ophthalmological surgery to compensate for this abnormality.

**Discussion**

Capillary hemangiomas are rare benign tumors that occur in 1 to 2.6% of live births, and 10% are found in the patient’s 1st year of life. The majority of capillary hemangiomas arise in the skin, scalp, or oral mucosa and typically appear within a few months of birth. These tumors are more common in female patients, and in adults may change size according to the menstrual cycle and pregnancy. Cavernous hemangiomas differ from capillary hemangiomas both pathologically and clinically. Capillary hemangiomas appear grossly as circumscribed, unencapsulated masses with lobules; microscopically, capillary-sized vascular spaces are seen. Cavernous hemangiomas are circumscribed red-brown masses with a firm or spongy surface; microscopically, large vessels are filled with blood and lined with flattened endothelial cells. Intracranial cavernous hemangiomas are usually found in older children and adults. It is possible that capillary hemangiomas progress to cavernous hemangiomas; however, unlike childhood capillary hemangiomas, cavernous hemangiomas may not involute spontaneously. Cavernous hemangiomas have
been reported to occur in the CS in adults. Pure capillary hemangiomas of the CS are rare.

In addition, capillary hemangiomas may be difficult to distinguish from intravascular papillary endothelial hyperplasia, which is believed to be a rare and unusual morphological manifestation associated with an organizing and recanalizing thrombus. According to Rosai intravascular papillary endothelial hyperplasia is identified by lack of necrosis, bizarre cells, and atypical mitoses. This lesion characteristically features a fibrinous and/or hyaline appearance of the papillary stalks and a frequent finding of residual organizing thrombi. For our two cases there was no evidence of necrosis, bizarre cells, or atypical mitosis. In addition, there was no evidence of thrombosis (pathological specimens obtained in Case 2 are presented in Fig. 7). The area or wall between endothelial cells shown in the figure had cell nuclei and was not as hyaline or fibrotic as one might expect in an organizing thrombus. The pathological findings in the two cases were those of a vascular malformation with thin-walled and delicate vessels, as one would find in capillaries.

Capillary hemangiomas are known to undergo a proliferative phase during the patient’s first 6 months of life, and it often continues up to 1 year of age. Gradual, spontaneous involution then occurs in approximately 75% of cases, with complete involution occurring by 5 to 10 years of age. These benign tumors may represent small, isolated lesions with minimal clinical significance or they may grow into

![Image](image1.png)

**Fig. 3.** Follow-up MR image revealing complete response with only minor residual contrast enhancement along the right CS (arrow). As of the latest follow-up images, there has been no progression.

![Image](image2.png)

**Fig. 4.** Left: Prebiopsy MR image demonstrating capillary hemangioma of the CS. Right: An MR image demonstrating significant increase in the size of the hemangioma before radiotherapy was administered.
large masses resulting in both short-term and long-term morbidity. In cases in which intervention is offered for symptomatic capillary hemangioma, the following modalities have been used: surgery, embo- lization or a combination of surgery and embolization, medical therapy (such as steroid medications, antifibrinolytic agents, or interferon), laser therapy, cryotherapy, sclerotherapy, and radiation therapy.

In cases in which capillary hemangiomas grow significantly and cause significant neurological deficits, continued observation may be fraught with uncertainty whether the lesion will spontaneously involute and whether further waiting will diminish the chances of neurological recovery. Although complete excision is an option for CS capillary hemangiomas, surgery is often difficult because of the proximity of critical structures (such as cranial nerves and the optic chiasm) and the tumor’s tendency to bleed.

Radiotherapy has been used in the treatment of hemangiomas. The optimal dosage is unknown, but in general, hemangiomas are radioresponsive. Before 1960, radiotherapy was given using radium/radon plaques, molds, or glass capsules applied to skin hemangiomas. Orthovoltage x-ray therapy and brachytherapy were also used. In that era, approximately 85% of hemangiomas resolved completely and 15% showed improvement. Infrequently, some hemangiomas failed to respond to radiation.

Current radiotherapy techniques include orthovoltage x-ray therapy, electrons for superficial lesions, conventional external megavoltage photon radiotherapy, and stereotactic radiosurgery or fractionated stereotactic radiation therapy for small intracranial hemangiomas. In terms of dose, childhood hepatic hemangiomas quickly regressed with doses as low as 400 cGy. Subglottic hemangiomas causing airway obstruction have been treated successfully with fractionated doses as low as 450 to 1800 cGy. Also, 1500 cGy of fractionated superficial radiotherapy has been used to treat hemangiomas of the eyelids and conjunctiva.

We report on two patients who presented with small (≤3 cm in maximum dimension) CS capillary hemangiomas. These lesions were ideal targets for stereotactic fractionated radiation therapy. The technique we used allows for normal tissue sparing by both dosimetric and radiobiological means. There is a rapid dropoff of dose around the target, thereby sparing normal tissue from high doses of radiation. Fractionation allows normal tissue to repair itself between fractions. The more rapidly proliferating capillary hemangioma cells are destroyed preferentially compared with the slower-growing normal cells. Although capillary hemangiomas may respond to lower doses, the exquisite response to radiation has been reported only in infants. The degree of radioresponsiveness in older children and young adults is unknown. A dose of 4500 to 5000 cGy was used in 180- to 200-cGy daily fractions in the two cases reported. This dose/fractionation schedule was adopted for these two cases because there is experience in using this radiation regimen in other benign central nervous system tumors such as benign meningioma, pituitary adenoma, or acoustic neuroma. This is a dose/fractionation schedule that is well tolerated and is associated with a small risk of late symptomatic radiation necrosis and/or edema according to our experience with patients treated in a similar manner for vestibular schwannoma.

Late radiation-induced malignancy is a concern for young patients treated for benign tumors. Furst, et al. investigated the possible association between absorbed dose and cancer risk in a cohort of 14,647 patients younger than 18 months of age who underwent radiation therapy for skin hemangioma between 1920 and 1959. There were 55 patients with breast cancer, 14 with thyroid cancers, 16 with brain tumors, and eight with tumors of the bone and soft tis-
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Fig. 7. A: Photomicrograph showing open and collapsed thin-walled vascular spaces surrounding a small nerve (asterisk). Hematoxylin-phloxine-saffron, original magnification × 100. B: Photomicrograph used for immunohistochemical studies for CD34, showing the vascular spaces to be lined by strongly immunoreactive endothelium (arrows). Original magnification × 200.

A statistically significant positive dose–response relationship for thyroid cancers and for bone and soft tissue tumors was observed. The possibility of late serious radiation sequelae has resulted in a diminishing role for radiotherapy in the treatment of this disease.

Given these concerns, radiotherapy may be offered only in selected cases of capillary hemangioma (for example, in situations in which other modalities are not feasible or if the hemangioma is recalcitrant to other modalities). The two patients in this report were offered stereotactic fractionated radiation therapy when the tumor demonstrated progression on neuroimages obtained after surgery or biopsy procedures, and when there was concern that further observation without treatment would lead to worsening and irreversible neurological sequelae. This decision was balanced with the small chance of radiation-induced injury to normal brain and cranial nerves, as well as the risk of late-radiation induced malignancy. Our experience with these two patients demonstrates that capillary hemangiomas can be radiosensitive. Both individuals achieved an excellent response to radiotherapy, which has endured as of the latest follow-up imaging (1.5 and 1.75 years postradiation). The first patient’s symptoms improved significantly after radiotherapy. Both patients, however, had persistent sixth cranial nerve palsy that was subsequently alleviated with ophthalmological surgery. During this short follow-up period neither patient has experienced radiation toxicity.

Conclusions

Fractionated stereotactic radiation therapy is a useful treatment in the management of capillary hemangiomas when they arise in regions of the brain or skull base where a complete resection cannot be accomplished.

References


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