Orbital Hemangiopericytoma

—Case Report—

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Abstract

A 43-year-old male presented with swelling involving the right eye. T₁-weighted magnetic resonance imaging demonstrated a round tumor in the lateral region of the right orbit, which was isointense relative to the cerebral gray matter and homogeneously enhanced by gadolinium-diethylenetriaminepenta-acetic acid with a flow-void signal area in the mass. The tumor was totally resected through the transcranial and fronto-orbitotemporal approach after embolization of feeding arteries arising from the external carotid artery. The histological findings were characteristic of hemangiopericytoma. No radiation therapy was administered. The transcranial and fronto-orbitotemporal approach provides a wide operative field with excellent exposure of the highly vascular orbital tumor.

Key words: hemangiopericytoma, orbit, magnetic resonance imaging, radiation, transcranial approach

Introduction

Hemangiopericytoma of the orbit is rare, with some cases described in the ophthalmological literature, but few reports of hemangiopericytoma involving the orbit in the neurosurgical literature.^{2,6-9,13-15)} The incidence of hemangiopericytoma in the orbit was 1.57% in one series.⁸⁾ Nearly 80% of the patients are between 20 and 70 years of age.⁵⁾ This tumor is difficult to resect completely because the diagnosis is not usually made preoperatively and the tumor is highly vascular.^{5,6)} We describe a patient with an orbital hemangiopericytoma which was completely resected after embolization of the feeding arteries.

Case Report

A 43-year-old male attended the eye clinic of Kurume University Hospital because of painless proptosis and swelling of his right eye lid January 27, 1994. The swelling involving the right eye was first noted in June 1993. A biopsy was attempted in the eye clinic but the procedure had to be abandoned due to massive hemorrhage from the tumor. The patient was admitted to the neurosurgical facility for removal of the tumor by the transcranial approach.

Physical examination showed upper lid swelling, forward proptosis, and upward displacement of the right eye (Fig. 1). Right visual acuity was normal. The fundus of the right eye was seen clearly. Ocular movements were normal other than restricted elevation. A soft, compressible mass with well-defined margins was palpable, temporal to the right eye. The mass was non-pulsatile. His left eye was entirely normal as were the other physical examination findings. Laboratory data were all within the normal ranges. Tumor markers, such as carcinoembryonic antigen, alpha-fetoprotein, and carbohydrate anti-

Fig. 1 Photograph showing lid swelling, proptosis, and upward displacement of the right eye.

Fig. 2 *left: T₁-weighted coronal magnetic resonance (MR) image showing an isointense mass with a flow-void signal area (arrow), located in the extraconal region of the right orbit. right: T₁-weighted MR image with gadolinium-diethylenetriaminepenta-acetic acid clearly revealing a mass with flow-void signal area (arrow).*

Fig. 3 *Right internal carotid angiogram (lateral view) showing the tumor staining in the area fed by the ophthalmic artery (arrow-head).*

gens 19-9 and 125, were all within the normal ranges.

Computed tomography showed a large intraorbital mass in the lateral region of the right orbit, with homogeneous enhancement after intravenous administration of contrast material. T₁-weighted axial magnetic resonance (MR) imaging showed the mass

Fig. 4 *Intraoperative photograph after the lateral orbital rim was removed (arrow) and the periorbital incision was made, showing the encapsulated tumor (T) with no adhesion to the periorbital tissue.*

to be isointense and homogeneously enhanced by gadolinium-diethylenetriaminepenta-acetic acid. Vascular channels were detected in the mass (Fig. 2). T₁-weighted coronal MR imaging showed the mass was located in the extraconal region of the right orbit (Fig. 2). Cerebral angiography showed a marked tumor stain fed by branches of the ophthalmic and internal maxillary arteries (Fig. 3).

The feeding arteries from the external carotid artery were successfully embolized using polyvinyl alcohol on September 7. The tumor stain became less intense. He underwent removal of the tumor via a fronto-orbitotemporal approach on September 11. After the frontotemporal craniotomy, the lateral orbital rim was removed. The great sphenoidal wing and orbital roof were removed with a rongeur. The periorbital incision was made. The tumor was encapsulated, and there was no adhesion to the periorbital (Fig. 4). The well-defined white tumor was soft, and elastic hard. Bleeding from the tumor was minimal.

Histological examination of the specimen revealed the typical features of hemangiopericytoma. The tumor cells were spindle-shaped with round-to-oval nuclei. No mitotic figures were identified. There were numerous small capillaries, which were dilated with a typical "staghorn" configuration (Fig. 5 upper, middle). Silver staining indicated that a large amount of silver staining reticulum had developed throughout the tumor that divided the tumor cells into small groups (Fig. 5 lower).

The postoperative course was uneventful except for transient mild restriction of right ocular movement. Postoperative MR imaging showed no residual tumor (Fig. 6). No radiation therapy was ad-

Fig. 5 Photomicrographs of the tumor specimen showing numerous small capillaries, which are dilated and show a typical "staghorn" configuration (upper: HE stain, $\times 60$); closely packed, small cells with hyperchromatic nuclei between numerous, thin-walled vessels (middle: HE stain, $\times 250$); and many capillaries, which are easily seen due to marking by the reticulin sheaths (lower: silver reticulin stain, $\times 250$).

ministered. Follow-up examination 2 years later confirmed the absence of recurrence.

Fig. 6 Postoperative T₁-weighted magnetic resonance image with gadolinium-diethylenetriaminepenta-acetic acid revealing no residual tumor.

Discussion

The characteristic symptom of orbital hemangiopericytoma is painless proptosis with downward displacement. Painless non-axial proptosis and intermittent upper lid swelling are suggestive of hemangiopericytoma.¹⁷⁾ Our patient also presented with lid swelling and painless proptosis. Hemangiopericytoma of the orbit has been described as a slow-growing tumor. The symptomatic duration ranges from one month to several years.^{5,6,8)} Many previous reports described tumor growth as causing relatively little disturbance of visual function.^{5,6,8)} The tumor tends to be non-tender and non-pulsatile. The differential diagnosis includes meningioma, metastatic tumor, and cavernous angioma.¹⁰⁻¹²⁾

The MR imaging features of the orbital hemangiopericytoma in our case were the same as those of intracranial hemangiopericytoma. MR imaging demonstrates homogeneous tumor enhancement with a flow-void signal area, which represents the feeding artery of the tumor and is one of the characteristic MR imaging features of hemangiopericytoma.^{4,10-12)} Cerebral angiography showed dense tumor staining, reflecting the blood supply from branches of the maxillary and ophthalmic arteries. However, feeding artery occlusion could be achieved via the interventional technique before radical resection of the tumor.

Complete surgical removal of the tumor is re-

quired for hemangiopericytoma. Various surgical routes can be used to expose orbital lesions.¹⁴⁾ Different patterns of tumor extension may influence the choice of the most appropriate approach. Usually the transcranial approach is used for intraorbital tumors located in the apex, optic canal, or both, and for intracranial tumors with extension into the orbit. The transcranial and fronto-orbitotemporal approaches provide the best exposure of the orbital cavity, and are indicated for tumors located in the orbital apex and deep medial orbit. The advantages of these approaches are a wide operative field and excellent cosmetic results. Lateral orbitotomy is the most frequently used surgical approach, which provides excellent exposure of the superior lateral and inferior anterior intraconal compartments. However, the lateral approach may be complicated by lateral rectus paresis.¹³⁾ We selected the transcranial and fronto-orbitotemporal approaches involving the removal of the lateral orbital rim. The tumor in the present case was hypervascular and located laterally, so a wider working space was needed for total removal. These approaches allow easy access to the superior and lateral orbit.

Incomplete removal of orbital hemangiopericytoma is apparently a factor contributing to eventual recurrence.⁶⁻⁸⁾ Four recurrent or metastatic cases occurred among 11 patients who were followed up for 11 years or longer.⁸⁾ The recurrences were frequent in case of incompletely excised tumors, but these recurrences might not manifest for up to 10 years after surgery.⁸⁾ Surgical removal followed by postoperative irradiation is the treatment of choice.^{8,17)} A patient with a recurrent tumor, treated with radiotherapy and chemotherapy, was tumor free 13.4 years later.⁵⁾ However, postoperative radiation therapy seriously damages visual function. We recommend that radiation therapy be used only in patients with an inoperable recurrence or metastasis. Recurrence and metastasis usually develop late in the clinical course of this disease.^{1,3,5,8,16,17)} Therefore, long-term follow-up for 10 years or more is essential.

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