Managing a long-recurring primary orbital hemangiopericytoma

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ABSTRACT

Hemangiopericytomas are uncommon vascular neoplasms that originate from the pericytes lining capillaries and postcapillary venules. Few cases of primary orbital hemangiopericytoma have been reported. We present a 61-year-old woman with longstanding, recurrent benign orbital hemangiopericytoma and results of its successful removal.

KEYWORDS Anterolateral thigh flap; conformer; hemangiopericytoma; oculocardiac arrest; orbital hemangiopericytoma; preoperative embolization

If improperly excised, hemangiopericytomas have a high local recurrence rate and can metastasize. Complete removal is required to prevent tumor recurrence.¹ This uncommon neoplasm occurs primarily in the lower extremities, pelvis, and retroperitoneum.² The localization of this tumor to the orbit is rare.¹

CASE PRESENTATION

A 61-year-old woman was known to have a recurrent, benign, left medial orbital hemangiopericytoma. Prior to her final resection, she had undergone six surgical procedures for tumor removal within the past 22 years. When she was 52 years old, during an attempt to completely resect the tumor, significant blood loss complicated by asystole from the oculocardiac reflex occurred. She was promptly treated with atropine and blood transfusion, but the surgery was aborted due to a significant asystolic episode and a dilated pupil. During the next few years, four more surgical debulking procedures were performed for symptom management and preservation of eyesight.

At age 61 the patient presented to us with a chief complaint of constant pain of her left orbit, marked proptosis, and severe visual debility. Additionally, she was experiencing chronic left frontal sinusitis, secondary to the obstruction caused by the neoplasm. Examination disclosed severe proptosis, optic pallor, and visual field abnormalities of her left eye, consistent with optic neuropathy. Hertel measurement

was 28 mm in her left eye and 18 mm in the right; visual acuity was 20/40 in the left eye and 20/20 in the right. She also had edema of the right and left eyelids, punctate keratitis from the secondary dry eye, conjunctival congestion, and mild chemosis (*Figure 1a*).

Orbital magnetic resonance imaging revealed an enhancing multilobular left orbital mass involving the intraconal as well as medial and superior extraconal spaces. It measured 3.8×3.7 cm axially and 3.1 cm craniocaudally, a significant increase compared to 2.6×2.5 cm axially and 2.1 cm craniocaudally the year before. Additionally, magnetic resonance imaging showed increasing mass effect on the optic nerve, laterally deviating it and splaying it along the inferolateral side of the tumor. The right orbit was unremarkable (*Figure 2*). Computed tomography of her sinuses showed tumor invasion of the frontal sinus. Given the size and recurrence of the tumor, the difficulty of preserving eyesight, and its effect on the patient's quality of life, we elected to proceed with a subtotal exenteration.

Preoperative embolization was done to obtain vascular control. The left internal carotid artery was successfully stented. Bead and coil embolization was achieved of the left ophthalmic artery and the internal maxillary artery, resulting in expected blindness of the left eye. The patient underwent surgical resection of the tumor the next morning.

The surgery began with enucleation of the left eye to decompress the orbit. Dissection proceeded around the edges

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Figure 1. The patient (a) preoperatively and (b) postoperatively with a prosthetic eye.

Figure 2. Orbital magnetic resonance imaging with contrast showing an enhancing multilobular left orbital mass involving the intraconal and medial and superior extraconal spaces.

of the tumor; the tumor was intimately involved with the superior, medial, and inferior rectus muscles and the superior oblique muscle, which were all removed. A portion of the medial maxilla bone was also excised to ensure a complete tumor margin with preservation of the nasolacrimal duct and infraorbital nerve. The upper and lower eyelids, orbital septum, and levator muscle were all preserved. The tumor bled minimally with an estimated blood loss of 10 cc.

Once the tumor had been removed, left frontal sinus trephination was performed through the orbital incision to alleviate the patient's sinusitis. A 12 French red rubber catheter was placed in the frontal sinus, drawn out to the nasal cavity, and appropriately sutured to serve as a stent during the healing of the free flap. A lateral canthotomy with bone flap removal of the lateral rim was then performed to make room for the anterolateral thigh flap, which was to be placed to fill the volume of the orbit. The free flap was placed into

the orbit and sutured to the residual conjunctival surface with a conformer placed. The patient tolerated the procedure well with no complications. The remainder of her hospital course was uneventful, and she was discharged on the third day after the surgery.

DISCUSSION

The patient's initial surgeries for recurrent benign hemangiopericytoma were unable to completely excise her tumor but had been appropriate for the mild to moderate proptosis and moderately sized tumor to preserve vision. Thus, debulking was able to buy the patient several years of good visual function.

The patient's history of a sensitive oculocardiac arrest and the vascular nature of hemangiopericytomas were taken into account in the treatment plan with preoperative embolization, allowing for easier resection with less bleeding. For the flap repair, volume needed to be restored to the orbit for aesthetic and functional reasons. The patient's levator muscles and eyelids were preserved to give her eyelid function once she was fit with a prosthetic eye. Thus, volume was necessary to maintain a prosthetic eye under the eyelids. This could only be done with a relatively large volume vascularized free flap. A temporalis muscle transfer would not have provided enough volume and the patient was too thin with a low basal metabolic index to allow a radial forearm flap to fill the volume; hence, an anterolateral thigh flap was harvested and implanted with vascular connection to the superficial temporal vessels. The patient subsequently underwent a planned debulking procedure of the anterolateral thigh flap with a frontalis suspension procedure to correct ptosis of the left eyelid. She has been fit for a prosthetic eye and has an excellent cosmetic result and no residual pain (*Figure 1b*).

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