



Case report

Persistent macular puckering following excision of causative orbital tumor

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ABSTRACT

Purpose: To describe the clinical course of a patient with a retrobulbar orbital tumor causing myopic shift and macular pucker.

Observation: Following complete surgical removal of a retrobulbar orbital cavernous hemangioma, the myopic shift improved but the macular pucker persisted even 3 years after orbital surgery, with no sign of tumor recurrence.

Conclusion and importance: Chorioretinal folds secondary to chronic mechanical force from an orbital tumor may persist long after the tumor is removed. This case may assist ophthalmologists in their discussions with, and counseling of, patients regarding visual prognosis following excision of orbital tumors that are causing retinal changes.

1. Introduction

Retrobulbar orbital tumors are common, and both intra- and extraconal tumors can compress the inner surface of the sclera to cause deformity of the choroid, Bruch's membrane, and retinal pigment epithelium and retina, leading to chorioretinal folds. Chorioretinal folds are caused by a relatively limited set of conditions, and hence have important value in the diagnostic workup of patients with vision changes and/or retinal findings. Symptoms and clinical findings usually improve after treatment of the causative condition. However, a recent article reported the persistence of chorioretinal findings even years after complete surgical removal of a retrobulbar cavernous hemangioma.¹ We report a second, similar case, suggesting that the chorioretinal changes secondary to chronic mechanical pressure from a cavernous hemangioma may lead to lasting retinal changes.

2. Case report

A 46-year-old woman presented with a 5 year history of progressively worsening hyperopic shift in the left eye. Refraction in the left eye had worsened significantly from +4.00 to +5.50 -0.25 × 165. Refraction in the right eye was +0.50 sphere. Her community ophthalmologist had been following a known macular pucker throughout that time, but felt that the recent acceleration of her vision changes warranted an evaluation by a retinal specialist. Upon initial retinal evaluation, visual acuity was 20/20 in the right eye, and 20/50 + 1 in the left eye. Macular striae with anterior displacement of the posterior

pole and choroidal and retinal folds, with no significant epiretinal membrane, were noted on ocular coherence tomography (OCT) (Fig. 1A, C). Subtle proptosis was also noted. These findings suggested a retrobulbar mass, and an oculoplastics evaluation was requested. On oculoplastics exam, exophthalmometry measured 19 mm on the right and 21 mm on the left. Ocular motility was full bilaterally. Ultrasound of the left orbit demonstrated a well-outlined, oval-shaped, intraconal lesion with maximal lateral dimension of 14.0 mm and maximal depth of 13.0 mm. The lesion was noted to be regularly structured and highly reflective, with marked sound attenuation and positive internal vascularity. An orbital CT scan demonstrated a heterogeneously enhancing oval shaped mass discrete from optic nerve and extraocular muscles, most consistent with a cavernous hemangioma (Fig. 1E). Given the progressive findings and visual compromise, a decision was made for surgical excision. A left lateral orbitotomy for orbital mass excision with cryo-extraction was performed uneventfully. Histo-pathology results confirmed the diagnosis of a cavernous hemangioma. During a six month follow-up visit, the patient's refractive error in the left eye improved by 1.25 diopters to +4.25 + 0.25 × 055 and the proptosis subsided. Refraction was unchanged on the right at +0.25 sphere. However, dilated fundus exam demonstrated persistence of prominent choroidal and retinal folds in the posterior pole of the left eye. Over the following two years, visual acuity did not improve further, and the macular changes noted above remained. A repeat CT scan demonstrated no evidence of recurrent tumor (Fig. 1F). Dilated fundus exam and repeat OCT showed residual macular striae in the left eye (Fig. 1B, D).

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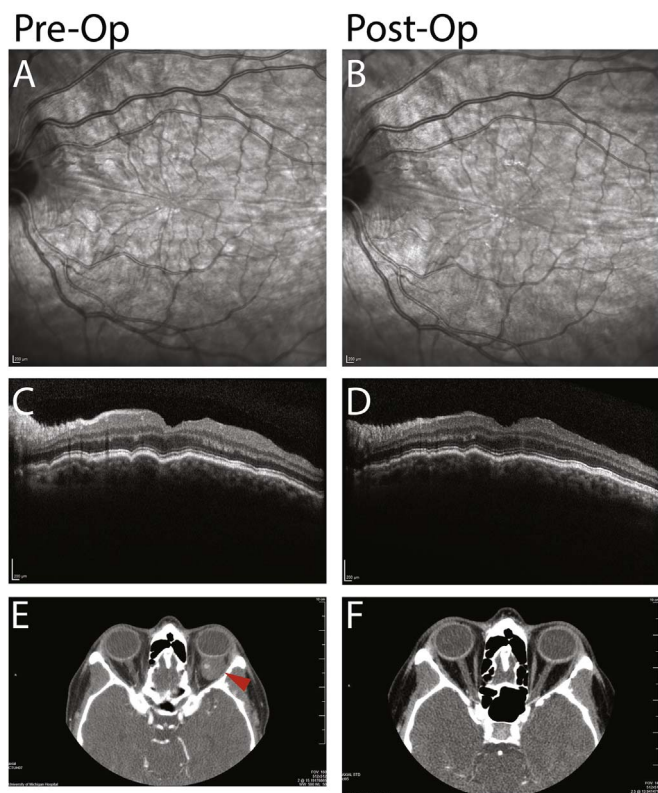


Fig. 1. Pre- and post-operative imaging demonstrating persistence of macular pucker despite complete excision of the retrobulbar orbital cavernous hemangioma with no sign of recurrence. A, B. Pre- and post-operative red-free retinal photo. C, D. Pre- and post-operative ocular coherence tomography scan of the macula. E, F. Pre- and post-operative orbital CT scan demonstrating intraconal orbital cavernous hemangioma abutting the globe, with no sign of recurrence 3 years following surgical excision.

3. Discussion

Studies have found that cavernous hemangiomas of the orbit represent roughly 6% of orbital masses.² Excision of these masses results in improved visual acuity in 32.4% of cases, while in 59.4% of cases, visual acuity remains unchanged.³ It is surmised that in this case, persistence of posterior pole findings represent gliotic and fibrotic changes involving all layers of the retina and choroid. This case highlights macular changes persisting for years after removal of the causative factor in posterior pole deformation.

4. Conclusion

Macular chorioretinal changes secondary to retrobulbar orbital tumors may persist following excision of the causative mass. This is the second example of persistent chorioretinal folds related to a retrobulbar cavernous hemangioma, which may be related to the slow, insidious growth of these tumors. Our findings may assist with counseling of patients with retrobulbar masses causing secondary hyperopic shifts and macular changes.

Patient consent

A written consent to publish this case report was obtained from the patient.

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Conflicts of interest

The following authors have no relevant financial disclosures: CH, DZ, AK.

Authorship

All authors attest that they meet the current ICMJE criteria for Authorship.

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References

- Jacobsen AG, Toft PB, Prause JU, Vorum H, Hargitai J. Long term follow-up of persistent choroidal folds and hyperopic shift after complete removal of a retrobulbar mass. *BMC Res Notes*. 2015;8:678.
- Shields JA, Shields CL, Scartozzi R. Survey of 1264 patients with orbital tumors and simulating lesions: the 2002 Montgomery Lecture, part 1. *Ophthalmology*. 2004;111:997–1008.
- Rootman DB, Heran MKS, Rootman J, et al. Cavernous venous malformations of the orbit (so-called cavernous haemangioma): a comprehensive evaluation of their clinical, imaging and histologic nature. *Br J Ophthalmol*. 2014;98:880–888.