

FIG 3. Orbital soft tissue histopathology slides. A, Microscopic examination showing a lipogranulomatous inflammatory reaction in the orbital soft tissue secondary to a reaction to silicone oil (hematoxylin, phloxine, saffron [HPS], original magnification $\times 50$). B, High-power view showing the multinucleated foreign body giant cells (arrow) that have engulfed the silicone oil (star; HPS, original magnification $\times 200$).

Silicone oil migration out of the vitreous cavity has been reported previously. Proposed mechanisms of oil extrusion include migration through incompletely healed sclerotomy sites^{2,3} and direct trans-scleral passage (especially if local scleromalacia with coinciding increased intraocular pressure)² or via the trabecular meshwork and episcleral veins.² These mechanisms would be more likely in cases of direct ocular trauma postoperatively. With the introduction of small-gauge mostly sutureless transconjunctival vitrectomy surgery,^{7,8} silicone oil migration has become more common. Even if the sclerotomy is presumed “watertight,” it is common for a small volume of silicone to escape as it is withdrawn and become trapped by overlying conjunctiva.

To our knowledge, this is the first report of silicone oil migration from the vitreous space into the periorbital space surrounding an extraocular muscle, after otherwise uncomplicated 23-gauge vitrectomy with intraocular silicone oil injection. Although the exotropia was unlikely secondary to silicone oil migration, the ocular discomfort may have been related to its presence, because the discomfort was relieved after strabismus surgery. The silicone oil migration was presumably due to sclerotomy site leakage, despite no other signs of leakage. Our case highlights the importance of meticulous sclerotomy site closure and the importance of a low threshold for

suturing sclerotomies, especially in pediatric vitrectomy surgery.

Literature Search

PubMed was searched on September 9, 2018, for English-language results using the following terms in combination: *silicone oil, extrusion, migration, complications, extraocular muscles, strabismus, sclerotomies, leakage, 23-gauge vitrectomy, and pediatric.*

References

1. Federman JL, Schubert HD. Complications associated with the use of silicone oil in 150 eyes after retina-vitreous surgery. *Ophthalmology* 1988;95:870-76.
2. Donahue SP, Friberg TR, Johnson BL. Intraconjunctival cavity inclusions of silicone oil complicating retinal detachment repair. *Am J Ophthalmol* 1992;114:639-40.
3. Ali Bautista S, Wladis EJ, Farber M, Falk NS, Phelps PO. Silicone oil migration into the orbit. *Orbit* 2018;37:368-70.
4. Deguchi Y, Maeno T, Hori Y, Hiruta N, Sasai D, Sato Y. Migration of intraocular silicone oil from the vitreous cavity into the upper eyelid causing ptosis. *Case Rep Ophthalmol* 2014;5:226-30.
5. Mathis S, Boissonnot M, Tasu J-P, Simonet C, Ciron J, Neau J-P. Intraventricular silicone oil: a case report. *Medicine (Baltimore)* 2016;95:e2359.
6. Cibis PA, Becker B, Okun E, Canaan S. The use of liquid silicone in retinal detachment surgery. *Arch Ophthalmol* 1962;68:590-99.
7. Eckardt C. Transconjunctival sutureless 23-gauge vitrectomy. *Retina* 2005;25:208-11.
8. Singh R, Kumari N, Katoch D, Sanghi G, Gupta A, Dogra MR. Outcome of 23-gauge pars plana vitrectomy for pediatric vitreoretinal conditions. *J Pediatr Ophthalmol Strabismus* 2014;51:27-31.

Episcleral racemose hemangioma

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Racemose hemangioma is a rare, benign vascular malformation. In the episclera, it appears as dilated, tortuous blood vessels that pass from the fornix over the globe surface to the limbal area, without capillary architecture, and then loop backward into the fornix.

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Fluorescein angiogram reveals the vascular malformation with rapid flow and confirms the episcleral vessels to be large in caliber and with a curvilinear, twisted configuration. We report the case of a 22-year-old man with nonhemorrhagic, coincidental racemose hemangioma of the episclera.

Case Report

A 22-year-old otherwise healthy man presented to the Wills Eye Hospital Ocular Oncology Service with a 2-year history of redness and irritation of the left eye. His symptoms were most bothersome in the morning and showed no improvement following topical antihistamines.

On examination, best-corrected visual acuity was 20/25 in each eye. The right eye was completely normal. The left eye revealed intense redness on the bulbar conjunctiva inferiorly, with dilated, curvilinear, tortuous episcleral vessels (Figure 1A). There was no evidence of inflammation, intraocular mass, retinal vascular abnormality, or hemorrhage. Anterior segment optical coherence tomography revealed no solid or cystic tumor or lymphangioma/lymphangiectasia.

Fluorescein angiography (FA) revealed vascular malformation characteristic of racemose hemangioma: rapid perfusion of convoluted blood vessels emanating from the fornix to loop at the limbus, without capillary architecture, returning to the inferior fornix (Figure 1B).^{1,2} The vascular complex comprised three separate areas of large-diameter tortuous vessels of at least 4-5 times normal caliber, looping and curvilinear within the episclera, with minimal overlying conjunctival vessels. At 30 seconds after injection, the arteriovenous complex was fully filled with fluorescein, and the late frames demonstrated no staining or leakage. Magnetic resonance imaging (MRI) of the brain and orbit revealed no abnormal findings. The final diagnosis was episcleral racemose hemangioma. Observation was advised.

Discussion

Racemose hemangiomas can appear in children; thus understanding the condition is relevant to formulating a differential diagnosis. There are several vascular tumors of the conjunctiva and episcleral/scleral region. In a comprehensive analysis of 140 consecutive cases of epibulbar vascular tumors, the final diagnoses included lymphangioma/lymphangiectasis (36%), pyogenic granuloma (22%), capillary hemangioma (14%), Kaposi sarcoma (7%), acquired sessile hemangioma (7%), racemose hemangioma (5%), varix (3%), cavernous hemangioma (2%), and glomangioma (<1%).¹ The racemose hemangiomas in that series were all of conjunctival origin; there were

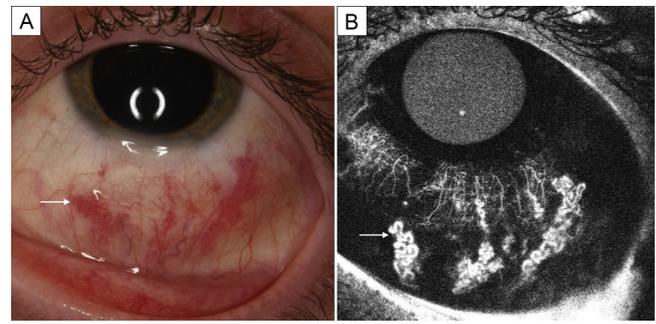


FIG 1. A, The left eye showing dilated, twisted, episcleral vessels inferiorly in three regions (medial [arrow], central, lateral), consistent with episcleral racemose hemangioma. B, Fluorescein angiography of episcleral racemose hemangioma revealing dilated, twisted vessels that pass from fornix to equator (medially [arrow]) or perilimbal region (laterally) without capillary architecture and with looping back to the fornix.

no cases of scleral or episcleral origin. A recent comprehensive analysis of 5,002 tumors of the conjunctival region yielded similar results, with the top three categories of melanocytic (52%), premalignant and malignant epithelial (14%), and lymphoid (9%) tumors.³ Vascular tumors were the 5th most common, representing 4% of cases, of which there were 4 cases (<1%) of conjunctival racemose hemangioma and none of scleral or episcleral racemose hemangioma.

There are notable differences between conjunctival and episcleral racemose hemangiomas. Conjunctival racemose hemangioma has the unique clinical feature of a nondilated vascular spiral or loop, often near the limbus and clinically asymptomatic. This lesion resides in the substantia propria and readily moves with the conjunctiva. Hemorrhage and hematoma are rare, and observation is often advised.⁴ The episcleral racemose hemangioma, by contrast, located deep within the episcleral, is prominent, dilated, twisted, and tortuous and produces mild symptoms of ocular irritation. These vascular lesions should be distinguished from enlarged sentinel vessels associated with ciliary body melanoma, hairpin-looped fine vessels of squamous neoplasia/papilloma, random fine intrinsic vessels within nevus, melanoma, and lymphoma, dilated corkscrew vessels of dural cavernous sinus fistula, and irregular ectatic vessels of lymphangioma.

To our knowledge, this is the first reported case of racemose hemangioma located in the episclera or sclera. In this case, fluorescein angiography demonstrated the vascular lesion to be comprised of deep, large diameter vessels within the episclera, and without staining or leakage of fluorescein, similar to retinal racemose hemangioma. Retinal racemose arteriovenous malformations can be associated with ipsilateral brain and intraosseous (mandibular) racemose hemangioma,

as in Wyburn-Mason syndrome.⁵ On the other hand, iris racemose hemangioma, conjunctival racemose hemangioma—and now episcleral racemose hemangioma—have not been associated with a systemic condition.⁶ In our case, there were no neurological findings, and brain MRI was unremarkable.

There are several episcleral/conjunctival vascular lesions that can resemble the episcleral racemose hemangioma including lymphangioma, varix, cavernous hemangioma, and capillary hemangioma. Conjunctival lymphangioma/lymphangiectasia presents with dilated randomly ectatic lymphatic vessels of variable diameter and with surrounding hemorrhage, occasionally with a deeper, diffuse orbital component.^{1,7,8} Conjunctival varix appears with a solitary dilation of a preexistent vein, rarely found isolated to the conjunctiva, and can undergo slow thrombosis with phlebolith formation.^{1,9} Conjunctival cavernous hemangioma occurs in the deep stroma as a red or blue multilobular mass, appearing like concord grapes, histopathologically composed of dilated congested veins separated by thick connective tissue septae.⁷ Finally, conjunctival capillary hemangioma typically presents in infancy, although acquired lesions in adults have been reported, and this lesion appears red, without hemorrhage, and vascular details are not resolvable.^{1,7}

Literature Search

PubMed was queried without date or language restriction on July 26, 2018, using the following terms: *sclera racemose hemangioma* and *episclera racemose hemangioma*.

References

1. Shields JA, Mashayekhi A, Kligman BE, et al. Vascular tumors of the conjunctiva in 140 cases. *Ophthalmology* 2011;118:1747-53.
2. Honavar S, Manjandavita F. Tumors of the ocular surface: a review. *Indian J Ophthalmol* 2015;63:187-203.
3. Shields CL, Alset AE, Boal NS, et al. Conjunctival tumors in 5002 cases: comparative analysis of benign versus malignant counterparts. *Am J Ophthalmol* 2017;173:106-33.
4. Shields CL, Shields JA. Tumors of the conjunctiva and cornea. *Surv Ophthalmol* 2004;49:3-24.
5. Shields CL, Shields JA. Phakomatoses. In: Schachat A, ed. *Ryan's Retina*. 6th ed. Philadelphia: Elsevier, Inc; 2016:2453-4.
6. Shields JA, Streicher TFE, Spirkova JHJ, et al. Arteriovenous malformation of the iris in 14 cases. *Arch Ophthalmol* 2006;124:370-75.
7. Shields JA, Shields CL. Vascular tumors and related lesions of the conjunctiva. In: Shields JA, Shields CL, eds. *EyeLid, Conjunctival, and Orbital Tumors: An Atlas and Textbook*. 3rd ed. Philadelphia, PA: Lippincott Walters Kluwer; 2016:349-66.
8. Saha K, Leatherbarrow B. Orbital lymphangiomas: a review of management strategies. *Curr Opin Ophthalmol* 2012;23:433-8.
9. Jakobiec F, Werdich X, Chodosh J, Freitag SK. An analysis of conjunctival and periocular venous malformations: clinicopathologic and immunohistochemical features with a comparison of racemose and cirroid lesions. *Surv Ophthalmol* 2014;59:236-44.

Mirror image hypermetropic anisometropia in a pair of monozygotic twins

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In monozygotic twins two embryos are formed from a single fertilized egg. In “mirror image twins” identical siblings have reverse asymmetric features in the right and left sides of the body. We report the case of twins with mirror-image hypermetropic anisometropia. They were referred to an ophthalmologist at the age of 3.5 years for amblyopia. Cycloplegic refraction of twin 1 was +1.00 in the right eye and +4.50 -0.75 ×180 in the left eye; of twin 2, +4.75 -1.00 ×180 and +1.25. Keratometry and axial length were measured with Lenstar LS 900 when the twins were 6 years of age. In twin 1, the axial length was 22.18 mm in the right eye and 20.97 in the left eye; in twin 2, 20.94 mm and 22.13. Keratometry of both eyes of these twins was relatively equal.

Case Report

Female monozygotic twins were referred to Shiraz University of Medical Sciences at the age of 3.5 years for evaluation of amblyopia. The twins shared physical characteristics (weight, height, head circumference, iris color, interpupillary distance), and their zygosity was assessed by the pediatrician and determined by questionnaire and standard genotyping. Twin 1 was left-handed and twin 2 was right-handed. Both twins underwent a complete ophthalmic examination, including visual acuity, ocular motility testing, cycloplegic refraction, and fundus examination. Cycloplegic refraction was performed 45 minutes after instillation of 2 drops of cyclopentolate 1% using the Topcon RM-800 autorefractometer (Topcon Medical System, Japan).

On examination, “mirror image” hypermetropic anisometropia was discovered. Twin 1 had anisometropia with hypermetropic astigmatism in the left eye. Her cycloplegic refraction was +1.00 in the right eye and +4.50 -0.75 ×180 in the left eye. Twin 2 had anisometropia with hypermetropic astigmatism in the right eye. Her cycloplegic refraction was +4.75 -1.00 ×180 in the right eye and +1.25 in the left eye. The best-corrected visual acuity was 20/100 in the amblyopic eyes and 20/25 in the

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