

Subtotal Excision With Adjunctive Sclerosing Therapy for the Treatment of Severe Symptomatic Orbital Lymphangiomas

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Orbital lymphangiomas are congenital malformations with abnormal and dead-end lymphatic channels and present management challenges to ophthalmologists and orbital surgeons. Recurrent hemorrhage and expansion can lead to vision loss and disfigurement. We report our technique that uses adjunctive intraoperative injection of sodium morrhuate, 5%, under direct visualization into lymphangioma channels prior to excision. We believe that in the hands of experienced orbital surgeons, and with appropriate preoperative evaluation and careful surgical technique, this procedure is useful in saving vision and avoiding complications from orbital lymphangiomas. *Arch Ophthalmol.* 2011;129(8):1073-1076

Orbital lymphangiomas represent a challenge to ophthalmologists and orbital surgeons. Lymphangiomas are congenital malformations with abnormal and dead-end lymphatic channels that change size with infection/inflammation. Bleeding into channels from feeder capillary networks causes "chocolate cysts." These malformations are integrated within surrounding tissues, so complete excision can cause collateral damage that is unacceptable within the orbit. Hence, observation and intermittent debulking are the standard of care, with persistent risks of disfigurement, pain, diplopia, and vision loss.^{1,2}

Ideal treatment of lymphangiomas entails removing abnormal tissues while preserving surrounding normal structures. For deep orbital lesions, sclerosing agents have been shunned because of tissue sensitivity and restricted orbital confines. However, several studies have used superficial sclerosing treatments, including a study showing involution of superficial lymphangiomas with sodium morrhuate.³ An important caveat is to exclude lymphangiomas that coexist with venous or arteriovenous malformations.^{4,5} In these cases, intravascular injection of sclerosing agents could damage normal vessels.

Hence, hemodynamic assessment of orbital vascular lesions is necessary before treatment of deep lymphangiomas.

We report our technique that uses adjunctive intraoperative injection of sodium morrhuate, 5%, under direct visualization into lymphangioma channels prior to excision. Sodium morrhuate is a sclerosing agent with prothrombotic properties. Its use improves hemostasis and causes intralésional sclerosis. Contrast-enhanced dynamic magnetic resonance angiography (MRA) is used to assess blood flow preoperatively to avoid direct intravascular injection.⁶ Because injected tissues are partially excised shortly after injection, our technique reduces deep orbital sclerosis and inflammation. Furthermore, the sclerosing agent remains in residual lymphangioma tissue, causing further sclerosis and involution. This leads to maximal debulking of severely symptomatic lymphangiomas that are refractory to traditional treatments. With appropriate preoperative evaluation and careful surgical technique, this procedure is useful in saving vision and avoiding severe complications from extreme cases of lymphangiomas.

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METHODS

A retrospective medical record review was performed for 2 patients with severe sympto-

matic orbital lymphangiomas who were initially evaluated for vision loss. Preoperative and postoperative photographs and examination measurements were compared.

In both cases, preoperative contrast-enhanced dynamic MRA (4-dimensional time-resolved MRA with keyhole, ie, 4D-TRAK; Philips Medical Systems, Best, the Netherlands) was assessed for vascular communication with surrounding circulations. These patients underwent a combined procedure of surgical debulking and adjunctive injection of the sclerosing agent.

The surgical approach was individualized based on tumor size and location. Patient 1 underwent anterior orbitotomy and transcranial orbitotomy through bicoronal incision and frontal craniotomy. The superior rim and orbital roof were removed for access to the superior and posterior orbit. Patient 2 underwent an anterior orbitotomy via transconjunctival approach.

In the orbit, blunt dissection was used to define the lymphangioma extent, which in case 1 was enmeshed in orbital tissues and in case 2, filled the orbit around the eye. Incremental volumes of 0.2 mL of sodium morrhuate were injected beneath the tumor surface, turning the surface dark from clotting. Injected tissues were bluntly and sharply excised. Cycles of dissection, injection, and excision were repeated until the majority of tumor bulk was removed. Total volume of injected sodium morrhuate was less than 5 mL. Hemostasis was maintained with judicious use of bipolar electrocautery.

In patient 1, the orbital rim was repositioned and fixated with titanium plates and screws. The craniotomy flap and scalp incisions were closed using standard neurosurgical techniques. In patient 2, the fornix incisions were closed in standard fashion.

RESULTS

Patient 1 was a 12-year-old girl whose right orbital lesion was biopsied several years earlier by a different surgeon, confirming clinical suspicion of lymphangioma. On presentation, she had almost daily bleeding episodes and she complained of decreased vision, persistent and worsening diplopia, and globe dystopia. Her best-corrected visual acuity (VA) at presentation was 20/60 OD and 20/20 OS. Exophthalmometry revealed 6.5 mm of exophthalmos in the right eye and

a large area of periocular hemorrhagic chemosis (Figure 1A). She was unable to perform visual field testing. There was clinical documentation of lesion progression. Imaging revealed a large lesion with fluid-filled cysts that infiltrated the orbit (Figure 1C and D). Contrast-enhanced dynamic MRA revealed no communication between the lymphangioma and orbitocranial vasculature (data not shown). The patient underwent transcranial orbitotomy to maximize exposure, facilitate injection of sodium morrhuate, and reduce risk of hemorrhage (Figure 1F-H). Pathology was consistent with lymphangioma. Her postoperative course was excellent, and her VA improved over 4 weeks. She continued to complain of double vision and displayed exotropia, despite improvement of proptosis and globe dystopia (Figure 1B). Postoperative computed tomographic scan was obtained at 6 months in preparation for strabismus surgery (Figure 1E). However, her family moved out of the area and she was lost to follow-up. Her final examination revealed VAs of 20/30 OD and 20/20 OS, and exophthalmometry showed 0.5 mm of right-sided proptosis.

Patient 2 was a 31-year-old woman. Her right orbital lymphangioma began growing around menarche, and she underwent multiple excisions over 20 years. Her most recent surgical attempt consisted of 3-wall orbital decompression with bone removal. The lesion continued to grow and at her first visit with us, her VA in the right eye was reduced with a relative afferent pupillary defect. Importantly, the left eye was amblyopic, and she was debilitated from vision loss in her better eye (Figure 2A-C). Her initial best-corrected VAs were 20/100 OU and exophthalmometry revealed 15 to 20 mm of right-sided proptosis. Orbital imaging with magnetic resonance imaging and computed tomography revealed an infiltrating lymphangioma with fibrosis and calcification (Figure 2G-H). Examination revealed right optic disc elevation, retinal striae, and macular folds. Humphrey visual field test results over 3 visits showed an enlarging paracentral scotoma. Because of le-

sion size, progression of symptoms, and gross deformity, she proceeded with surgical excision with adjunctive sodium morrhuate injection. Preoperative dynamic MRA revealed no communication between the lymphangioma and orbitocranial vasculature (data not shown). She underwent an anterior orbitotomy with excision of the infiltrative lesion (Figure 2I). Adjunctive sodium morrhuate injection was used to reduce the risk of hemorrhage and sclerose elements of the lesion left behind in the deep orbit. Her postoperative course was excellent with improvement of vision and afferent pupillary defect. Because of the absence of a bony orbital floor from prior decompression surgery, she developed globe dystopia (Figure 2D-F), but this was mild and no further intervention was recommended. On her most recent examination at 1 year postsurgery, her VAs were 20/60 OD and 20/100 OS. Exophthalmometry showed 3 mm of right-sided exophthalmos. Her fundus examination revealed mild pigmentary changes but was otherwise unremarkable.

COMMENT

Orbital lymphangiomas represent a unique treatment challenge. These infiltrative lesions are prone to bleeding and can result in disfigurement, vision loss, and diplopia. Complete surgical excision is typically unattainable without collateral damage to surrounding structures.

The propensity of lymphangiomas to bleed seems counterintuitive, since they contain dead-end lymphatics with no vascular connection. However, capillary networks, which feed the lesion, can break and bleed into empty lymphatic channels because of expansion and contraction of the lymphangioma. Blood can accumulate, forming "chocolate cysts" or leading to channel rupture with external bleeding. With bleeding episodes, scar tissue forms, causing further enlargement and infiltration.

Thus, lymphangiomas are considered incurable and not amenable to complete surgical excision. Interest in sclerosing therapy has a long history, and several scler-

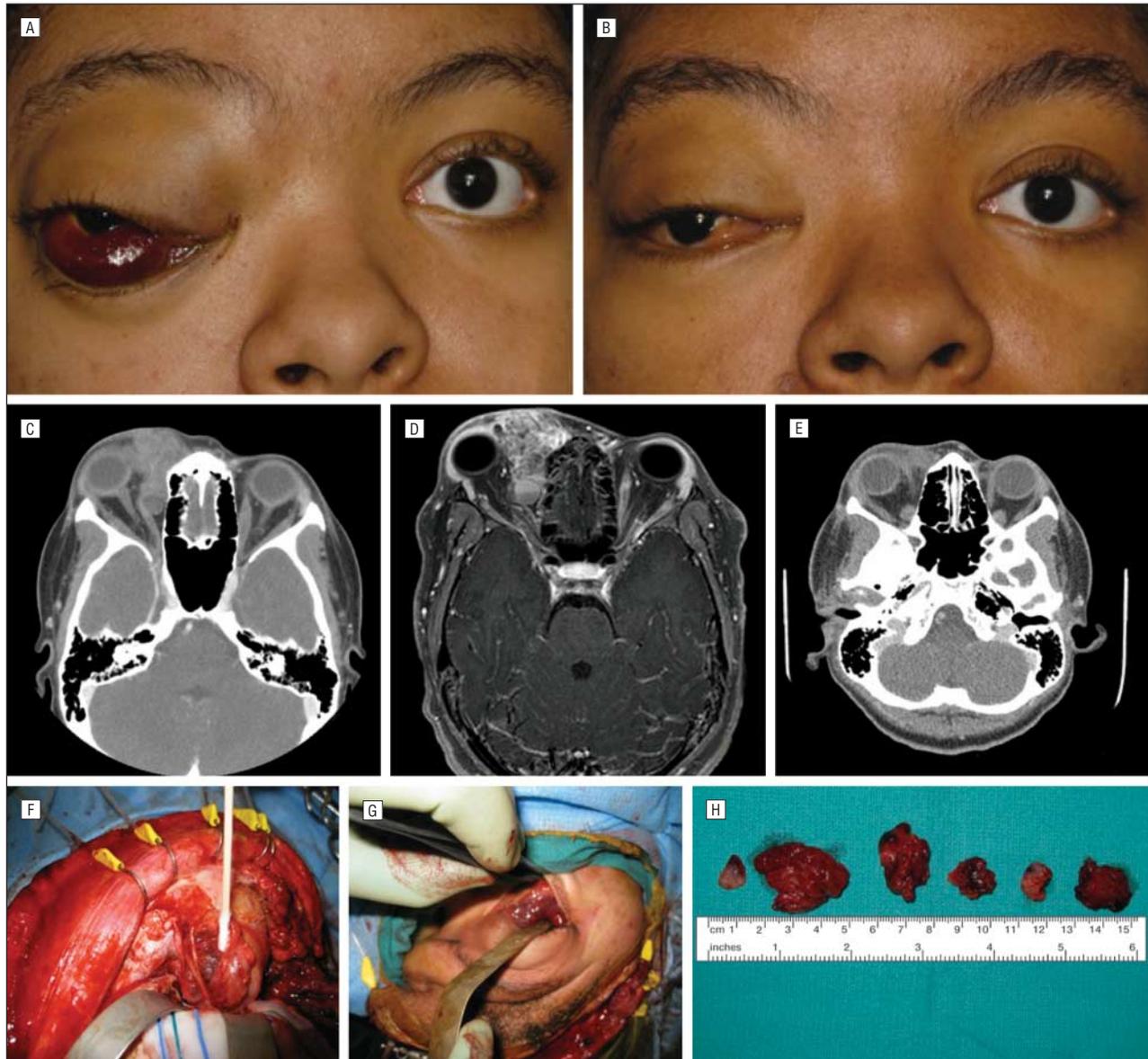


Figure 1. Patient 1 had decreased vision and orbitofacial deformity, with recurrent orbital hemorrhages (A). Computed tomography and magnetic resonance imaging revealed a large, infiltrative lymphangioma (C-E). Orbitocranial surgical debulking with sodium morrhuate injection was performed (F-H), resulting in significant improvement in both vision and deformity (B). A craniofacial surgical approach allowed optimal surgical exposure; note the transcranial posterior view (F) and anterior orbital view (G).

rosing agents have been studied throughout the world. Sodium morrhuate, 5% formulation, is available in the United States and approved by the Food and Drug Administration for sclerosing therapy of simple varicose veins with competent valves. Sodium morrhuate is made from sodium salts of saturated and unsaturated fatty acids from saponified cod liver oil. The injected solution has a pH of approximately 9.5 and causes fibrosis and vascular obliteration by intimal inflammation and thrombus formation (www.rxlist.com/morrhuate-sodium-drug.htm).

Schwarz and colleagues³ limited use of sodium morrhuate to superficial orbital lymphangiomas because of the potential for severe deep orbital inflammation and fibrosis. Our approach builds on their innovation by combining sclerotherapy under direct visualization with surgical excision of accessible tissue. A major benefit is reduced bleeding through direct thrombosis. Furthermore, lymphangioma remnants contain residual sclerosing agent, which causes fibrosis and involution. Because lymphangiomas lack communication with surrounding tissues, residual sclerosing agent remains

within the injected lesion and mostly spares normal tissues. Indeed, our patients did not experience prolonged orbital inflammation post-operatively.

Because injecting sclerosing agents into the orbitocranial circulation can have devastating effects, it is important to confirm that the lymphangioma is isolated, rather than mixed, such as a varix-lymphangioma.^{4,5} We achieved this using contrast-enhanced dynamic MRA, which is a new imaging modality that captures contrast flow with rapid-acquisition magnetic resonance imaging while maintain-

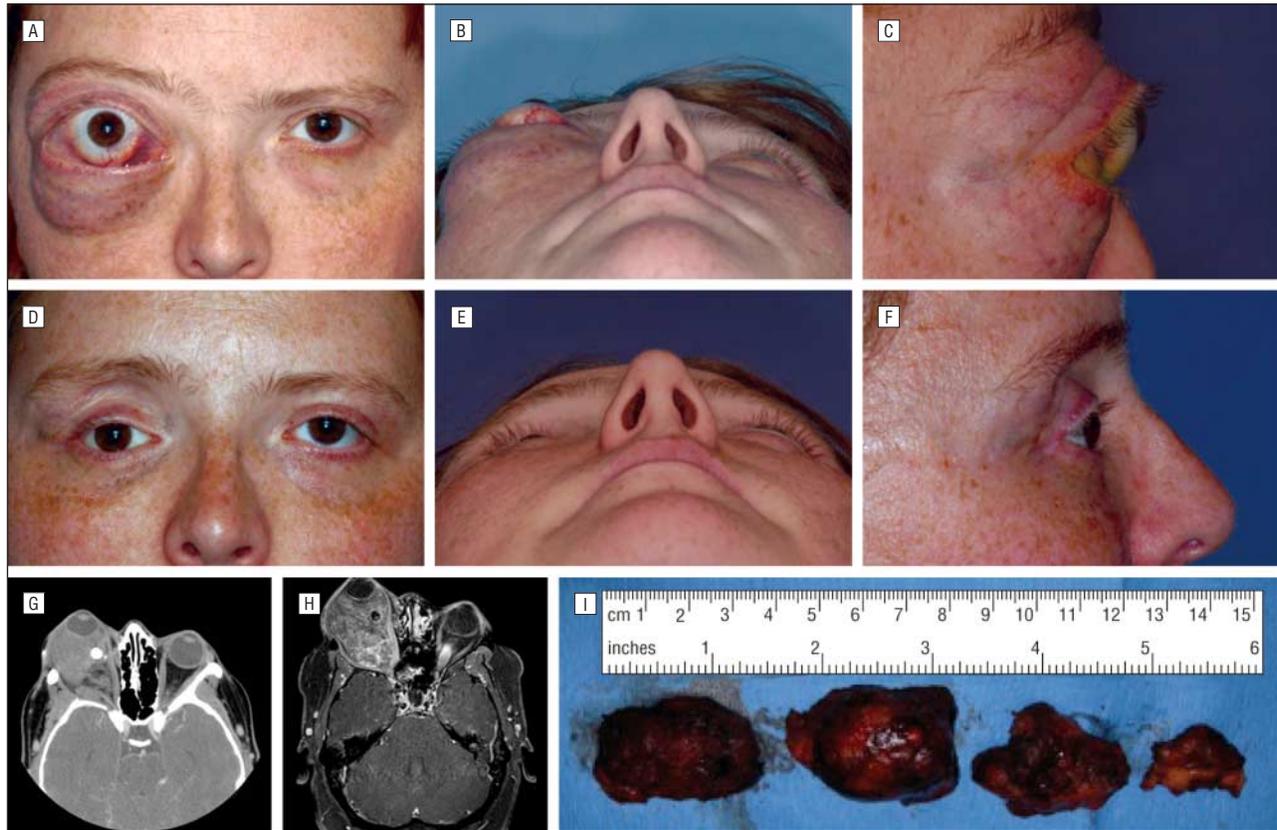


Figure 2. Patient 2 had long-standing severe orbital lymphangioma with persistent, progressive vision loss (A-C). Imaging revealed an infiltrative right orbital lymphangioma and signs of prior orbital decompression surgery (G-H). Surgical debulking (I) with adjunctive sodium morrhuate injection was performed, and 1 year postoperatively, the patient's condition was greatly improved (D-F).

ing excellent soft tissue resolution. The combination of vascular and soft tissue imaging is helpful in surgical planning. For patient 1, we chose a transcranial approach because of the extent of the lymphangioma. For patient 2, an anterior approach was chosen because her prior orbital decompression surgery provided ample space.

In conclusion, severe symptomatic orbital lymphangiomas that cannot be controlled by other means, and do not communicate with surrounding vasculature, may be candidates for subtotal excision with adjunctive sclerosing therapy using sodium morrhuate, 5%. Excellent knowledge of surgical anatomy is required, as is experience with deep orbital surgery and its potential complications. In experienced hands, this

procedure can preserve visual function. Additional research is needed to assess long-term results as well as applicability to less severe lymphangioma lesions.

Submitted for Publication: July 27, 2010; final revision received November 8, 2010; accepted November 9, 2010.

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Financial Disclosure: None reported.

Additional Contributions: We acknowledge generous support from a Research to Prevent Blindness Career Development Award (Dr Kahana) and a Knights Templar Eye Foundation award (Dr Bohnsack).

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