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.

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METHODS

A retrospective medical record review was performed for 2 patients with severe symptomatic orbital lymphangiomas. 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 intralesional sclerosis. Contrast-enhanced dynamic magnetic resonance angiography (MRA) is used to assess blood flow preoperatively to avoid direct intravascular injection. 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.
atic 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 fixed 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.

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 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 periorcular 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 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 staining of proptosis. Exophthalmometry showed 3 mm of right-sided exophthalmos. Her fundus examination revealed mild pigmentary changes but was otherwise unremarkable.

**RESULTS**

**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 scle-
Sclerosing 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 colleagues’ 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 postoperatively.

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 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).
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.

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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).