Chapter 8
Low-Flow Vascular Malformations of the Orbit: Evaluation and Management

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8.1 Introduction

Orbital lymphatic and venous malformations are low-flow vascular anomalies that commonly occur in the head and neck region [1, 2]. In most studies, they represent less than 2 % of all orbital and periorbital lesions, occurring most commonly in the first decade of life [1, 2]. A significant number of patients with these malformations present with intralesional bleeding or infection [3, 4]. Without appropriate therapy, approximately 40 % of patients develop reduced vision and 7 % develop blindness [3].

8.2 Clinical Presentation

Periorbital and orbital swelling is the most common clinical presentation of orbital venous and lymphatic malformations [4]. Intralesional bleeding or infection can result in sudden periorbital swelling and proptosis with or without pain (Fig. 8.1a). Lesions that extend anteriorly under the conjunctiva can present with mild to severe chemosis. Orbital involvement may also cause binocular diplopia that is not always appreciated due to associated mechanical ptosis. Another important sequel of mechanical ptosis in early childhood is the development of amblyopia. In addition, exposure keratitis and corneal erosion are other causes of vision loss in these patients [3, 5].

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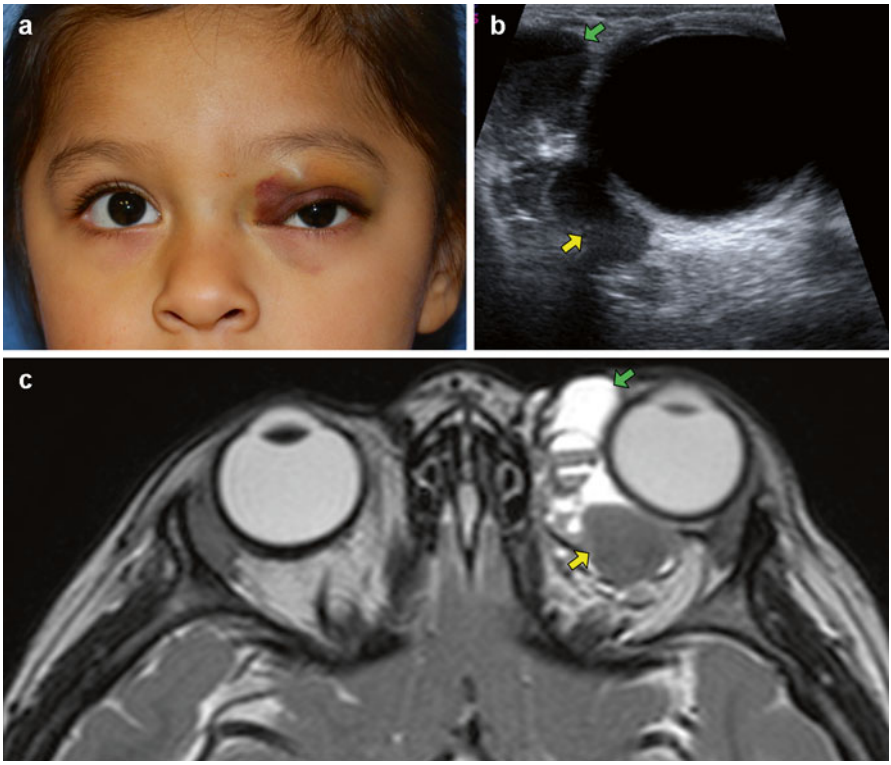

Fig. 8.1 A 3-year-old girl presenting with acute painful proptosis and ecchymosis with normal visual acuity and IOP. (**a**) Clinical photograph showing left eyelid ecchymosis and periorbital swelling. (**b**, **c**) Ultrasound and axial T2W MRI of the left orbit showing intraconal (*yellow arrow*) and pre-septal (*green arrow*) lymphatic microcysts with fluid-fluid levels suggestive of internal hemorrhage

8.3 Evaluation

8.3.1 Clinical

Complete ophthalmic examination including clinical photographs, visual acuity, pupillary response, extra-ocular motility, slit-lamp examination, dilated funduscopic examination, and intraocular pressure (IOP) measurements is performed.

8.3.2 Imaging

The anatomical distribution and characterization of the malformations are evaluated using pre- and post-contrast MRI of the face and orbits, with T2-weighted imaging being most valuable. Ultrasonography is performed by the interventional radiologist for treatment planning (Fig. 8.1).

8.4 Management

Current options for management of low-flow orbital vascular malformations include observation, sclerotherapy, and surgical debulking with partial to total excision. Our institutional protocol for evaluation and management of low-flow orbital vascular malformations is summarized in Fig. 8.2.

Incidentally detected asymptomatic malformations can be managed conservatively, but malformations that present with acute proptosis and compressive optic neuropathy secondary to intralesional bleeding usually require emergent management by image-guided lymphatic cyst aspiration or surgical decompression. Malformations that present with repeated intralesional hemorrhage, infection, chronic proptosis, retro-orbital discomfort, ophthalmoplegia, headaches, or visual impairment may be managed by either surgical excision [5] or sclerotherapy [6–10].

8.4.1 Surgical Therapy

Low-flow orbital malformations are often very infiltrative, and total excision is virtually impossible because of the associated morbidity [11]. However, well-delineated extraconal lesions have been resected with low complication and recurrence rates [12]. In some long-term studies, however, these malformations have a tendency to recur with recurrence rates ranging between 58 % at a mean of 3.4 years [13] and 71 % at a mean of 7.2 years [5]. Therefore, less invasive techniques such as sclerotherapy have emerged as a treatment option.

8.4.2 Sclerotherapy

Sclerotherapy is a well-established management option for low-flow orbital vascular malformations [6–10]. The primary challenge for sclerotherapy is safe access into the retrobulbar space. The secondary challenge is management of orbital compartment syndrome that can occur from the inflammatory response post sclerotherapy [14]. Sclerotherapy requires multidisciplinary management involving ophthalmology, oculoplastic surgery, interventional radiology, and pulmonology services.

8.4.3 Sclerotherapy Agents

A variety of agents used for sclerotherapy including their advantages and disadvantages have been described in the literature [15]. At our institution, we use bleomycin for lymphatic malformations and a sequential injection of 3 % sodium tetradecyl

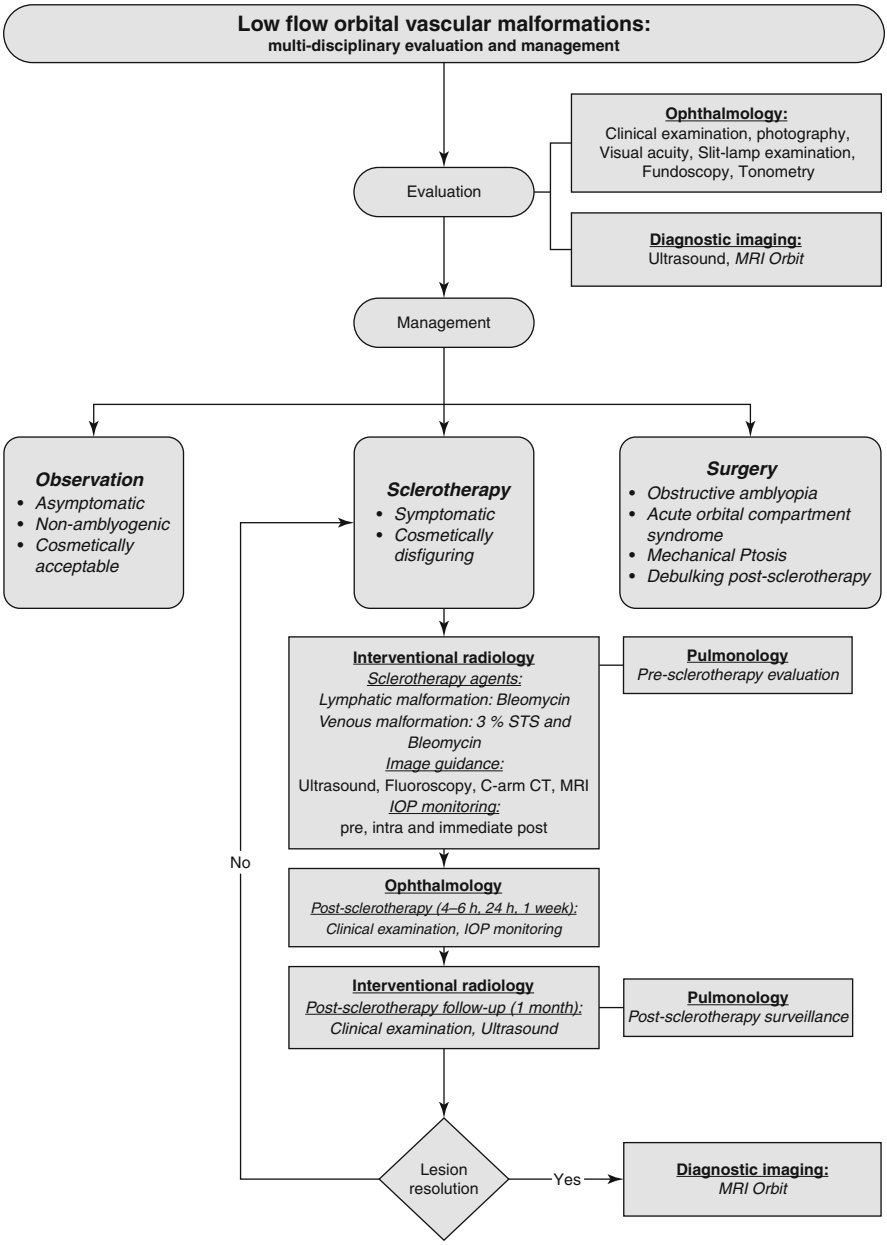

Fig. 8.2 Algorithm for evaluation and management of low-flow orbital vascular malformations

sulfate (STS) and bleomycin for venous malformations. The maximum bleomycin dose per session is 0.5 units/kg for <1-year-old and 15 units for >1-year-old [16].

Cumulative toxicity of systemic bleomycin during treatment of malignancy has been associated with late-onset pulmonary fibrosis [17]. Although no case of pulmonary fibrosis secondary to intralesional injection of bleomycin has been reported thus far, a pulmonary fibrosis surveillance program is followed at our institution. Children older than 7 years undergo exercise tolerance test and pulmonary function tests including spirometry, plethysmography, and diffusion capacity of carbon monoxide. Children younger than 7 years old are screened using exercise tolerance, and chest imaging is performed only if necessary. The clinical and functional pulmonary evaluations are repeated at 6 and 12 months post sclerotherapy and yearly thereafter for 10 years.

Skin pigmentation after intralesional bleomycin has been reported in a few cases [14, 18, 19]. The following precautions are followed to reduce the risk of skin trauma and pigmentation secondary to bleomycin: (i) avoiding tape application to the skin; (ii) using alcohol-free skin barrier film prior to tape application; (iii) securing the endotracheal tube without tape by using a Dale tube holder, a Thomas tube holder, or silk sutures; and (iii) retaining of EKG pads for 48 hours.

8.4.4 Sclerotherapy Technique

The procedure is performed under general anesthesia with oral or nasal intubation. The intraocular pressure (IOP) is measured bilaterally using applanation tonometry, and a corneal protector is placed over the eye.

Lymphatic malformations are accessed under ultrasound guidance using 20G, 21G, and 25G needles for cysts <2 cm and 5 or 7 F pigtail catheters for cysts >2 cm (Figs. 8.3 and 8.4). Whenever ultrasound imaging is limited,

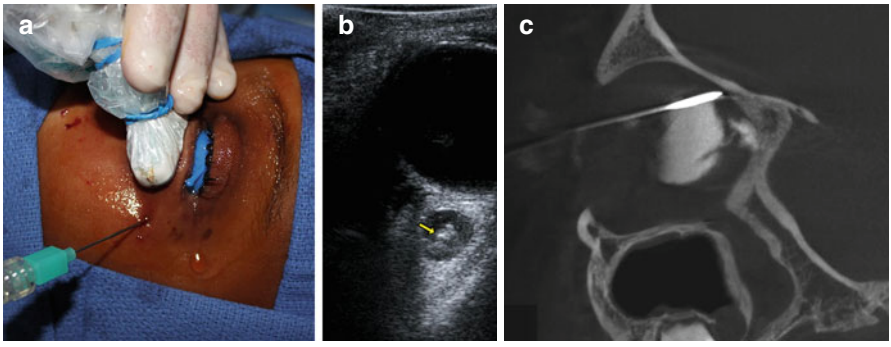

Fig. 8.3 Image-guided access of lymphatic malformation. (**a**, **b**) Ultrasound-guided needle access of retrobulbar lymphatic microcyst (*yellow arrow - needle tip*). (**c**) C-arm cone-beam CT image-guided access of retrobulbar lymphatic macrocyst and visualization of bleomycin-contrast mixture within the cyst

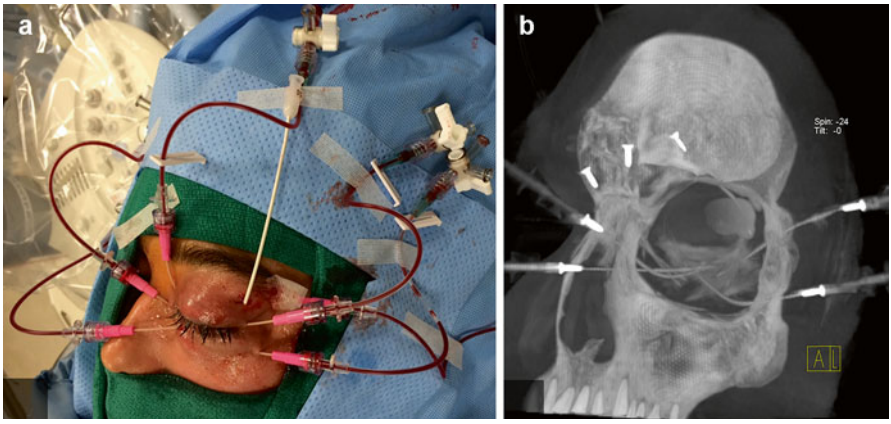

Fig. 8.4 An 18-year-old boy with infiltrative mixed macro-microcystic lymphatic malformation. (**a**) Lesion access using multiple 20G angiocatheters and a pigtail catheter. (**b**) 3D reconstructed C-arm cone-beam CT image showing distribution of bleomycin-contrast mixture within the malformation

particularly in the retrobulbar space, additional intra-procedural C-arm cone-beam CT guidance is utilized to supplement ultrasound. Cysts are aspirated whenever possible prior to injection of bleomycin mixed with 10 % contrast (Omnipaque 300 mg/ml) in a concentration of 1–3 units/ml. The total intra-orbital sclerosant volume is restricted to 5 ml to reduce the risk of rise in IOP post sclerotherapy.

Injection of bleomycin is monitored using visualization of moving microbubbles under ultrasound and 2D fluoroscopy (Fig. 8.5a, b). The detailed anatomical distribution of bleomycin in relationship to critical orbital structures is further evaluated using intra-procedural C-arm cone-beam CT [20] (Fig. 8.5c, d). Image fusion of C-arm cone-beam CT images with pre-procedural T2W MRI is performed to evaluate any untreated areas of the malformation (Fig. 8.5e, f). These areas are accessed and treated in the same or separate session.

Venous malformations are accessed using a similar technique followed by evaluation of their venous drainage using digital subtraction roadmap venography. Then, they are treated with a sequential injection of 3 % sodium tetradecyl sulfate foam followed by bleomycin.

8.4.5 Intraocular Pressure Monitoring

IOP is monitored at the end of the procedure while the patient is still intubated. Ophthalmic examination is performed 4–6 h and 24 h following the procedure. The patients are discharged from the hospital if no concerning findings are identified.

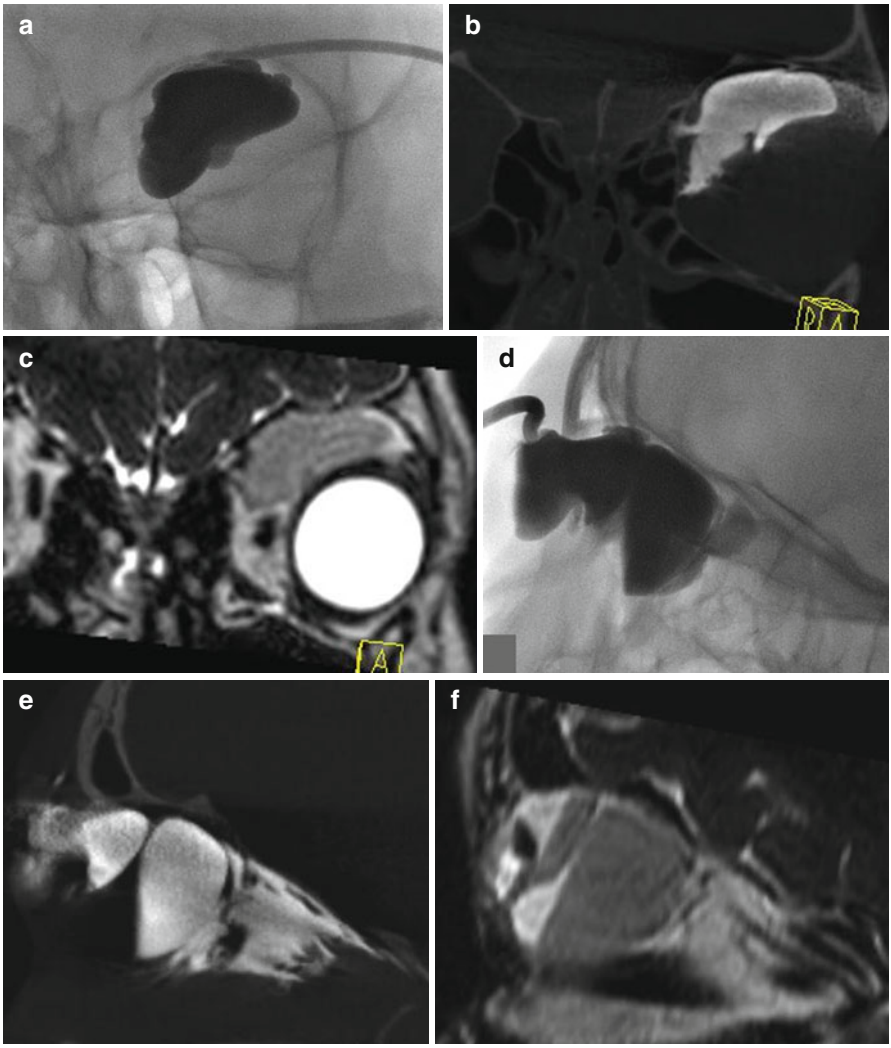

Fig. 8.5 A 13-year-old boy with mixed macro-microcystic lymphatic malformation of the left orbit. (**a**, **b**) Frontal and lateral 2D fluoroscopic images showing distribution of bleomycin-contrast mixture within the malformation. (**c**–**f**) Co-registered images showing good correlation between bleomycin distribution in C-arm CT (**c** coronal, **d** sagittal) and lesion distribution in pre-sclerotherapy MRI (**e** coronal, **f** sagittal)

8.4.6 Management of Post-sclerotherapy Complications

Fever up to 101 F within 24 hours post sclerotherapy and lasting for 24–48 hours is a systemic response to bleomycin and does not require additional investigation or treatment.

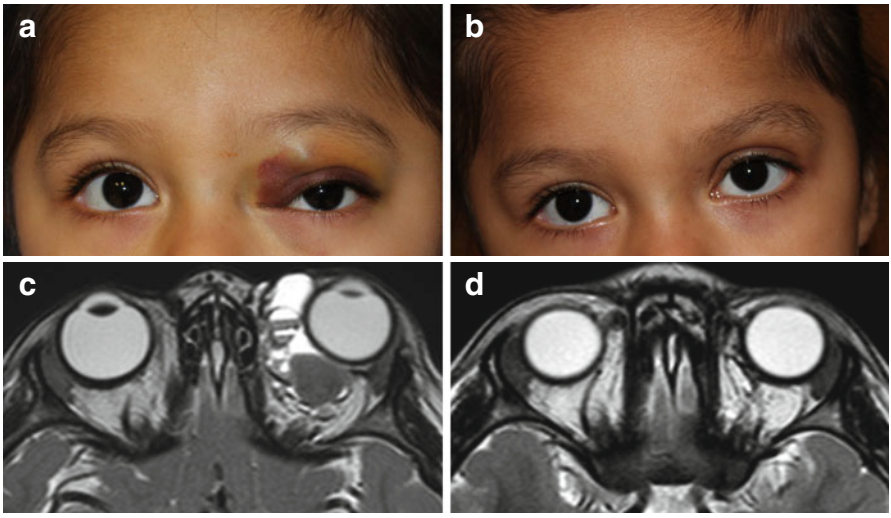

Fig. 8.6 A 3-year-old girl with microcystic lymphatic malformation of the left orbit: comparison of pre- and post-sclerotherapy images. (**a**, **b**) Clinical photographs showing resolution of proptosis and ecchymosis. (**c**, **d**) Axial T2W MRI showing complete resolution of malformation after one sclerotherapy session

One of the most problematic complications of sclerotherapy is elevated IOP resulting in orbital compartment syndrome [14, 18, 21, 22]. Our protocol for management of raised IOP is as follows: A rise in IOP above 35 is managed with topical beta-blockers and alpha-2 agonists. If the IOP rises above 40, systemic steroids are added to reduce swelling secondary to inflammation. In addition, ultrasound or CT imaging is performed to diagnose intralesional bleeding as a cause of raised IOP that can be managed by ultrasound-guided drainage. Surgical decompression is rarely necessary with appropriate monitoring and management of raised IOP.

8.4.7 Follow-Up

Ophthalmic examination is repeated at 1 week post procedure and interventional radiology evaluation including ultrasound examination at 1 month post procedure. If necessary, further sclerotherapy sessions are performed at 6-week intervals. MRI examination is performed at 3 months after the conclusion of sclerotherapy. Post-sclerotherapy MR images are co-registered with pre-sclerotherapy MR images to correlate imaging outcome with clinical resolution (Figs. 8.6 and 8.7).

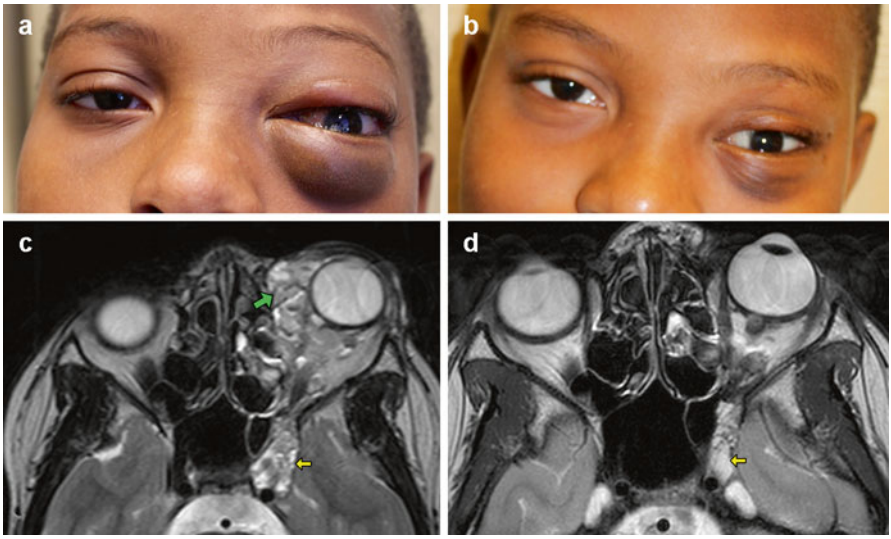

Fig. 8.7 A 9-year-old boy with microcystic lymphatic malformation of left orbit. (**a**, **b**) Clinical photographs showing resolution of proptosis, chemosis, and ecchymosis. Note post-sclerotherapy hyperpigmentation of the lower eyelid secondary to bleomycin. (**c**, **d**) Axial T2W MRI showing complete resolution of intra-orbital malformation (*green arrow*) and partial resolution of malformation within the wall of cavernous sinus (*yellow arrow*) after two sclerotherapy sessions

8.5 Conclusion

Low-flow orbital vascular malformations manifest as cosmetically disfiguring lesions associated with amblyopia, diplopia, ptosis, and even loss of vision. Many of these lesions, particularly those located in the intraconal and extraconal orbital space, are very difficult to treat surgically and often recur. Sclerotherapy has evolved as a minimally invasive treatment option for these malformations with good clinical outcome. The best management of these uncommon, yet vision-threatening lesions is achieved with a multidisciplinary collaborative team effort.

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