Vascular Malformations of the Orbit

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Introduction

Orbital vascular malformations, like vascular malformations elsewhere in the body, can be categorized by vessel and flow type: arteriovenous malformation, venous malformation, and lymphatic malformation. Together they comprise approximately 15 % of all orbital lesions [1]. The appearance and behavior differ among these categories, from bright red to deep blue, from well-demarcated to infiltrative into orbital and periorbital tissues. Each has different clinical presentation, imaging characteristics, natural history, genetic etiology, and management strategies [2]. Clinical impact can vary greatly even within a single histologic category, and diagnosis is not always straightforward. Therefore, thorough clinical evaluation and specific ancillary testing are essential in making the diagnosis and eventually guiding the therapeutic alternatives. Current approaches range from serial observation to radical resection with many intermediate alternatives.

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Clinical Evaluation

Patients with orbital vascular malformations come to clinical attention for a variety of reasons. Among these, exophthalmos, pain, and diplopia are most common. Patients presenting with these symptoms must undergo a thorough orbital and ophthalmic history and examination. Every patient should have a complete eye exam in order to document the type of the tumor and determine its clinical impact.

Careful history taking is one of the most important tools available to the ophthalmic clinician. Chief complaint typically includes periocular pain or retro-orbital headache, consistent or intermittent visual loss, double vision, or disfigurement. The duration, timing, and character of the complaint should be determined: Is the pain mild or severe? Is it intermittent or does it occur continuously? Is it exacerbated by supine or inverted positioning, physical activity, or extraocular motility? If proptosis exists, is it sudden onset or slowly progressive? Is it associated with pain? Is the proptosis exacerbated by Valsalva, sneezing or cough, physical activity, or inversion? Does the lesion change with ambient temperature or menstruation? On presentation is redness of the eye present? Does the patient hear a bruit or internal pulsations? All these questions

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aid the clinician in determining the type of lesion and indications for intervention.

Comprehensive orbital evaluation begins with visual acuity, but there are four critical parameters of optic nerve health that should always be measured whenever an orbital process is suspected: visual acuity, pupil function, color vision, and visual field. Pupils are inspected with the swinging flashlight for a relative afferent pupillary defect. Color vision is tested using standardized color plates, and visual field is inspected for scotomas using automated (static) or manual (dynamic) testing devices. Color vision is felt to be the most sensitive indicator of optic nerve compromise, while visual field assessment has greater capacity to indicate the nature of the optic nerve insult. In the absence of formal visual field testing, confrontational visual fields can help detect an optic neuropathy caused by orbital tumor. Pupillary function is an easy test to perform that provides general insight into optic nerve conditions. Other important parameters in cases of suspected orbital disease include extraocular motility, exophthalmometry, diplopia mapping, and intraocular pressure (a proxy measurement of orbital pressure). Clinical photos are strongly recommended to document appearance at presentation and for future comparison.

The eye examination should consist of the best corrected visual acuity and intraocular pressure detected by applanation tonometry (normal pressure raging between 9 and 21 mmHg). Slit lamp examination is used to inspect the conjunctiva for any dilated blood vessels, tortuosity, and chemosis (conjunctival swelling). "Corkscrew" vessels are indicative of carotid-cavernous fistula.

Inspection of the eyelids includes measuring the height of the palpebral fissure (distance between the upper and lower lids), the margin to corneal light reflex distance, and binocular comparison for asymmetry; orbital tumors can produce either eyelid ptosis or eyelid retraction. Exophthalmometry quantifies protrusion of the eyeball. Measurement above 21 mm or more than 2 mm difference between the eyes is considered abnormal. Ocular motility should be evaluated in all gazes: up, down, right, and left. One common scale is the 5-point scale with 4 indicating full motility in that direction and 0 indicating no movement at all. Extraocular motility will be affected depending on the specific location of the lesion within the orbit. Ocular motility disturbance can produce diplopia in various gazes or in primary position (looking straight ahead). Dilated fundus exam is another important component of the examination, allowing direct inspection of the retina and optic nerve. Pupils can be dilated safely using phenylephrine 2.5 % or tropicamide 1 %. The optic nerve head should be examined to evaluate its margins. Hazy borders, swelling, or hyperemia indicate acute or subacute processes; optic nerve pallor or atrophy indicates chronic disease. Choroidal folds or retinal striae indicate the presence of a space-occupying lesion in the orbit or posterior compression of the eyeball.

When a vascular lesion is suspected within the orbit, it is essential to inspect for pulsatile proptosis or the presence of a thrill. This can be done by simple auscultation. Examining the patient in sitting position and then asking her/him to cough or bend can help to highlight venous lesions that are sensitive to changes in venous pressure.

Important ancillary tests include ultrasonography, computed tomography (CT), magnetic resonance imaging (MRI), CT angiogram, MR angiogram, and dynamic CT. Each of the three major orbital vascular malformations has its own clinical characteristics, imaging modalities, and pathognomonic findings.

Arteriovenous Malformation (AVM)

Blood flows directly from the arterial circulation into the venous one without passing through intervening capillaries. Intraorbital AVMs are congenital anomalies with slow growth patterns [3]. They are supplied by branches of both the external and internal carotid artery. AVMs of the orbit can result from trauma or can appear spontaneously.

Diagnosis

Patients with orbital AVMs typically present with pulsatile proptosis. When the lesion is anterior in

the orbit, a blue pulsating subcutaneous mass can be seen on physical exam (Fig. 42.1a, b). Other signs include epibulbar congestion and audible bruit.

Nonenhanced CT is helpful in visualizing the small foci of calcification often associated with arteriovenous malformations and foci of hemorrhage if any exist. Contrast-enhanced CT reveals enhancement of the vascular channels, whereas MRI demonstrates these lesions as vascular channels that appear as flow voids (Fig. 42.1c, d). More adequate evaluation is usually obtained by a magnetic resonance angiography or CT angiography, but the gold standard remains traditional angiography [4] (Fig. 42.1e).

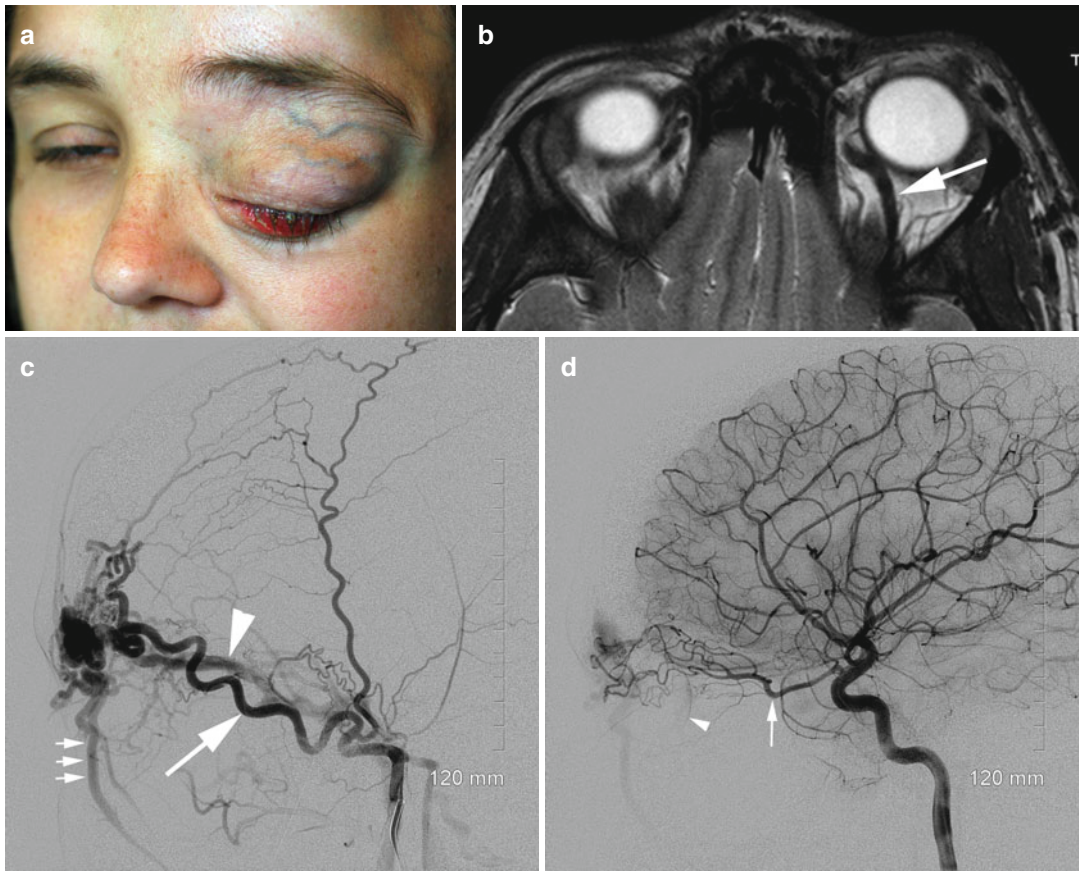


Fig. 42.1 Arteriovenous malformation of the orbit and eyelids. (**a**) Clinical photo demonstrates anterior displacement of the eyeball (proptosis), with upper eyelid ptosis and conjunctival swelling (chemosis). Arterialized veins can be seen in the eyelid. (**b**) Axial views of this T2-weighted MRI in a different patient with orbital AVM demonstrates multiple serpiginous flow voids in the preseptal soft tissues of the left orbit with a dilated left superior ophthalmic vein (*arrow*). (**c**) Left distal EXTERNAL carotid artery angiogram shows two branches of the superficial temporal artery, one of which is supplying blood to the AVM (*arrow*). Venous drainage can be seen posteriorly along the superior ophthalmic vein (*arrow head*) and inferiorly along the facial vein (*small arrows*). (**d**) Left INTERNAL carotid artery angiogram shows AVM with blood supply from supraorbital and ethmoidal branches of the ophthalmic artery (*arrow*). Choroidal blush can be seen identifying the outline of the retina (*arrow head*). (**e**) Left EXTERNAL carotid artery angiogram post-embolization shows complete elimination of the malformation with preservation of all the normal arteries including the maxillary branch (*arrow*) of the external carotid and two major branches of the superficial temporal artery (*arrow heads*). (**f**) Left INTERNAL carotid artery angiogram post-embolization. The supply to the AVM from the ophthalmic artery (*arrow*) has been completely eliminated. Choroidal blush remains (*arrow head*), indicating retained blood supply to the retina

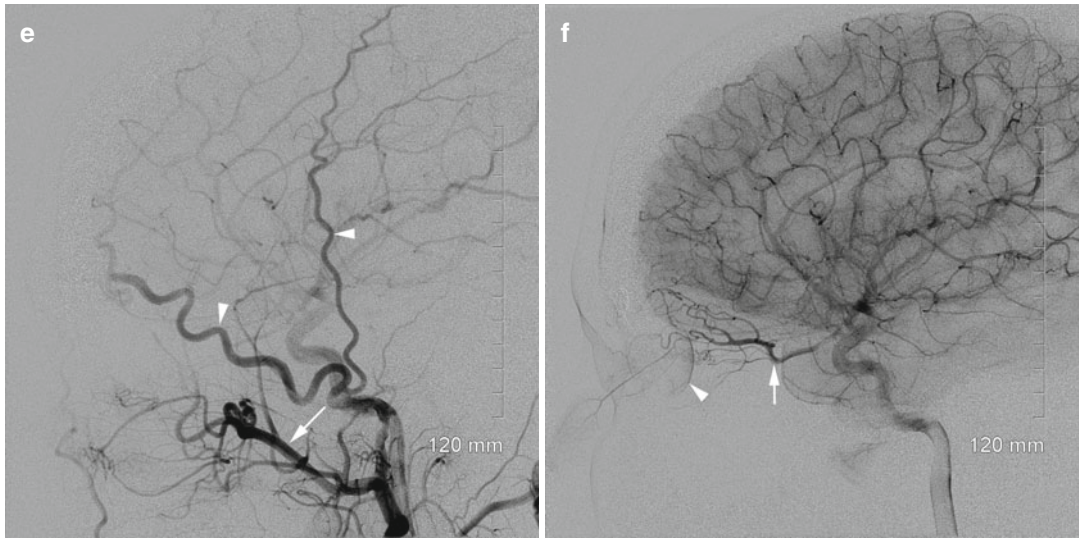


Fig. 42.1 (continued)

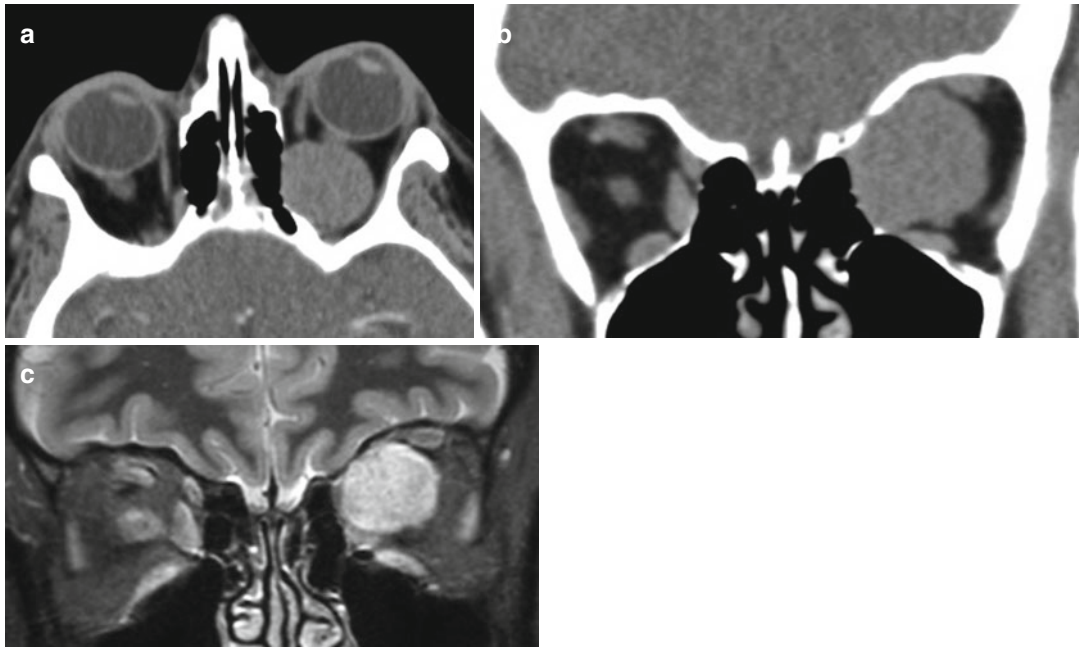
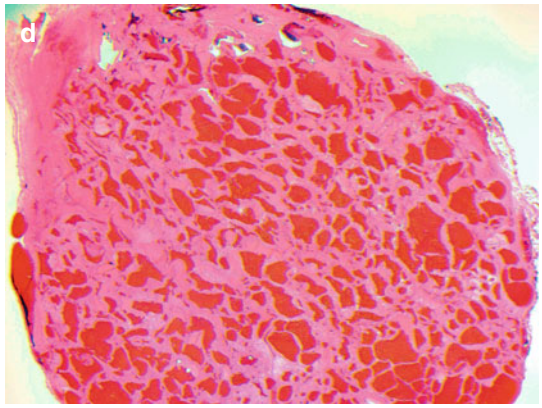


Fig. 42.2 Encapsulated venous malformation of the orbit. (**a**) Axial view of computed tomograph (CT) of the orbit without contrast demonstrates an ovoid, clearly delimited lesion of approximately 2 cm in the intraconal space. (**b**) Coronal view of the same CT shows the lesion abutting the medial wall and roof of the orbit in the superomedial quadrant of the orbit. (**c**) This coronal view of this T1-weighted, gadolinium-enhanced, fat-suppressed MRI demonstrates numerous septations within the lesion but fails to highlight the relationship of the lesion to the thin bones of the orbit. Additional findings include displacement of the extraocular muscles and optic nerve sheath complex. (**d**) Histopathologic section of the lesion above demonstrates the general architecture of the lesion with numerous thick fibrous septa (hematoxylin and eosin, 20×). (**e**) Higher magnification view demonstrates large vascular spaces filled with erythrocytes, with flattened endothelial cells (hematoxylin and eosin 100×). D2–40 stain for lymphatic endothelial cells was negative (not shown)



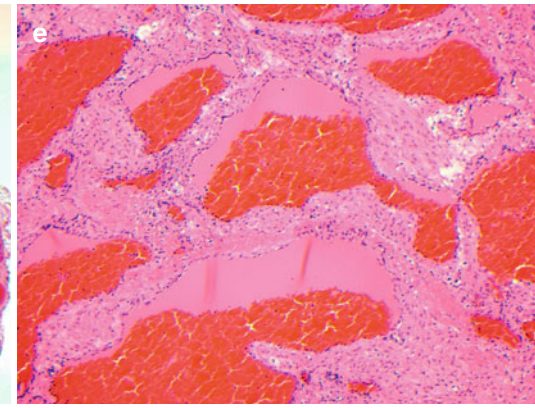


Fig. 42.2 (continued)

Treatment

Angiographic embolization can occlude the nidus and major bleeding vessels in an orbital AVM [4, 5] (Fig. 42.1f). The most serious risk of this treatment is ophthalmic artery or central retinal artery occlusion and is stratified according to specific location of the AVM. The effect of embolization is often temporary, however. Surgical resection, in contrast, can be definitive. Without embolization, surgical resection is possible but difficult due to the risk of massive bleeding. A combination of embolization and surgical excision offers better control and ultimately an optimal outcome. Gamma knife radiotherapy, according to Liscak et al., resulted in obliteration of AVMs in 74 % of cases after the first cycle and 69 % after the second [6].

Carotid-Cavernous Fistula

A carotid-cavernous fistula is an abnormal shunt between the carotid artery and cavernous sinus, or a directly related ophthalmic vein. These fistulae are sometimes categorized as arteriovenous malformations in the ophthalmic literature.

Direct carotid-cavernous sinus fistulae can be caused by trauma or spontaneous rupture of an aneurysm of the intracavernous carotid or an atherosclerotic artery [7]. Indirect carotid-cavernous sinus fistulae are characterized by arterial blood that flows through the meningeal branches of the internal or the external carotid arteries indirectly into the cavernous sinus [7].

Diagnosis

Carotid-cavernous sinus fistulae usually present with epibulbar injection, pulsatile proptosis, and a bruit or thrill. Blepharoptosis and hemorrhagic chemosis can also be seen. The patient can experience limitations of extraocular movements and complain of decrease in vision, headache, and eyelid swelling. The presence of increased intraocular pressure is typically caused by elevation in episcleral venous pressure, limiting outflow of aqueous humor. Anterior segment ischemia, characterized by corneal edema, iris atrophy, and rubeosis iridis, can be seen. Retinal hemorrhages are possible.

CT and MRI can detect a prominence of the superior ophthalmic vein. An arterial angiography helps in making the definitive diagnosis.

Treatment

In most cases, observation allows spontaneous resolution of the fistula. Selective embolization to occlude the fistula is usually done if visual impairment becomes progressive or in case of severe proptosis or if the patient is complaining of intolerable headache or bruit [8].

Venous Malformations

Venous malformations (VM) can be encapsulated or distensible. The former have been incorrectly called "cavernous hemangiomas," while the latter are sometimes referred to as "varices."

Encapsulated venous malformations are the most common orbital lesions found in adults. They are benign, slow-growing vascular lesions of the orbit [9].

Diagnosis

Patients with encapsulated venous malformations typically present with axial proptosis described as painless, slowly progressive protrusion of the eye associated with mild eyelid swelling. Axial proptosis (direct forward displacement of the eyeball) is produced most commonly, as these lesions tend to be found in the intraconal space defined by the four rectus muscles. Less commonly, the encapsulated venous malformation is seen extraconally, in which case the eyeball may be displaced vertically or horizontally. With direct observation alone, this can be difficult to distinguish from strabismus, a condition in which the eye is *rotated* vertically or horizontally. Alternate cover testing reveals the difference; in the strabismic condition, the uncovered eye rapidly rotates back into primary fixation, whereas the eye does not move back from horizontal or vertical translation. When located close to the orbital apex, encapsulated venous malformations can cause compressive optic neuropathy and decreased visual acuity. Diplopia, when present, is explained by distortion or distension of the extraocular muscles rather than by direct intramuscular involvement [1, 10]. Differential diagnosis usually includes schwannoma because of the similarity in size, shape, and location [11].

As with all orbital conditions, imaging studies are critically important. First, an orbital ultrasound, with the advantage of being a noninvasive technique, can detect a uniformly high-echogenic lesion with well-defined borders. A color Doppler US helps in detecting the blood flow and mapping the vasculature. Computed tomography should be performed with contrast. It shows an oval- or round-shaped, sharply demarcated homogenous lesion with slow contrast enhancement. On MRI, the lesion is seen isointense on T1 and hyperintense on T2; few internal septa can be seen, with a progressive accumulation of the contrast on late-phase images [12] (Fig. 42.2a).

Treatment

The treatment of orbital venous malformations is mostly conservative, but an intervention is needed in case of high orbital pressure or unbearable pain, deep orbital hemorrhage causing drop in vision, and cosmetic deformity [13].

In the cases in which surgical excision is indicated, the approach is dictated by the location of the lesion within the orbit. Most cavernous hemangiomas are found within the intraconal space between the extraocular muscles and the optic nerve. When the lesion is located lateral to the optic nerve, a lateral approach is used, either with or without temporary removal of the orbital rim. Inferior or inferomedial lesions can be removed endoscopically [14]. The medial intraconal space can also be accessed transconjunctivally, sometimes requiring temporary disinsertion of the medical rectus; occasionally the lateral orbital rim is removed to allow retraction of the eyeball during medial tumor removal. Inferiorly located lesions can be approached with a subciliary incision or transconjunctivally, while superiorly located lesions can be removed via an eyelid incision. Superior-posterior lesions may require craniorbitotomy for complete removal (Fig. 42.2b).

Distensible Venous Malformations

In contrast to encapsulated venous malformations, distensible venous malformations are meandering and poorly demarcated. They are more likely located outside the muscle cone and also tend to involve the eyelids. These lesions are not encapsulated and do not respect anatomic boundaries. Whereas the encapsulated lesions are almost always seen in isolation, the distensible

lesions are more commonly associated with venous malformations of the hemiface including scalp, airway, buccal space, and masseter. These lesions have been described as a "bag of grapes" [1]. The most striking feature of distensible venous malformations, a feature starkly contrasted in encapsulated venous malformations, is a dramatic volumetric response to changes in venous pressure.

Diagnosis

Patients with distensible venous malformations of the orbit tend to complain of intermittent orbital pain, intermittent exophthalmos, or enophthalmos (sunken eye). The clinical scenario is dictated by the degree to which native orbital fat has been replaced by the malformation. When present, the pain is associated with physical activity and concomitant with exophthalmic episodes. One imagines that the pain results from rapid physical distortion of orbital soft tissues and venous congestion that raises the pressure in the orbit. Alternatively, direct neural tension or compression may be the painful stimulus. Importantly, when present, the exophthalmos is non-pulsatile and not associated with bruit. It is easily demonstrated with inversion, Valsalva maneuver, or other methods of raising venous pressure (Fig. 42.2a,b).

The patient experiences this change during athletic or vocational activities that may bring the lesion to medical attention. In cases with eyelid involvement, coughing or other raised venous pressure produces unsightly, externally visible irregularities of the lid structure and veins. The conjunctiva may house significant extensions of the primary lesion or may occur in complete isolation from an orbital venous malformation. Alternatively, conjunctival vessels may be secondarily enlarged and tortuous.

While most cases can be diagnosed clinically, imaging studies are important in order to define which anatomical structures are directly or indirectly involved, the specific anatomic location of the lesion, and to determine treatment alternatives planning. Orbital ultrasound can be extremely useful in detecting and measuring orbital masses. In the case of vascular lesions, ultrasound and color Doppler imaging can detect not only location of the lesion but also blood flow. In venous malformations, reversal of flow can be seen during Valsalva maneuver. (No such flow can be identified in cases of encapsulated venous malformations.) On computed tomography, distensible venous malformations may be entirely invisible, especially when the scan is performed in the usual supine position. In many cases an indistinct, small lesion may be visible with contrast-enhanced imaging. If the scan is performed in the prone position, or if the venous pressure is artificially raised during scanning, the lesion typically becomes obvious and potentially enormous within and around the orbit. Phleboliths within the lesion are pathognomonic. Any imaging of the orbit (computed tomography or nuclear magnetic resonance) must provide thin, 1–3 mm slices, with direct views in the axial and coronal planes. Sagittal views can also be helpful in some cases. While computed tomography is unsurpassed for anatomic location and, therefore, surgical planning, magnetic resonance imaging is preferred to identify tissue types and lesion characteristics. Magnetic resonance imaging, however, reveals the bony orbit only as unimaged space, and phleboliths can appear as flow voids. Magnetic resonance images should be ordered with gadolinium enhancement, and as in computed tomography, the venous pressure must be raised. The venous malformation appears with intermediate signal intensity on T1 and high signal intensity on T2, and it enhances strongly after the administration of a contrast material (Fig. 42.2c,d). This lesion, which intermingles with orbital fat, can be seen more easily with T1 fat suppression.

Treatment

Treatment of orbital venous malformations ranges from observation to complete surgical extirpation. Superficial lesions that involve primarily the eyelid can be treated with YAG laser, sclerotherapy, or surgical excision. Surgery is made easier

if the lesion is pretreated with endovascular embolism. Laser treatment of the conjunctival component must be undertaken with extreme caution; the epibulbar conjunctiva should not be treated with laser. In many cases, surgical removal is more straightforward, safer, and definitive. Indeed, small conjunctival lesions can be cauterized using bipolar electrocautery.

More significant, space-occupying orbital lesions require greater treatment planning. In cases where exophthalmos is the chief complaint, a two-stage approach is needed. Endovascular sclerotherapy has become an excellent first-stage treatment to eliminate the mass of the malformation. Bleomycin is a safe and effective agent. Critical to successful treatment is containment of the sclerosant away from the cavernous sinus, where most of these lesions drain. Successful first-stage treatment is likely to produce enophthalmos, which requires surgical augmentation with soft tissue or alloplastic implants.

Surgical excision of orbital venous malformation alleviates any risk related to sclerosing agents but often still requires a two-stage approach. Excision of untreated lesions risks truly significant orbital hemorrhage that is difficult to control without risking injury to the optic nerve or other neural structures transiting the orbital apex. Therefore, preoperative endovascular embolization is preferred. These lesions can be embolized percutaneously or directly with surgical exposure of the lesion. Thrombin-gelatin slurries have been effective agents. The lesion is effectively thrombosed, making surgical removal technically simpler and safer. Bipolar electrocautery is indispensible during these orbital surgeries.

Lymphatic Malformations

Until recently, lymphatic malformations have been called "lymphangiomas" in the ophthalmic literature. These lesions represent between 1 and 8 % of all orbital lesions [15]. Periocular lymphatic malformations can involve the orbit, eyelids, and conjunctiva. They can be seen in isolation or associated with hemifacial lymphatic malformations. Another typical distribution involves the anterolateral scalp down to the superior orbit and eyelid. Within the orbit, these lesions do not respect tissue planes and tend to involve both intraconal and extraconal spaces [16, 17]. Rapidly expanding cystic segments due to hemorrhage have been called "chocolate cysts" and produce acute-onset exophthalmos, pain, and optic neuropathy.

Diagnosis

Lymphatic malformations of the orbit most commonly present in children ages birth to puberty. They may include visible components in the eyelid or conjunctiva. Diagnosis in these cases may be straightforward. Conjunctival involvement, however, can mimic the "salmon patch" typical of conjunctival lymphoma. Confusion can usually be resolved, however, by patient age; lymphoma is very rare in children, and new onset lymphatic malformation is exceedingly rare in adults. Anterior lesions appear as several soft bluish masses in the upper nasal quadrant associated with a cystic conjunctival element. Eyelid involvement can mimic venous malformation especially when the blue hue arises from intracystic hemorrhage. Deeper lesions, without a superficial manifestation, may be obvious at birth or may remain quiescent for years then suddenly become symptomatic, particularly at the onset of puberty [15]. Posterior orbital lesions usually induce slowly progressive proptosis of the eye, but sudden painful proptosis can be caused by spontaneous hemorrhage (Fig. 42.3a,b). Subacute presentation is seen in cases of upper respiratory infection or other immunologic stimuli. Rhabdomyosarcoma must be excluded with any rapidly evolving orbital lesion in the pediatric age group [18].

Ultrasound evaluation of orbital lymphatic malformations demonstrates large cystic areas in macrocystic areas and irregular lesions in microcystic regions. Flow is absent in these lesions, making color Doppler less useful. Computed tomography is the favored imaging technique in most orbital pathology, as it demonstrates the relationship of the lesion to the surrounding bony orbit and specific anatomic location of the lesion that is critical in surgical planning. Magnetic

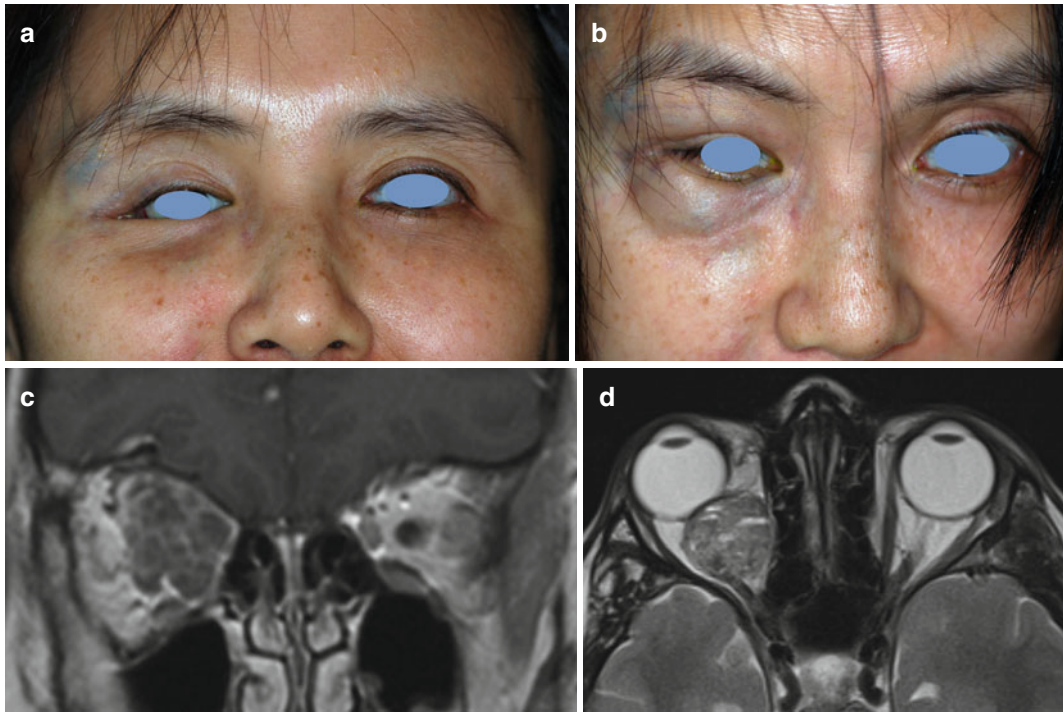


Fig. 42.3 Distensible venous malformation. (**a**) With the patient in the upright position, the right eyeball appears sunken (enophthalmos), and *blue discoloration* can be seen in the upper lateral eyelid. The lower lid shows incipient entropion that often accompanies severe enophthalmos. (**b**) With the patient in the inverted position, the lower eyelid component of the lesion is appreciated, the eyeball has moved forward, and the position of the lower eyelid has stabilized against the eyeball. The venous nature of the lesion can be appreciated in the increase blue discoloration of the upper eyelid. (**c**) This T1-weighted MRI in the coronal plain demonstrates the irregular nature of the lesion, with irregular septa and cystic spaces extending from the intraconal to the extraconal space. (**d**) The T2-weighted axial view demonstrates more discrete boarders with irregular internal cavities in the medial, retrobulbar orbit

resonance, however, is the gold standard for imaging orbital lymphatic malformations, as the tissue type, fluid type (e.g., lymph, new blood, old blood), and nature of the cysts are most exquisitely demonstrated with this technology. Hemorrhagic cysts appear hyperintense on T1-weighted images (Fig. 42.3c).

Treatment

Surgical excision of extensive orbital lymphatic malformations has been fraught with difficulty; the lesions tend to hemorrhage due to delicate vessels in the cyst walls, and complete extirpation is frequently impossible because of the meandering and stealthy projections of lesion well beyond the visible borders. "Recurrence" is better described as "continued proliferation" and may be anticipated in cases of incomplete resection [19] (Fig. 42.3d). One area in which surgical resection continues to be the optimal treatment is the conjunctival component. Quadrantic resection allows adequate healing and may involve amniotic reconstruction of the ocular surface [20].

More recently, endovascular sclerotherapy has become the treatment of choice for extensive or surgically inaccessible orbital lymphatic malformations. These lesions are extremely sensitive to bleomycin treatment. OK-432 is strongly discouraged due to uncontrolled inflammation and swelling that can produce an orbital compartment syndrome [21]. In this case, massive pressure develops from inflammatory swelling, with subsequent compression or stretching of the optic nerve. In the emergent situation, lateral canthotomy

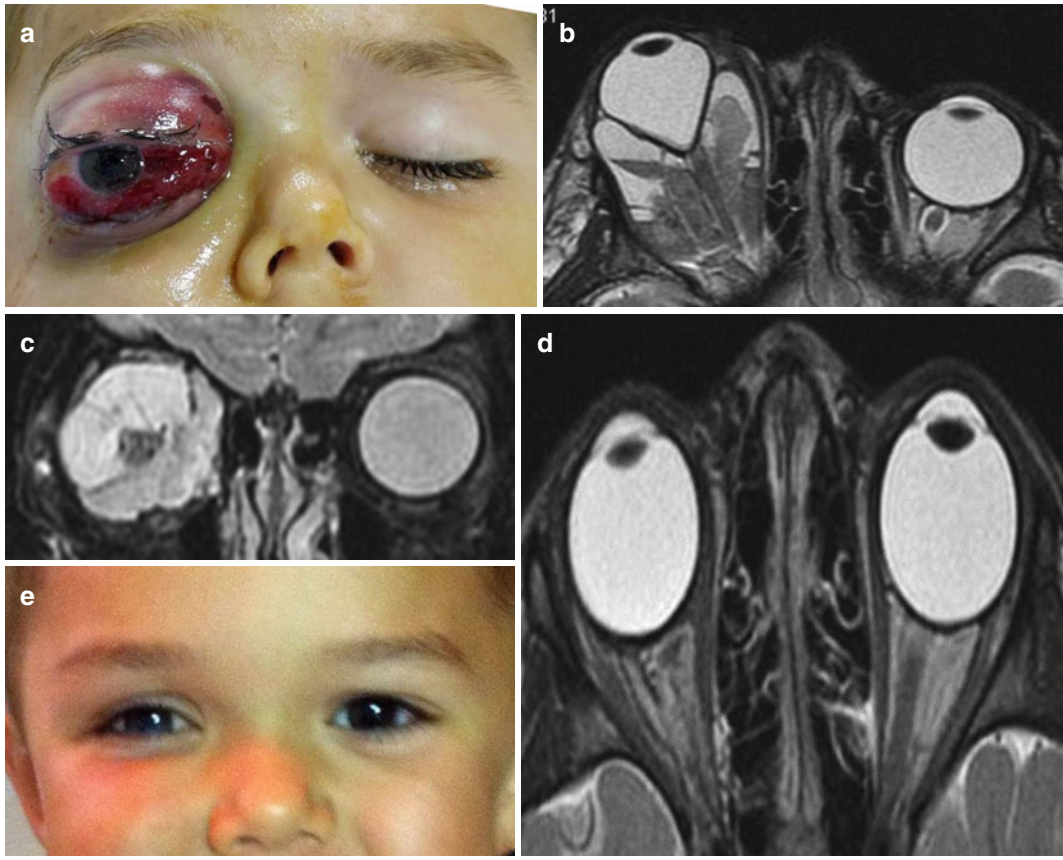


Fig. 42.4 Lymphatic malformation of the eyelids and orbit. (**a**) Severe anterior displacement of the eyeball (proptosis) of the right eye with exposed, hemorrhagic conjunctiva. (**b**) Axial views of this gadolinium-enhanced, T2-weighted MRI with fat suppression demonstrate massive proptosis of the eyeball with dramatic deformity of the globe and stretching of the optic nerve. The intra- and extraconal spaces are occupied with a multicystic lesion demonstrating characteristic fluid levels. (**c**) Coronal view of the same MRI demonstrates the lesion completely filling the orbit. (**d**) Axial view of this posttreatment, T2-weighted MRI demonstrates nearly complete resolution of the lesion. (**e**) One year after initial treatment, the eyeball has returned to normal position

and cantholysis can be vision-saving maneuvers. Alcohol sclerosants have reportedly been used effectively, but these agents are neurotoxic and also produce inflammation that is difficult to control [16] (Fig. 42.4).

Conclusion

Orbital vascular malformations including arteriovenous malformation, venous malformation, and lymphatic malformation demonstrate a wide spectrum of clinical presentation. The most urgent concern is protection of the optic nerve and visual acuity, followed by preservation of periocular functions such as eyeball and eyelid movements. Depending on the type and location of the lesion, and guided by the specific ancillary tests, optimal management can be achieved.

References

1. Rodgers R, Grove SA (2000) Vascular lesions of the orbit. In: Albert D, Jakobiec F, Azar D et al (eds) Principles and practice of ophthalmology, vol 4, 2nd edn. WB Saunders Company, Philadelphia, pp 3144–3154
2. Yadav P, De Castro DK, Waner M, Meyer L, Fay A (2013) Vascular anomalies of the head and neck: a review of genetics. Semin Ophthalmol 28(5–6):257–266
3. Kaufman Y, Cole P, Dauser R et al (2007) Intraorbital arteriovenous malformation: issues in surgical management. J Craniofac Surg 18:1091–1093

4. Hayes BH, Shore JW, Westfall CT et al (1995) Management of orbital and periorbital arteriovenous malformations. Ophthalmic Surg 26(2):145–152
5. Dmytriw AA, Ter Brugge KG, Krings T et al (2014) Endovascular treatment of head and neck arteriovenous malformations. Neuroradiology 56(3):227–236
6. Liscak R, Vladyka V, Simonova G et al (2007) Arteriovenous malformations after Leskell gamma knife radiosurgery: rate of obliteration and complications. Neurosurgery 60:1005–1014
7. Kanski J (2007) Orbit. In: Kanski J (ed) Clinical ophthalmology: a systemic approach, 6th edn. Butterworth-Heinemann/Elsevier, Edinburgh;/New York, pp 180–185
8. Williamson R, Ducruet A, Crowley W et al (2012) Transvenous coil embolization of an intraorbital arteriovenous fistula: case report and review of the literature. Neurosurgery 72:130–134
9. Osaki TH, Jakobiec FA, Mendoza PR, Lee Y, Fay AM (2013) Immunohistochemical investigations of orbital infantile hemangiomas and adult encapsulated cavernous venous lesions (malformation versus hemangioma). Ophthal Plast Reconstr Surg 29(3):183–195
10. Jakobiec FA, Zakka FR, Papakostas TD, Fay A (2012) Angiomyofibroma of the orbit: a hybrid of vascular leiomyoma and cavernous hemangioma. Ophthal Plast Reconstr Surg 28(6):438–445
11. Andreoli CM, Hatton M, Semple JP, Soukiasian SH, Fay AM (2004) Perilimbal conjunctival schwannoma. Arch Ophthalmol 122(3):388–389
12. Lewin JS (2004) Low-flow vascular malformations of the orbit: a new approach to a therapeutic dilemma. AJNR Am J Neuroradiol 25(10):1633–1634
13. Rootman J (1988) Diseases of the orbit: a multidisciplinary approach. JB Lippincott, Philadelphia, pp 553–557
14. Chhabra N, Wu AW, Fay A, Metson R (2014) Endoscopic resection of orbital hemangiomas. Int Forum Allergy Rhinol 4(3):251–5
15. Wiegand S, Eivazi B, Bloch L et al (2013) Lymphatic malformations of the orbit. Clin Experim Otorhinolaryngol 6:30–35
16. Illif WJ, Green WR (1979) Orbital lymphangiomas. Ophthalmology 86(5):914–929
17. Vavvas D, Fay A, Watkins L (2004) Two cases of orbital lymphangioma associated with vascular abnormalities of the retina and iris. Ophthalmology 111(1):189–192
18. Fay A, Fynn-Thompson N, Ebb D (2003) Klippel-Trénaunay syndrome and rhabdomyosarcoma in a 3-year-old. Arch Ophthalmol 121(5):727–729
19. Eivazi B, Ardelean M, Baumier W et al (2009) Update on hemangiomas and vascular malformations of the head and neck. Eur Arch Otorhinolaryngol 266(2):187–197
20. Mehta M, Waner M, Fay A (2009) Amniotic membrane grafting in the management of conjunctival vascular malformations. Ophthal Plast Reconstr Surg 25(5):371–375
21. Suzuki Y, Obana A, Gohto Y et al (2000) Management of orbital lymphangioma using intralesional injection of OK-432. Br J Ophthalmol 84(6):614–617