*Case Report*

# Ocular Arteriovenous Malformation Manifesting as Proptosis

**Urgent message:** Proptosis may be caused by a variety of vascular, infectious, endocrine, and neoplastic diseases that may threaten vision or cause serious systemic complications or death. Providers should be familiar with causes and work-up, including which findings suggest a need for immediate imaging and consultation.

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## Introduction

This report describes a case of unilateral proptosis due to an intraorbital arteriovenous malformation. Proptosis can be caused by a variety of vascular, infectious, endocrine, and neoplastic diseases that may threaten vision or cause serious systemic complications or death. Accurate diagnosis and timely and appropriate referral from the urgent care setting can prevent these complications. Thus, it is important for urgent care providers to understand the disease processes that can present with proptosis and include them in the differential diagnosis when evaluating eye problems.

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## Case Presentation

A 32-year-old man presented to an urgent care clinic with a 5-year history of right-eye pain and gradual

swelling. There had been no inciting trauma, fever, or hyperthyroid symptoms. He had no blurry or double vision. There was no family history of ocular problems.

### *Physical Examination*

On initial presentation, the patient's vital signs were as follows:

- Temperature: 36.5°C
- Blood pressure: 134/90 mm Hg
- Pulse: 83 beats/min
- Respiratory rate: 19 breaths/min
- Oxygen saturation: 99% on room air

On physical examination, the patient appeared to be well. He demonstrated right-eye injection, proptosis, and upper-lid swelling. His pupils were equally round and reactive to light; extraocular movements were intact. His dentition was unremarkable, and his sinuses were not tender. His visual acuity was 20/20 bilaterally without correction.

### *Imaging*

The patient was referred for magnetic resonance imaging (MRI).

### *Diagnosis*

The diagnosis was arteriovenous malformation (AVM), as seen in **Figures 1–5**, rather than carotid-cavernous fistula.

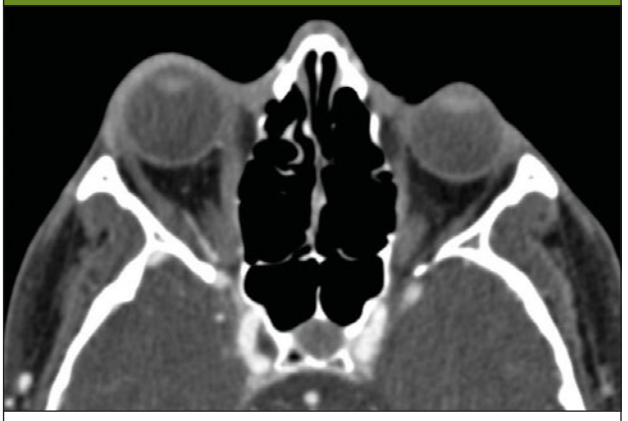
## Discussion

Proptosis is an uncommon presentation or examination finding, with an incidence of 16 cases/100,000 persons per year among women and 2.9 cases/100,000 persons per year among men.[1] Proptosis can be subtle, and thus it may go unnoticed by the patient if it develops gradually. In the evaluation of eye presentations, especially with concerning features such as diplopia, vision loss, and restriction of extraocular movements, proptosis is an important physical examination finding that can help direct the work-up. Proptosis is easily missed, but looking for it can lead to timely imaging and treatment before the underlying disease irreversibly damages the patient's eyesight.

### *Anatomy*

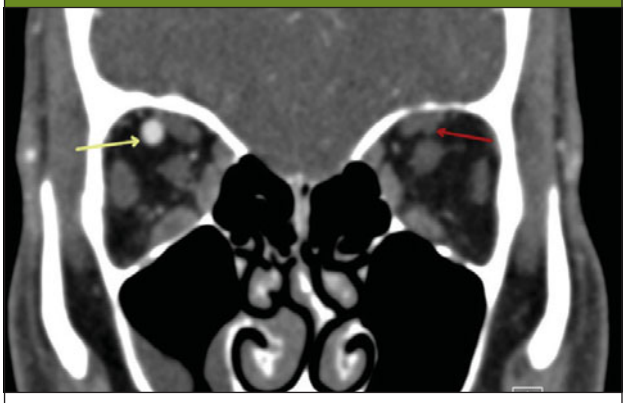
Blood flow to the eye is supplied by the internal carotid artery, which gives rise to the ophthalmic artery, which then branches into the central retinal artery and other smaller vessels. Venous drainage occurs through the central retinal vein and vortex veins; these drain into the cavernous sinus, the pterygoid venous plexus, and the facial vein.[2] AVMs occur when the embryonic vasculature does not differentiate in one or more locations, leading to a mass of tangled arteries and veins. The resulting high-pressure shunt between the arterial and venous systems causes congestion in the veins draining the AVM, leading to mass effect, tissue edema, and aneurysm formation.[3] In both fistulas and malformations, blood bypasses the capillaries and

**Figure 1.**



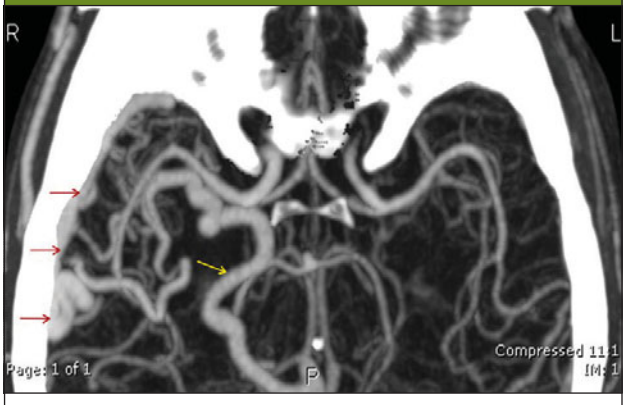
Right orbital proptosis.

**Figure 2.**



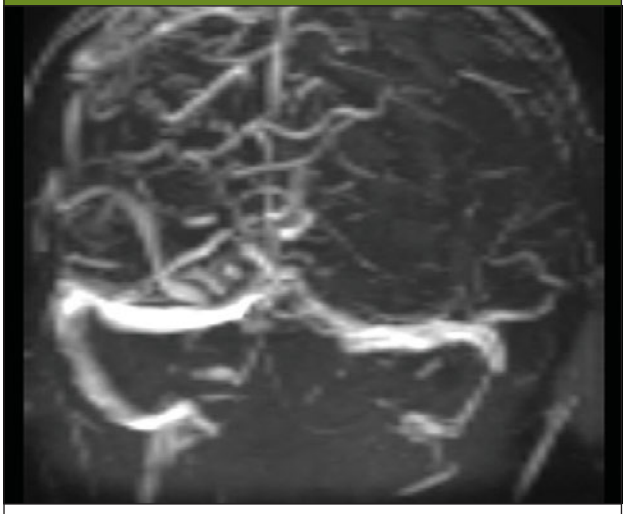
Coronal computed tomography angiogram through the orbits shows asymmetrically enlarged right superior ophthalmic vein (*yellow*) compared with the nondilated left superior ophthalmic vein (*red*).

**Figure 3.**



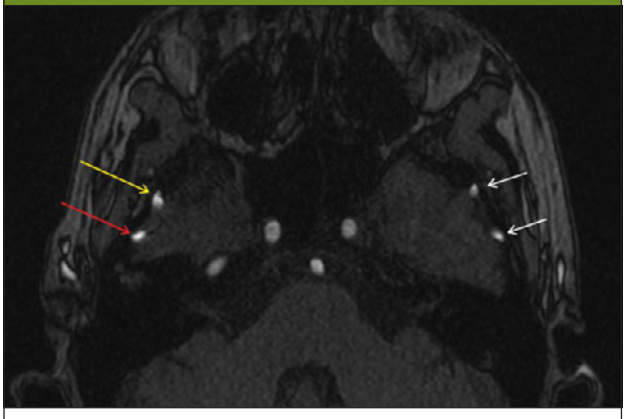

Enlarged, tortuous right cortical veins (*red*) and enlarged right basal vein of Rosenthal (*yellow*).

**Figure 4.**



Head magnetic resonance venogram coronal maximum-intensity projection image showing enlarged, tortuous right cerebral veins.

**Figure 5.**



Asymmetrically enlarged right external carotid artery branches: middle meningeal artery branches (anterior branch, *yellow arrow*; posterior branch, *red arrow*) and occipital artery (*green arrow*). The left middle meningeal artery branches are also enlarged, but less than on the right (*white arrows*).

tissues: Arteriovenous fistulas involve a single connection between one artery and one vein, whereas AVMs involve multiple connections.[4]

### *Diagnosis and Disposition*

When evaluating proptosis, clinicians must consider a broad differential diagnosis, ranging from benign to sight- and life-threatening conditions (**Table 1**). Patients

*"Sudden onset of severe pain, nausea and vomiting, worsening proptosis, decreased visual acuity, and restriction of extraocular movements in the setting of a known or suspected AVM may represent orbital hemorrhage and retrobulbar hematoma. Bleeding within the confined space of the orbit leads to an orbital compartment syndrome and retinal ischemia, causing irreversible vision loss in as little as 60 to 100 minutes."*

with unilateral proptosis should have same-day or urgent orbital imaging with intravenous contrast. If imaging suggests a vascular lesion, follow-up imaging should include contrast evaluation of cranial vasculature. Initial treatment of AVMs can be done medically, including symptomatic management of headache and control of hypertension and diabetes mellitus.[5] Once identified, AVMs should be assessed in a neurosurgery department or a neuro-interventional radiology department for definitive surgical treatment.[6]

***Treatment***

Surgical options for the treatment of intraorbital AVM include microsurgical resection to remove the nidus of the AVM, endovascular embolization with thrombosing agents to reduce the size of the shunt, and stereotactic radiosurgery, which also reduces the shunt via radiation-induced thrombosis. Endovascular embolization is the preferred, least-invasive procedure, with a high cure rate and minimal risks. These three modalities may also be used in combination to maximize effect or reduce risk of hemorrhage; usually preoperative embolization is followed by surgical excision.[7] Whichever treatment strategy is chosen, long-term follow-up is recommended to assess for regrowth of the lesion.

***Complications***

Major complications of intraorbital AVMs include intraorbital hemorrhage, headaches, eye pain, glaucoma, and visual disturbances such as decreased visual acuity (if cranial nerve II is affected) and diplopia (if cranial nerves III, IV, and/or VI are affected).[6] The mortality rate for AVM-related hemorrhage is 6% to 14%, and the annual risk of bleeding for patients with an AVM is 1% to 2%.[8]

Sudden onset of severe pain, nausea and vomiting, worsening proptosis, decreased visual acuity, and restriction of extraocular movements in the setting of a known or suspected AVM may represent orbital hemorrhage and retrobulbar hematoma. Bleeding within the confined space of the orbit leads to an orbital compartment syndrome and retinal ischemia, causing irreversible vision loss in as little as 60 to 100 minutes.[9] There are few reported cases detailing the treatment of acute spontaneous hemorrhage of AVMs; lateral canthotomy and cantholysis have been used in postoperative AVM bleeds.[10] Decompression by lateral canthotomy should be considered when there is evidence of intraocular compartment syndrome,

**Table 1. Proptosis Differential Diagnosis and Evaluation**

| Diagnosis | Pathophysiology | Presentation | Work-Up |
|---|---|---|---|
| ***Endocrine/Oncology*** | | | |
| Graves ophthalmopathy[11] | Autoimmune process causing expansion of orbital soft tissues, seen in association with hyperthyroidism | Heat intolerance, unexplained weight loss, palpitations, sweating, exophthalmos, upper eyelid retraction | TSH, total $T_3$, free $T_4$, CT, MRI |
| Horner syndrome[12] | Interruption of sympathetic innervation to the eye and surrounding structures | Ptosis, meiosis, anhydrosis | CT/CTA, MRI/MRA, CXR |
| IgG4-related orbital disease[13,14] | Autoimmune lymphocytic infiltration by IgG4+ plasma cells | Painless eyelid swelling, proptosis | Biopsy and histology showing absolute IgG4+ cell count >10–100 cells/HPF or IgG4+/IgG+ ratio >0.4 |
| Leukemia/lymphoma[15] | Lymphoid malignancy causing mass effect within the orbit as well as systemic symptoms | General weakness, bleeding, petechiae, ecchymosis | CBC, LDH, peripheral smear, bone marrow biopsy |
| Meningioma, sphenoid wing[16] | Tumor arising from the dural covering of the brain | Bulging temporal bone, anosmia, oculomotor palsy, altered mentation due to elevated intracranial pressure | CT, MRI |
| Orbital neoplasm[17] | Includes primary orbital tumors (melanoma, pleomorphic adenoma, adenoid cystic carcinoma, optic nerve glioma, schwannoma, neurofibroma) and metastatic tumors (most commonly breast, prostate, lung, and skin) | Mass effect within the orbit | CT, MRI, biopsy |
| Pseudotumor (orbital inflammatory syndrome)[18] | Idiopathic inflammation of orbital structures | Proptosis, restriction of extra-ocular movements, pain, erythema, chemosis | CT, MRI, MRI with DWI |
| Sinus mucocele[19,20] | Cystic lesions originating in the sinuses due to blockage of sinus drainage, which may cause gradual expansion of the sinus and erosion of its bony walls. | Proptosis, diplopia, cranial nerve III palsy, vision loss, retro-orbital headache | CT, MRI |
| ***Infectious*** | | | |
| Mucormycosis[21] | Aggressive fungal infection occurring in immunocompromised patients | Fever, eschar of nares, black nasal discharge, facial pain/swelling, hyposmia, decreased vision | CT, MRI, biopsy of necrotic lesions |
| Dacryoadenitis[22] | Inflammation of the lacrimal gland due to infectious, autoimmune, lymphoproliferative, or idiopathic causes | Pain, erythema, and edema in superior and lateral region of the orbit (lacrimal gland), may be acute or chronic | CT with contrast, smear and culture, biopsy |
| Orbital cellulitis[18] | Infection of tissue posterior to the orbital septum | Diplopia, pain with extraocular movements, proptosis, chemosis, fever, history of sinusitis | CT with contrast, MRI, blood cultures |

*(continues)*

as evidenced by a tonometric measurement of intraocular pressure >40 mm Hg.[9]

## Take-Home Points

The patient presented in this case had slowly progressive unilateral proptosis without fever or clinical evidence of cancer, making infection or malignancy less likely. Normal findings on thyroid studies excluded Graves ophthalmopathy. Structural or vascular etiologies remained prominent on the differential diagnosis; a vascular etiology was confirmed by contrast-enhanced MRI. The patient was referred to a neuro-interventional radiology department for evaluation.

For ill-appearing patients with proptosis, indications for transfer to an emergency department include intraorbital hemorrhage, infection, and trauma. Rapid progression of symptoms, severe pain, and decreased vision may indicate intraorbital hemorrhage and retrobulbar hematoma, which should be managed by lateral canthotomy and by transfer to facilities with ophthalmology services. Patients for whom there is high suspicion for infection, including rapid progression, pain, fever, or risk factors for mucormycosis, may be transferred with intravenous antibiotics or antifungals.

In contrast, patients with slowly progressive proptosis not suggestive of infection or trauma can safely be discharged for semi-urgent imaging for vascular or structural abnormalities as long as close follow-up can be ensured. Ultimately, these clinically well patients may need referral to ophthalmology, neurosurgery, or neuro-interventional radiology departments, depending on imaging results. ■

**References**

1. Bartley GB, Fatourechi V, Kadrmas EF, et al. The incidence of Graves' ophthalmopathy in Olmstead County, Minnesota. *Am J Ophthalmol*. 1995;120:511–517.

2. Hayreh SS. Orbital vascular anatomy. *Eye*. 2006;20:1130–1144.

**Table 1. Proptosis Differential Diagnosis and Evaluation *(continued)***

| Diagnosis | Pathophysiology | Presentation | Work-Up |
|---|---|---|---|
| ***Trauma*** | | | |
| Orbital fracture[23] | May occur at orbital floor, walls, or roof; often leads orbital swelling and entrapment of extraocular muscles/fat | Trauma, periocular swelling, diplopia, ecchymosis, chemosis, subconjunctival hemorrhage | CT, extra-ocular movement assessment, eye pressure, vision examination |
| ***Vascular*** | | | |
| Anterior cranial fossa dural arteriovenous fistula[24] | Fistula through the dura, connecting branches of dural arteries to dural veins or a venous sinus | Proptosis, diplopia, cranial nerve palsies (due to mass effect from venous congestion in superior ophthalmic vein) | CTA, MRA, angiography, color Doppler |
| Carotid-cavernous fistula[25,26] | Direct connection between carotid artery and cavernous sinus | Proptosis, chemosis, cephalic bruit, pain, elevated intra-ocular pressure/glaucoma, diplopia, vision loss, intracranial hemorrhage | CT, MRI, CTA, MRA, angiography, color Doppler |
| Cavernous hemangioma[27] | Dilated cluster of veins forming a slow-growing tumor, leading to mass effect within the orbit | Proptosis, visual deficits, restriction of extra-ocular movements, papilledema | CT (often missed), MRI |
| Cavernous sinus thrombosis[28] | Spread of infection from extra-cranial structures to the cavernous sinus through emissary veins. Local endothelial damage and bacteria-derived prothrombotic substances lead to thrombosis and venous congestion. | Proptosis, chemosis, periorbital edema, headache, sepsis | CT, MRI |
| Cirsoid scalp aneurysm[29] | Arteriovenous fistula between superficial temporal artery and scalp veins causing venous congestion | Scalp swelling, bleeding, tinnitus, headache, bruit | CTA, angiography |
| Intraorbital aneurysm[30] | Aneurysm of the intraorbital segment of the ophthalmic artery | Cranial nerve palsies, vision loss, hemorrhage, proptosis | CTA, MRI/MRA |
| Intraorbital arteriovenous malformation[8,31] | Direct connection between intraorbital arteries and veins | Orbital bruit | CTA, MRI/MRA |
| Orbital varices[19] | Congenital venous malformation consisting of thin-walled and distensible veins. These veins have low flow and are prone to thrombosis. | Proptosis (usually intermittent and reversible), periorbital pain, acute vision loss | CT, MRI, color Doppler |

CT, computed tomography; CTA, computed tomography angiography; DWI, diffusion-weighted imaging; HPF, high-power field; Ig, immunoglobulin; LDH, lactate dehydrogenase; MRA, magnetic resonance angiography; MRI, magnetic resonance imaging; $T_3$, triiodothyronine; $T_4$, thyroxine; TSH, thyroid-stimulating hormone.


3. Biousse V, Mendicino ME, Simon DJ, et al. The ophthalmology of intracranial vascular abnormalities. *Am J Ophthalmol*. 1998;125:527–544.
4. Rootman J, Heran MKS, Graeb DA. Vascular malformations of the orbit: classification and the role of imaging in diagnosis and treatment strategies. *Ophthal Plast Reconstr Surg*. 2014;30:91–104.
5. Mohr JP, Parides MK, Stapf C, et al; international ARUBA investigators. Medical management with or without interventional therapy for unruptured brain arteriovenous malformations (ARUBA): a multicenter, non-blinded, randomised trial. *Lancet*. 2014;383:614–621.
6. Warrier S, Prabhakaran VC, Valenzuela A, et al. Orbital arteriovenous malformations. *Arch Ophthalmol*. 2008;126:1669–1675.
7. Rootman J, Heran MKS, Graeb DA. Vascular malformations of the orbit: classification and the role of imaging in diagnosis and treatment strategies. *Ophthal Plast Reconstr Surg*. 2014;30:91–104.
8. Moin M, Kersten RC, Bernardini F, et al. Spontaneous hemorrhage in an intraorbital arteriovenous malformation. *Ophthalmology*. 2000;107:2215–2219.
9. Rowh AD, Ufberg JW, Chan TC, et al. Lateral canthotomy and cantholysis: emergency management of orbital compartment syndrome. *J Emerg Med*. 2015;48:325–330.
10. Levin MH, Moss HE, Pineles SL, et al. Orbital congestion complicating treatment of cerebral vascular anomalies. *World Neurosurg*. 2014;82(1/2):239.e13–239.e17.
11. Bahn RS. Graves' ophthalmopathy. *N Engl J Med*. 2010;362:726–738.
12. Walton KA, Buono LM. Horner syndrome. *Curr Opin Ophthalmol*. 2003;14:357–363.
13. Andrew N, Kearney D, Selva D. IgG4-related orbital disease: a meta-analysis and review. *Acta Ophthalmol*. 2013;91:694–700.
14. Islam AD, Selmi C, Ditta-Mitra A, et al. The changing faces of IgG4-related disease: Clinical manifestations and pathogenesis. *Autoimmun Rev*. 2015;14:914–22.
15. Bernardini FP, Bazzan M. Lymphoproliferative disease of the orbit. *Curr Opin Ophthalmol*. 2007;18:398–401.
16. Whittle IR, Smith C, Navoo P, et al. Meningiomas. *Lancet*. 2004;363:1535–1543.
17. Tailor TD, Gupta D, Dalley RW, et al. Orbital neoplasms in adults: clinical, radiologic, and pathologic review. *Radiographics*. 2013;33:1739–1758.
18. Pakdaman MN, Sepahdari, AR, Elkhamary SM. Orbital inflammatory disease: pictorial review and differential diagnosis. *World J Radiol*. 2014;6:106–115.
19. Pahwa S, Sharma S, Das CJ, et al. Intraorbital cystic lesions: an imaging spectrum. *Curr Probl Diagn Radiol*. 2015;44:437–448.
20. Fernandes R, Pirgousis P. Longstanding giant frontal sinus mucocele resulting in extra-axial proptosis, orbital displacement, and loss of vision. *J Oral Maxillofac Surg*. 2010;68:3051–3052.
21. Lemos J, Eggenberger E. Neuro-ophthalmological emergencies. *Neurohospitalist*. 2015;5:223–233.
22. Mombaerts I. The many facets of dacryoadenitis. *Curr Opin Ophthalmol*. 2015;26:399–407.
23. Boyette JR, Pemberton JD, Bonilla-Velez J. Management of orbital fractures: challenges and solutions. *Clin Ophthalmol*. 2015;9:2127–2137.
24. Hacein-Bey L, Konstas AA, Pile-Spellman J. Natural history, current concepts, classification, factors impacting endovascular therapy, and pathophysiology of cerebral and spinal dural arteriovenous fistulas. *Clin Neurol Neurosurg*. 2014;121:64–75.
25. Korkmazer B, Kocak B, Tureci E, et al. Endovascular treatment of carotid cavernous sinus fistula: a systematic review. *World J Radiol*. 2013;5:143–155.
26. Rose GE, Verity DH. Acute presentation of vascular disease within the orbit—a descriptive synopsis of mechanisms. *Eye*. 2013;27:299–307.
27. Scheuerle AF, Steiner HH, Kolling G, et al. Treatment and long-term outcome of patients with orbital cavernomas. *Am J Ophthalmol*. 2004;138:237–244.
28. Desa V, Green R. Cavernous sinus thrombosis: current therapy. *J Oral Maxillofac Surg*. 2012;70:2085–2091.
29. Goel V, Verma AK, Singh S, et al. Cirsoid aneurysm of scalp: demonstration on CT angiography (CTA). *BMJ Case Rep*. Published online: 2013. Accessed: 16 January 2016. doi: 10.1136/bcr-2013-202459.
30. Della Pepa GM, Sabatino G, La Rocca G, et al. Intraorbital and intracanalicular ophthalmic artery aneurysms. Literature review and report of a case. *Br J Neurosurg*. 2014;28:787–790.
31. Hieu PD, Besson G, Roncin S, Nonent M. Successful surgical treatment of intraorbital arteriovenous malformations: case report. Neurosurgery. 1997;40:626–631.