

INCREASED EPISCLERAL VENOUS PRESSURE AS AN ETIOLOGY OF GLAUCOMA

Advice on differential diagnosis and management.

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CASE PRESENTATION



Patient No. 1

A 54-year-old Asian woman presented after 1 month of redness in her right eye accompanied by swollen eyelids but minimal pain. The patient had no significant prior medical or ocular history.

On examination, her visual acuity was 20/25 OD and 20/20 OS. We noted no afferent pupillary defect. Hertel measurements demonstrated 5 mm of proptosis

(Figure 1A), a slight abduction deficit, and mild periorbital edema in the right eye. IOP measured 32 mm Hg OD and 18 mm Hg OS. The slit-lamp exam showed dilated and tortuous conjunctival and episcleral vessels in the right eye (Figure 1B). The dilated fundus exam was unremarkable, and we noted no significant cup-to-disc asymmetry (cup-to-disc ratio = 0.3 OU). Gonioscopy revealed open angles in both eyes and blood in Schlemm canal of the right eye. Computed tomography (CT) of the orbits with contrast revealed a dilated superior ophthalmic vein (Figure 1C), leading us to suspect a carotid-cavernous fistula.

We started the patient on timolol in her right eye, and the IOP decreased to 22 mm Hg. She underwent cerebral angiography, which identified a right dural carotid-cavernous fistula that was treated by embolization. At postoperative month 2.5, the patient had a visual acuity of 20/20 OU. The proptosis, motility deficit, and dilated episcleral vessels had resolved. Her IOP measured 18 mm Hg OD and 13 mm Hg OS without medication.

Patient No. 2

A 61-year-old Asian man was referred to our glaucoma clinic for elevated IOP in his right eye. The patient had no history of other ocular conditions and no significant past medical history.

On exam, his visual acuity was 20/20 OU. No significant proptosis, ptosis, or chemosis was observed, and motility was normal. We noted an afferent pupillary defect in the right eye. IOP measured 27 mm Hg OD and 21 mm Hg OS. The slit-lamp and dilated fundus exams were remarkable for slightly dilated episcleral vessels in the right eye (Figure 2A) and a significant cup-to-disc asymmetry (0.9 OD and 0.6 OS). Gonioscopy revealed open angles in both eyes and blood in Schlemm canal of the right eye. Visual field testing found significant visual field loss in the right eye (Figure 2B), whereas the left visual field was normal and intact. Computerized tomographic angiography was unremarkable.

The patient's right eye was treated with brimonidine and a fixed combination of dorzolamide and timolol. The IOP has since been controlled in the low teens.

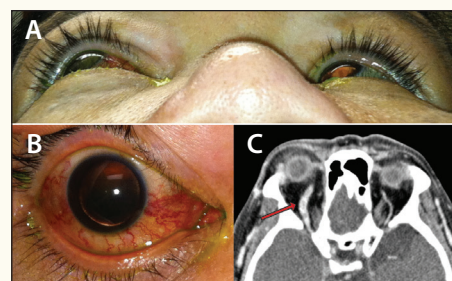


Figure 1. Elevated episcleral venous pressure (EVP) secondary to dural fistula. Right eye exhibits 5 mm of proptosis (A) as well as dilated corkscrew episcleral and conjunctival vessels (B). Axial CT orbit demonstrates dilated right superior ophthalmic vein (arrow, C).

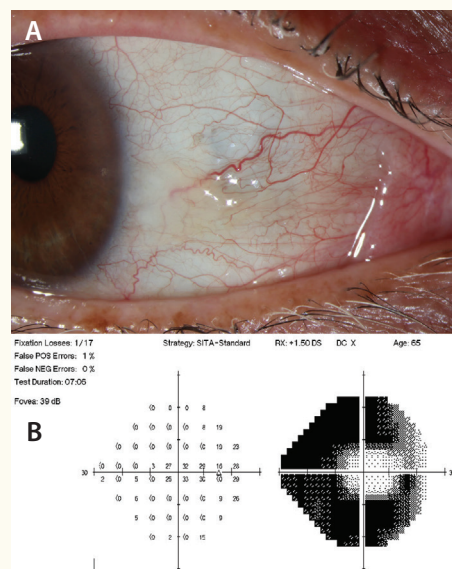


Figure 2. Idiopathic elevated EVP in the right eye. Subtle dilation of episcleral vessels (A) and severe visual field loss (B).

TABLE 1. CAUSES OF ELEVATED EVP

Venous Congestion	Arteriovenous Fistulas	Idiopathic (Radius-Maumenee Syndrome)
Thyroid-associated ophthalmopathy	Carotid-cavernous sinus fistula	Diagnosis of exclusion
Superior vena cava syndrome	Dural arteriovenous shunts	
Retrobulbar tumors	Orbital varices	
Cavernous sinus thrombosis	Sturge-Weber syndrome	
Orbital amyloidosis		

BACKGROUND AND PATHOPHYSIOLOGY

Normal EVP ranges from 8 to 10 mm Hg, and for every 1 mm Hg of elevation in EVP, the mean IOP is thought to rise 1 mm Hg.¹ The underlying causes of elevated EVP can be divided into three categories: (1) venous congestion, (2) arteriovenous fistulas, and (3) idiopathic (Table 1).

CLINICAL PRESENTATION

The most common sign of elevated EVP is dilation and increased tortuosity of the episcleral and conjunctival vessels. Depending on the underlying cause of the increased EVP, additional findings may include proptosis, chemosis, and orbital bruit (Table 2). Gonioscopy typically reveals an open angle; in some cases, Schlemm canal may contain blood, thought to be present as a result of blood reflux from the episcleral veins. The IOP is elevated, and depending on the chronicity and severity of this increase, there may be evidence of glaucomatous optic neuropathy.

DIAGNOSIS

It is important to identify any underlying causes of elevated EVP. Issues to explore include a history of thyroid disease, amyloidosis, superior vena cava syndrome, Sturge-Weber syndrome, or carotid-cavernous sinus fistula, a mechanism made more likely by a history of trauma. Additional diagnostic workup for these EVP elevation causes may include orbital ultrasound, CT, and/or magnetic resonance imaging to rule out intraorbital or intracranial etiologies. Cases are only considered idiopathic when no underlying etiology is identified.

Commercial instruments to measure EVP are unavailable. Accordingly, the diagnosis of glaucoma secondary to idiopathic elevated EVP is made based on clinical findings.²

TABLE 2. DIFFERENTIATING IDIOPATHIC ELEVATED EVP FROM ARTERIOVENOUS FISTULA

	Idiopathic Elevated EVP	Arteriovenous Fistula
Onset	May be congenital	Often acquired
Predisposing factors	Possibly familial	Trauma, hypertension
Symptoms	None	Pulsatile bruit/tinnitus, pain, diplopia
Neurological signs	None	Unilateral or bilateral cranial nerve VI paresis, cranial nerve III paresis
External signs	Dilated episcleral vessels Conjunctival vessels can be normal	Dilated corkscrew episcleral and conjunctival vessels Chemosis, eyelid edema
Orbital signs	None	Bruit, proptosis
Imaging	Normal	Dilated superior ophthalmic vein or cavernous sinus
Catheter angiography	Normal	Arteriovenous fistula

Adapted from Foroozan et al.³

MANAGEMENT

Initial management should be directed at reducing IOP, followed by the treatment of any underlying cause of the elevated EVP. Consultations with oculoplastics, neurology, neurosurgery, and/or interventional radiology may be required. Because aqueous outflow is compromised, aqueous suppressants are more effective than are medications that enhance outflow for the medical management of IOP resulting from elevated EVP. Apraclonidine in particular has been shown to be effective for lowering IOP in cases of elevated EVP, because the drug causes arterial vasoconstriction, which decreases the amount of blood flow to the eye.⁴

If the IOP remains uncontrolled despite medical management, then surgical options are considered. Several case reports point out that trabeculectomy in these eyes can be complicated by intraoperative or postoperative uveal effusions, even at nonhypotonous IOPs.⁵⁻⁷ Some physicians have recommended “tight” trabeculectomies, with multiple sutures surrounding the scleral flap, to allow gradual titration of the IOP in the postoperative period.⁸ In one case report of an eye with idiopathic elevation of EVP, the ophthalmologist avoided the complication of postoperative uveal effusions by performing non-penetrating filtering surgery followed by goniotomy



AT A GLANCE

- Elevated episcleral venous pressure can result from a number of underlying etiologies that require systematic diagnostic studies followed by appropriate treatment.
- In some cases, treatment of the underlying cause of the increased episcleral venous pressure can lead to resolution of the associated IOP elevation.
- Medical management with aqueous suppressants is preferred but often inadequate.
- Surgical management is often fraught with complications such as intraoperative or postoperative choroidal effusions, and failure is common. Before intervening surgically, it is important to make the patient aware of the guarded prognosis.

(a staged trabeculectomy).⁹ Nonpenetrating deep sclerectomies have also been attempted, resulting in fewer complications, but they have been less successful at maintaining long-term IOP control.¹⁰ Before intervening surgically, it is important to make the patient aware of the guarded prognosis. ■

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