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Unilateral Proptosis, Redness, Diplopia, and Numbness in a Young Woman

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A woman in her 30s with no significant medical history presented with a 1-month history of worsening “bulging,” redness, and mild pain in her right eye. She also reported intermittent, binocular diplopia. Results from a review of systems were otherwise negative, and she had not experienced recent trauma. She was started on topical antihypertensive drugs for elevated intraocular pressure (IOP) in the affected eye (30mmHg) and referred to our institution. On examination, visual acuity was 20/20 in both eyes with subjective red desaturation OD. Pupils were equal and without afferent pupillary defect. Confrontation visual fields were full. Intraocular pressures were 24 mm Hg OD and 12 mm Hg OS. Hertel exophthalmometry showed 4.5 mm of relative proptosis OD. She had limitation of her extraocular muscles in nearly all fields of gaze OD, with the most obvious limitation occurring in abduction. She had decreased sensation to light touch in the right cheek (cranial nerve V2). There were no orbital bruits. Ocular examination revealed diffuse conjunctival injection OD with tortuous, dilated, corkscrew-shaped blood vessels (Figure, A). Dilated fundus examination revealed mild optic nerve hyperemia and mild vascular tortuosity OD. Vital signs and results from complete blood cell count and thyroid function tests were normal. Computed tomography and magnetic resonance imaging, angiography, and venography of the brain and orbits revealed extraocular muscle enlargement and enhancement and a dilated superior ophthalmic vein OD. There was no mass, abscess, or arteriovenous fistula (Figure, B).

Diagnosis

Carotid-cavernous fistula

What to Do Next

D. Order catheter cerebral angiography

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Discussion

When treating a red, proptotic eye, it is important to consider the possibility that the pathologic abnormality may be intracranial rather than intraorbital. Dilation of the superior ophthalmic vein on imaging and impairment of the maxillary (V2) branch of the trigeminal nerve, as in this case, localize abnormality to the cavernous sinus (which contains cranial nerves III, IV, V1, V2, and VI), causing secondary orbital signs and symptoms. This is in contrast to primary orbital disease (eg, orbital cellulitis, orbital inflammatory syndrome, thyroid eye disease, orbital tumor). The constellation of findings in our patient, including V2 impairment, elevated intraocular pressure, dilated and tortuous conjunctival vessels, and evidence of orbital congestion, including proptosis, extraocular muscle enlargement, and a dilated superior ophthalmic vein, led us to strongly suspect a carotid-cavernous fistula (CCF), despite the absence of this finding on magnetic resonance angiography. This patient therefore underwent catheter cerebral angiography (answer choice D), which confirmed an indirect, type D, CCF characterized by an abnormal communication between the cavernous sinus and meningeal branches of both the internal and external carotid arteries. Answer choices A, B, and C are inadequate given that the clinical and radiographic findings did not strongly support a neoplastic, inflammatory, or infectious process.

Carotid-cavernous fistula is a condition describing an abnormal communication between the carotid artery and the cavernous sinus. It is divided into 2 broad categories: (1) direct, or type A, and (2) indirect, dural, or types B, C, and D.¹ Direct CCFs are high-flow fistulas and most commonly occur after trauma. Indirect CCFs are low-flow fistulas and most commonly occur spontaneously. Indirect CCFs may present quite variably or insidiously and are often initially misdiagnosed as other conditions, including thyroid eye disease, orbital inflammatory syndrome, or chronic conjunctivitis.

Direct and indirect CCFs are capable of producing similar clinical manifestations, although indirect CCFs generally tend to have less severe and acute presentations. Possible findings include mild pain, unilateral (or less frequently, bilateral) proptosis, pulsatile exophthalmos,² orbital bruit,³ abnormal ocular motility (secondary to impaired function of cranial nerves III, IV, or VI), exposure keratopathy, neurotrophic keratopathy from V1 neuropathy, conjunctival chemosis and injection (manifesting as dilated, tortuous, corkscrew-like vessels), elevated IOP, glaucoma, optic disc swelling, venous stasis retinopathy, retinal vascular occlusions, and choroidal effusion.⁴ The gold standard for diagnosis is catheter cerebral angiography, although less invasive neuroimaging may also be helpful. Importantly, as shown by this case, certain CCFs, particularly indirect CCFs, may be missed unless catheter cerebral angiography is performed.

Endovascular embolization is the treatment of choice for both direct CCFs and those indirect CCFs that fail to spontaneously close or are vision threatening.^{2,5,6} After treatment, elevated IOP often returns to baseline within 2 days, but orbital congestion and ophthalmoparesis often require 1 to 4 weeks to return to normal.²

Patient Outcome

Our patient successfully underwent endovascular embolization at the time of her cerebral angiogram. Within 2 days of treatment, her vision subjectively improved, IOP returned to normal, proptosis decreased, conjunctival injection decreased, and ophthalmoparesis had markedly improved.

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WHATWOULD YOU DO NEXT?

- A.** Orbital biopsy
- B.** Start systemic corticosteroids
- C.** Start systemic antibiotics
- D.** Order catheter cerebral angiography

Figure.

A, External photograph demonstrating diffuse conjunctival injection and proptosis of the right eye. B, Contrast-enhanced, fat-suppressed, T1-weighted magnetic resonance imaging of the orbits demonstrating a dilated superior ophthalmic vein and extraocular muscle enlargement of the right eye.