Case 37

History of Present Illness

A 51-year-old sheet metal worker developed photophobia and bilateral eye ache 2–3 months ago. The pain is constant, which she describes as 5 out of 10. The pain worsens when she immerses herself in cold water. She states that the pain is localized to the eyes and, overall, is not getting better or worse. Two weeks after the pain began, her right eye became red. She denies tearing, discharge, or foreign body sensation. She denies blurred vision, double vision, or ptosis. She does not wear contact lenses and has been using Visine without improvement in the redness or pain. The redness is worse upon awakening. She has heard a ringing in his left ear for the past month, which is constant.

Past medical and ocular history Steroid-responsive glaucoma (given for pink eye in the past) Wears glasses	*Past surgical history* None
Medications Artificial tears PRN Tetrahydrozoline (Visine) PRN	*Family history* Mother had brain cancer Father had glaucoma but not severe
Social history Works with sheet metal Social drinker Has never smoked	*Review of systems* Per HPI

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Examination
Acuity with correction Right eye: 20/20 Left eye: 20/25
Pupils Equal in size, Round shape, briskly reactive, no APD
Intraocular pressure Right eye: 23 mmHg Left eye: 23 mmHg
External exam No ptosis, 1 mm of proptosis RE, no eyelid edema
Motility and Eye alignment Normal
Slit lamp examination RE: Normal cornea, large tortuous conjunctival vessels, no papillae or follicles. The vessels do not blanch with neosynephrine (see Fig. 37.1a) LE: normal
Visual field Normal
Fundus examination 0.4 CDR RE/0.5 CDR LE Normal
Neurologic examination Normal facial sensation Otherwise unremarkable

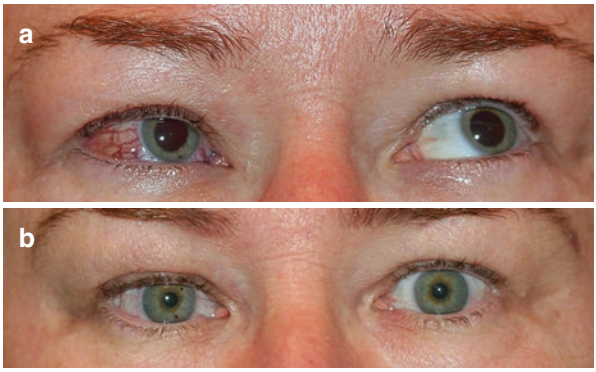

Fig. 37.1 Color photos show a red right eye. (**a**) Notice the corkscrew type vessels typical of dural cavernous fistulas. (**b**) After treatment the redness resolved

Discussion

Ophthalmic Perspective—Dr. Lee

Although her initial complaint was eye pain, I cannot ignore the red eye (I doubt this is a red herring since it is ipsilateral to the pain). I will say that I have no idea why her pain is worse when going under water. This story is not consistent with conjunctivitis since there is no discharge or tearing. Additionally, the 2-month history is

unusual for conjunctivitis (Case 13). Generally, the pain of scleritis gets worse over time. She does not have any other stigmata of inflammation such as conjunctival edema (chemosis), and she describes pain in both eyes but only her right eye is red. Metal workers can get a metallic foreign body in their corneas, but her corneas were clear. An intraocular foreign body typically causes a decline in vision and ipsilateral, unilateral pain. It is not unusual to have 1 mm of relative proptosis in normal individual but this may represent a real orbital finding.

I would like to know more about the ringing in her ear. Is it high-pitched or does sound like rushing water that pulses with his heartbeat pulse synchronous tinnitus (PST)? If you look closely at the redness, there are areas of relative "white" sclera between the big red vessels, which are veins. Note also that these beefy veins start at the limbus (junction of the cornea and conjunctiva). These are characteristic vessels that we would see in a carotid cavernous (C-C) fistula.

C-C fistulas are abnormal connections between the carotid artery and the venous flow in the cavernous sinus. There are two basic kinds of C-C fistula: high-flow and low-flow. High-flow fistulas are usually traumatic and dramatic. The eye looks very proptotic with dramatic eyelid edema, conjunctival edema, and proptosis. Basically, it looks like the eye is popping out of the head, which is not the case here. Low-flow fistulas typically present in elderly women and PST is common. If the venous flow is toward the eye, then the patient gets arterialized conjunctival veins as seen here (if the venous flow is more posterior, then the eye looks white and quiet). Some patients can develop diplopia or facial pain from CN 3,4,5,6 involvement. Occasionally, patients have a defect between cavernous sinuses and can develop bilateral findings despite a unilateral fistula. The diagnosis is supported by a CT or MRI that shows enlargement of the superior ophthalmic vein (SOV) but may require conventional catheter angiography to confirm. I would start with a CT/CTV or an MRI/MRV to evaluate this patient.

Neurologic Perspective—Dr. Digre

I saw a case like this 2 days ago in my clinic, so I know these cases are not rare. First, she is of the right sex and age. Individuals with the indirect fistulas are usually middle or older aged women. The dull ache is really typical of these indirect dural fistulas. I do not understand the worsening of pain with water either—unless cold water could cause cold-induced headache. Sometimes having the patient bend over will make it worse. Morning worsening is definitely a typical headache feature. See the ICHD 3 beta criteria for dural arteriovenous fistula (Table 37.1). The cork-screw like vessels on the conjunctivae either unilaterally or bilaterally are also really typical—and like this lady, they do not blanch or go away with a topical sympathomimetic. The red did not come out with Visine (which has a topical sympathomimetic). The elevated intraocular pressure is also typical—and sometimes the pressure is high enough that it too requires therapy.

When fistulas are posteriorly draining into the petrosal sinuses, you do not see the red eye, but can get facial numbness from trigeminal neuropathy, facial weakness from a seventh nerve palsy, or diplopia from usually a third nerve palsy. The posteriorly draining dural fistulas often cause a painful eye, but there will be no

Table 37.1 ICHD 3 beta criteria for dural arteriovenous fistula

(A) Any new headache fulfilling criterion C
(B) A dural arteriovenous fistula (DAVF) has been diagnosed
(C) Evidence of causation demonstrated by at least two of the following:
1. Headache has developed in close temporal relation to other symptoms and/or clinical signs of DAVF, or has led to the diagnosis of DAVF
2. Either or both of the following:
(a) Headache has significantly worsened in parallel with other symptoms or clinical or radiological signs of growth of the DAVF
(b) Headache has significantly improved after treatment of the DAVF
3. At least one of the following:
(a) Headache is accompanied by pulsatile tinnitus
(b) Headache is accompanied by ophthalmoplegia
(c) Headache is both progressive and worse in the morning and/or during coughing and/or bending over
4. Headache is localized to the site of the DAVF
(D) Not better accounted for by another ICHD-3 diagnosis, and intracerebral hemorrhage and cerebral venous thrombosis have been excluded by appropriate investigations

Headache Classification Committee of the International Headache Society. The International Classification of Headache Disorders: 3rd edition (beta version). *Cephalalgia*. 2013;33:629–808

red eye. Risk factors to getting these fistula include: female sex, older individuals, hypertension, and connective tissue disorders.

Besides getting MRI or CT with contrast, orbital color Doppler can be helpful since it may detect the abnormal direction of flow. Occasionally angiography is required to find the fistula since these fistulas can be made up of branches from the internal or external carotid artery. We usually watch and admire if there are few symptoms since 20–50% of these can close spontaneously. Air plane travel, using the opposite hand to intermittently occlude the carotid artery on the side of the fistula—in this case she would use her left hand to occlude her right artery—often will assist in closure. She is not on anti-platelet therapy, but if she were, you could have her stop it. If she chooses to observe, she should have her intraocular pressure followed. Sometimes chemosis can progress and further treatment may be necessary. These procedures include endovascular therapy (most common), direct surgery, and radiotherapy. Rarely, these fistulas can recur.

Non-ophthalmic/Non-neurologic Perspective

Common things being common, history, and exam should help us rule out conjunctivitis and foreign body (see above discussion). Intraocular inflammation is harder to see unless you are facile with a slit lamp. Generally speaking, a 2–3 month history without worsening of pain or vision does not warrant an emergent consultation no matter the cause. This particular kind of red eye is pattern recognition and if PST is present, then the suspicion becomes very high. If you obtained a scan, then you would direct the radiologist to look carefully at the SOV.

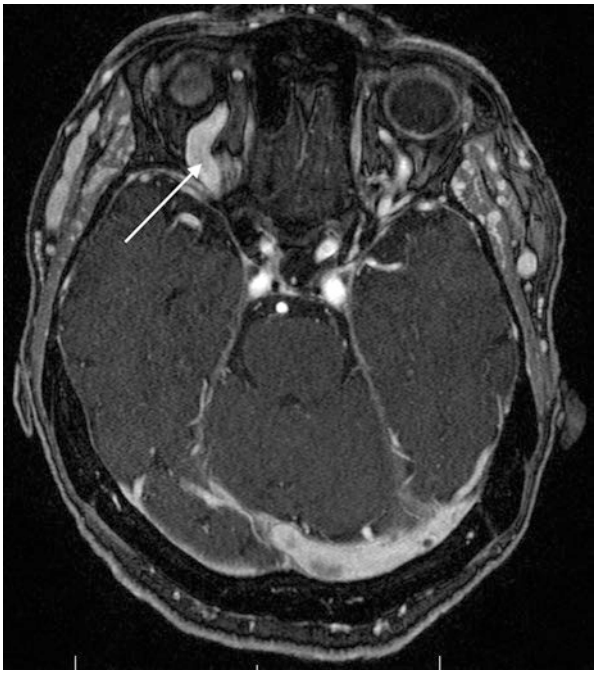

Fig. 37.2 Axial T1 MRI shows a dilated and tortuous right superior ophthalmic vein (*arrow*)

Follow-up

The patient endorsed PST. MRI (Fig. 37.2) and MRV showed enlargement of the right SOV and several atypical flow voids in the right cavernous sinus. Approximately 50% of low-flow fistulas can spontaneously resolve over a year, and observation is reasonable in the absence of a defect or deficiency. Angiography may be required to secure a diagnosis in some individuals and is also used to close fistulas. This patient chose to undergo angiography and coiling. The redness and pain were gone at 6-week follow-up (Fig. 37.1b). *Final diagnosis: Low-flow, Cavernous-carotid fistula (dural cavernous fistula)*.

For Further Study

1. Miller NR. Dural carotid-cavernous fistulas: epidemiology, clinicalpresentation, and management. Neurosurg Clin N Am. 2012;23(1):179–92.
2. Subramanian PS, Williams ZR. Arteriovenous malformations and carotid-cavernous fistulae. Int Ophthalmol Clin. 2009;49(3):81–102.