# Case 17

## History of Present Illness

A 62-year-old man developed a severe burning pain around the right eye and the right side of his head. Initially, he thought it was due to sinus infection, which he had in the past. Two years before, he had a bicycle accident and dislocated his shoulder but denied head trauma. The pain was continuous and not associated with light or sound sensitivity or nausea or vomiting; occasionally the pain is bad enough that he will lie down. As a youngster, age 12, he was shot with a b-b gun and still has a copper BB behind his right eye and near the optic nerve.

| | |
|---|---|
| *Past medical and ocular history*<br>Fatigue and muscle aching—Worked up for polymyalgia rheumatica but this was negative<br>Previous sinus infection<br>Barrett's esophagus<br>Hypothyroidism<br>Essential hypertension | *Past surgical history*<br>Knee surgery twice; tonsillectomy |
| *Medications*<br>Metoprolol 100 mg each day<br>Multiple vitamin<br>Omeprazole 20 mg<br>Vitamin D 1000 I.U. | *Family history*<br>Diabetes in father and sister<br>Hypertension in brother and mother |
| *Social history*<br>No smoking or alcohol; married<br>Social worker for LDS church | *Review of systems*<br>Occasional fatigue |


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| Examination |
|---|
| *Acuity with correction*<br>Right eye: 20/25<br>Left eye: 20/20-2 |
| *Pupils*<br>OD: 2 mm light and 4 mm darkness<br>OS: 3 mm light and 6 mm darkness<br>Dilation lag on the right (Fig. 17.1) |
| *Color vision*<br>9/9 BE Ishihara |
| *Stereo vision*<br>Reduced; 1/3 animals on Titmus test |
| *Intraocular pressure*<br>Right eye: 23 mmHg<br>Left eye: 23 mmHg |
| *External exam*<br>No ptosis |
| *Eye motility and alignment*<br>Full excursions<br>Comitant 10 prism diopter exophoria (long standing) |
| *Slit lamp examination*<br>Normal |
| *Visual field*<br>Normal |
| *Fundus examination*<br>Normal disks 0.3 cup to disk ratio in both eyes |
| *Neurologic examination*<br>Normal facial sensation<br>The rest of the exam was normal |

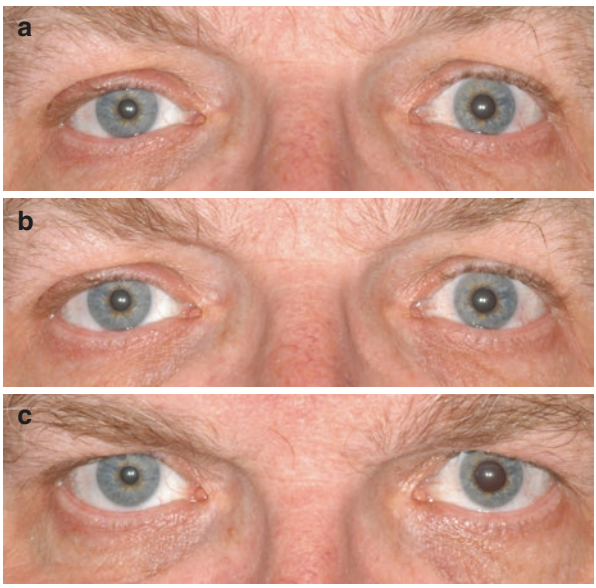


**Fig. 17.1** External photographs show very mild ptosis on the right. Note that the palpebral fissure is smaller on the right. There is anisocoria at (**a**) 5 s after turning off the lights, which becomes less obvious at (**b**) 15 s. This difference is known as dilation lag. (**c**) Thirty minutes after instillation of 5% cocaine eye drops, the anisocoria becomes more obvious. This is a positive cocaine test, diagnostic of a Horner syndrome

## Discussion

### *Neurologic Perspective: Dr. Digre*

In every patient with eye pain, I look carefully at the pupils to see if there is a Horner's syndrome (small pupil, ptosis, upside down ptosis, and dilation lag) since many headache syndromes are associated with a Horner's or partial Horner's syndrome. To look for a Horner's syndrome I look for three things: First—assess the size of the pupil in light and darkness. Typically, the Horner's patient will have more anisocoria in darkness than in light. Second, I look for a dilation lag—you can do this in a dimly lighted room with a flashlight below the chin and turn the overhead light on and off, looking to see if the smaller pupil is slow to dilate. Third, I look for ptosis on the side of the smaller pupil—both superior lid ptosis but also upside down ptosis (lower eyelid is higher). In this case the Horner's syndrome was subtle since he really did not have any appreciable ptosis.

This man fits the syndrome of paratrigeminal oculosympathetic syndrome (sometimes called Raeder's syndrome)—a painful Horner's syndrome. It is typified by pain in V1 or V2 and a Horner's pupil. See Table 17.1 for the ICHD 3 beta definition of paratrigeminal oculosympathetic syndrome. This is a new headache in someone who does not have headaches.

Painful Horner's syndrome can occur in an acute Cluster headache attack—and I have seen this several times—a painful Horner's in the ER and after all testing, it was the first attack of cluster headache. Imaging is critical since carotid dissection or middle cranial fossa lesions (e.g., cavernous sinus disease) could also present this way.

Sometimes, the Horner's is so subtle that I am not sure it is a Horner's; then I perform pharmacologic testing. Here photographs of the pupils before and after drop testing are very helpful. I will apply cocaine (which blocks norepinephrine reuptake and therefore dilates the normal pupil) 5% when available to each eye and wait 30–60 min to see if both pupils dilate equally—if one pupil fails to dilate, it is a positive test. If there is more than 0.8 mm of anisocoria after the testing, Horner's pupil is diagnosed.

**Table 17.1** ICHD 3 Beta: Paratrigeminal oculosympathetic syndrome (Raeder's syndrome)

Diagnostic criteria:

A. Constant, unilateral headache fulfilling criterion C
B. Imaging evidence of underlying disease of either the middle cranial fossa or of the ipsilateral carotid artery
C. Evidence of causation demonstrated by both of the following:
    1. Headache has developed in temporal relation to the onset of the underlying disorder
    2. Headache has either or both of the following features:
        (a) Localized to the distribution of the ophthalmic division of the trigeminal nerve, with or without spread to the maxillary division
        (b) Aggravated by eye movement
D. Ipsilateral Horner's syndrome
E. Not better accounted for by another ICHD-3 diagnosis

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

Sometimes cocaine is not available and the apraclonidine test is used. In this pharmacologic test, we take advantage of adrenergic hypersensitivity since apraclonidine is a weak adrenergic agent. In this case, a drop is instilled in each eye. After waiting for 30–40 min, you will see a reversal of anisocoria (that is the Horner's pupil will appear larger than the non-Horner's pupil).

When available, I like to localize the Horner's as a second- or third-order neuron Horner's pupil. It helps me order the appropriate imaging. In this test hydroxyamphetamine (which releases any norepinephrine from the iris terminal which causes dilation in a normal pupil and in a first or second order Horner's pupil) is instilled in a separate session (usually 1–2 days apart from the previous testing). You wait 30–40 min and a third-order Horner's will show NO dilation to hydroxyamphetamine. When the Horner's syndrome is acute, you may want to skip this test, since getting to imaging is critical to prevent stroke.

To determine if the Horner's is acute—the history is helpful—sudden onset of pain and changes in the eyelid or pupil. I usually look at old photographs—if a patient has a driver's license and has had a smaller pupil on that side for years, the emergency of the pupil is lessened. If the Horner's pupil is acute, I order imaging—usually MRI and MRA since I am most worried about a carotid dissection. In this man's case, he had a BB to the eye, so only a CT could be done, but a CTA was also ordered.

## *Ophthalmic Perspective: Dr. Lee*

In the ophthalmologist's office, a technician often evaluates patients and then dilates the patient prior to seeing the physician. In cases of eye pain, it is critical that the technician evaluates the pupil and eyelid, because if they dilate the patient, then you will never find the Horner syndrome and may miss the carotid dissection. Since this is an acute oculosympathetic disruption, some patients may have other autonomic signs or symptoms such as ipsilateral conjunctival injection, tearing, lower intraocular pressure, nasal congestion, or an abnormally close near point of accommodation. These other symptoms are transient and may only be there for hours to days. Patients with autonomic symptoms can sometimes be mistaken for conjunctivitis or trigeminal autonomic cephalgia. Keep in mind that 20–40% of folks can have physiologic anisocoria. If your cocaine test is negative (both pupils dilate equally) then the patient likely has headache plus physiologic anisocoria.

If the neuroimaging is normal and this is not part of an autonomic headache, the Horner syndrome is generally idiopathic and permanent. Patients can undergo eyelid surgery (Mullerectomy) for repair. Because the pupil and eyelid are supersensitive to alpha 1 agonists, I have asked individuals to try over-the-counter tetrahydrozoline (Visine) to reverse the lid and pupil findings for cosmesis.

As with all things medicine, differing opinions exist. Personally, I do not localize a Horner syndrome with hydroxyamphetamine as described by Dr. Digre. I usually

image the entire oculosympathetic axis. If the scan is normal then I do not pursue further workup, since idiopathic Horner syndrome is not uncommon.

## *Non-ophthalmic/Non-neurologic Perspective*

If you suspect a painful Horner's syndrome and it is acute, it is a relative emergency to prevent a stroke from occurring. Imaging will be critical to ensure that this is not a carotid dissection. Most patients do not have the full triad and will not endorse the anhidrosis. The radiologist needs to know that you are looking for a carotid dissection, or they may not protocol the scan properly.

## *Follow-Up*

Normally, I would have ordered an MR on this patient, but because of the BB shot, a CT scan was done. The CT showed the expected B-B pellet behind the right eye and a double lumen sign consistent with a carotid artery dissection (Fig. 17.2). He was placed on aspirin 81 mg and did well without further problems. Horner's syndrome can be the presenting sign of carotid dissection along with headache. *Final diagnosis: Horner syndrome secondary to carotid artery dissection*.

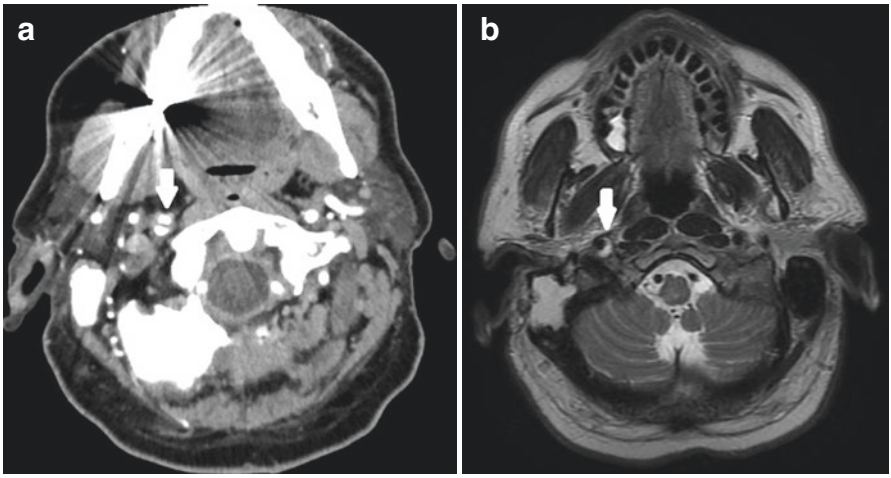


**Fig. 17.2** (**a**) CTA in this case showing a carotid dissection with a double lumen (*arrow*). (**b**) Axial T1 MRI of another case shows classic hyperintense crescent typical of a carotid dissection (*arrow*)

## For Further Study

1. Davagnanam I, Fraser CL, Miszkiel K, Daniel CS, Plant GT. Adult Horner's syndrome: a combined clinical, pharmacological, and imaging algorithm. Eye (Lond). 2013;27(3):291–8.
2. Lemos J, Eggenberger E. Neuro-ophthalmological emergencies. Neurohospitalist. 2015;5(4):223–33.
3. Sheikh HU. Headache in intracranial and cervical artery dissections. Curr Pain Headache Rep. 2016;20(2):8.