# Case 29

## History of Present Illness

Five months ago, a 66-year-old man with no previous headache history developed pain behind both eyes while running. Since then, he has noted that the pain occurs every time he coughs, sneezes, or picks up something heavy. It intermittently occurs when he bends his head below his waist, strains on the toilet, or clears his throat. It can occur if he gets up from his chair or goes up the stairs too quickly, but if he goes slowly it does not happen. If he does not Valsalva, then it would not happen. He describes it as a pressure pain, rating 5–6 out of 10, and lasting only seconds. He has been getting chiropractic manipulation of his neck and he thinks this is helping. He had a CT and an MRA brain done, which were read as normal. He denies other visual symptoms and migraine accompaniments. He denies any change to his appearance.

| | |
|---|---|
| *Past medical and ocular history*<br>Hypothyroidism<br>Seasonal allergies<br>Irritable bowel syndrome<br>Atrial fibrillation | *Past surgical history*<br>Inguinal hernia repair 15 years ago |
| *Medications*<br>Levothryoxine<br>Fish oil capsules<br>Multivitamin<br>Flucatisone nasal spray<br>Olapatadine<br>Aspirin | *Family history*<br>Father—irritable bowel syndrome<br>Mother—stroke, heart disease, macular degeneration<br>2 brothers—hypertension |
| *Social history*<br>Former smoker none for 27 years<br>2–3 drinks, 2–3 times per week<br>No drug use<br>Married | *Review of systems*<br>Post nasal drip<br>Knee and hip pain<br>Mild scalp tenderness<br>No weight loss, jaw claudication, malaise |

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| Examination |
|---|
| *Acuity with correction*<br>Right eye: 20/20<br>Left eye: 20/20 |
| *Pupils*<br>Equal, brisk, no afferent pupillary defect |
| *Intraocular pressure*<br>Right eye: 18 mmHg<br>Left eye: 24 mmHg |
| *External exam*<br>No swelling, normal temporal arteries, no tenderness to palpation of trochlea, supraorbital or infraorbital foramina |
| *Eye alignment*<br>Normal |
| *Slit lamp examination*<br>Normal, no cells, deep anterior chamber |
| *Visual field*<br>Normal |
| *Fundus examination*<br>Epiretinal membrane RE, single microaneurysm LE |
| *Neurologic examination*<br>Normal |

## Discussion

### *Ophthalmic Perspective—Dr. Lee*

At first glance, it seems like he is describing headaches brought on by movement, which might make one think of migraine (Case 19), which is often made worse with movement. However, he is older to have a first migraine, migraines do not last seconds, and he denies other symptoms that accompany migraine. Low intracranial pressure can lead to headaches when a patient is sitting or standing but resolve with lying down. Usually, the headache persists while the patient is upright and that is not what he is describing. Pain lasting seconds might be consistent with neuropathic pain. Classically, neuropathic pain is described as electric shock like, but he describes an ache. He notes that chiropractic manipulation has made him better, so I would palpate over his greater occipital nerve (Case 23). It may be a red herring. He notes mild scalp tenderness, but no other symptoms of GCA (Case 32). I might consider a sed rate and CRP, if I felt that his story were strong enough after talking to him. If the values were markedly elevated, then I would pursue a temporal artery biopsy. If they were normal or mildly elevated, then I would probably observe from that perspective. If they were markedly high, then I would start prednisone and pursue a temporal artery biopsy. His eye pressure is elevated, but not to the degree that this would cause pain. Also, angle closure glaucoma (Case 5) would not be bilateral and simultaneous and would last for longer periods of time.

Boiling it down, he really sounds like it is more commonly associated with effort or Valsalva. There is an entity known as cough headache. This can be a benign phenomenon or relate to an Arnold Chiari malformation (Fig. 29.1), posterior fossa lesions, or aneurysms. Typically, these lesions would give more significant headache

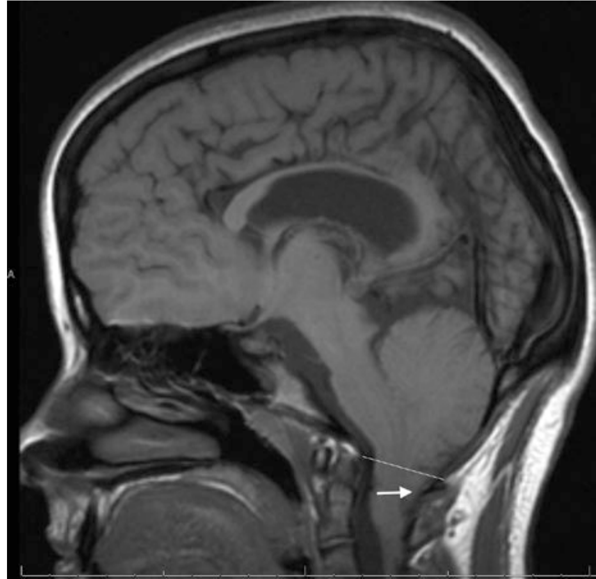

**Fig. 29.1** Sagittal T1 MRI showing significant tonsillar herniation. Note the line signifying the foramen magnum. The tonsils (*arrow*) are well below this level consistent with an Arnold Chiari malformation

symptoms—worse pain, longer durations. I would look to see if his imaging shows any issues. If he has a lesion, then I would consider referral to neurosurgery.

## *Neurologic Perspective—Dr. Digre*

This is a short headache brought on by cough or straining in an older individual with presumably NO previous headache disorder. This is a headache to pay attention to. Traction on the dura can cause these new headaches. The headaches are brief—too short for many different headache types—maybe like ice pick headache or SUNCT (this SUNCT while brief has conjunctival injection and tearing, Case 28). See Table 24.1 where we go through the differential diagnosis of short headaches.

One of the prominent features of this headache is that it is precipitated by coughing or Valsalva maneuver. There is a primary headache disorder (meaning there is no other cause to it) called Primary Cough Headache! It is sometimes also known as Valsalva maneuver headache (see Table 29.1). While primary cough headache is a rare headache disorder it can localize around the eyes. It is most often bilateral and sometimes posteriorly and interestingly this headache hits people over 40 or 50.

However, there is caution with cough headache. Look for a secondary cause! The one diagnosis you do not want to miss with cough headache is a Chiari malformation since up to 40% of individuals presenting with cough headache can have this diagnosis. Other posterior fossa lesions like tumors should be ruled out. So this guy deserves an MR not a CT scan to look at the cranial cervical junction. In addition, be careful not to miss intracranial hypotension (Case 31) that can look a lot like a Chiari 1. Other causes of a cough headache include pinealoma, basilar impression, subdural hematoma, brain tumor, midbrain cyst, and pituitary tumor. Treatment of these secondary or symptomatic cough headaches is dictated by the underlying pathology.

**Table 29.1** ICHD-3 beta Primary Cough Headache

| *Diagnostic criteria:* |
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| (A) At least two headache episodes fulfilling criteria BD |
| (B) Brought on by and occurring only in association with coughing, straining, and/or other Valsalva maneuver |
| (C) Sudden onset |
| (D) Lasting between 1 s and 2 h |
| (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

Treatment of primary cough headache has been indomethacin—sometimes in higher doses. Acetazolamide, amitriptyline, naproxen, and propranolol have been reported to be helpful in some.

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

This is an uncommon cause of headache, but requires a careful history. The patient should have an MRI and MRA to evaluate for Chiari malformations, posterior fossa lesions, or aneurysm. We would recommend giving contrast, since one can also look for thickening and enhancement of the dura mater seen in spontaneous intracranial hypotension.

### *Follow-up*

I did not have his scans, and so I called radiology where he had it done and asked them to look at the source images for an Arnold Chiari malformation or other posterior fossa lesions. There were none present. Therefore, this clinical presentation would be consistent with primary cough headache (formerly known as benign cough headache). This entity often lasts seconds at a time, is more common in men, and does not occur under the age of 40 years. His description is a little different in that most patients describe sharp pain. It is associated with a normal MRI and mild symptoms. It may benefit from the use of indomethacin or acetazolamide, but the patient was not interested. This often spontaneously resolves over the course of several years. The patient was given reassurance and will follow-up with his primary eye doctor regarding the epiretinal membrane, elevated eye pressure, and single microaneurysm in the LE. *Final diagnosis: Primary cough headache*.

## For Further Study

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