Case 31

History of Present Illness

A 32-year-old body builder and car salesman lifted weights every morning. He had no previous headache or eye problems. One day while at work in his usual routine, he developed a relatively acute, severe headache with pressure behind his eyes. He noticed if he would lie down, it would abate, but within minutes of standing up, he had a severe headache and the pressure feeling behind his eyes would begin. He also complains of neck pain. He has minimal light sensitivity and mild sound sensitivity—and some muffling to his hearing, but no nausea. The pain does worsen with activity—and he stopped lifting weights. He tried to sleep it off, but when it did not resolve after 2 weeks, he began missing work and was referred for further evaluation. Recently, he has noted fleeting diplopia—which is side by side, and only present in the distance—and not all of the time.

Past medical and ocular history History of MVA at age 18—no loss of consciousness	*Past surgical history* None
Medications None	*Family history* Non-contributory
Social history Married; used to smoke but quit years ago. Drinks socially on occasion	*Review of systems* Since the headache started he has been trying to sleep more

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Examination
Acuity with correction Right eye: 20/20 Left eye: 20/20
Pupils Equal and no RAPD
Intraocular pressure Right eye: 14 mmHg Left eye: 14 mmHg
External exam Normal
Eye alignment He has a mild comitant esophoria of 4–8 diopters on right and left lateral gaze
Slit lamp examination Normal
Visual field Normal to confrontation
Fundus examination Normal
Neurologic examination Normal

Discussion

Neurologic Perspective—Dr. Digre

This is a new onset of a daily headache in someone who has not had headaches in the past—he requires further work up and questioning. While this could be "new daily persistent headache"—a diagnosis of a headache starting one day and never going away, the real clue in this case is the positional nature of his headache. Anytime there are positional headaches, one has to consider a low cerebrospinal fluid volume or low intracranial pressure. The most common cause is after a lumbar puncture, but these positional headaches can start spontaneously at any time. Eye pain does occur with these headaches as well, but is often a dull ache—and not the primary complaint. Other things can cause a positional headache too. Individuals who have had a Chiari malformation surgery, a large dural sac, colloid cyst of the third ventricle, post-coital headache, cardiac cephalgia (eye pain and headache with upright position and exertion, relieved by rest), and postural orthostatic tachycardia syndrome (POTS) (usually seen in young women with striking tachycardia after standing up for a while) can have positional headaches. These headaches are commonly mis-diagnosed as tension-type headache. The diplopia is not rare—probably coming from tugging on the sixth cranial nerve as the brain stem slumps downward toward the foramen magnum.

While trauma is obviously a risk factor for developing these headaches, sports such as golf and weight lifting, coughing from a upper respiratory infection, chiropractor

Table 31.1 ICHD 3 beta: Intracranial hypotension

Description: Orthostatic headache caused by low cerebrospinal fluid (CSF) pressure of spontaneous origin. It is usually accompanied by neck stiffness and subjective hearing symptoms. It remits after normalization of CSF pressure
Diagnostic criteria:
(A) Any headache fulfilling criterion C
(B) Low CSF pressure (<60 mm CSF) and/or evidence of CSF leakage on imaging
(C) Headache has developed in temporal relation to the low CSF pressure or CSF leakage, or has led to its discovery
(D) 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

manipulation, fishing and even yoga have been reported with its development. Even minor trauma like sitting on a 4-wheeler can induce it in certain individuals. It is thought that people with connective tissue problems like Marfan's and Ehlers Danlos syndromes are more susceptible.

The diagnosis is usually made by history and with imaging (Table 31.1). MRI shows a sagging brain including cerebellar tonsillar herniation, optic nerve and chiasmal downward displacement, and striking meningeal enhancement. The diffuse meningeal enhancement seen on MRI, reported just over 10 years ago, has become one of the key diagnostic imaging features. Since meningeal enhancement alone, however, can be a sign of other conditions causing headache (meningitis, meningeal carcinomatosis, neurosarcoidosis, subarachnoid hemorrhage), look for other findings. The other clear feature is the descent of the brain and brain stem, including: cerebellar tonsillar herniation, reduced size of the pre-pontine cistern, inferior descent of the optic chiasm, and descent of the iter (or the aqueduct opening). Ventricular size is on the smaller size in some and reverts to normal after normalization of pressure. Enlargement of the pituitary gland has also been shown. Engorgement of the venous plexus and spinal veins also occurs.

In this disorder, the spinal fluid pressure is not invariably low. Less than 50% of individuals in many had pressures of less than 40 mm CSF—and many are normal—so reliance on pressure alone is inadequate. There can be lymphocytic pleocytosis and increased CSF protein (up to a 1000 mg/dL in some cases). Once the diagnosis has been made—treatment with a blind blood patch is often recommended since many individuals will have complete resolutions of their symptoms.

However, if that fails, then looking for the leak is the next step and this is sometimes tricky. The first imaging test can be high-resolution MRI, and rarely a leak can be found. A CT myelogram is usually recommended, from the base of the skull through the lumbar sacral regions. MR gadolinium myelogram has also been recommended.

Often medical treatment with IV fluids, corticosteroids, and IV caffeine has been successful in treating the headache. Most frequently a directed blood patch is required. The cause of the headache is more likely to be related to the volume and not the pressure. Many complications of untreated intracranial hypovolemia or hypotension occur including: subdural hematomas, stroke, central herniation syndrome with stupor and coma.

Ophthalmic Perspective—Dr. Lee

I think it is important to note that the diplopia is not positional in intracranial hypotension—meaning the double vision does not vary as quickly as the headache. Additionally, not all patients have double vision or a sixth nerve palsy. So, if the patient merely has eye pain or headache, then the diagnosis is really predicated on the history of positional headache. Unless of course, you have an MRI and this shows the dural thickening and enhancement, then the diagnosis may be suggested by the radiologist.

Orbital varices can cause proptosis that varies with head position or Valsalva, but these are not typically painful. Patients with autonomic dysfunction and pre-syncope may develop transient bilateral visual loss with standing. Associated symptoms can include lightheadedness, photopsias, and ataxia. Headache would be very unusual in this circumstance. Patients with Arnold Chiari malformations could develop headache with Valsalva or cough (Case 29), but this should be easily distinguishable from the positional headache of low ICP.

When confronted with this diagnosis, it is best to send the patient for a blood patch or to neurology to identify a leak using the imaging tools Dr. Digre mentioned. These patients most often have a leak from the area of a spinal root.

Non-ophthalmic/Non-neurologic Perspective

These positional headaches can occur in the primary care provider office and the history is not always straight forward. I have had patients come to my clinic—and when I walk in the room they are lying on the floor or on the examining table because they are more comfortable. This is a great clue to this disorder. The new headache is really a tip off too—and always asking about positional changes helps to make this diagnosis. Sometimes if the pain has been present for a long time, ask about what the pain was like at the very onset of the new daily persistent headache; after a long period of time, the positional aspect of this pain can go away.

If the patient has diplopia—have them look at the end of the room at a target and see if by cross covering their eyes you pick up an eso deviation (i.e., the eye moves from in to out) and especially look for an eso in right and left gaze. Also check for downbeating nystagmus because this can occur with this type of headache and eye pain.

Follow-up

Imaging showed classic signs of intracranial hypovolemia with slumping of the posterior fossa and tonsillar herniation, enlarged pituitary gland, axial images showed compression of the midbrain, and smooth dural enhancement (see Fig. 31.1).

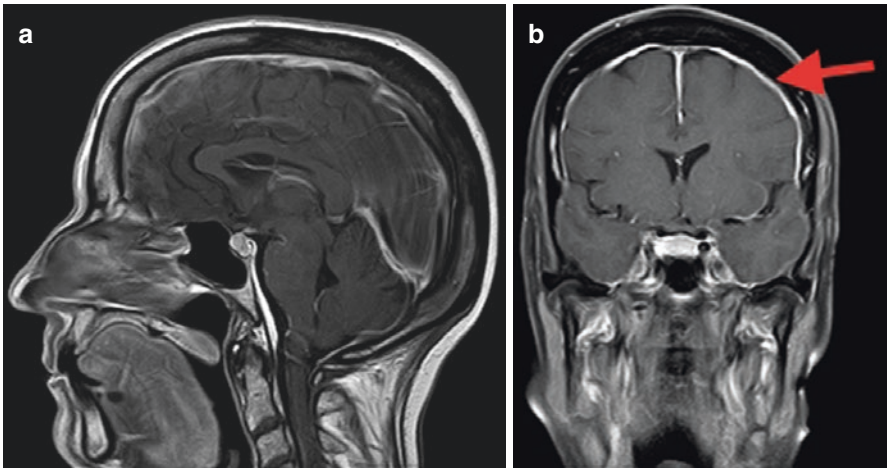

Fig. 31.1 (**a**) Sagittal T1 MR scan showing tonsillar descent, dilated superior sagittal sinus and enlarged pituitary; (**b**) coronal postgadolinium T1 shows marked thickening and enhancement of the dura (*arrow*)

We did a blind blood patch with good relief of the headache and eye pain, but since he still had intermittent pain, we repeated the blood patch with a larger volume and the symptoms resolved. We recommended that he hold off on weight lifting for about 3–6 months so that he did not have a recurrence. *Final diagnosis: spontaneous intracranial hypotension.*

For Further Study

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2. Mokri B. Spontaneous low pressure, low CSF volume headaches: spontaneous CSF leaks. Headache. 2013;53(7):1034–53.
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