Case 36

History of Present Illness

A 28-year-old woman was referred for eye pain and headaches. She had been to the emergency room in 3 different hospitals for headache and eye pain. Each time she was treated with a migraine cocktail without success. She was referred now for a dilated examination to look for papilledema. She had a family history of migraine, and she herself had occasional migraines. However, this headache and eye pain started rather abruptly and she thinks it is different than her migraines. She has some pain with eye movement and a pressure feeling behind her eyes. The headache is holocranial with pain into her neck and between her shoulder blades. She occasionally has a whooshing noise in her head—especially at night when it is quiet.

Past medical and ocular history Hypothyroidism Obesity	*Past surgical history* Tonsillectomy at age 8
Medications Multiple vitamin Synthroid	*Family history* Mother has migraine
Social history She does not smoke or drink	*Review of systems* Trouble sleeping at night

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Examination
Acuity with correction Right eye: 20/20 Left eye: 20/20
Pupils Equal pupils, No Relative afferent pupillary defect
Intraocular pressure Right eye: 15 mmHg Left eye: 15 mmHg
External exam Normal
Eye alignment Normal
Slit lamp examination Normal
Visual field Normal to confrontation; Formal visual fields show enlarged blindspot and scattered defects
Fundus examination Bilateral optic disc swelling
Neurologic examination BMI is 45 Normal neurological

Discussion

Neurologic Perspective—Dr. Digre

This young woman has a change in her headache and has eye pain. What is more, she has been to the emergency room three times for help and no one bothered to look at her optic discs. I have seen this frequently, in fact, if I hear someone has been to the emergency room several times with a headache or eye pain that does not go away, I immediately consider increased intracranial pressure and idiopathic intracranial hypertension (IIH). Neurologists often miss papilledema since they are not comfortable using an ophthalmoscope and they rarely dilate the patient. Another problem is overdiagnosing IIH especially if someone has anomalous optic discs and headache.

Of course with papilledema, one first has to consider other causes of intracranial hypertension and I would order an MRI and MRV or CTV. The MR can be helpful since it may show signs of increased intracranial pressure as well. These include: an empty sella, dilated optic nerve sheaths, flattening of the posterior globe, and often dilated spaces around the foramen ovale as well as Meckel's cave. It also will exclude a mass lesion. The venogram is also critical since individuals with venous thrombosis can present identically to the primary or idiopathic intracranial pressure; the treatment of venous sinus thrombosis is different and therefore important to exclude.

After imaging, a lumbar puncture, measuring the opening pressure is important as well as checking the fluid to be sure there is no evidence of meningitis. The opening pressure is also important to measure correctly—usually in the lateral decubitus

Table 36.1 IIH headache according to the ICHD3 beta

Headache caused by idiopathic intracranial hypertension (IIH), usually accompanied by other symptoms and/or clinical signs of IIH. It remits after normalization of cerebrospinal fluid pressure
Diagnostic criteria:
(A) Any headache fulfilling criterion C
(B) Idiopathic intracranial hypertension (IIH) has been diagnosed, with CSF pressure >250 mm CSF (measured by lumbar puncture performed in the lateral decubitus position, without sedative medications, or by epidural or intraventricular monitoring)
(C) Evidence of causation demonstrated by at least two of the following:
1. Headache has developed in temporal relation to IIH, or led to its discovery
2. Headache is relieved by reducing intracranial hypertension
3. Headache is aggravated in temporal relation to increase in intracranial pressure
(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

position with legs outstretched—being sure to have the patient relaxed and not having a Valsalva maneuver.

Most neurologists do not have the capability of performing visual fields. It is extremely important if someone has papilledema to get a formal visual field. Visual acuity alone is not enough to follow these individuals. We get optic disc photos to follow the papilledema and sometimes OCT is also helpful in order to see if the swelling is decreasing.

To diagnose the headache associated with IIH see the ICHD 3beta (Table 36.1). There are many interesting caveats about diagnosing IIH-related headache. First, when papilledema is not present, can you still call it IIH if the pressure is only mildly elevated? I see many patients with clear migraine and elevated pressures being called IIH. I think IIH without papilledema is not common. I suggest that we do not worship the opening pressure alone. Second, notice in the definition that the headache leads to the discovery of the IIH—such as is in this case. Also, the finding of the headache going away with the lumbar puncture is not 100% either. Many individuals with migraine say that their headaches improve after lumbar puncture.

Treatment of IIH is usually weight loss—and this woman is obese. I recommend five vegetables a day and walking 10,000 steps or swimming as exercise. For some, a dietician is very helpful in setting up a weight loss program. We also use acetazolamide. The recently completed Idiopathic Intracranial Hypertension Treatment Trial (IIHTT) showed that acetazolamide along with weight loss was more efficacious over weight loss alone.

Treating the eye pain and headache in IIH can really be a challenge, since just lowering the pressure alone may not treat the headache and eye pain. The best treatment for the headache is usually acetazolamide and a migraine preventative such as topiramate, amitriptyline (although weight gain can occur with this), or other migraine preventive. This woman also has a history of underlying migraine—individuals with IIH have a higher incidence of migraine (60%)—which is much higher than in the general public (18% of women). The migraine headache can be treated acutely with triptans or other migraine-specific medications.

Ophthalmic Perspective—Dr. Lee

You can titrate the acetazolamide up to 4 g/day according to the IIHTT. If you are not comfortable with that, then perhaps go up to 2.5 g/day (in divided doses). If that does not help the eye or head pain, then I would send the patient to a neurologist to help manage headache but the acuity, field, and nerves need to be monitored by an ophthalmologist.

Generally, I do not advocate for a shunting procedure in patients with severe headache alone. Although uncommon, there is risk to having a shunt, so I typically reserve a shunt for those with moderate to severe visual loss. I have seen patients go to a neurosurgeon for a shunt for the pulse synchronous tinnitus and headache despite my protests.

In some cases, the patient complains of persistent headache despite the papilledema resolving. They insist that this is due to the intracranial pressure being high. Sometimes, I will repeat the lumbar puncture to demonstrate to the patient that the pressure is not high. In other cases, the patient is willing to visit a neurologist to manage headache. I also have several patients who just want to stay on low-dose acetazolamide. I do not have a problem with this, since we know that this was used to treat glaucoma and has a reasonable safety profile over many years of treatment. It also may help some people with migraine.

If a patient becomes pregnant, topiramate is contraindicated. Acetazolamide is category C, but there have been a number of patients who have taken acetazolamide while pregnant. We ask patients on acetazolamide to avoid pregnancy. If they become pregnant, we may discuss stopping the medication for the first trimester or staying on it. Finally, patients with a sulfa antibiotic allergy do not have cross reactivity with sulfa diuretics, but anyone with an allergy to one medication may be allergic to another.

Non-ophthalmic/Non-neurologic Perspective

Finding papilledema can be challenging for the primary care physician too. Dilating the patient with 0.5% tropicamide is helpful in better seeing the optic disc. Recently photographs with a non-mydriatic camera have helped primary care and emergency room physicians see the optic discs better. There are also new phone apps and devices that allow one to take a picture of the fundus.

If papilledema is seen—MR and MRV or CTV is important to order along with a lumbar puncture. A primary care physician may be the best place to assist the patient with weight loss strategies that are known to be helpful along with acetazolamide.

Headache and eye pain with IIH is so common. Over 50% of patients with IIH had previously diagnosed migraine headache. It is also the headache that causes a reduced quality of life. So treating the migraine is really important in the treatment of IIH.

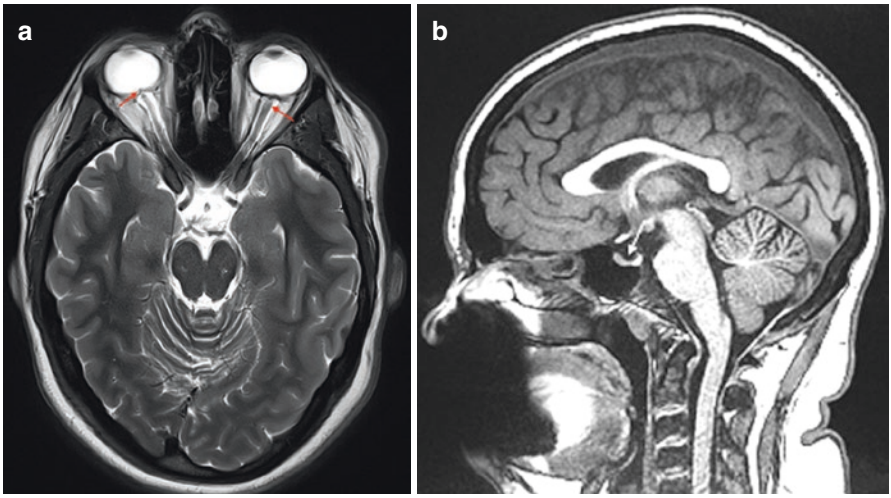

Fig. 36.1 (**a**) Axial T2 MRI shows evidence of increased intracranial pressure including flattened posterior globe and dilated optic nerve sheaths (*arrows*). (**b**) Sagittal T1 MRI shows a partially empty sella (*arrow*)

Follow-up

This woman had a normal MR and MRV except for an empty sella and other findings of intracranial pressure (see Fig. 36.1). We started her on 500 mg acetazolamide twice daily and encouraged weight loss. Her most acute headache improved after the lumbar puncture, and she now is followed with resolution of the papilledema, but her headaches persist. I have kept her on acetazolamide and topiramate. That combination can cause lowered bicarbonate levels, tingling, taste changes, and weight loss, but by and large it is well tolerated. If intolerable, then sodium bicarbonate 1300 mg twice daily can improve these side effects. Another consideration is electrolyte replacement drinks. I follow electrolytes, which so far have been normal. *Final diagnosis: idiopathic intracranial hypertension*.

For Further Study

1. Fisayo A, Bruce BB, Newman NJ, Biousse V. Overdiagnosis of idiopathic intracranial hypertension. Neurology. 2016;86:341–50.
2. NORDIC Idiopathic Intracranial Hypertension Study Group Writing Committee, Wall M, McDermott MP, Kieburtz KD, Corbett JJ, Feldon SE, Friedman DI, Katz DM, Keltner JL, Schron EB, Kupersmith MJ. Effect of acetazolamide on visual function in patients with idiopathic intracranial hypertension and mild visual loss: the idiopathic intracranial hypertension treatment trial. JAMA. 2014;311(16):1641–51.
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