Idiopathic Intracranial Hypertension in a 24-Year-Old Woman

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A 24-year-old woman presented with a history of severe headache and blurry vision.

Case

A 24-year-old woman presented to the ED for evaluation of a 3-week history of worsening headache and a 5-day history of increasingly blurry vision. The patient stated that she had initially contacted her primary care physician, but instead presented to the ED because he had no open appointments until the following week and recommended that she go to the ED.

The patient described her headache as a pulsating and throbbing pain over her entire head, which only mildly improved after taking over-the-counter (OTC) ibuprofen. She further noted that her headache was somewhat worse when lying down, and reported the sensation of hearing her own pulsating heartbeat in her ears.

The patient had no personal or family history of migraines, tension headaches, aneurysms, clotting disorders, bleeding disorders, or renal disease, and stated that she had never experienced this type of headache before. She denied photophobia, phonophobia, neck stiffness, fever, vomiting, cough, numbness or weakness in her extremities, or pain anywhere else in her body.

Over the past 5 days, the patient noticed her vision had become increasingly blurry. She was not on any prescription

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Table. Differential Diagnosis for Increased Opening Pressure on Lumbar Puncture

Meningitis/encephalitis
Cerebral edema
Intracranial mass lesion
Subarachnoid hemorrhage
Venous sinus thrombosis
Pseudotumor cerebri
Guillain-Barré syndrome
Jugular vein compression (from neck mass or postsurgical scarring)
Meningeal carcinomatosis
Choroid plexus tumor (causes overproduction of cerebrospinal fluid)

medications, stating the only medication she used was occasional OTC ibuprofen. She had no known allergy to medications and denied smoking or recreational drug use; she admitted to occasional alcohol consumption.

The patient resided with her husband, who had no similar symptoms. Physical examination showed an obese woman (height, 5 ft 6 in; weight, 195 lb; body mass index, 32 kg/m^2), lying supine in apparent discomfort. Vital signs at presentation were all normal, and oxygen saturation was normal on room air.

A bedside ocular examination showed 20/100 in both eyes while using glasses; no visual field cuts or obvious central scotoma was present. The patient was alert and oriented to time and place. The neurological examination showed intact cranial nerves, 5/5 strength in all extremities, intact sensation in all extremities, no pronator drift, negative Romberg test, and a normal gait. Fundoscopic examination revealed mildly blurred medial optic discs bilaterally. The rest of the physical examination was normal.

Discussion

Pseudotumor cerebri, more commonly referred to as idiopathic intracranial hypertension (IIH), is characterized by increased intracranial pressure (ICP) with no explanatory findings on imaging studies or in cerebrospinal fluid (CSF) analysis, and may be accompanied by symptoms of chronic headache, tinnitus, papilledema and progressive vision loss caused by optic nerve damage.[1] Though historically IIH was referred to by several other names, including "benign intracranial hypertension," the condition is not benign—when untreated, IIH can cause chronic disabling headaches and permanent vision loss.[1]

Clinical Course

The clinical course of IIH is unpredictable: In some patients, vision loss occurs gradually over the course of several weeks, while in others, loss occurs over a several month period. There are also patients with IIH who do not experience any alteration or loss of vision. Furthermore, some patients will experience permanent resolution of symptoms after a single lumbar puncture (LP); others have symptom recurrence after less than 24 hours; and some patients spontaneously remit on their own with no treatment whatsoever.[1-4]

Etiology

In the United States, IIH is a rare cause of headache, occurring in just 1 person per 100,000 annually.[1] Though 90% of IIH cases occur in obese women of childbearing age, the etiology of IIH is unknown. Lumbar puncture usually alleviates the patient's headache, but the CSF pressure typically returns to its pre-tap levels after a few hours.[4,5] Neither CSF overproduction nor insufficient CSF resorption is responsible for causing IIH. One theory on the etiology of IIH proposes its cause to be due to a congenital malformation of the

venous sinuses. This theory would explain why the symptoms so closely mimic those of venous sinus thrombosis, and why some IIH patients experience relief of symptoms after placement of a venous sinus stent.[2]

Symptoms

As noted previously, the most common symptom of IIH is headache, which patients usually describe as pressure-like and throbbing, and often involving retro-ocular pain. One feature in over half of patients is pulse-synchronous tinnitus (ie, hearing their own heartbeat in their ears). Eye pain, photophobia, blurry vision, and nausea/vomiting are all common symptoms in IIH, but these symptoms are also present in other causes of headache. The IIH headache might be relapsing and remitting, and can last from a few hours to weeks.[2-4,6]

Diagnosis

Imaging Studies. Noncontrast computed tomography (CT) imaging studies do not typically demonstrate any abnormal findings.[1] Magnetic resonance imaging (MRI) studies show some inconsistent and subtle findings, such as flattening of the backs of the eyeballs, empty sella, or tortuous optic nerves.[1]

Lumbar Puncture. On LP, a very high opening pressure is a hallmark of IIH. An opening pressure <20 cm H_2O is generally considered normal, 20 cm to 25 cm H_2O is "equivocal," and >25 cm H_2O is abnormal.[7] Patients presenting with IIH commonly have an opening pressure that exceeds 200 cm H_2O.[1-3] Extremely high pressures, however, are not required for the diagnosis, but some elevations in opening pressure will always be present.[2,5] With the exception of a high opening pressure, the patient's CSF analysis is normal.

Differential Diagnosis

Idiopathic intracranial hypertension is essentially a diagnosis of exclusion, one that is made after exclusion of all other potential causes of increased ICP (**Table**). Since contrast CT and MRI can identify subtle anatomical deformities and small lesions, their absence on these studies can help establish a diagnosis of IIH.

Venous Sinus Thrombosis. Venous sinus thrombosis is a rare but devastating condition that also cannot be diagnosed from a noncontrast CT but must always be considered in the differential diagnosis of IIH.[8-10] Venous sinus thrombosis is characterized by a clot in one of the large venous sinuses that drain blood from the brain; the clot causes pressure to back up into the smaller cerebral vasculature, eventually inducing either a hemorrhagic stroke from a stressed vessel rupturing, or an ischemic stroke from lack of blood flow to the affected area of the brain. This condition is even more rare than IIH (0.5 cases per 100,000 population), but it can be devastating if missed, carrying a mortality rate as high as 15% in some studies.[11]

Risk Factors

Risk factors known to cause cerebral venous clots include genetic thrombophilias, pregnancy or recent pregnancy, oral contraceptive use, inflammatory bowel disease, severe dehydration, local infection/trauma, and substance abuse. Regardless of risk factors, the most recent guidelines of the American Heart Association/American Stroke Association recommend imaging studies of the cerebral venous sinuses for any patient presenting with new-onset symptoms suggestive of IIH (Class 1, Level of Evidence C).[11] The two imaging options for evaluation of the cerebral venous sinuses are CT venography or MR venography. Since the 2013 American College of Radiology Appropriateness Criteria do not indicate a preference of one modality over the other, the choice of can be left to your radiologist.[12]

Patient Disposition

Patients with IIH typically do not require inpatient admission. Only about 3% of IIH patients will have a fulminant course of rapid-onset of vision loss, but even

the most severe and acute cases will deteriorate over weeks, not hours or days.[13] Nevertheless, close neurology follow-up is essential. If rapid and thorough outpatient neurological care is unavailable, admission is required.

Management

Not every patient with IIH experiences amelioration or resolution of symptoms following an LP; moreover, there is no clear way to differentiate patients who will experience therapeutic effects from LP from those who will not. Serial LPs as treatment for IIH have been discussed in the literature, but a ventriculoperitoneal shunt is a more practical approach in patients who do not respond to an initial LP.[2,14]

CSF Volume. The volume of CSF that can be removed safely may be 15 to 25 mL or more. A 1974 paper by Johnston and Paterson[15] described five pseudotumor patients whose CSF was drained until their pressure had normalized; the amount removed varied from 15 to 25 mL, without adverse effects. A 1975 case series by Weisberg[6] described safe removal of up to 30 mL of CSF in pseudotumor patients—the precise amount removed was determined by that which was necessary to lower the CSF pressure into the normal range. In 2007, a case report by Aly and Lawther[16] of a pregnant woman with IIH describes twice weekly LP drainage of 30 mL.

There is nothing in the current literature to suggest that removing 10 to 30 mL of CSF instead of the 4 to 8 mL typically drawn in a diagnostic LP is going to pose any risk to the patient. The main complication associated with therapeutic LP is post-LP headache.[5,17,18] There are currently no studies documenting outcomes after specific amounts of CSF removal.

Lifestyle Modifications: Weight Loss. No prospective, randomized controlled trials have proven weight loss to be effective in ameliorating the symptoms of IIH; however, several studies have found that rapid weight loss—whether through aggressive dieting or gastric bypass surgery—can improve symptoms dramatically within several months.[19,20] One small study by Johnson et al has suggested that a 6% weight reduction is associated with marked improvement in papilledema.[21]

Pharmacotherapy. The accepted first-line medication to alleviate symptoms of IIH is acetazolamide, and its use is supported by a recent randomized controlled trial conducted by the Neuro-Ophthalmology Research Disease Investigator Consortium (NORDIC).[22] Most neurologists will administer a starting dose of acetazolamide 500 mg twice a day, and then increase the dose until symptoms are controlled or adverse effects appear (eg, fatigue, nausea/vomiting/diarrhea, electrolyte abnormalities, kidney stones) that contraindicate further dosage increases. In the NORDIC trial, patients were given up to 4 g of acetazolamide daily.[22]

Other medications, including loop diuretics and corticosteroids, should not be

used except under the direct supervision of a neurologist.[2,14]

Refractory Cases

A patient who fails conservative treatment should be referred to a neurosurgeon for placement of a CSF shunt, optic nerve sheath fenestration, or placement of a venous sinus stent.[23]

Case Conclusion

After a noncontrast CT of the head was interpreted as completely normal, an LP was performed with the patient in the left lateral recumbent position. The opening CSF pressure exceeded 55 cm H_2O (the upper limit of the manometer). The CSF was clear, and opening pressure was rechecked after each 5 mL draw. After 15 mL had been removed, the patient reported a sudden, dramatic disappearance of her headache and clearing of her vision. After 19 mL of CSF had been removed, the CSF pressure had dropped into the normal range (<20 cm H_2O), and the procedure was ended.

To definitively rule out venous sinus thrombosis, a CT venogram was performed in the ED, and interpreted as normal. All other CSF results (cell count, protein, glucose, and gram stain) were normal. After complete resolution of the patient's symptoms, she was discharged home with a prescription for acetazolamide 500 mg twice daily and instructions to follow-up with a neurologist within 48 hours. At discharge, the patient also received weight-loss counseling and was instructed to return immediately to the ED if her headache recurred or if she experienced any new neurological symptoms.

Summary

Idiopathic intracranial hypertension, also referred to as pseudotumor cerebri, is a rare but potentially vision-threatening cause of headache. Patients with signs and symptoms of IIH often initially present to the ED for evaluation and management. While the etiology of IIH is poorly understood, its clinical picture is unique: elevated ICP (sometimes markedly so) with no other significant findings on noncontrast head CT or CSF analysis. Venous sinus thrombosis, a life-threatening mimic of IIH, must always be included in the differential diagnosis.

Idiopathic intracranial hypertension is initially treated with rapid weight loss and acetazolamide. Many patients experience instant, though sometimes only transient, symptom relief from LP. No definitive studies to support any specific approach, including "therapeutic lumbar punctures." The condition is rarely fulminant, and hospital admission is not typically required as long as urgent outpatient neurology follow-up is available.

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