CLINICAL VIGNETTE

Idiopathic Intracranial Hypertension: A Case Review

Suruchi Kelly, M.D.

Case Presentation

A 19-year- old female presents with 3-month history of gradually worsening left sided headache described as throbbing in nature and ranging from 6/10 to 10/10 in severity. She experiences it daily and it can be accompanied by lightheadedness and blurry vision. She has no history of recent head injury. She has no accompanying photophobia but does get nausea and vomiting with the headaches.

Her family history is significant for her father who had a hemorrhagic stroke due to uncontrolled hypertension. She is a nonsmoker and overweight but otherwise healthy.

Her BP is 116/86, pulse 86, temp 98.1 and resp 18. Her height 5 ft 8 inches, weight 260 lbs and BMI 39.5.

The general exam was unremarkable. She was alert and oriented and her neurological exam including cranial nerves 2-12 was grossly normal without evidence of papilledema.

An MRI/MRA showed no mass lesion, infarct or hemorrhage, and no significant intracranial stenosis or aneurysm.

Twelve days later, the patient presented to the ER with worsening headache and underwent lumbar puncture. Opening pressure was 26 cm. She reported 80% improvement in her headache after the lumbar puncture. Cell counts and cultures on CSF fluid were negative.
She was seen by Neurology and started on acetazolamide for idiopathic intracranial hypertension.

Discussion

Idiopathic intracranial hypertension (IIH) is a rarely occurring diagnosis in the setting of headache but can be associated with obesity, most commonly in women of childbearing age. The etiology is still unknown. By definition, the disease is defined by isolated raised intracranial pressure (ICP) that is unrelated to an intracranial disorder, meningeal process or cerebral venous thrombosis. It is a preferred term over pseudotumor cerebri, which can include patients with other causes of raised ICP such as cerebral venous thrombosis. Because of the rising prevalence of obesity, the economic costs of this disease are becoming more significant[1]. IIH is diagnosed using the Modified Dandy Criteria[2].

1) headache, nausea and vomiting, transient visual obscurations, papilledema
2) lack of focal neurologic signs, except possible sixth nerve paresis
3) cerebrospinal fluid opening pressure >26 cm of water, without abnormal cell counts or cytology.
4) Normal neuro-imaging excluding cerebral venous thrombosis

Some subsets that have an increased risk of visual loss include men, African-Americans, those with obstructive sleep apnea and those with a fulminant course. Certain medications have been thought to precipitate IIH, including tetracycline and its derivatives, cyclosporine, lithium, nalidixic acid, nitrofurantoin, oral contraceptives, levonorgestrel, danaxol and tamoxife[1]. It is unclear whether the association with obstructive sleep apnea is a common pathophysiologic link with obesity or an independent risk factor[2].

Classically, IIH was not associated with any imaging abnormalities but in more recent years, progress in radiologic imaging has led to a change in the description of IIH. Findings indicative of IIH include flattening of the posterior poles of the eyes, dilation and tortuosity of the optic nerve sheaths, empty sella turcica and stenosis of one or both of the transverse cerebral venous sinuses. Although MRI findings may suggest the presence of elevated intracranial pressure and the diagnosis of IIH, they are not predictive of visual outcomes[3].

Treatment options include the initial diagnostic lumbar puncture, which has been in shown to be therapeutic in some IIH patients[1]. Weight loss is a critical intervention as even a 5-10% loss of weight

can resolve symptoms of headache or visual loss, and maintaining the weight loss minimizes risk of recurrence. For those that are unsuccessful at weight reduction by conservative options, bariatric surgery has been shown to successfully lower intracranial pressure and symptoms[1,4].

Carbonic anhydrase inhibitors, such as acetazolamide, have been shown to decrease CSF production and are therefore the treatment of choice in IIH. A recent clinical trial studied acetazolamide in IIH patients maintained on a low-sodium weight reduction diet and was found to modestly improve visual field function but there are no controlled trials to measure effect on headaches[5]. Acetazolamide can have common side effects of paresthesias, reduction of carbon dioxide levels, altered taste sensation and lethargy.

Topiramate and oral steroids have been used as alternative medication options but are less desirable[1].

Surgical options include lumboperitoneal (LP) or ventriculoperitoneal (VP) shunts. Both techniques are considered effective in reducing intracranial pressure but the LP shunts have a lower failure rate and the VP shunts have a lower revision rate. They are usually performed in patients with imminent catastrophic visual loss. Stenting for patients with IIH caused by transverse venous sinus stenosis can also reduce intracranial pressure, but complications such as stent migration, venous sinus perforation, in-stent thrombosis or subdural hemorrhage limits this procedure to patients with bilateral transverse sinus stenosis with refractory symptoms or who cannot undergo more conventional surgical treatments. Optic nerve sheath fenestration has been used to reduce visual symptoms but does not address headache[1].

Conclusion

Our patient had significant reduction in headache and visual blurring on acetazolamide and is pursuing weight loss strategies. She did develop paresthesias in her lips, fingers and toes on the medication as well as metallic taste in the mouth and reduction in her carbon dioxide blood levels. She is currently tolerating acetazolamide 250 mg three or four times daily. She had mild if any papilledema noted by Ophthalmology.

REFERENCES

1. **Biousse V, Bruce BB, Newman NJ**. Update on the pathophysiology and management of idiopathic intracranial hypertension. *J Neurol Neurosurg Psychiatry*. 2012 May;83(5):488-94. doi: 10.1136/jnnp-2011-302029. Epub 2012 Mar 15. Review. PubMed PMID: 22423118; PubMed Central PMCID: PMC3544160.
2. **Thurtell MJ, Trotti LM, Bixler EO, Rye DB, Bliwise DL, Newman NJ, Biousse V, Bruce BB**. Obstructive sleep apnea in idiopathic intracranial hypertension: comparison with matched population data. *J Neurol*. 2013 Jul;260(7):1748-51. doi: 10.1007/s00415-013-6858-6. Epub 2013 Feb 15. PubMed PMID: 23412355; PubMed Central PMCID: PMC3707935.
3. **Saindane AM, Bruce BB, Riggeal BD, Newman NJ, Biousse V**. Association of MRI findings and visual outcome in idiopathic intracranial hypertension. *AJR Am J Roentgenol*. 2013 Aug;201(2):412-8. doi: 10.2214/AJR.12.9638. PubMed PMID:23883223; PubMed Central PMCID: PMC4048553.
4. **Fridley J, Foroozan R, Sherman V, Brandt ML, Yoshor D**. Bariatric surgery for the treatment of idiopathic intracranial hypertension. *J Neurosurg*. 2011 Jan;114(1):34-9. doi: 10.3171/2009.12.JNS09953. Epub 2010 Jan 22. Review. PubMed PMID: 20095788.
5. 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 Apr 23-30;311(16):1641-51. doi: 10.1001/jama.2014.3312. PubMed PMID: 24756514.

Submitted on September 2, 2014