Idiopathic Intracranial Hypertension: Papilledema and Neuro-Ophthalmology Referral Patterns

Given the relatively small number of neuro-ophthalmology specialists in the US, it is incumbent upon neurologists to diagnose and manage IIH.

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DIAGNOSIS AND MANAGEMENT OF IIH

To expand on this subject, we must first address the question of what aspects of a patient history drive clinical suspicion of IIH. Clinical suspicion tends to increase as an increasing number of the following signs/symptoms are present:

1. The patient is an obese female.
2. The headaches tend to be orthostatic with the worst pain occurring upon waking up in the morning, worsening of pain with recumbence, and improvement of pain while upright.
3. The headaches involve transient visual obscurations (TVO). TVO are episodes of sudden loss of vision lasting less than 30 seconds. TVO can occur in one or both eyes, and are followed by full visual recovery. TVO can occur in 68 percent of IIH patients. In addition to TVO, diplopia can also at times be appreciated.
4. The headaches involve vomiting, which is often projectile. After vomiting, many patients experience improvement of their headaches, and patients at times endorse trying to make themselves vomit to hasten headache pain relief.
5. The headaches involve pulsatile tinnitus, which can occur in 52 percent of IIH patients.

The previously used term for IIH is pseudotumor cerebri because these headaches behave similarly to headaches that are induced by a tumor. As such, it is not inappropriate to proceed with MRI imaging to rule out a mass lesion based on clinical suspicion. Insurance companies would be hard pressed not to cover the expense of an MRI of the brain for a patient with new onset (often worsening) headaches that involve characteristics of headaches caused by a brain tumor, pulsatile tinnitus, and unexplained visual disturbances. IIH neuroimaging findings include empty sella turcica, distension of the perioptic subarachnoid space, flattening of the posterior sclerae, protrusion of the optic nerve papillae into the vitreous, and transverse cerebral venous sinus stenosis. Like the absence of papilledema on examination, the absence of these MRI findings should not dissuade further testing and treatment in the appropriate clinical setting.

The diagnostic characteristics for IIH were first outlined by Dandy in 1937, and formulated into a set of criteria by Smith in 1985, known as the modified Dandy Criteria. It has since been updated and then more recently revised. The diagnostic criteria continue to evolve as more is learned about this disorder, especially with the advancement of imaging techniques. Depending on the version of the Dandy Criteria used, papilledema and/or a strict CSF opening pressure cut off may or may not be required to establish the diagnosis. Even with the most recent and strict revised criteria, a diagnosis of IIH can still be established in the absence of papilledema, which is helpful for the subset of patients with otherwise typical IIH who do not have papilledema. Given the various iterations of the diagnostic criteria and the provision to diagnose probable IIH if certain required criteria are not met, the diagnostic interpretation of a given clinical presentation may differ from one clinician to another.

When making a diagnosis of IIH, most headache specialists refer to the International Classification of Headache Disorders 3rd Edition, Beta Version (ICHD-3b) diagnostic criteria, which are listed in Table 1.

According to the ICHD-3b criteria, a diagnosis of IIH can only be made once a lumbar puncture (LP) has demonstrated elevated opening pressure over 250 mm CSF. As such, an LP should not be considered an unnecessary, invasive procedure in the setting of high clinical suspicion of IIH. Again, high clinical suspicion should not solely be an obese woman with headaches. In addition to diagnostic value, an LP can be of therapeutic benefit when performed in the setting of a severe headache. As such, performing routine lumbar punctures in the absence of a headache should be avoided, as the intervention will not be of any therapeutic benefit, and the opening pressure may not be at peak elevation as can occur with a severe headache. Due to the unpredictable timing of flares, it is at times recommended that patients proceed to the emergency department for an urgent diagnostic/therapeutic LP the next time they have a severe headache. A mildly elevated CSF opening pressure should not be dismissed, as CSF pressure can vary over time. Continuous CSF pressure monitoring studies have demonstrated that intracranial pressure elevations tend to occur over a 24-hour period, especially during sleep, which may not be noted with a ‘spot’ lumbar puncture over minutes.

Prior to performing MRI imaging and an LP, it is reasonable to proceed with empiric IIH treatment based on clinical
Article at a Glance

Idiopathic intracranial hypertension (IIH) is a syndrome that involves headaches, visual disturbances, and papilledema on examination. A funduscopic examination should be performed on all patients presenting with suspected IIH, but the ophthalmoscope is a valuable diagnostic tool that is heavily underutilized throughout the practice of medicine. The presence of papilledema supports the diagnosis of IIH, but is not a formal part of the diagnostic criteria for IIH. Empiric treatment with medication to lower intracranial pressure should also be instituted based on clinical suspicion. Given the long wait times for a neuro-ophthalmology consultation and a lack of neuro-ophthalmologists in many parts of the United States, delaying appropriate diagnostic and therapeutic interventions for IIH is not only impractical, but can seriously compromise patient care with at times grave consequences including blindness.

CONCLUSION

A funduscopic examination should be performed on all patients presenting with suspected IIH, and the ophthalmoscope is a valuable diagnostic tool that is heavily underutilized throughout the practice of medicine. For this reason, proper use of the ophthalmoscope is a skill that should be emphasized with all trainees. The presence of papilledema certainly supports the diagnosis of IIH, but is not a formal part of the ICHD-3b diagnostic criteria for IIH. As such, the absence or inability of a provider to appreciate papilledema should not delay further evaluations. In a clinical presentation consistent with IIH (more features than just an obese female with headaches), ruling out an intracranial mass lesion with imaging studies and performing a diagnostic/therapeutic lumbar puncture are appropriate in making the correct diagnosis. Empiric treatment with a medication to lower intracranial pressure should be instituted based on clinical suspicion. Given the long wait times for a neuro-ophthalmology consultation and a lack of neuro-ophthalmologists in many parts of the United States, delaying appropriate diagnostic and therapeutic interventions for IIH is not only impractical, but can seriously compromise patient care with at times grave consequences including blindness.