

Misdiagnosing idiopathic intracranial hypertension

You've got some nerve

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The diagnosis of idiopathic intracranial hypertension (IIH) can be challenging and relies heavily on the proper interpretation of ophthalmoscopy. For many patients, the diagnosis can be straightforward when the typical demographic features, fundus findings of papilledema, neuroimaging signs, and CSF findings are present. In other situations, the diagnosis of IIH may be difficult, particularly when fundus findings are normal, subtle, or complicated by pseudopapilledema (as can occur with optic nerve head drusen). Occasionally, true papilledema may coexist with pseudopapilledema. We must also recognize that a small percentage of patients with IIH will not develop papilledema.^{1,2}

In their retrospective study, Fisayo et al.³ found overdiagnosis of IIH to be common in the large neuro-ophthalmology tertiary referral practice they sampled. In a matter of 8 months, the authors evaluated 165 patients with either a preexisting diagnosis of IIH or with a question about the possibility of an IIH diagnosis. Among patients with the preexisting diagnosis of IIH at the time of referral, 39.5% (34/86) were misdiagnosed with IIH. Inaccurate ophthalmoscopy interpretation and “thinking bias” (e.g., headache in an obese woman is IIH) were the most common errors. In fact, 20% of the patients in this study were given the diagnosis of IIH without an ophthalmoscopic examination. The overdiagnosis of IIH had its consequences in the study cohort, leading to unnecessary tests, medication use, and surgical procedures. Strengths of the study include the large number of patients evaluated by true experts in the field and the ability of the neuro-ophthalmologists to obtain the referring doctors' records in every case.

The article by Fisayo et al. raises a number of important issues about the dying art of the routine performance and accurate interpretation of ophthalmoscopy. In previous articles, the authors of the current study emphasized that the failure of properly performed ophthalmoscopy can lead to diagnostic errors, with misattribution of patients' signs and symptoms to a variety of neurologic and systemic

disorders.^{4–6} This is a particular risk in the emergency department setting. The technical difficulties inherent in using the direct ophthalmoscope, and the challenges associated with interpreting the fundus findings, have largely led to the abandonment of the ophthalmoscope. Negative attitudes by senior clinicians regarding use of the ophthalmoscope have exacerbated this problem.

While one solution may be to have a neuro-ophthalmologist on every street corner, such is not going to be practical or possible. New tools have emerged that may mitigate the misinterpretation and technical challenges of the ophthalmoscopic examination. Fundus photography is now much easier to obtain; the introduction of nonmydriatic cameras has also revolutionized our ability to assess accurately the optic nerve and macula region. In fact, medical students and others greatly prefer nonmydriatic photography to using the ophthalmoscope, noting that it enhances their diagnostic capabilities.⁷ Since it seems unlikely that we are going to have a sudden return to the routine use of the ophthalmoscope by every practicing neurologist, the advances afforded by nonmydriatic fundus photography offer every clinician the opportunity to see the pertinent findings. This technique should be widely adopted, and may now even enable use with handheld/smartphone applications, which enable a dynamic view of the ocular fundus by a video feed. Finally, the development of optical coherence tomography (OCT) has revolutionized our ability to examine the optic nerve head and macula. However, despite the existence of OCT and other new tools, traditional ophthalmoscopy is still not done and has become a lost art within the neurologic examination. The study by Fisayo et al. is a humbling reminder that the culture of relying on MRI scans and cursory neurologic examinations is certain to fail in some cases.

The article has the limitations inherent in a retrospective study.³ It is difficult to judge by just reviewing records what the confidence of a particular diagnosis is by the referring doctor. Therefore, referral bias of the most challenging cases may be likely. In

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some patients, disc swelling may resolve over time and with treatment. There are patients with IIH who do not manifest disc swelling; these patients may be more frequent than once recognized, particularly among those with headache. Vascular headaches commonly occur in the setting of IIH, and many of these patients have chronic daily headaches.^{8,9} There may be variability in the interpretation of neuroimaging findings and some of the misdiagnosis of IIH may reside with the neuroradiologists themselves.

Where do we go from here? It is clear that we need multicenter prospective studies to better understand the true frequency of IIH misdiagnosis. Effects of referral bias may be magnified in a single-center study by virtue of practices and other factors unique to a given geographical area. The emergence of telemedicine might bring neuro-ophthalmologists and other experts in ophthalmoscopy closer to practicing neurologists. Reimbursement and compensation for telemedicine are issues that still need to be worked out. The digital world is bringing us closer together, and is leveling the playing field for certain technical skills. Nonetheless, fundus interpretation will continue to require basic clinical expertise that ultimately trumps technology.

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