Internet Journal of Allied Health Sciences and Practice

Volume 21 | Number 3 Article 6

June 2023

Papilledema in Patients with Presumed Idiopathic Intracranial Hypertension Diagnosed Without a Lumbar Puncture: A Case Report

Marlon J. Monelyon-Demeritt
Nova Southeastern University, dmarlon@nova.edu

Beata I. Lewandowska
Nova Southeastern University, blewandows@nova.edu

Papilledema in Patients with Presumed Idiopathic Intracranial Hypertension Diagnosed Without a Lumbar Puncture: A Case Report

Abstract

Purpose: The purpose of this case report is to present two cases of papilledema secondary to presumed IIH and medically managed with an oral diuretic without first obtaining cerebrospinal fluid opening pressure and composition. **Method:** A 31-year-old female presented for evaluation of blurry vision in both eyes and a history of transient vision loss with postural changes. A 22-year-old female presented for evaluation of blurry vision in both eyes and headaches that started during her pregnancy. She reported double vision that was relieved when she closed one eye. This is a case report of two overweight females who presented with bilateral optic disc edema and were referred for neuro-ophthalmological evaluation. Both patients were treated with oral acetazolamide without lumbar puncture to confirm elevated intracranial pressure or abnormal cerebrospinal fluid components. **Result:** The features associated with idiopathic intracranial pressure may include papilledema, a headache, pulsatile tinnitus, blind spot enlargement on visual field testing, and diplopia. **Conclusion:** Although the diagnosis of IIH has historically been contingent on cerebrospinal fluid opening pressure of 25 cm H_2O measured by a lumbar puncture, the health professions community should be aware that some patients are being managed with oral acetazolamide for presumed IIH-related signs and symptoms without the evidence of elevated intracranial pressure or cerebrospinal fluid abnormalities.

Author Bio(s)

Marlon Monelyon-Demeritt, OD, MBA, FAAO is an Associate Professor and Attending Optometrist at Nova Southeastern University College of Optometry and The Eye Care Institute.

Beata Lewandowska, OD, MS, Diplomate ABO is an Associate Professor and Attending Optometrist at Nova Southeastern University College of Optometry and The Eye Care Institute.

IJAHSP™
The Internet Journal of Allied Health Sciences and Practice
Dedicated to allied health professional practice and education
Vol. 21 No. 3 ISSN 1540-580X

Papilledema in Patients with Presumed Idiopathic Intracranial Hypertension Diagnosed Without a Lumbar Puncture: A Case Report

Marlon J. Monelyon-Demeritt
Beata I. Lewandowska

Nova Southeastern University

United States

ABSTRACT
Purpose: The purpose of this case report is to present two cases of papilledema secondary to presumed IIH and medically managed with an oral diuretic without first obtaining cerebrospinal fluid opening pressure and composition. **Method:** A 31-year-old female presented for evaluation of blurry vision in both eyes and a history of transient vision loss with postural changes. A 22-year-old female presented for evaluation of blurry vision in both eyes and headaches that started during her pregnancy. She reported double vision that was relieved when she closed one eye. This is a case report of two overweight females who presented with bilateral optic disc edema and were referred for neuro-ophthalmological evaluation. Both patients were treated with oral acetazolamide without lumbar puncture to confirm elevated intracranial pressure or abnormal cerebrospinal fluid components. **Result:** The features associated with idiopathic intracranial pressure may include papilledema, a headache, pulsatile tinnitus, blind spot enlargement on visual field testing, and diplopia. **Conclusion:** Although the diagnosis of IIH has historically been contingent on cerebrospinal fluid opening pressure of 25 cm H_2O measured by a lumbar puncture, the health professions community should be aware that some patients are being managed with oral acetazolamide for presumed IIH-related signs and symptoms without the evidence of elevated intracranial pressure or cerebrospinal fluid abnormalities.

Keywords: optic nerve, papilledema, lumbar puncture

INTRODUCTION

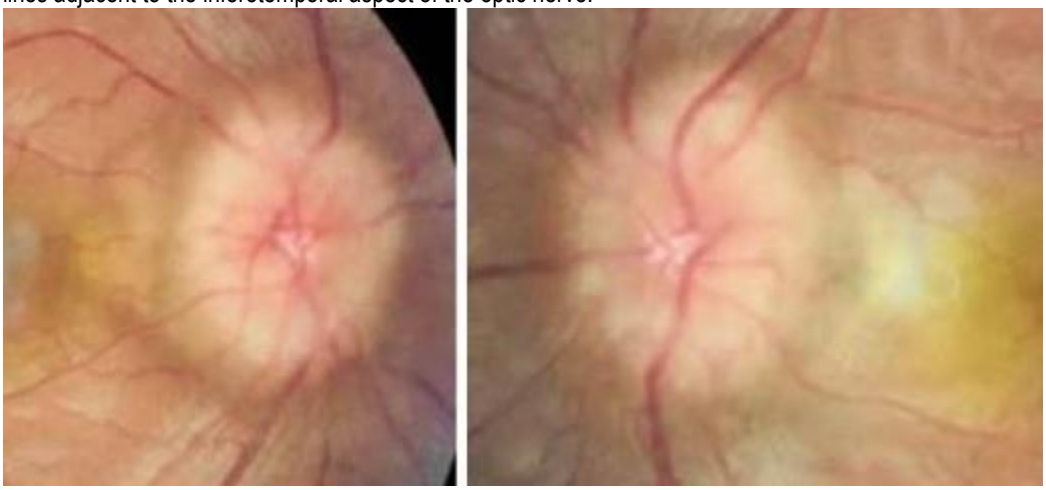
Papilledema is defined as optic disc edema that occurs secondary to elevated intracranial pressure (ICP). It can be an alarming sign of many life or vision-threatening conditions, including space-occupying intracranial lesions and inflammatory or infectious processes that affect the brain and meninges.[1] Papilledema is prevalent in overweight females of childbearing age. The most common cause of papilledema, especially in patients under the age of 50, is idiopathic intracranial hypertension (IIH).[1] The clinical findings associated with IIH are optic disc edema with normal to near-normal visual function, transient visual obscurations, pulsatile tinnitus, nausea, vomiting, and headaches exacerbated by coughing, straining, and Valsalva maneuvers. Magnetic resonance imaging (MRI) of the brain and orbits is performed to exclude non-idiopathic causes. Historically, the diagnostic criterion for IIH was a cerebrospinal fluid opening pressure (CSFOP) greater than 25 cm H_2O measured by a lumbar puncture (LP), which also allows for the examination of cerebrospinal fluid (CSF) composition.

We describe two cases of patients who presented to the clinic with bilateral optic disc edema and were referred to neuro-ophthalmology for evaluation and management. Each patient had an MRI, but neither of them had a lumbar puncture (LP) to measure the CSFOP or look at the composition of the CSF. They were diagnosed with presumed IIH and started on oral acetazolamide. The purpose of this case report is not to debate the necessity of an LP but to present cases of papilledema secondary to presumed IIH and medically managed with an oral diuretic.

CASE REPORT #1

A 31-year-old female presented complaining of blurry vision in her left eye. She reported a history of oral contraceptive use for 3 months, but she discontinued it 2 months prior to her eye exam due to headaches, nausea, and a weird taste. She also reported transient vision obscuration with posture changes. At the time of the exam, she was not taking any medications and denied suffering from any allergies. She presented with best-corrected visual acuities (BCVA) of 20/20 in the right eye (OD) and 20/30-2 in the left eye (OS). Pupils were equal, round, and reactive to light, with no afferent pupillary defect (APD) in both eyes (OU). Confrontation visual fields were full to finger counting OD, and constricted ST and IT OS. Ocular motility was full range of motion OU. Biomicroscopy of the anterior segment was unremarkable OU. Dilated fundus examination (DFE) of the right and left eye revealed grade 4+ disc edema, with circumferential retinal folds (Paton lines) inferotemporal to the disc OS (Figure 1). The vitreous, macula, vessels and peripheral were all unremarkable OU. The patient was referred for neuro-ophthalmological evaluation. The patient had an MRI with and without contrast, which confirmed optic disc edema OU without the presence of a space-occupying lesion. A lumbar puncture was not performed. She was subsequently started on oral acetazolamide 500 mg po BID. Unfortunately, the patient was lost to follow-up.

Figure 1. Fundus images depict stage 4 edema of both optic nerves. Note the complete elevation of the optic discs and Paton lines adjacent to the inferotemporal aspect of the optic nerve.

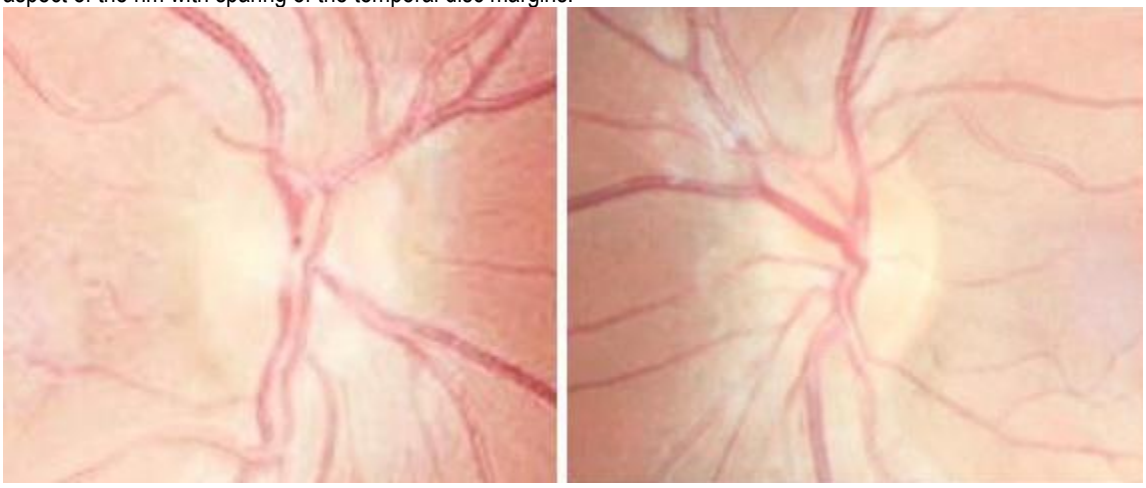


CASE REPORT #2

A 22-year-old female presented complaining of intermittent blur in both eyes. She reported the symptoms started towards the end of her pregnancy, and progressively got worse after she gave birth 2 months ago. She also reported transient vision obscuration with posture changes, horizontal diplopia that is relieved with monocular occlusion and headaches that started during her pregnancy. At the time of the exam, she was taking nifedipine 10 mg for systemic hypertension. She denied suffering any allergies.

She presented with BCVA of 20/20-1 OD and 20/20-2 OS. Pupils were equal, round, and reactive to light, with no APD OU. Confrontation visual fields were full to finger counting OD and OS. Ocular motility was full range of motion OU. Quantifying ocular alignment was difficult due to constant fluctuations, but she exhibited an esodeviation each time she reported diplopia in-office. Biomicroscopy of the anterior segment was unremarkable OU. DFE of the right and left eye revealed pink optic nerves with indistinct margins OU (Figure 2). The vitreous, macula, vessels, and periphery were all unremarkable OU. The patient was referred for neuro-ophthalmological evaluation. The patient had an MRI of the brain and orbits with and without contrast, which confirmed optic disc edema OU without the presence of a space occupying lesion. A lumbar puncture was not performed. She was subsequently started on oral acetazolamide 500 mg po BID. Unfortunately, the patient was lost to follow-up.

Figure 2. Fundus images depict stage 1 edema of both optic nerves. Note the indistinct margins on the superior, inferior, and nasal aspect of the rim with sparing of the temporal disc margins.



DISCUSSION

Papilledema is a potentially blinding condition most commonly associated with IIH. The diagnosis of IIH is based on history, physical examination, MRI, and LP. Neuroimaging studies are performed before the consideration of an LP to rule out any space occupying lesions. The decision to perform or to not perform an LP on a patient suspected of having IIH has been a highly debated topic amongst neuro-ophthalmology experts.

There are some members of the neuro-ophthalmology community, who consider that an LP is not indicated in patients who undergo high-quality MRI with venography and contrast enhancement.[2] These patients fall within the typical IIH demographic of a female of childbearing age with increased body mass index who presents with the typical symptoms of transient visual obscurations, tinnitus, dizziness, headaches, nausea and vomiting, where the visual function is preserved, and the expected MRI findings associated with IIH are present (such as the absence of an intracranial mass, hemorrhage or cerebral edema, protrusion of the optic nerves with a flattened posterior sclera, optic nerve enhancement, distention of the perioptic subarachnoid space, vertical tortuosity of the optic nerves, and empty sella).[3] It is likely that an elevated ICP and normal CSF composition will be found during an LP. There is also a question of whether LP provides additional clinically relevant information in patients with a typical IIH demographic and with typical IIH symptoms that are otherwise systemically healthy. Additionally, the rationale for not performing an LP on these individuals is that the opening pressure during the LP does not correlate with severity of the disease nor visual prognosis.[4,5]

Patients who undergo an LP may develop unwanted side effects. Post lumbar puncture headaches occur in up to 40% of patients.[6,7] Additional, but rarer side effects include meningitis, nerve root pain, infection at the skin site and rarely an epidural abscess.[8]

There are various reasons why an LP may not be valuable, and clinicians should be cognizant of the fact that the CSFOP obtained during an LP is prone to error.[2,9,10] If the opening pressure is under 25 cm H_2O in a typical IIH patient with bilateral optic nerve edema with indirect neuroimaging signs of ICP it can be assumed that the results are misleading, or the normal value is not the same for all individuals and treatment is offered to the patient.[11] Cases of patients with normal pressure IIH have been reported in literature.[11,12] The mechanism responsible for this IIH variant has not been elucidated, but it has been proposed that patients may have different susceptibilities to ICP levels.[11]

CONCLUSION

Patients presenting with bilateral optic disc edema should be evaluated for elevated blood pressure and referred for neuroimaging. Therapeutic options need to be considered and initiated for patients diagnosed with papilledema associated with IIH due to the potential of vision loss. Patients who fit the typical demographic for IIH (present with typical symptoms, typical signs are found on clinical evaluation and neuroimaging) are being offered by some neuro-ophthalmologists to forgo a LP and start oral acetazolamide. The health professions community should be aware that patients with presumed IIH diagnosed without an LP may be medically managed with oral acetazolamide under careful observation by specialists in neuro-ophthalmology.

REFERENCES

1. Xie JS, Donaldson L, Margolin E. Papilledema: A review of etiology, pathophysiology, diagnosis, and management. Surv Ophthalmol. 2022 Jul-Aug;67(4):1135-1159.
2. Moss HE, Margolin EA, Lee AG, Van Stavern GP. Should Lumbar Puncture Be Required to Diagnose Every Patient with Idiopathic Intracranial Hypertension? J Neuroophthalmol. 2021 Sep 1;41(3):379-384.
3. Vaphiades MS, Brodsky MC. Neuroimaging signs of elevated intracranial pressure. Contemporary Neurosurgery. 2001 Jul 15;23(14):1-7.
4. Bahnasy, WS, El-Heneedy, YAE, Elhassanien, MEM, Sharaf, AF, Khalid, HA. Neuro-ophthalmological biomarkers of visual outcome in newly diagnosed idiopathic intracranial hypertension. *Egypt J Neurol Psychiatry Neurosurg* **55**, 26 (2019).
5. Friedman DI, Quiros PA, Subramanian PS, Mejico LJ, Gao S, McDermott M, Wall M; and the NORDIC IIHTT Study Group. Headache in Idiopathic Intracranial Hypertension: Findings from the Idiopathic Intracranial Hypertension Treatment Trial. Headache. 2017 Sep;57(8):1195-1205.
6. Dieterich, M, Perkin, GD. Postlumbar puncture headache syndrome. *Neurological Disorders*. Academic Press, 2003. 77-82.
7. Evans RW. Complications of lumbar puncture. Neurol Clin. 1998 Feb;16(1):83-105.
8. Baer ET. Post-dural puncture bacterial meningitis. Anesthesiology. 2006 Aug;105(2):381-93.
9. Lee SC, Lueck CJ. Cerebrospinal fluid pressure in adults. J Neuroophthalmol. 2014 Sep;34(3):278-83.
10. Bellamkonda VR, Wright TC, Lohse CM, Keaveny VR, Funk EC, Olson MD, Laack TA. Effect of spinal needle characteristics on measurement of spinal canal opening pressure. Am J Emerg Med. 2017 May;35(5):769-772.
11. Green, JP, Newman, NJ. Normal pressure pseudotumor cerebri. *Journal of Neuro-Ophthalmology* 17.4 (1997): 280.
12. Abdelfatah MA. Normal Pressure Pseudotumor Cerebri: A Series of Six Patients. Turk Neurosurg. 2017;27(2):208-211.