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A Comparison of Idiopathic Intracranial Hypertension With and Without Papilledema

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Abstract

Objective—To compare clinical features, visual characteristics, and treatment of idiopathic intracranial hypertension patients with and without papilledema.

Background—Idiopathic intracranial hypertension does not often occur without papilledema. This study estimates the prevalence and compares the clinical characteristics of idiopathic intracranial hypertension patients with and without papilledema.

Methods—We performed a cross-sectional analysis of all idiopathic intracranial hypertension patients diagnosed at the University of Utah Neuro-Ophthalmology Unit between 1990 and 2003. Patient records were reviewed for presence of papilledema and other signs, symptoms, and treatment characteristics. Each patient without papilledema was matched to the patient with papilledema who was closest to his/her age and sex. McNemar's and Wilcoxon-signed rank sum tests were used to compare characteristics between matched pairs.

Results—Among all patients (n = 353), the prevalence of those without papilledema was 5.7% (n = 20). Patients without papilledema reported photopsias (20%), and were found to have spontaneous venous pulsations (75%) and non-physiologic visual field constriction (20%) more often than did those with papilledema. Mean opening pressure, although above normal, was lower in patients without papilledema (mean = 309 mm cerebrospinal fluid) compared with those with papilledema (mean = 373 mm cerebrospinal fluid, $P = .031$). Idiopathic intracranial hypertension patients without papilledema had more frequent diagnostic lumbar punctures than did patients with papilledema. Visual acuities and treatment were similar between groups.

Conclusions—The clinical presentation of idiopathic intracranial hypertension without papilledema is only somewhat different from that of idiopathic intracranial hypertension with

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papilledema. The lower opening pressure in patients without papilledema may explain variations in symptoms and signs between the 2 groups. When there are visual field changes in idiopathic intracranial hypertension without papilledema, non-physiologic visual loss should be considered.

Keywords

idiopathic intracranial hypertension; headache; visual loss; visual fields; pseudotumor cerebri; functional visual loss

Idiopathic inracranial hypertension (IIH), also known as pseudotumor cerebri, is a condition of high intracranial pressure not associated with any identifiable etiology. The prevalence of IIH in the general U population is about 1 in 100,000; the prevalence rises to 19/100,000 in obese women of childbearing age.[1]

The modified Dandy criteria used to diagnose IIH include: headache, increased intracranial pressure (>250-mm water) with normal cerebrospinal fluid (CSF) constituents, no focal neurological deficits, normal neuroimaging results in an awake and alert patient, and papilledema (swelling of the optic disc due to increased intracranial pressure).[2,3] One variant of this clinical presentation is idiopathic intracranial hypertension *without* papilledema (IIHWOP). IIHWOP was recognized in the International Headache Society classification criteria,[4] and it has been documented in case reports and series.[5–16] The Dandy criteria were recently amended so that presence of papilledema is no longer a requirement for IIH diagnosis.[17] The prevalence of IIHWOP in a series of patients with chronic migraine has been estimated to be 5–14%.[12,18,19] The prevalence of IIHWOP in a series of patients with chronic tension-type headache with transverse sinus stenosis was similar (9%).[20] The overall prevalence of IIHWOP, however, remains unknown.

Headaches, visual findings, and other aspects of the clinical presentation of IIH have been evaluated.[2,17,21,22] However, the clinical features characteristic of IIHWOP have not been directly compared with the more commonly observed idiopathic intracranial hypertension with papilledema (IIHWP). The main objective of this study was to compare the clinical features, visual characteristics, and treatment of IIH patients with and without papilledema. We also sought to estimate the prevalence of IIHWOP within a population of IIH patients seen at an academic medical center.

METHODS

This study was approved by the University of Utah Institutional Review Board and is in accord with Health Insurance Portability and Accountability Act regulations.

We performed a cross-sectional analysis of IIH patients diagnosed at the University of Utah Neuro-Ophthalmology Unit from 1990 to 2003. Patient records were reviewed to identify all patients diagnosed with IIH according to the modified Dandy criteria. All new patients fill out a standardized questionnaire in the neuro-ophthalmology unit as part of their initial evaluation. IIH patients with stage 0 optic nerve swelling on the Lars Frisén staging scale[23] were designated as IIHWOP. To confirm the presence or absence of papilledema or secondary optic atrophy, all IIH patients were examined by a University of Utah neuro-

ophthalmologist. Dilated binocular indirect ophthalmoscopy was performed to exclude minor disc swelling or subtle papilledema. A dilated examination of both discs, using indirect ophthalmoscopy and in some cases stereo disc photographs, confirmed the absence of papilledema. We confirmed that lumbar punctures (LPs) were performed in the lateral decubitus position. Visual fields were judged to be "Non-physiologic" if the person had non-physiologic constriction of the visual field (eg, the 3-meter visual field was inside the 1-meter visual field at the tangent screen). Either Goldmann kinetic perimetry or static Humphrey perimetery was reviewed for each case. We analyzed review of systems questionnaires to determine the incidence of vertigo or dizziness reported by each patient.

Each IIHWOP patient was matched as closely as possible by gender and age at diagnosis to an IIHWP patient diagnosed by the same Neuro-Ophthalmology Unit during the same time period. Patients with an opening pressure (OP) of less than 250 mm were excluded from both groups.

Statistical Analyses

Patient groups were compared regarding the following characteristics: body mass index (BMI), OP, history of migraines, history of depression, symptoms at initial presentation (including vertigo/dizziness, transient visual obscurations and pulse synchronous tinnitus, and others), visual acuity, visual fields (static and kinetic), headache characteristics, medical or surgical treatment of intracranial hypertension, and headache therapy. The Wilcoxon-signed rank sum test, the nonparametric form of the paired *t*-test, was used to compare continuous characteristics between matched pairs. McNemar's test was used to compare categorical characteristics between matched pairs. All statistical analyses were performed by a biostatistician (WL) using STATA® version 8.2 for Windows (StataCorp LP, College Station, TX, USA).

RESULTS

We identified 353 patients with a confirmed diagnosis of IIH based on the modified Dandy criteria. Twenty (5.7%) of these patients met our criteria for IIHWOP, including 18 women and 2 men. We were unable to identify a gender-and age-matched comparison subject for one of the male IIHWOP patients; therefore, he was matched by age to a female IIHWP subject.

Patients in both groups had similar ages of symptom onset, BMI, and migraine history (Table 1). However, the mean OP was lower for IIHWOP patients (mean = 309-mm CSF) than for IIHWP patients (mean = 373-mm CSF; $P = .031$). Twelve of the 20 IIHWOP patients underwent more than one LP (total of 33 LPs). All 12 patients had at least one additional pressure that was ≥250-mm CSF. The mean pressure of these 33 was 273-mm CSF. Only 6 of the 33 pressures were less than 250-mm CSF. Four IIHWP patients underwent a total of 6 additional LPs. The mean pressure of these 6 was 315.5-mm CSF.

The most common symptom reported for both groups was headaches (Table 2). Photopsias ($P = .046$) and spontaneous venous pulsations ($P = .025$) were more common in IIHWOP

patients than in IIHWP patients, while sixth nerve palsy was less common in patients with IIHWOP (P = .059).

Approximately 60% of patients in both groups experienced bilateral headaches on a daily basis (Table 3). Auras were more frequently reported by the IIHWOP patients (P = .035). Vertigo with or without dizziness was reported by 75% of patients in the IIHWOP group and by 55% of the patients in the IIHWP group (P = .206).

Visual acuity was similar in both groups, both at the initial and the last visit (Table 4). Loss of visual acuity was not common in either group. However, IIHWOP patients were more likely than IIHWP patients to have normal visual fields at presentation (P = .002). Those IIHWOP patients with visual field defects were more likely to have non-physiologic visual field constriction, whereas enlarged blind spots were observed more frequently in patients with IIHWP.

Medical therapy with a diuretic, alone or with a migraine medication, was effective for the majority of patients in both groups (Table 5). Headache therapy was similar in both groups; carbonic anhydrase inhibitors were most often used for treatment. Overuse of analgesics was documented in only 3 IIHWOP patients and one IIHWP patient. The majority of patients in both groups reported benefits from prophylactic use of anticonvulsants, a carbonic anhydrase inhibitor such as acetazolamide, or another diuretic.

There were 6 patients who underwent surgery for increased intracranial pressure or papilledema: 4 IIHWOP patients and one IIHWP patient had at least one lumbar or ventricular shunt placement; 2 patients with IIHWP had optic nerve sheath fenestration (ONSF). No IIHWOP patient underwent ONSF (Table 5).

DISCUSSION

This study found that 5.7% of patients diagnosed with IIH do not have papilledema. A previous report from a headache center documented 15% of patients with chronic headache (n = 85) had increased intracranial pressure (ICP above 250-mm CSF) and no papilledema.[12] More recently, Vieira et al reported that 10% of their 62 chronic migraine patients had elevated pressure (ICP above 200 mm CSF) and that obesity was a predictor of increased pressure.[19] In another study, 24-hour intracranial pressure monitoring was performed in 10 patients suspected of having IIHWOP.[16] All were examined by a neuro-ophthalmologist and found not to have papilledema. All 10 patients were found to have B waves (monitored oscillations associated with elevated intracranial pressure),[24] and 9 had plateau waves (a sign of loss of pressure volume compensatory reserve).[25,26] These findings suggest that perhaps the loss of pressure volume compensatory reserve plays a role in the development of increased ICP.

Making the diagnosis of IIHWOP can be difficult. A careful observation of the optic disc for subtle signs of swelling by trained observers is important to exclude subtle true swelling.[2] Carefully measured OP with a relaxed patient in the lateral decubitus position must ensue. Strict adherence to the criteria of IIH must be met.[17] Failure to follow these guidelines can lead to a false diagnosis.[27]

It should not be surprising that a disc can remain normal, without swelling, in the face of elevated intracranial pressure. In clinical settings in which the cause of increased ICP is known, such as brain tumors, head trauma, intracranial hemorrhage, endocrine disorders, or other systemic disorders, papilledema is not uniformly observed.[28–31] Animal studies have also shown that papilledema need not accompany elevated ICP.[32]

There is no known explanation for the occurrence of intracranial hypertension without papilledema. However, asymmetric papilledema (where one nerve is severely swollen, and the other one is normal or minimally swollen) has been reported.[33–35] A change in brain compliance,[33] congenital variation of the canalicular optic nerve,[34] or intermittent rises in pressure[35] are mechanisms proposed to explain why one disc may not swell. These explanations may pertain to IIHWOP as well.

Another study found that even after papilledema resolved in 10 IIH patients, 5 still had significantly increased ICP.[36] More recently, another long-term follow-up study showed that IIH may be a chronic condition that can recur even after a period of stability.[37] This suggests that IIH can be a chronic disease in which papilledema (if present) can disappear despite persistent elevated intracranial pressure. This may also be an explanation for IIHWOP.

As expected, the majority of patients in this series reported headaches, consistent with previous studies.[2,14,19] Depression was present in about half the patients in both groups; this finding is also consistent with previous studies.[38] Pulsatile tinnitus was a common symptom in both IIHWP patients (48%) and IIHWOP patients (33%); this was consistent with previous findings.[14] A significant number of IIHWOP patients in this series reported auras. This may indicate that a significant number of IIHWOP patients are also migraine sufferers. Indeed, it has been observed that most patients with IIH have migrainous headaches unrelated to increased intracranial pressure.[39] Some may argue that IIHWOP is merely migraine in obese individuals. However, all of our IIHWOP patients underwent carefully performed measurements of OPs or they were not included in the study. In addition, 65% of our patients had 2 or more elevated reading on more than one occasion. Furthermore, 11 of our patients with IIHWOP had one or more other symptoms of increased ICP (besides headache). Finally, 4 of our IIHWP patients also had aura symptoms with migraine in addition to intracranial hypertension.

Vertigo was more frequently present in IIHWOP although nearly half of the IIHWP subjects also endorsed dizziness or vertigo on a review of systems. In a study reviewing complaints of patients with IIH, vertigo and dizziness are more common when clinicians routinely query for them.[40] Children with IIH frequently complain of dizziness and are ataxic.[41]

Although both groups of patients had mean OPs above 250 mm, the mean OP for the IIHWOP group was significantly lower than that for the IIHWP group. This difference may explain, in part, why the symptoms and signs differed in the 2 groups. We also found that visual fields were more likely to be normal in IIHWOP than in IIHWP. This finding is expected because higher grade visual field defects are usually associated with increased amounts of papilledema.[42] What we did not expect was that IIHWOP patients are more

likely to manifest non-physiologic constriction of the visual field. Clinicians should be alerted to review constricted visual fields in IIHWOP patients carefully to be sure that the constriction is physiologic. Kathol et al found that functional constriction of the visual field does not usually change over time;[43] we had similar findings.

The reason for the increased prevalence of constricted visual fields is not known. This constriction appeared to be non-physiologic in our study. However, it is conceivable that other mechanisms, such as posterior nerve compression, may be present.[44] In addition, if patients have unrecognized IIHWOP for prolonged periods of time and are not optimally managed, the protracted course alone may lead patients to adopt strategies to amplify their symptoms. One recent study found that almost 20% of those with non-physiologic visual changes had underlying organic disease.[45] In another study, non-physiologic visual field disorders were observed in some patients with both psychiatric and non-psychiatric disease.[43]

The few treatment guidelines that exist for IIHWP-associated headaches may not optimally address IIHWOP-associated headaches. Headaches in IIH patients are more refractory to treatment than are headaches in patients without increased ICP.[14] Furthermore, IIH-associated headaches may continue even after the pressure has normalized, indicating that there may be other factors contributing to IIH headache.[39] The IIHWOP patients in this series received medical treatments that were similar to those prescribed for the IIHWP patients. Consistent with previous studies, the combination of a diuretic and migraine preventative medication was the most successful medical treatment in both IIHWP and IIHWOP patients.[12] However, 4 IIHWOP patients in our series, compared with none of our IIHWP patients, underwent at least one CSF shunting procedure to treat intractable headache (one IIHWP patient underwent LP shunting to prevent further visual loss, but not to treat headache). This observation suggests that IIHWOP-associated headache may be even more refractory to standard medical treatment than IIHWP-associated headache. Three of the 4 patients with IIHWOP did have some improvement of headache. Depression and anxiety, both of which severely affect quality of life, are also known to be present in patients with IIH.[38] Successful management of symptoms in these patients depends upon adequately addressing these issues in addition to headache management.

There are limitations to this study. As a cross-sectional review of existing patient records, only association, not causality, should be inferred between the various characteristics evaluated and whether a patient with IIH does or does not have papilledema. While our intake form provides much uniformity in collecting data, some headache-specific questions were not uniformly addressed in all patients. In addition, these 2 groups were matched on sex and age, but other confounders could affect results. We would expect at least 2 of the 50 tests of significance to occur by chance alone. With 9 statistically significant results obtained, however, larger studies appear warranted.

Taken together, our data and previous reports indicate that while less common than IIHWP, IIH can occur in the absence of papilledema. The alert clinician should not dismiss the possibility of IIH in a patient with symptoms and signs of IIH, even if the patient does not

have papilledema. If there are changes in the visual fields in patients with IIHWOP, the alert clinician will look for non-physiologic visual loss.

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Abbreviations

BMI	body mass index
CSF	cerebrospinal fluid
ICP	increased intracranial pressure
IIH	idiopathic intracranial hypertension
IIHWOP	idiopathic intracranial hypertension without papilledema
IIHWP	idiopathic intracranial hypertension with papilledema
LP	lumbar puncture
LP	shunt lumbar peritoneal shunt
ONSF	optic nerve sheath fenestration
OP	opening pressure

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Table 1

Demographic and Clinical Characteristics of Patients With Idiopathic Intracranial Hypertension With (IIHWP) and Without Papilledema (IIHWOP)

Characteristic	IIHWOP (n = 20)	IIHWP (n = 20)	*P* value
Mean age presented for evaluation (range)	32 (10–49)	32 (15–56)	n/a
Mean age of symptom onset (range)	27 (8.5–45)	29 (13–48)	.134
Women	18 (90%)	19 (95%)	n/a
Mean body mass index (kg/m^2) (range)	34 (16–50)	36 (18–56)	.941
Mean opening pressure (mm CSF) (range)	309 (260–420)	373 (260–550)	.031
Number patients with more than 1 LP performed	12	4	.021
Migraine history	13 (65%)	9 (45%)	.248
Depression history	10 (50%)	9 (45%)	.706

CSF = cerebrospinal fluid; LP = lumbar puncture; n/a = not applicable; the groups were matched for sex and age.

Table 2

Symptoms and Signs at Presentation of Patients With Idiopathic Intracranial Hypertension With (IIHWP) and Without Papilledema (IIHWOP)

Symptom/sign	IIHWOP (n = 20) (%)	IIHWP (n = 20) (%)	*P* value
Headache	19 (95)	19 (95)	1.000
Transient visual obscurations	5 (25)	11 (55)	.109
Visual loss (decreased vision, blurry vision)	5 (25)	8 (40)	.257
Tinnitus	7 (35)	10 (50)	.366
Photopsias	4 (20)	0 (0)	.046
Diplopia	4 (20)	9 (45)	.166
No symptoms	0 (0)	1 (5)	.317
Sixth nerve palsy	1 (5)	6 (30)	.059
Spontaneous venous pulsations†	12 (75)	0 (0)	.025

† Data regarding spontaneous venous pulsations were only available for 16 IIHWOP patients and 9 IIHWP patients.

Table 3

Headache Characteristics of Patients With Idiopathic Intracranial Hypertension With (IIHWP) and Without Papilledema (IIHWOP)

Characteristic	IIHWOP (n = 20) (%)	IIHWP (n = 20) (%)	*P* value
Unilateral	8 (40)	9 (45)	.739
Bilateral	12 (60)	11 (55)	.739
Quality: pulsating	15 (75)	15 (75)	1.000
Pressure/ache	5 (25)	5 (25)	1.000
Frequency: daily	12 (60)	12 (65)	.706
Duration: constant	10 (50)	14 (70)	.206
Nausea/vomiting	13 (65)	11 (60)	.739
Photo/phonophobia	14 (70)	11 (55)	.366
Auras reported	11 (55)	4 (20)	.035
Vertigo/dizziness	15 (75)	11 (55)	.206
Neck stiffness	1 (5)	3 (15)	.157
Worse on awakening	8 (40)	3 (15)	.059

Table 4

Visual Function Characteristics of Patients With Idiopathic Intracranial Hypertension With (IIHWP) and Without Papilledema (IIHWOP)

Characteristic	IIHWOP (n = 20)	IIHWP (n = 20)	*P* value
Mean (range) of follow-up	25 months (0–6 years)	25.6 months (1–8 years)	
Visual acuity (mean [SD] logMAR)			
Initial	0.05 (0.10)	0.08 (0.26)	.430
Final	0.04 (0.10)	0.12 (0.50)	.694
Visual field (Humphrey – mean [SD] defect)			
Initial	−3.71 (5.47)	−3.21 (2.86)†	.861
Final	−3.41 (5.17)	−2.20 (1.26)	.875
Visual field interpretation: initial‡ (% per visual fields)			
Visual fields	40	39†	
Normal	29 (73%)	5 (13%)	.002
Enlarged blind spot	0 (0%)	23 (59%)	<.001
Nasal defects	2 (5%)	4 (10%)	.317
Constriction	0 (0%)	4 (10%)	.083
Non-physiologic constriction	8 (20%)	0 (0%)	.046
Altitudinal/arcuate/temporal	1 (3%)	3 (8%)	.317
Visual field interpretation: final‡ (% per visual fields)			
Visual fields	40	38§	
Normal	28 (70%)	19 (50%)	.480
Enlarged blind spot	0 (0%)	11 (29%)	.008
Nasal defects	3 (8%)	4 (11%)	.414
Constriction	0 (0%)	3 (8%)	.157
Non-physiologic constriction	8 (20%)	0 (0%)	.046
Altitudinal/arcuate/temporal	1 (3%)	1 (3%)	1.000

†One patient had a congenital glaucoma in one eye.

‡Significance test for having the characteristic in at least one visual field.

§One patient had such poor vision that the visual field could not be reliably tested.

SD = standard deviation.

Table 5

Medical and Surgical Treatment of Patients With Idiopathic Intracranial Hypertension With (IIHWP) and Without Papilledema (IIHWOP)

Characteristic	IIHWOP (n = 20)	IIHWP (n = 20)	*P* value
Effective medical therapy			
Diuretic alone (%)	6 (30)	11 (55)	.096
Diuretic + migraine medication (%)	7 (35)	6 (30)	.706
Migraine medication alone (%)	4 (20)	2 (10)	.414
No response to any therapy (%)	3 (15)	1 (5)	.317
Surgical therapy			
VP or LP shunt (%)	4 (20)	1 (5)	.179
Of these, those whose symptoms improved (%)	3 (75)	0 (0)	.083
Nerve sheath decompression (%)	0 (0)	2 (10)	.157
Of these, those whose symptoms improved (%)	N/a	1 (50)	n/a

LP = lumbar puncture; n/a = not applicable; VP = ventricular peritoneal shunt.