Reasonable Doubt: Can High Intracranial Pressure Occur Without Papilledema?

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Jonathan C. Horton
hortonj@vision.ucsf.edu
http://www.ucsf.edu/hortonlab

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Papilledema can be a Neurological Emergency, Causing Preventable Blindness
70-year-old man complaining of “foggy vision” for a month

OPHTHALMOLOGY
Refracting Physician: Charles M. D.
Patient Name: David

Ophthalmic evaluation reveals the following diagnosis:
- Cataract
- Diabetic Retinopathy
- Diabetes without retinopathy
- Macular Degeneration
- Retinal vascular disease
- Glaucoma


New glasses Rx given

Encore 30-2 print-out
Severe Papilledema Can Look Mild in the Presence of Optic Atrophy
15-year-old with 6 week history of blurred vision and headache

Acuity: 20/200
Acuity: 20/40
Normal Optic Discs
Can be Confused
with Papilledema

39-year-old woman with history of migraines for 8 years
Neurologist's Examination and Plan

Yesterday the patient saw an ophthalmologist, Dr. [Redacted], who diagnosed her with bilateral papilledema.

PHYSICAL/NEUROLOGICAL EXAMINATION
Pleasant, cooperative woman who is depressed and was crying frequently during the evaluation. Blood pressure 123/78, pulse 71 and regular. There are no carotid bruits.

Cranial Nerves Evaluation: Pupils 0.4 cm, reactive to light and accommodation. Fundoscopic evaluation reveals possible bilateral papilledema of a mild degree. All other cranial nerves are normal.

PLAN
I tried to schedule a brain MRI and was unable to get an appointment for the next one or two days. I decided that the patient most likely will need an urgent spinal tap and I thought it would be reasonable for her to go to the emergency room. I called the emergency room doctor at St. Mary’s Hospital first and then at St. Francis Hospital. The doctor at St. Francis Hospital agreed to see the patient and order a head imaging study and spinal tap if
Refractive Error May Confound Evaluation of the Optic Discs

39-year-old woman referred for “swollen optic discs”
Don’t be Fooled by Pseudopapilledema

5-year-old boy with headaches, not doing well in school
CT scanning reveals optic disc drusen; MR does not

Beware the Pitfall of
Optical Coherence Tomography
54-year-old woman shunted in 2010
Referred for persistent headache & papilledema
Ms. Doe returned to my office today for follow-up. This is a 54-year-old woman with pseudotumor cerebri on Diamox 250 mg twice daily with no side effects. She denies any loss of vision or blurred vision. She experienced occasional headaches at 5/10 intensity. At the peak of the headache, she sometimes gets blurry vision. No restriction in the visual field. She continues to have lower back pain. She thinks Cymbalta 30 mg current dose twice daily has helped some. She is no longer on Kepros. She has used Celebrex for breakthrough headaches. She could not tolerate. She is unable to tolerate Norco, as well.

She is awake, alert, interactive, in no apparent distress. Grossly intact cranial nerve examination, cerebellar function, stance and gait. I do not see significant papilledema.

Encounter Date: 06-11-2012

Ms. Doe returned to my office today for follow-up. This is a 54-year-old woman with pseudotumor cerebri who was last seen in my office in June. She was later seen by ophthalmologist, Dr. Lee, was found to have worsening papilledema. Her dose of Diamox was increased from 250 mg twice daily to 500 mg twice daily.

She has been on it for a month. She denied any loss of vision or restriction in the visual field currently. She gets headaches off-and-on.

She continues to suffer from lower back pain. She is currently on Cymbalta 60 mg daily. She uses Tylenol #3 for breakthrough headaches.
Lumbar Puncture Opening Pressure 160 mm H₂O

Routine Fundus Photography without Eyedrops

Ask to See Fundus Photographs
Absence of Papilledema
tells you nothing about intracranial pressure if the optic nerves are atrophic
(Imageing can fool you too)

33-year-old pregnant woman shunted at age 12 for congenital aqueductal stenosis, now complaining of worsening vision OD.

20/400 right eye, light perception only left eye
Two weeks later

Shunt failure without ventriculomegaly proclaimed by ophthalmic findings

David M. Katz, M.D., Jonathan D. Thore, M.D., Karin M. Muraszko, M.D.,
Robert C. Dauer, M.D.

Department of Ophthalmology, W. K. Kellogg Eye Center; and Departments of Neurology and
Surgery (Neurosurgery), University of Michigan, Ann Arbor, Michigan

Four patients who developed increased intracranial pressure from ventricular shunt failure suffered a delay in
diagnosis because magnetic resonance imaging of the brain did not show ventriculomegaly and because ophthalmic
findings were initially overlooked or misinterpreted. None of the patients had the conventional manifestations of
shunt failure: severe headache, nausea, vomiting, and depressed consciousness. Three patients suffered marked, per-
manent vision loss from chronic papilloedema. These cases affirm that increased intracranial pressure may occur in
shunt dependency without producing either conventional clinical symptoms or signs on imaging of the brain.
Because ophthalmic manifestations may be the major clues to diagnosis, and because irreversible loss of vision is
possible if these clues are overlooked, consideration should be given to periodic ophthalmological examination of
shunt-dependent patients.

Fig. 5. Case 3. A: Axial computerized tomography scan performed before shunt revision when the patient complained
of worsening vision, showing no enlarged ventricles. B: Scan performed 2 weeks later, when the patient complained of
headache, confusion, and total vision loss, showing enlarged ventricles.

Ventricular Size Can Be Misleading
55-year-old man with history of headache

6 June 2011
Spinal Tap at Sutter Medical Center, Sacramento
L3/L4 Opening Pressure: 98 mm water
4 weeks after right frontal VP shunt. Still has headache, taking 6 Hydrocodone tablets per day.

Most Recent Progress Note (12 May 2012)

Overall assessment and plan: As you know the last MRI showed no significant problems with the patient’s ventricles. His exam is completely and totally normal. I see no evidence that the patient’s current complaints are related to his shunt. Therefore I have asked the patient to go back to see his neurologist to see if there is any other possible etiologies for these headaches. To me quite frankly they seem like they are in a tension headache type category. Again I do not think anything further needs to be done with his shunt. I have not scheduled him for followup visit but would be happy to reevaluate the patient on an as-needed basis. I’d be happy to discuss his case with any of the concerned parties. Very truly yours.
Headache after Shunting

If You Did Not Perform the Lumbar Puncture, Doubt the Opening Pressure

44-year-old woman on disability because of headache (242 lbs)
Positioned prone on the CT table, a suitable trajectory for access to the thecal sac was identified. The overlying skin was prepped and draped in the usual aseptic fashion and then anesthetized via local infiltration at approximately 5 cc 1% lidocaine. Under CT guidance, a 19-gauge 10 cm coaxial needle was advanced just proximal to the thecal at the L2/3 level through which a 20-gauge 17 cm needle was advanced into the thecal sac. Upon return of clear-colored CSF, a total of 6 cc was removed over a 30 minute period which included aspiration and changes in patient positioning. More CSF could not be obtained and the needles were removed. The opening pressure was 20 mmHg. A coaxial approach was used as there was extensive scar tissue within the subcutaneous soft tissues overlying the L2/3 level making it difficult to advance a single 20-gauge needle.
A Partially Empty Sella is Usually Present in Patients With Chronic Intracranial Hypertension

85% with pseudotumor cerebri have a partially empty sella*

Measurement of the Sagittal Area of the Sella Turcica In 48 Patients with PTC and 48 Control Subjects

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Mean sella area = 65 ± 16 mm²
Mean gland area = 42 ± 13 mm²

The sella turcica is 65% (42/65) full in control subjects.

Mean sella area = 90 ± 30 mm²
Mean gland area = 34 ± 14 mm²

The sella is 38% (34/90) full in pseudotumor cerebri patients.

Does the Pituitary Gland Also Shrink?

- We calculated that enlargement of the sella from 65 mm² to 90 mm² would reduce the gland’s cross-sectional profile from 42 mm² to 38.6 mm² (keeping the gland’s spherical segment volume constant).

- In our pseudotumor patients, mean gland area was smaller (34 mm²) than the value (38.6 mm²) predicted by the assumption of no gland shrinkage. Therefore, we cannot exclude the possibility of some gland shrinkage.
Normal Range of Intracranial Pressure is Uncertain

Lumbar Puncture Opening Pressure in 1,033 Patients (94% between 70 and 180 mm H₂O)

Merritt & Fremont-Smith (1938)
Avery et al. 2010:
No effect of age on opening pressure

Prior literature indicates lower ICP in young children

Avery et al, Neurology May 2011
Opening Pressure of 220 mm Water and No Papilledema

Figure 3. CSF opening pressure (cm CSF) according to four categories of body mass index (BMI). Boxes represent point estimates of median CSF opening pressure, and error bars represent 95% reference intervals. The area of each box is proportional to the sample size of each group.
Intracranial Pressure May Fluctuate Widely

Avery et al, Neurology May 2011

Opening Pressure of 220 mm Water and No Papilledema

N = 41 in each group
Mean age = 13

The only subject in our study diagnosed with intracranial hypertension who had an OP below 28 cm H2O had undergone renal transplantation and developed ONHE following withdrawal of long-term oral corticosteroids. Her LP demonstrating an OP of 22 cm H2O was performed after restarting her oral corticosteroids. She still had papilledema, but most symptoms of IBH had improved following resumption of corticosteroid therapy.

Avery et al, Neurology May 2011

Opening Pressure of 300 mm Water and No Papilledema

N = 41 in each group
Mean age = 13

Intracranial Pressure May Fluctuate Widely

Long-term intracranial pressure recording in the management of pseudotumor cerebri

M. Gómez-García, M.D., PH.D., AND LAWRENCE v. THOMAS, PH.D.

Department of Neurological Surgery, The Johns Hopkins Hospital, Baltimore, Maryland, and The Applied Physics Laboratory of The Johns Hopkins University.

Fig. 3. Case 1. Left: Intracranial pressure (ICP) recorded by sensor for 24 hours on December 21, 1977. The patient was fully withdrawn from corticosteroids and had recurrence of visual symptoms and headache. The ICP shows great variability ranging from 30 to 400 mm H2O. Right: Eye grounds showing papilledema.
Ford and Murphy 1939

“One determination of intracranial pressure in a case of disease of the central nervous system is no more instructive than one determination of the patient's temperature during the course of a fever”

Chronic Intracranial Hypertension Can Occur Without Papilledema (Rarely)

52-year-old man with intracranial tumor and transient obscurations
The Challenge:

- Headache patients are common.
- Rarely, papilledema can be subtle or absent in patients with chronic elevation of intracranial pressure.
- Lumbar puncture measurement of intracranial pressure is often difficult or inaccurate.
Indications for Invasive ICP Monitoring:

- Pseudotumor cerebri profile, headache, no papilledema, and equivocal LP pressure measurements.

- Pseudotumor cerebri profile, headache, optic atrophy, and equivocal LP pressure measurements.

- Shunted Pseudotumor cerebri patients, with persistent headache, no papilledema, and negative shunt evaluation. (Make sure patient is not over-shunted)