

Papilledema and idiopathic intracranial hypertension



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Clinical case

A 34 year-old overweight female is sent to her ophthalmologist by the neurologist for evaluation, as part of work-up for chronic headache of recent onset. She has no past medical history, no medication and does not complain of any visual symptoms. The headache is diffuse, variable during the day, worsening during postural changes. When specifically asked, the patient recalls that she has experienced some mild bilateral tinnitus during the last weeks. The neurological examination was found to be normal.

At examination, visual acuity and confrontation visual fields are normal bilaterally; examination of the orbits, eyelids, pupils and of the anterior segment discloses no abnormality. There is no evidence for an ocular misalignment and intraocular pressure is within normal limits. Fundoscopy discloses mild, asymmetric bilateral papilledema (Figure 1), with no associated retinal findings; venous pulsations are not detected.

At this point, secondary intracranial pressure is suspected, but appropriate neuroimaging rules out an intracranial mass or venous sinus thrombosis. A lumbar puncture in supine position is performed: the opening pressure is 350 mmH₂O, with normal cerebrospinal fluid composition. Since no underlying condition is found, the patient is

diagnosed with idiopathic intracranial hypertension.

Comment

Idiopathic intracranial hypertension (IIH) is a condition defined by elevated intracranial pressure but no clinical, laboratory or neuroimaging evidence of space occupying lesion, hydrocephalus, infection or vascular abnormality. It is therefore a diagnosis of exclusion. The typical clinical presentation of IIH, is that of a young, obese woman, usually complaining of suggestive symptoms (severe constant headache, pulsatile tinnitus, transient visual obscurations, sometimes horizontal diplopia, rarely blurred vision). Work-up of bilateral, sometimes asymmetric

and even isolated papilledema can lead to diagnosis of IIH, after ruling out an underlying intracranial mass lesion, and if the cerebral spinal fluid is documented to have an elevated opening pressure and normal composition. However, clinical evaluation of unilateral or bilateral papilledema always starts with ruling out other mimicking conditions (pseudopapilledema), such as optic disc drusen and elevated discs in small, hyperopic eyes (Figure 2).

In patients with early papilledema related to IIH, visual acuity and color perception are generally preserved. A common visual symptom is occurrence of monocular or binocular transient visual obscurations, several times a day, after positional changes or a Valsalva manoeuvre. Visual field testing may disclose more subtle, non-specific abnormalities such as blind spot enlargement, nasal defects, arcuate scotomas and concentric constrictions. Sixth nerve palsy with horizontal diplopia is the most common

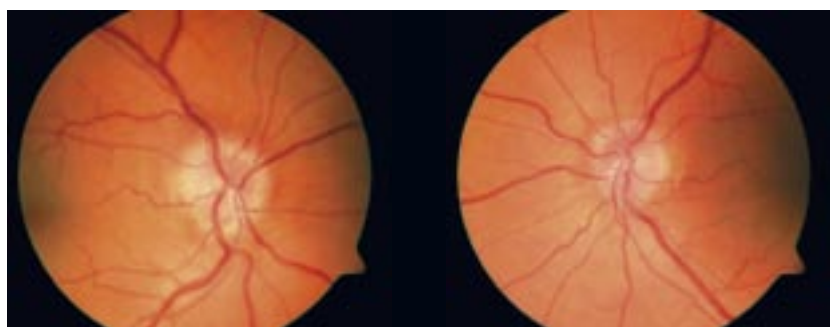


Figure 1. Bilateral papilledema, leading to diagnosis of idiopathic intracranial hypertension.

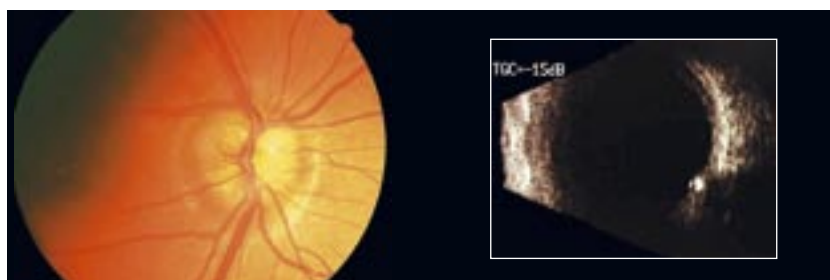


Figure 2. The main differential diagnosis of papilledema is optic drusen (Fig 2a), best diagnosed by ultrasound, disclosing calcification (Fig 2b).

Fortsættes side 6

TABLE 1. The diagnostic criteria of idiopathic intracranial hypertension (International Headache Classification - IHCD-II)

1. Normal neurological examination. The only accepted abnormalities are :
 - (a) Papilloedema
 - (b) Enlarged blind spot
 - (c) Visual field defect
 - (d) Sixth nerve palsy
 2. Increased CSF pressure (>200 mmH₂O in the non-obese, >250 mmH₂O in obese)
 3. Normal CSF chemistry and cellularity.
 4. Absence of intracranial disease (including venous sinus thrombosis) ruled out by appropriate investigation.
 5. No metabolic, toxic or hormonal causes of intracranial hypertension.
- IIH-related headache should be progressive with at least one of the following:
- (a) Daily occurrence
 - (b) Diffuse and/or constant non-pulsating pain
 - (c) Aggravated by coughing or straining

neurological objective abnormality, other cranial nerves functions being rarely impaired.

Headache is often the presenting symptom, although asymptomatic papilledema can also lead to this diagnosis. On the contrary, absence of papilledema is a very infrequent situation in confirmed IIH. Headache usually develops over weeks to a daily pain of moderate intensity, typically aggravating in the morning and during physical activity or postural changes. The quality and location of the headache are non specific, but may resemble a fairly severe chronic migraine associated with daily visual non-aura symptoms. Associated pulsatile bruit-like tinnitus, either unilateral or bilateral is common, disappearing after normalization of the cerebrospinal fluid pressure. IIH is not always associated with headache, visual or auditory disturbances, making appropriate diagnosis more difficult.

Ophthalmoscopically, papilledema consists of blurring of the disc borders, absent spontaneous venous pulsations, distended retinal veins and eventually protrusion of the optic disc and peripapillary haemorrhages and exsudates. Parapapillary retinal folds may involve the macular region and cause visual loss and metamorphopsiae. In chronic courses of intracranial hypertension, optic nerve atrophy may occur at late stages, which is the reason why the term of «benign » intracranial hypertension should be avoided. Indeed, « ma-

lignant » intracranial hypertension may at times develop, consisting of rapid significant visual field loss and marked papilledema, whereas visual loss may occur rapidly over days to weeks. There are no ophthalmoscopic signs that could differentiate primary from secondary raised intracranial hypertension, making neuroimaging (at best by MRI and venous phase MRI) mandatory to rule out an intracranial mass lesion or a sinus thrombosis. Since certain conditions (anemia, hypothyroidism) and some medications have been associated with IIH (tetracyclines, vitamin A, cyclosporine, amiodarone, etc) any associated disease and potential iatrogenic effect should be considered as a possible curable situation.

Diagnosis of IIH implies documented raised cerebrospinal fluid pressure, measured by an appropriate procedure in a patient in lateral supine position with legs extended and as relaxed as possible. It is considered that pressure above 250 mmH₂O in obese and above 200 mmH₂O in non-obese individual (IHCD-II criteria) is consistent with the diagnosis of IIH, and less than 200 mmH₂O is normal.

Treatment of IIH is only symptomatic, since little is known about the pathophysiological mechanisms of the disease. Its aim is to lower the pressure of the cerebrospinal fluid, and to avoid morbidity related to visual loss and chronic headache. Various medical and surgical interventions can be offered, but no

comparative randomized, prospective studies of the treatment strategies currently exist.

Weight loss in obese patients has beneficial effects on IIH reduction and a supervised weight loss program should be encouraged whenever is possible. Oral acetazolamide (Diamox) at increasing doses (from 250 mg/day up to 1250 mg/day) is commonly used as the first-line medication although other diuretics (Thiazide, Furosemide) or antiepileptics with carbonic anhydrase inhibitory effect (Topiramate), also can be used to control visual field loss and headache. Corticosteroids should not be used since their side effects (weight gain) and their withdrawal may worsen the condition and the associated visual loss. If the medical treatment fails (worsening of the visual fields, vision and/or of headache), repeated lumbar punctures can be an alternative, although not very used option. In IIH patients with progressive visual loss and no headache, optic nerve sheath fenestration is an interesting indication. Neurosurgical shunting is proposed in patients refractive to medical therapy and if headache is a dominant feature, although complications are not uncommon.

The ophthalmic follow-up of these patients usually includes regular monitoring of visual acuity, static automated perimetry and optic disc photos, at least every 6 months. New evaluation methods, such as peripapillary OCT measurements may play a role in the future for a better follow-up.

In conclusion, IIH appears to have an increasing incidence in the western countries, due to the recent increase of obesity and diet habits. Its diagnosis and management are best provided by a close collaboration between ophthalmologists and neurologists. Since there is no clear consensus of therapeutic intervention, new studies in dedicated centres are needed for a better understanding of the mechanisms of the disease and its appropriate treatment.

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