Idiopathic intracranial hypertension: What the radiologist should know

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Learning objectives

The purpose of this educational exhibit is to:

- Learn about possible imaging findings in patients with idiopathic intracranial hypertension (IIH) and to highlight the necessity of correlating those findings with clinical symptoms and the results of other diagnostic procedures.
- Explain the role of neuroimaging in the exclusion of the IIH in case when some identifiable causes of raised intracranial pressure are found.
Background

**Idiopathic intracranial hypertension** (also known as pseudotumor cerebri and benign intracranial hypertension) is a disorder of increased cerebrospinal fluid (CSF) pressure of unknown cause. Previously it was commonly known as pseudotumor cerebri because of common clinical signs of intracranial hypertension without tumoral causes. The overall incidence of idiopathic intracranial hypertension is approximately between 2 and 19 per 100,000. It mostly occurs in young women in childbearing age who are obese (with a body mass index above 25).

**Pathogenesis and etiology of idiopathic intracranial hypertension**

The pathophysiology of this condition is poorly understood. Several pathophysiological mechanisms regulating cerebrospinal fluid (CSF) pressure have been postulated. Some of the proposed mechanisms are: inflammatory factors in correlation to obesity, increased cerebral blood volume, excessive CSF production, venous outflow obstruction and compromised CSF resorption. Obesity and female gender are the strongest and most consistent risk factors of IIH. Moreover, some studies suggest that weight gain, as opposed to initial BMI, is the leading factor of poor visual outcome. This finding accentuates cross-links between weight gain and high cerebral venous pressure in the pathogenesis of IIH. One of the possible pathophysiological mechanisms for raising intracranial pressure in obese patients could be the increased intraabdominal pressure resulting in reduced cerebral venous drainage. Furthermore, chronic inflammation associated with obesity can lead to a prothrombotic state which contributes to development of IIH. On the other hand, adipose tissue is an actively secreting endocrine tissue. One of the long-standing hypotheses for the pathogenesis of IIH cites abnormal vitamin A metabolism as well as elevated levels of retinol found in CSF in some patients with IIH. Therefore, there is a possible correlation between adipose tissue, vitamin A production and development of IIH. Other possible agents in pathogenesis of IIH are sex hormones because of preferential occurrence of the disorder among postpubertal, pre-menopausal women, and the absence of a gender preference before puberty.

The transverse sinus stenosis (TSS) theory is widely spread and much discussed, but it is still unclear whether it could be a cause or a consequence of IIH. This theory supports obstruction of the intracranial venous drainage as a main pathophysiological mechanism for IIH.

ICP can be raised due to secondary causes such as:
• Cerebral venous abnormalities - dural venous sinus thrombosis, bilateral jugular vein thrombosis, superior vena cava syndrome, arteriovenous fistula
• Decreased CSF absorption from a previous intracranial infection or subarachnoid hemorrhage
• Exposure to or withdrawal from certain exogenous substances - tetracycline, antibiotics, amiodarone, levodopa, ketoprofen, lead, lithium, vitamin A, retinoids, anabolic steroids, withdrawal from chronic corticosteroids
• Endocrine disorders (Addison disease, hypoparathyroidism), metabolic disorders, systemic disease

Clinical presentation

IIH is characterized by increased intracranial pressure (ICP) leading to headache, papilledema, visual symptoms and signs without any lateralizing findings in the neurological examination, and normal CSF findings.

The most prominent clinical finding in patients with IIH is papilledema which may be bilateral, asymmetrical, or even unilateral. However, in some cases IIH can occur in the absence of papilledema. These patients, without papilledema may experience a headache profile similar to chronic daily headache with migrainous features, responding well to specific antimigraine agents. The patients with papilledema can develop serious visual impairment and eventually blindness. The condition must be taken seriously and adequate treatment must follow the diagnosis.

Typical IIH clinical presentation and findings are:

- Headache
- Transient visual obscurations
- Photopsia, diplopia, visual disturbance (acuity and/or field loss) and blindness
- Sudden visual loss
- Symptoms of increased intracranial pressure
- Retro-orbital pain
- Pulsatile tinnitus
- Radicular pain

Headache is one of the leading symptoms. It is usually progressive with at least one of the following characteristics: daily occurrence, diffuse and/or constant (non-pulsating) and aggravated by coughing or straining. In some cases the headache may be accompanied
with allodynia, typically in unilateral V1 distribution. Since the headache can mimic migraine the diagnosis of IIH can be difficult.

Transient visual obscurations occur in most patients as a symptom of papilledema. The disturbance can last up to 30 seconds and is described as a dimming or blackout of vision in one or both of the eyes. They are often predominantly orthostatic.

Diplopia is typically horizontal caused by sixth nerve palsy.

Sudden visual loss may happen due to intraocular hemorrhage secondary to peripapillary subretinal neovascularization related to chronic papilledema.

Radicular pain is an uncommon symptom, typically localized in the arms.

Friedman et al., proposed recently updated Dandy criteria for the diagnosis of IIH which are presented in Table.1.
The optimal management usually creates problems for clinicians. It ranges from simple dietary measures to neurosurgical and endovascular procedures. Not all patients require treatment. In general, severe headache and evidence of optic nerve involvement are reasons to initiate the treatment. Since the disease can lead to severe visual impairment and eventually blindness the main therapeutic goal is to preserve optic nerve function. First line treatment is weight loss in obese patients. The dietary measurements are usually accompanied with pharmacologic therapy - acetazolamide.
and furosemide for lowering ICP; amitriptyline, propranolol for primary headache prophylaxis; and, in some cases corticosteroids for lowering ICP.

The therapeutic role of repeated lumbar punctures which were popular in the past is now questioned due to its difficulties for the patient and lack of evidence showing any long lasting effects.

Surgical interventions include optic nerve sheath fenestration and CSF diversion procedures including either lumboperitoneal or ventriculoperitoneal shunting. In patients with bilateral TSS unilateral TSS stenting appears to be a safe and effective treatment.
Findings and procedure details

**Brain MR** (magnetic resonance) is considered to be the best imaging tool in IIH. When MR is not available, a contrast-enhanced CT (computer tomography) scan should be sought. The preferred imaging is brain MR with T2 coronal images and fat-sat (FS) images of orbit accompanied by MRV (magnetic resonance venography).

It is very important to understand the main roles of neuroimaging:

- **to exclude other pathologies** causing intracranial hypertension
- if possible, **to detect** findings associated with IIH.

The cause of raised ICP in IIH is unknown and, in contrast to cases where the role of radiologist is to find specific radiologic signs to confirm the proposed diagnosis, in this case the main radiologic task is to exclude other causes of raised ICP. The causes for secondary raised intracranial pressure (secondary pseudotumor syndrome) that mustn't be overlooked on neuroimaging scans are dural venous sinus thrombosis and space-occupying lesions (Table 2). Recognition of a secondary cause of raised ICP like venous sinus thrombosis or the diagnosis of IIH has crucial therapeutic and prognostic implications.

The differentiation of central venous thrombosis and IIH may be difficult even on venographic studies.

| 1. Increased blood volume | a. occlusion (e.g. venous thrombosis)  
b. blood outside vessels (e.g. hemorrhagic stroke) |
|---------------------------|--------------------------------------------------|
| 2. Increased brain volume | a. intracranial mass (e.g. tumor)  
b. cerebral edema (e.g. chronic meningitis) |
| 3. Increased CSF volume | a. increased production (e.g. choroid plexus papilloma)  
b. decreased absorption/obstruction to CSF outflow (e.g. obstructive hydrocephalus) |

**Table 2**

*References:* University Hospital, Sisters of Charity University Hospital - Zagreb/HR

Brain MR scan showing no abnormalities is a frequent finding in IIH. However, subtle radiologic findings suggestive of IIH have emerged with modern neuroimaging and the radiologist should know them thoroughly in order to perform an accurate reading of the scans.
The possible MR findings in IIH are:

- posterior globe flattening (Fig.1)
- intraocular protrusion of the optic nerve head (Fig.2)
- optic nerve sheath enlargement (Fig.3)
- vertical optic nerve tortuosity (Fig.3)
- empty or partially empty sella (Fig.4)
- enhancement (with gadolinium) of the prelaminar optic nerve
- venous sinus stenosis
- enlarged Meckel cave (Fig.5)
- meningoceles, typically within the temporal bone and sphenoid wing (Fig.5)
- slit-like ventricles (uncommon finding)

These findings are not diagnostic of IIH, and their presence is not obligatory for the diagnosis of definite IIH. Their discovery on brain imaging should not prompt invasive procedures, unless clinical signs of IIH, such as papilledema, are present.
Fig. 2: Axial T2 MR scans through orbits of the patient suffering from IIH show posterior globe flattening with mild bulging of the optic nerve disc head due to papilledema.

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Fig. 1: A) Sagittal T1 midline brain MRI scan of 43 year old woman presenting with headache and diplopia and B) Axial T2 MRI orbits scan of 40 year old obese woman with papilledema and visual disturbance. Both scans are showing posterior globe flattening.
Fig. 3: Coronal STIR orbit MR scans show dilated optic nerve sheaths and optic nerve tortuosity in a patient with papilledema and suspected IIH.
Fig. 4: Sagittal T1 midline brain MRI scan shows partially empty sella in a female patient with suspected IIH.

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Fig. 5: MR brain scans of obese 38 year old female patient with clinical symptoms of IIH. A) Sagittal T1 midline scan shows partially empty sella, B) Axial T2 scan shows enlarged Meckel cave bilaterally asymmetrical, C) Coronal STIR scan shows trans-ethmoid meningocele.

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Conclusion

When IIH is suspected, neuroimaging can help confirm the diagnosis and distinguish it from other conditions such as venous thrombosis and brain masses.
References