Etiology and Diagnosis of Intracranial Hypertension: Using MRI to Narrow an Index Patient’s Differential

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Objectives

Using an index case of a patient with intracranial hypertension (IH), this presentation will provide the skills and knowledge necessary to:

- Recognize the signs of IH on MRI
- Understand the logical basis for the differential diagnosis of IH
- Develop a foundation for systematically approaching MR scans in patients with IH
Our Index Patient: Chief Complaint, History, and Physical

- 34 year-old woman with chronic headache (recently worsened) and blurred vision referred to an ophthalmologist
- Complicated medical history, including 50 lb weight gain in the past 1-2 years
- Physical exam: optic disc swelling, i.e. papilledema (see figure)
- Referred by the ophthalmologist for an MRI to look for the potential cause of the patient’s signs and symptoms

(Bienfang, DC; UpToDate)
Before looking at our patient’s MRI, we’ll review

(1) a few points about CTs and MRIs; and
(2) a few points about different MRI sequences
Menu of Tests: CT vs. MRI in Patients with Papilledema

- Depending on the suspected cause of a patient’s papilledema, physicians may choose to order a CT and/or an MRI
  - For example, as we will explore later in the presentation, subarachnoid hemorrhage is one potential cause of papilledema; in a patient with acute subarachnoid hemorrhage, CT is usually the test of choice
  - MRI, on the other hand, is preferable to CT for diagnosis of many other causes of papilledema, e.g. soft tissue lesions
- This presentation concentrates only on diagnosis using MRI
Distinguishing MRI Sequences

- “T1” = T1 weighted image (*fluid is dark, i.e. low intensity*)
- “T2” = T2 weighted image (*fluid is bright, i.e. high intensity*)
- “C+” = Image enhanced with gadolinium (*i.e. IV contrast was administered*)
- “FLAIR” = Fluid attenuated inversion recovery (*a sequence that nulls signals from fluids to allow visualization of lesions whose signals are normally obscured by the signal from cerebrospinal fluid*)
- “MP-RAGE” = Magnetization prepared rapid acquisition gradient echo; designed for rapid image acquisition (*a T1 weighted scan*)
Now let’s look at our patient’s MRI
Our Patient: Evidence of Intracranial Hypertension on MRI

Findings
- Increased fluid in optic nerve sheath bilaterally (bright signal)
- Mild flattening of posterior globes
- Tortuous optic nerves
What fluid could be responsible for the high intensity signal surrounding the optic nerve within the optic sheath?
Review of CSF Flow

- CSF can track along potential space surrounding optic nerve

CSF Production and Flow

CSF in Optic Nerve Sheath

(Cummings, B; IUPUCAnatomy) (Corkrey O'Hara, JK website)
CSF and Intracranial Hypertension

- Increased pressure within the intracranial compartment is necessary for CSF to track along the optic sheath

- Sometimes intracranial hypertension causes other findings, such as hydrocephalus

Now let’s review some basic principles of intracranial pressure to understand what could underlie our patient’s MRI findings that are suggestive of intracranial hypertension
Compliance and Intracranial Pressure

- Compliance is equal to change in volume divided by change in pressure \((C = \Delta V / \Delta P)\)
  - The cranial compartment is incompressible (i.e. it has a low compliance)
  - If compliance is low, then for a given increase in volume, there will be a large increase in pressure \((\Delta P = \Delta V / C)\)

- Therefore, any change in volume of the structures or fluids within the cranium will produce large changes in pressure
As a result, to generate a differential diagnosis for intracranial hypertension (and therefore, to find a cause for the patient’s papilledema and orbital MRI findings), let’s consider what inside the intracranial compartment could be increased in volume
Major Physical Components of the Intracranial Compartment

1. Blood
2. Brain
3. CSF
Causes of Intracranial Hypertension: Differential Diagnosis

1. Increased blood volume
2. Increased brain volume
3. Increased CSF volume
Causes of Intracranial Hypertension: Differential Diagnosis

1. Increased blood volume
   a. occlusion (e.g. venous thrombosis)
   b. blood outside vessels (i.e. hemorrhagic stroke)

2. Increased brain volume

3. Increased CSF volume
Causes of Intracranial Hypertension: Differential Diagnosis

1. Increased blood volume
   a. occlusion (e.g. venous thrombosis)
   b. blood outside vessels (i.e. hemorrhagic stroke)

2. Increased brain volume
   a. intracranial mass (e.g. tumor)
   b. cerebral edema

3. Increased CSF volume
Causes of Intracranial Hypertension: Differential Diagnosis

1. Increased blood volume
   a. occlusion (e.g. venous thrombosis)
   b. blood outside vessels (i.e. hemorrhagic stroke)

2. Increased brain volume
   a. intracranial mass (e.g. tumor)
   b. cerebral edema

3. Increased CSF volume
   a. increased production (e.g. from choroid plexus papilloma)
   b. decreased absorption/obstruction to CSF outflow
Let’s look at our patient’s MRI more closely to consider each of these potential causes of our patient’s IH.

We will also look at MRIs from other patients (whom we will call “companion patients”) for comparison.
Companion Patient 1: Venous Thrombosis on MRI (Differential 1a)

Venous Thrombosis
(filling defect in superior and straight sagittal sinuses)

Index Patient: Normal

Sagittal C+ T1 MRI
(Poon, CS, et al; AJROnline)

Sagittal C+ T1 MRI
(PACS BIDMC)
Companion Patient 2: Subarachnoid Hemorrhage on MRI (Differential 1b)

Subarachnoid Hemorrhage
(high intensity signals in basal cisterns)

Index Patient: Normal

Axial FLAIR MRI
(Medscape)

Axial FLAIR MRI
(PACS BIDMC)
Companion Patient 3: Metastatic Tumor on MRI (Differential 2a)

Tumor
(heterogeneously enhancing lesion in the lateral ventricle: a metastasis from the lungs)

Index Patient: Normal

Axial C+ T2 MRI (PACS BIDMC)

Axial C+ T1 MRI (PACS BIDMC)
Companion Patient 4: Cerebral Edema on MRI (Differential 2b)

Cerebral Edema
(enhancing fluid suggestive of vasogenic edema)

Index Patient: Normal

Axial C+ FLAIR MRI
(PACS BIDMC)

Axial FLAIR MRI
(PACS BIDMC)
Companion Patient 5: Choroid Plexus Papilloma on MRI (Differential 3a)

Choroid Plexus Papilloma
(heterogeneous lesion in the lateral ventricle)
(hydrocephalus from increased CSF production)

Index Patient: Normal

Axial T2 MRI
(RadsWiki)

Axial T2 MRI
(PACS BIDMC)
Companion Patient 6: Obstructive Hydrocephalus on MRI (Differential 3b)

**Obstructive Hydrocephalus**
(large vestibular schwannoma)
(hydrocephalus due to obstruction)

**Index Patient:** Normal

Sagittal MP-RAGE MRI
(PACS BIDMC)

Sagittal MP-RAGE MRI
(PACS BIDMC)
Having examined MRIs of our patient juxtaposed with MRIs of relevant companion patients, we now return to our differential
In our Patient’s MRIs, Did We See Any Evidence for the Following Diagnoses in Our Differential?

1. Increased blood volume
   a. occlusion (e.g. venous thrombosis)
   b. blood outside vessels (i.e hemorrhagic stroke)

2. Increased brain volume
   a. intracranial mass (e.g. tumor)
   b. cerebral edema

3. Increased CSF volume
   a. increased production (e.g. from choroid plexus papilloma)
   b. decreased absorption/obstruction to CSF outflow
We Saw No Evidence of Increased Blood Volume

1. Increased blood volume
   a. occlusion (e.g. venous thrombosis)
   b. blood outside vessels

2. Increased brain volume
   a. intracranial mass (e.g. tumor)
   b. cerebral edema

3. Increased CSF volume
   a. increased production (e.g. from choroid plexus papilloma)
   b. decreased absorption/obstruction to CSF outflow
We Saw No Evidence of Increased Brain Volume

1. Increased blood volume
   a. occlusion (e.g. venous thrombosis)
   b. blood outside vessels

2. Increased brain volume
   a. intracranial mass (e.g. tumor)
   b. cerebral edema

3. Increased CSF volume
   a. increased production (e.g. from choroid plexus papilloma)
   b. decreased absorption/obstruction to CSF outflow
And We Saw No Evidence of Increased CSF Volume

1. Increased blood volume
   a. occlusion (e.g. venous thrombosis)
   b. blood outside vessels

2. Increased brain volume
   a. intracranial mass (e.g. tumor)
   b. cerebral edema

3. Increased CSF volume
   a. increased production (e.g. from choroid plexus papilloma)
   b. decreased absorption/obstruction to CSF outflow
We created a differential diagnosis for our patient by logically working through the components of intracranial pressure and intracranial-compartment volume.

Yet we saw no evidence in our patient of any of the diagnoses on the differential.

Therefore, we have no etiologic explanation for the patient’s intracranial hypertension.

What do we call diseases for which we have no etiologic explanation?
Causes of Intracranial Hypertension: A More Complete Differential Diagnosis

1. Increased blood volume
   a. occlusion (e.g. venous thrombosis)
   b. blood outside vessels

2. Increased brain volume
   a. intracranial mass (e.g. tumor)
   b. cerebral edema

3. Increased CSF volume
   a. increased production (e.g. from choroid plexus papilloma)
   b. decreased absorption/obstruction to CSF outflow

4. Idiopathic Intracranial Hypertension (i.e. Pseudotumor Cerebri)
Pseudotumor Cerebri: A Diagnosis of Exclusion

Idiopathic Intracranial Hypertension (IIH) = Pseudotumor Cerebri
(two names for the same diagnosis)

Dandy Criteria:

- Symptoms and signs of increased intracranial pressure (e.g. headache, transient visual obscurations, pulse synchronous tinnitus, papilledema, visual loss)
- No other neurologic abnormalities or impaired level of consciousness
- Elevated intracranial pressure with normal cerebrospinal fluid (CSF) composition
- A neuroimaging study that shows no etiology for intracranial hypertension
- No other cause of intracranial hypertension apparent
Pseudotumor Cerebri: Clinical Presentation and Epidemiology

**Presentation**
- Headache (92% of patients)
- Transient visual obscurations (72%)
- Intracranial noises (pulsatile tinnitus) (60%)
- Photopsia (54%)
- Retrobulbar pain (44%)
- Diplopia (38%)
- Sustained visual loss (26%)

**Epidemiology**
- Annual incidence: 1-2 per 100,000 people
- Higher in obese women ages 15-44 years old: 4-21 per 100,000
Pseudotumor Cerebri: Presentation on MRI

- Flattening of the posterior sclera (80% of patients)
- Empty sella (70%)
- Distension of perioptic subarachnoid space (50%)
- Enhancement (with gadolinium) of the prelaminar optic nerve (45%)
- Vertical tortuosity of the orbital optic nerve (40%)
- Intraocular protrusion of the prelaminar optic nerve (30%)
Our Patient: Diagnosis and Follow-Up

- Following MRI, lumbar puncture showed elevated opening pressure and normal CSF, confirming suspected diagnosis

- Treated with diuretics; advised to lose weight

- Presented at ED a week later with diplopia and headache
Our Patient: Intracranial Hypertension on MRI at Follow-Up

- Presented with same findings as on previous MRI
- Treated with lumbar-puncture drainage of CSF
- Prescribed medication for headache prophylaxis
- Treatment will be ongoing and will be aimed at alleviation of symptoms and prevention of permanent vision loss

Index Patient: Follow-Up
(Can you identify the same 3 findings identified on our patient’s MRI on slide 8?)

Axial T2 MRI (PACS BIDMC)
Summary

- Understanding the basic anatomy and physiology of the intracranial compartment allows for the logical formulation of a differential diagnosis for intracranial hypertension.

- Radiologic imaging modalities such as MRI are used not only to identify patients with characteristic “Aunt Minnie” findings indicative of a particular disease, but also to make diagnoses of exclusion, such as pseudotumor cerebri (i.e. idiopathic intracranial hypertension).
References

Bienfang, DC. Images of papilledema. UpToDate (accessed July 2009).
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