UCNS Review Course: High Yield Images

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Disclosures

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Learning Objectives

• recognize normal and abnormal findings on diagnostic imaging in patients presenting with headache
• contextualize the significance of imaging findings to potential diagnoses in patients with headache
• identify visible findings on physical or neurologic exam that aid in diagnosis of patients with headache
Depressed, Demented Dolores

- 64 y.o. woman referred for depression and dementia
- Migraines since her 20s
- Memory troubles since her late 40s
- In her early 50s had left-sided weakness for a few weeks which resolved
- Was told she has multiple sclerosis
Depressed, Demented Dolores

- Increasing headache frequency
- Further cognitive decline over the past few years
- Depressed and at times agitated
- One period with psychosis
- Father had “Alzheimer’s”
- Multiple family members also with migraine, and a paternal aunt diagnosed with MS
Is this her MRI?
Is this her MRI?

No. That looks like UBOs.
Or is this her MRI?
Or is this her MRI?

No. That looks like MS.
No, this is. Diagnosis?
CADASIL

(Cerebral Autosomal Dominant Arteriopathy with Subcortical Infarcts and Leukoencephalopathy)

Gene: NOTCH3
Chromosome: 19
Weary, One-Eyed Wanda

- 85 y.o. woman 7 months of slowly progressive right-sided headache
- Throbbing temporal/parietal
- Ipsilateral photophobia, lacrimation, nasal congestions
- Nocturnal exacerbations
- Treated with CBZ for TN – no benefit
Weary, One-Eyed Wanda

- One week ago, increased tearing and eye swelling, given steroid eye drops after normal ophtho exam
- Three days ago, abrupt ptosis and diplopia
- PMHx: DM, HTN, cardiomyopathy
Cavernous sinus aspergillosis in uncontrolled diabetes

45 year-old man, 8 months of progressive headache with migrainous features including n/v, also blurry vision

Normal exam except mild convergence insufficiency

Initial MRI with ventricular asymmetry but normal CSF flow at skull base

CT shows?
Unilateral hydrocephalus (obstructive)

Intrathecal dye fills only one side

• This man has right-sided cluster attacks and just finished strenuous exercise
• What does the photo illustrate?
• Harlequin syndrome
• Sympathetic dysfunction on right (ipsilateral to cluster)

Familial hemiplegic migraine (FHM)

- **FHM1** (19p13): *CACNA1A* encodes $\alpha_1$ subunit of voltage-gated neuronal $Ca_v2.1$ (P/Q) $Ca^{2+}$ channel
  - 50% of cases
- **FHM2** (1q23): *ATP1A2* encodes $\alpha_2$ subunit of Na+/K+ pump
- **FHM3** (2q24): *SCN1A* encodes $\alpha_1$ subunit of neuronal voltage-gated Na+ channel $Na_v1.1$
Positional headache and this MRI?
• Marfanoid or Ehlers-danlos
• At risk for spinal CSF leak
• Might not have positional headache
• Best initial step to localize is spine MRI
• May need CISS sequences (heavily T2-weighted) with thin cuts
• Myelogram if can’t get MRI or it’s unrevealing
• Extravasation of dye through nerve sheath diverticula into paraspinal spaces
• Radionuclide study another option
Normal radioisotope cisternogram

Slow-flow CSF leak

Reduced activity at convexities at 24 hr

High-flow CSF leak

Early activity in kidneys and bladder

What happened here?

before

after
Prior Images
Misdiagnosed as Chiari, worse after decompression
A bit of herniated brain...
Very fast leak that I still haven’t found. He might have a CSF-venous fistula (hard to diagnose).

4 hours  
22 hours
MRI findings in high CSF pressure/volume states
Grey arrows: optic disc edema

White arrows: excess CSF in optic sheaths
• Flattened pituitary
• Or frank empty sella
• Can have downward displacement of rounded cerebellar tonsils
Bilateral transverse sinus stenosis
Frontal headache aggravated by reading. What is it?
Frontal headache aggravated by reading. What is it?

Trochleitis. How is it treated?
Local injection of steroid and anesthetic
<table>
<thead>
<tr>
<th>Disorder</th>
<th>Duration</th>
<th>Frequency</th>
<th>Gender (F:M)</th>
<th>Acute Treatment</th>
<th>Preventive/Bridge Treatment</th>
</tr>
</thead>
<tbody>
<tr>
<td>Cluster</td>
<td>15-180 min</td>
<td>Every other day to 8/day</td>
<td>1:3-7 (trending towards women)</td>
<td>Oxygen, SC sumatriptan, NS sumatriptan or zolmitriptan</td>
<td>verapamil, topiramate, lithium, methylergonovone, corticosteroids</td>
</tr>
<tr>
<td>PH</td>
<td>2-30 min</td>
<td>1-40/day</td>
<td>2-3:1 (trending towards men?)</td>
<td>None</td>
<td>indomethacin</td>
</tr>
<tr>
<td>SUNCT/SUNA</td>
<td>1-600 sec</td>
<td>Dozens to hundreds per day</td>
<td>2:1 SUNCT</td>
<td>None</td>
<td>lamotrigine, topiramite, gabapentin, indomethacin?</td>
</tr>
<tr>
<td>HC</td>
<td>Constant with spikes</td>
<td>Few to many per day</td>
<td>2:1 or less</td>
<td>None</td>
<td>indomethacin</td>
</tr>
</tbody>
</table>
Thunderclap Headache

- Severe headache
  - Sudden onset
  - Peak intensity in < 1 minute

- Must rule out acute neurologic event

- Often primary migraine
Thunderclap Headache

10 things that cause thunderclap headache?
**Subarachnoid Hemorrhage**

- Sudden and dramatic: “a blow on the head”
- Severe unilateral headache
  - Becomes generalized
  - Spreads to back of head
  - May have backache
- Photophobia, neck stiffness, Kernig’s sign, focal neurologic signs, and alterations in consciousness
- CT/LP usually diagnostic
Imaging Subarachnoid Hemorrhage

- CT – positive in 98% within 24 hours
- MRI FLAIR
  - Equally sensitive as CT
  - More sensitive than CT 3-40 days after ictus
- MRA Sensitivity for aneurysm: 70-100%
- CTA Sensitivity for aneurysm: 85-98%
## CSF Xanthochromia post-SAH

<table>
<thead>
<tr>
<th>Time post SAH</th>
<th>Probability (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>&lt;6 hours</td>
<td>?</td>
</tr>
<tr>
<td>12 hours</td>
<td>100</td>
</tr>
<tr>
<td>One week</td>
<td>100</td>
</tr>
<tr>
<td>Two weeks</td>
<td>100</td>
</tr>
<tr>
<td>Three weeks</td>
<td>~70</td>
</tr>
<tr>
<td>Four weeks</td>
<td>~40</td>
</tr>
</tbody>
</table>
9 more things that cause thunderclap headache?
## Thunderclap Headache

### Vascular
- Subarachnoid hemorrhage
- Aneurysmal thrombosis or expansion
- Cerebral hemorrhage
- Cervical arterial dissection
- Cerebral venous thrombosis
- Hypertensive crisis
- Reversible cerebral vasoconstriction syndrome
- Pituitary apoplexy

### Non-Vascular
- Spontaneous Intracranial Hypotension/Hypovolemia
- Colloid cyst of the third ventricle
- Meningitis
- Sinusitis (especially sphenoid)
- Primary cough, sexual, and exertional headache
- Primary thunderclap headache (idiopathic)
Some cerebrovascular causes of TCH

Cerebral hemorrhages: 5-10% of all TCH (possible in infarcts)

Dissection: TCH in 5%

Venous thrombosis: TCH in 3%

RCVS

Temporal arteritis

Pituitary apoplexy

Aneurysmal warning leak
Other disorders causing TCH

- Tumors: 3d ventricle, pituitary, cerebellum
- Spontaneous intracranial hypotension
- TCH in 15%
- Sinusitis
- Meningitis
- Myocardial ischemia (cardiac cephalgia)
- Heart

Spontaneous intracranial hypotension TCH in 15%
A 32-year-old woman who gave birth to her first child 3 weeks ago reports a history of four severe headaches over the past 2 weeks. Each headache reached maximum intensity almost instantly and was located over the back of her head bilaterally. Her medical history is otherwise significant for depression, treated with fluoxetine, and migraine headaches that she states are very different from this new type of head pain. A CT scan without contrast and an MRI with and without contrast are both normal. Which of the following is the most likely diagnosis in this patient?

A. dural venous sinus thrombosis
B. internal carotid artery dissection
C. intracranial aneurysm
D. intracranial arteriovenous malformation
E. reversible cerebral vasoconstriction syndrome (RCVS)
A. dural venous sinus thrombosis
B. internal carotid artery dissection
C. intracranial aneurysm
D. intracranial arteriovenous malformation

E. reversible cerebral vasoconstriction syndrome (RCVS)

The preferred response is E (reversible cerebral vasoconstriction syndrome [RCVS]). The recurrent nature of this patient’s thunderclap headaches together with her history of recently giving birth and a history of migraines would suggest the diagnosis of RCVS. Taking a serotonergic medication (fluoxetine) would also put her at risk for this condition. For more information, refer to page 1064 of the Continuum article “Thunderclap Headache.”
Reversible Cerebral Vasoconstriction Syndrome

Multifocal segmental stenosis (beading) of the major intracranial arteries

Calabrese, L.H., et al., Narrative review: Reversible cerebral vasoconstriction syndromes. *Annals of internal medicine, 2007*
Reversible Cerebral Vasoconstriction Syndrome

- RCVS: multiple areas of cerebral arterial constriction (string and beads)
- Recurrent thunderclap headache, +/- neurologic symptoms or signs
- Idiopathic, pregnancy, puerperium, idiosyncratic drugs reaction (licit and il-), pheocromocytoma
- CSF normal, angiography diagnostic
- Treatment: observation, calcium channel blockers (nomodipine, verapamil), steroids?
Reversible Cerebral Vasoconstriction Syndrome

Stenosis (white arrows) and dilatation (black arrows) which resolved after treatment with calcium channel blocker

Calabrese, L.H., et al., Narrative review: Reversible cerebral vasoconstriction syndromes. *Annals of internal medicine, 2007*
Brain Imaging in RCVS

Legend: (A) CT scan showing a small cSAH, (B) MRI (FLAIR sequence) showing a small cSAH, (C) CT scan showing an occipital intracerebral haemorrhage, (D) MRI showing sequelae of bilateral occipital infarcts and left frontal-parietal infarct, (E) MRI (FLAIR) showing hyperintensities and (F) MRI in the same patient after 28 days showing resolution.

FSPGR MPR coronal reconstructed image (a) revealed the trigeminal nerve in its cisternal tract, bilaterally (arrows); 3D-TOF MR angiography sequence MPR coronal reconstruction (b) showed the contact between left superior cerebellar artery and the upper surface of the nerve (arrowhead). FSPGR: fast spoiled gradient echo; MPR: multi-planar reconstruction; 3D-TOF MR: three-dimensional time-of-flight magnetic resonance.
FISP MPR coronal reconstructed image (a) revealed the trigeminal nerve in its cisternal tract, bilaterally (arrows); 3D-TOF MR angiography sequence MPR coronal reconstruction (b) and MIP reconstruction (c) showed the contact between right superior cerebellar artery and the upper surface of the nerve (arrowhead). FISP: fast imaging with steady-state precession; MPR: multiplanar reconstruction; 3D-TOF MR: three-dimensional time-of-flight magnetic resonance; MIP: maximum intensity projection.
Questions?