Secondary Headaches

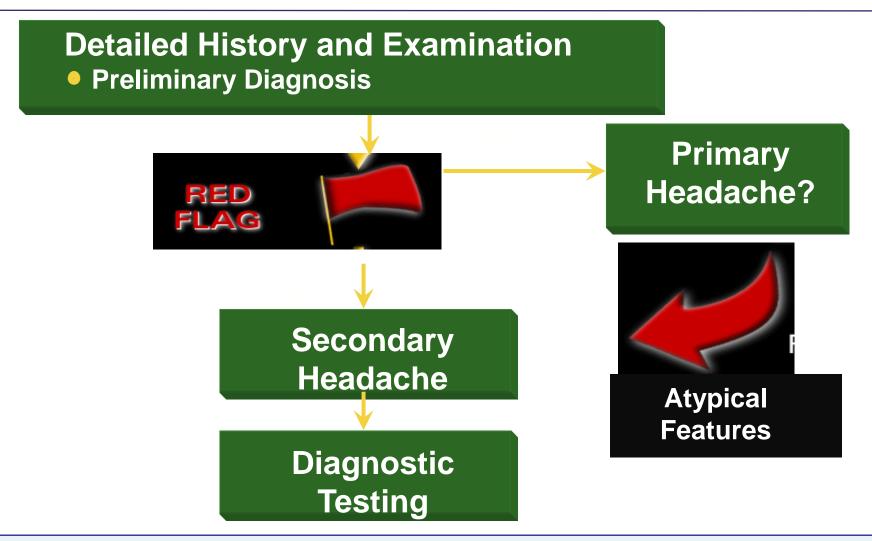
- Amir Rezaei M.D.
 - Neurologist

Learning Objectives

- 1. To recognize "red flags" for headaches with a discernable cause
 - Is it time to rename "secondary headaches"?
- 2. To reinforce understanding on how to diagnose, manage and treat dangerous and rapidly progressive headaches due to specific causes.
- 3. To review headaches that we may not encounter on a regular basis.
 - How much of our practice is primary headache disorders due to access reasons?
 - Who is actually taking care of these patients?

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DIAGNOSIS AND TESTING





The Best 2 Tools We Have

History

- Headache onset
- Duration
- Location
- Severity and Quality
- Associated Features
- Aggravating Factors
- Exam
 - General exam!
 - Neurologic exam



SNOOP (or some variation...)

- Systemic disease or Symptoms (or Secondary risk factors)
 - Malignancy
 - Fever
 - Weight loss
- Neurologic signs or symptoms
- Onset that is sudden (acute or thunderclap)
- Onset "later in life". (Older age of onset) Use the epidemiology!!!
- Previous headache history with new or different headache features
- Progressive
- Postural
- Precipitation by Valsalva or exertion (distinguished from simply worsening with movement)
- Pregnancy.... Papilledema....

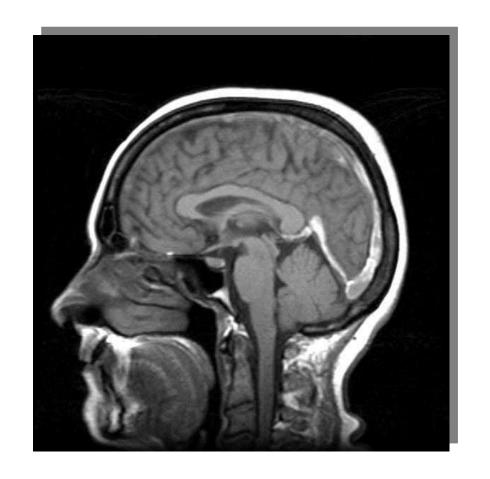


Organic Causes for Headache Missed by CT Scanning

- Cerebrovascular
 - Arterial dissection (MRA)
 - Cerebral venous sinus thrombosis (MRV)
 - CNS vasculitis
- Meningoencephalitis
- Tumors
 - Posterior fossa
 - Pituitary
 - Leptomeninges
- High and low intracranial pressure syndromes

MRA = Magnetic resonance angiography

MRV = Magnetic resonance venography



Headache Diagnoses Missed by CT

Vascular

- Saccular aneurysms
- SAH
- AVM
- Carotid or vertebral dissection
- Stroke
- Cerebral venous sinus thrombosis
- Vasculitis
- RCVS

Neoplastic

- Parenchymal and extra-axial neoplasms (esp in posterior fossa)
- Meningeal carcinamatosis
- Pituitary tumor/hemorrhage
- Brain metastasis

Infectious

- Meningoencephalitis
- Cerebritis and brain abscess
- Cervicomedullary lesions
 - Chiari malformation
 - Foramen magnum meningioma
 - Acoustic neuroma
- Other
 - CSF leak/intracranial hypotension
 - Intracranial hypertension
 - Dural and leptomeningeal disease

What to discuss...



Too Many Headaches to Cover!!!

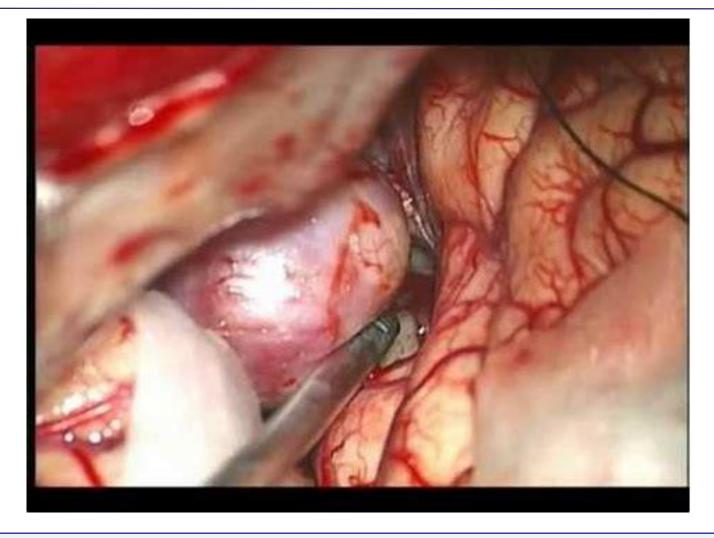
Will focus on selected disorders:

- 1. Headaches secondary to Vascular Disorders
- 2. Headaches associated with Spinal Fluid Disorders
- 3. Headaches associated with CNS Infections

4. Potpourri

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Vascular Headaches...

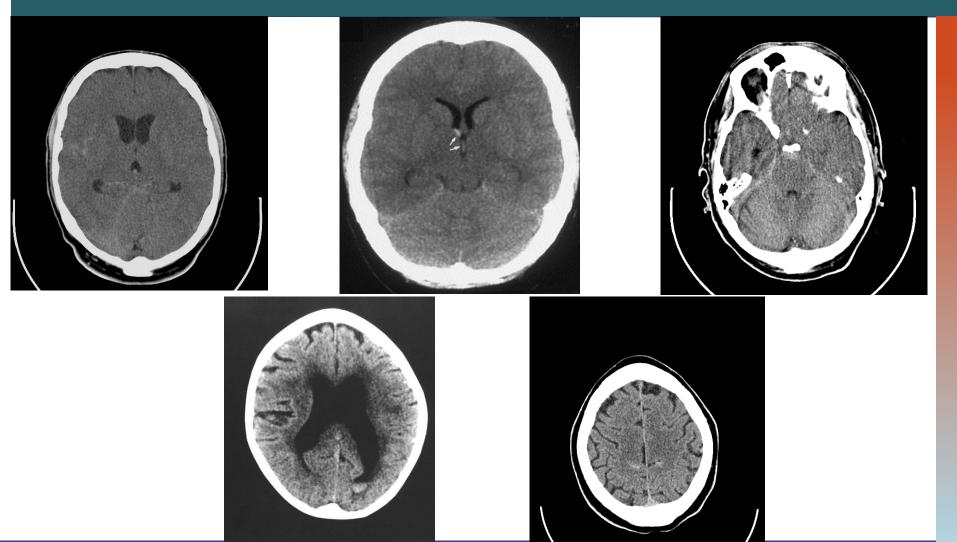


Secondary Headache: Subarhachnoid Hemorrhage

- First or worst Headache
- Often missed
- Maximum headache in <60sec
- LOC, focal findings, seizures, cardiac death
- Early CT +in > 90%, LP + early in 100%



Subarachnoid Hemorrhage: Beware of Blood in the PITS



Parenchymal

ntraventricular

runcal

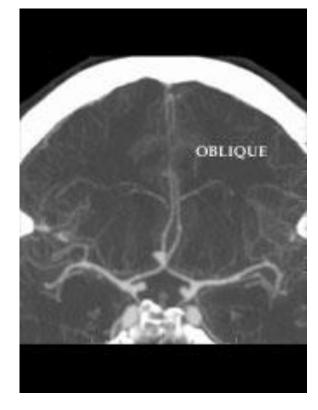
Sulcal



Subarachnoid Hemorrhage

- Risks: rebleed and vasospasm
- Early surgery,then increase perfusion
- 10% have multiple aneurysms
- Best evidence: control blood pressure!





Intracerebral Hemorrhage

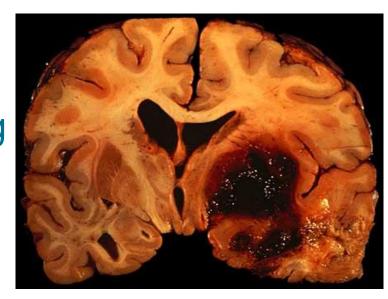
- Focal deficits plus headache
- Progresses quickly
- Difficult to arouse
- If hemosiderin staining

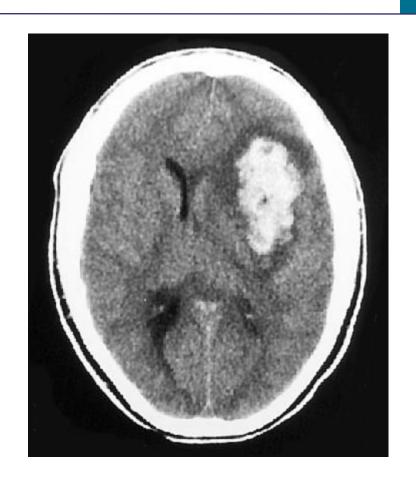
And young

Think cavernoma

If older

Think Amyloidosis



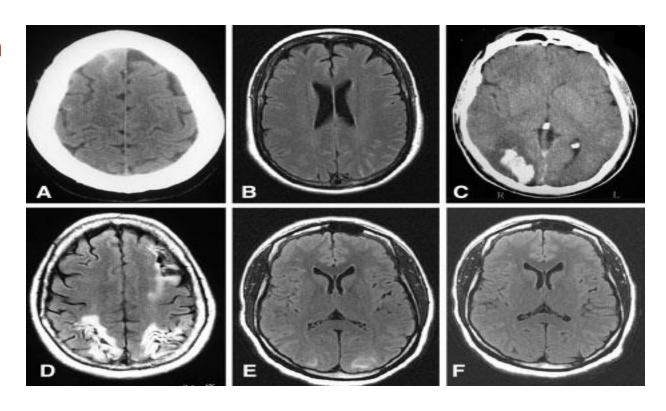




Reversible Cerebral Vasoconstriction Syndrome (RCVS)

RCVS

- Most commonly precipitated post-partum or with the use of vasoactive substances
 - Cannabis
 - Cocaine
 - SSRIs
 - Binge drinking
 - Nasal decongestants
- May result in:
 - SAH
 - ICH
 - Stroke
 - PRES
- Treated with IV/PO calcium channel blockers





Temporal/ Giant Cell Arteritis

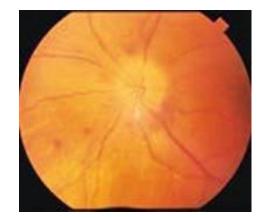
- Age >60 with subacute onset of headaches
- High risk to involve ophthalmic artery, posterior ciliary arteries, or branch of external carotid
- Less common: cerebral and coronary arteries
 - Could be considered subset of primary CNS vasculitis
- Symptoms:
 - visual loss (arteritic anterior ischemic optic neuropathy)
- -Also
 - Temporal artery tenderness, weight loss, malaise, fever, chills, and jaw claudication
 - Polymyalgia rheumatica common

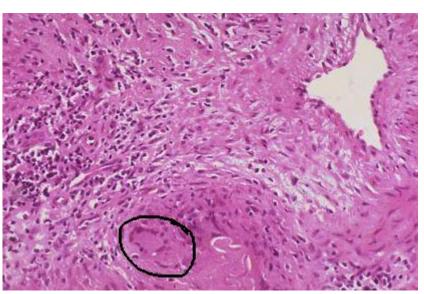


Giant Cell Arteritis

- Lab: ↑↑ ESR
 - Also, Anemia, leukocytosis and elevated liver enzymes
- Diagnosis: Need temporal artery bx although increasing evidence for ultrasound
- Treatment: High dose steroids (sometimes other immune suppressants
- CD4 Lymphocytes and macrophages







From blood vessels to spinal fluid...

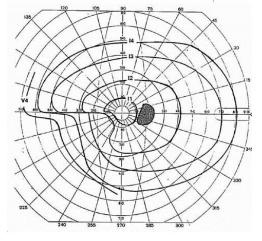
Pressure Disorders:

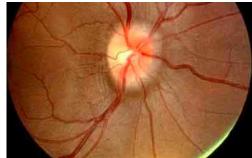


Pseudotumor

- Headache: Unremarkable daily headache
- May have coocurrence with migraine
- Pulsatile tinnitus, TVOs
- Papilledema
 - Enlargement of blind spot
 - Loss of inferonasal field
 - other
- 6th Nerve palsy in 10 20%
- Empty Sella
- Lateral Sinus Abnormality







Pseudotumor

- Risk factors include female gender and obesity
- Not all cases are idiopathic
 - Increased risk of thrombosis with cigarette smoking and with the use of oral contraception.
- NORDIC IIHT trial demonstrated safety and efficacy of acetazolamide for pseudotumour
 - 38/86 tolerated the 4 grams per day in study
- There is mixed evidence for the use of stenting
- Shunts are possible, but complications common
- Optic Nerve Fenestration is option to preserve vision

PSEUDOTUMOR CEREBRI LAWSUIT

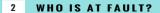


Women who have used the IUD birth control product Mirena and have been diagnosed pseudotumor cerebri, may be entitled to compensation.

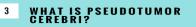




Hundreds of pseudotumor cerebri lawsuits have been filed by women who used Mirena and developed migraine headaches, vision loss, blindness, and more.



Bayer, the manufacturer of the Mirena IUD, is accused of failing to warn about the risk.



Pseudotumor Cerebri is a condition that causes progressively higher pressure inside the skull when there is too much cerebrospinal fluid.

The fluid builds up and is not released or absorbed properly, putting pressure on the brain — just like a growing brain tumor, but it is not actually a tumor.

4 WHO SHOULD FILE A LAWSUIT

Women who were diagnosed with pseudotumor cerebri while using Mirena should talk to a lawyer about filing a lawsuit against Bayer.

As of January 2018, over 400 pseudotumor cerebri lawsuits nationwide have been centralized in the U.S. District Court for Southern New York

5 WHERE CAN I LEARN MORE?

If you are interested in learning more about the pseudotumor cerebri and the Mirena lawsuit, please visit Advocacy For Patients™

On our website, we provide important news, information, and resources regarding this urgent Heath Alert.

www.advocacyforpatients.org









The information provided by Advocacy For Patients™ is not a substitute for and should not be taken as medical or legal advice. Viewers are encouraged to speak with a medical professional or lawyer regarding their individual situation.



7.2 Headache attributed to low cerebrospinal fluid pressure



7.2.1 Post-dural puncture headache

- A. Headache worsens within 15 minutes after sitting or standing and improves within 15 minutes of lying and has at least 1 of the following
 - Neck stiffness, tinnitus, hypacusia, photophobia, nausea
- B. Dural puncture has been performed
- C. Headache develops within 5 days after dural puncture
- D. Headache resolves either:
 - Spontaneously within 1 week
 - Within 48 hours after effective treatment of the spinal fluid leak (usually by blood patch)
 - (True in 95% of cases)

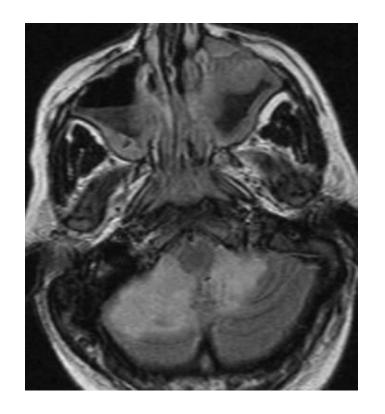


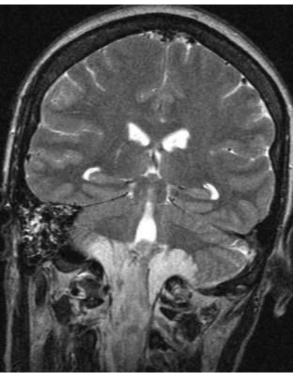
7.2.3 Headache Attributed to Spontaneous (Idiopathic) CSF Leak

- A. Diffuse or dull headache that worsens within 15 minutes after sitting or standing with at least one of the following:
 - Neck stiffness, tinnitus, hypacusia, photophobia, nausea
- B. At least one of the following:
 - Evidence of low CSF pressure on MRI
 - Evidence of CSF leakage on myelography, CT myelography or cisternography
 - CSF pressure <60mm H2O in sitting position
- C. No history of dural puncture or other causes of CSF Fistula
- Headache resolves within 72 hours after epidural blood patching



Complications





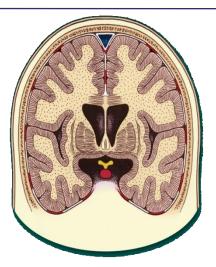


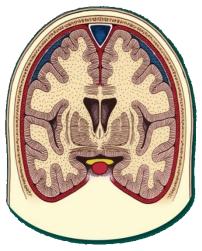
Patient with known intracranial hypotension who rapidly deteriorated shows cerebellar, brainstem & cord infarctions.

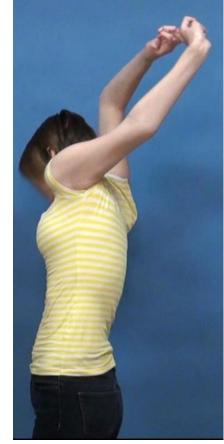
Spinal Fluid Leaks

SEEPS

- Subdural fluid collection
- Enhancement of meninges
- **E**ngorgement of veins
- Pituitary hyperemia
- Sagging of brain



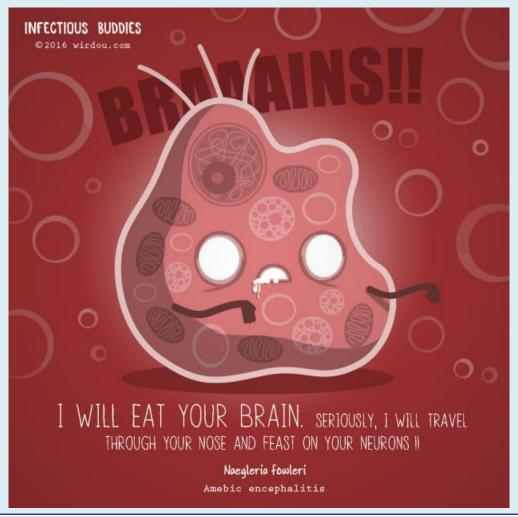




Hypermobile joints sometimes seen in those with SIH

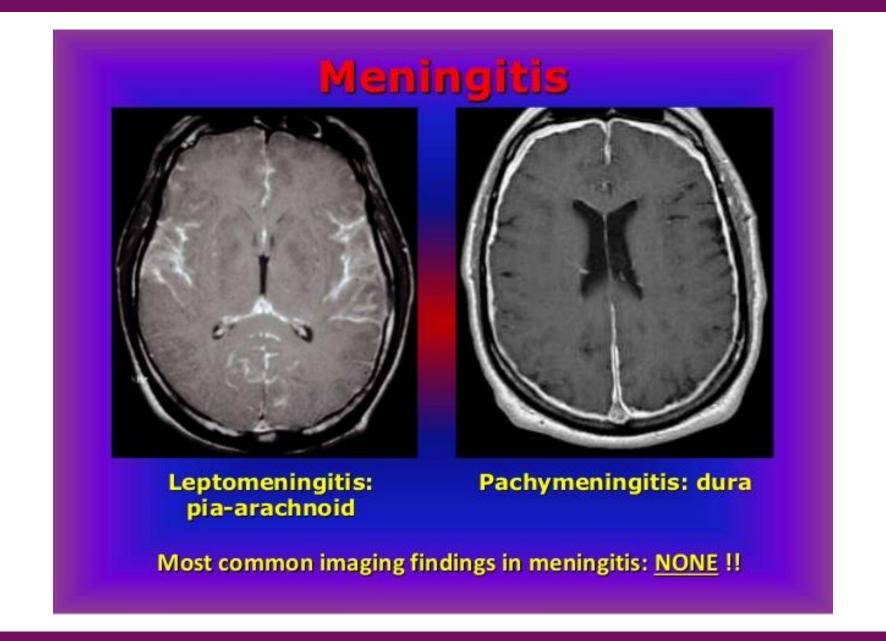


CNS Infections



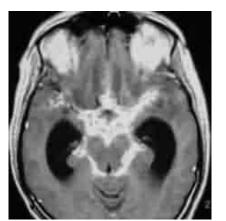
Meningitis

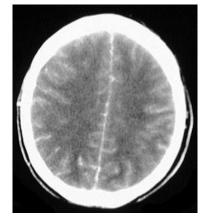
- Headache, fever, stiff neck, confusion, decreased consciousness and cranial neuropathies
- Bacterial: Rapidly declining... CT, LP, antibiotics ASAP
- Viral/Aseptic: Slower onset, "less sick", lesson from Mollaret
- Chronic/Fungal/TB: Cranial neuropathies more common, stiff neck less common. Often requires high volume tap to obtain positive cells.



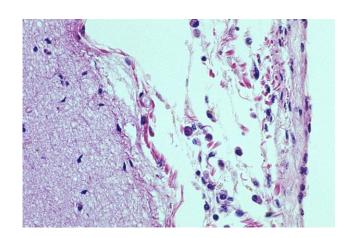
Meningitis







Meningeal enhancement in Bacterial meningitis.





Acute Meningitis

Bacterial Meningitis:

- LP shows elevated PMN's, elevated protein and reduced glucose.
- treat adults with Ampicillin and Ceftriaxone pending cultures.

Viral Meningitis/ Encephalitis:

- LP shows mild increase of lymphocytes, increased protein and normal glucose
- Can include enteroviruses, lymphocytic choriomeningitis virus, HIV, as well as many others.

Chronic Meningitis

Tuberculosis:

- LP shows increased lymphocytes, elevated protein and low glucose. Large volume tap gives 50% yield of acid fast mycobacteria.
- Treat with four drugs for 1st 2 months until sensitivity is known.

Neurosyphilis:

- Primary, Secondary, Tertiary
- Gummas, aortitis, chorioretinitis
- Three major syndromes: tabes dorsalis, Argyll Robertson pupils, general paresis of the insane.
- LP for VDRL- may consider FTA-ABS in late syphilis.
- Treat with Penicillin 2.4 million units IM weekly for 3 weeks if non neurologic. Otherwise treat Penicillin G 2-4 million units IV q4 for 10 days.

Chronic Meningitis

Lyme Disease:

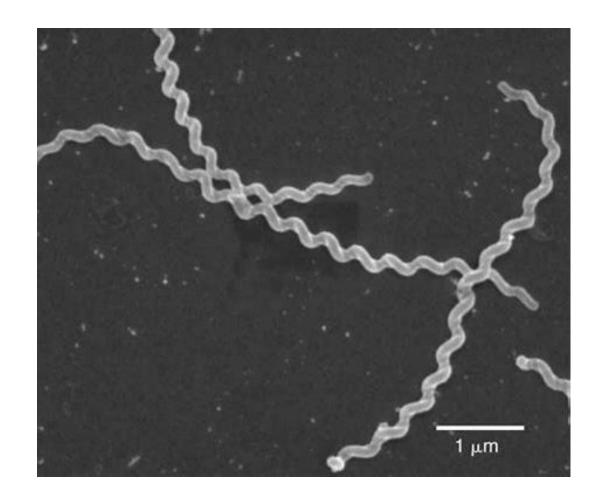
- Mononuclear pleocytosis with increase protein. Can send PCR for Borrelia or detect IgG antibodies via ELISA or Western Blot.
- Ceftriaxone 2 g IV qday or Penicillin G 4 million units IV q 4 for 2-4 weeks.

Fungal Meningitis

- Can include Cryptococcus, Coccidioides, Candida, Histoplasma, Blastomyces
- LP shows mononuclear increase, elevated protein and near normal glucose.
- Treat with Amphotericin B or some with fluconazole.

Leptospirosis

- Spirochetal disease most often Leptospira Interrogans from cats, dogs, etc.
- Fever, chills, myalgia, nausea, diarrhea, meningitis, hepatitis, renal failure.
- CSF shows mononuclear pleocytosis with elevated protein
- Treat with doxycycline 100mg IV q12 or Penicillin G 5 million units IV q6 for 7 days.



Brain Abscess

- Bacterial Abscess
 - Subacute progressive headache (75%), altered mental status(70%), focal neurological signs(50%), fever (50%)
 - Ring enhancing lesion; pathogen by CSF 10%, by biopsy 80%
 - Look for extradural cause
 - Treat empirically with oxacillin 2g IV q4 (or penicillin G), Metronidazole and Ceftriaxone. Consider Amphotericin B
- Subdural Empyema
- Cranial and Spinal Epidural Abscess
 - High dose dexamethasone 60 to 100mg IV push followed by 10-20 mg q6 (and call surgery!).
 - Vancomycin and Ceftriaxone
- Toxoplasmosis
 - Sulfadiazine and pyrimethamine, Clindamycin
- Cysticercosis



Viral Encephalitis

- Herpes Simplex Encephalitis
 - Acyclovir 10-12.5 mg/ kg IV q8 for 2-3 weeks.
 - Phenytoin
 - Consider Steroids
- Mumps, Enterovirus, Arbovirus (Equine, St. Louis, California, colorado tick virus, West Nile, Zika, Chikungunya, Dengue)
- Measles virus
- Rabies Virus
- Epstein Barr virus
- Cytomegalovirus
- Whipple's Disease
- Elizabethkingia



Approach to the patient (The Basics)

- Think- Fever, Headache, Neurologic Sign
- History and Exam
 - Acute or Chronic
 - Predisposing Factors (diabetes, EtOH abuse, Malignancy, Steroids, chemotherapy, AIDS)
 - Systemic Infection (endocarditis, pneumonia, osteomyelitis, skull fracture, otitis media, tick bite, animal bites)
 - Exam should include: signs of fever, headache, change in mental status, focal weakness and back pain, as well as: papilledema, meningismus, skin rash, sinus tenderness, spine tenderness.
- Lumbar Puncture



Lumbar Puncture

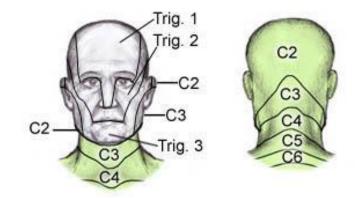
Fever, Headache, Change in Mental Status

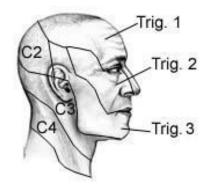
 Consider CT imaging if: Papilledema, Focal Neurologic Deficit (especially brainstem), Known Intracranial Mass Lesion, AIDS, Lethargy, Stupor or Coma

Don't Forget Your Opening Pressure!!

Cervicogenic Headache

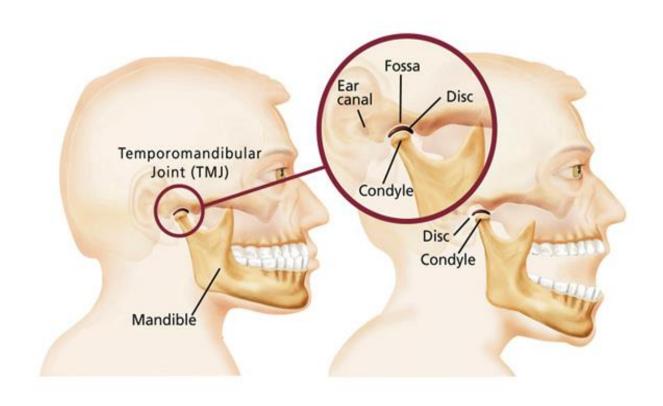
- Headache caused by disorder of cervical spine (bone, disc and soft tissue) demonstrated by clinical, laboratory and/ or imaging evidence
- Need at least 2 of 4:
 - Headache developed in temporal relation to onset of disorder
 - Headache significantly improved or resolved in parallel with improvement of cervical disorder
 - Cervical range of motion reduced and headache worsened with manoeuvers
 - Headache abolished following diagnostic blockade of cervical structure or nerve supply
- Not better accounted by another diagnosis!
 - HA attributed to cervical myofascial pain
 - HA attributed to upper cervical radiculopathy





Temporomandibular Joint Disorder

- Temporomandibular joint dysfunction is an umbrella term covering pain and dysfunction of the muscles of mastication (the muscles that move the jaw) and the temporomandibular joints
- More common in young adults
- Pain in jaw and ear, restricted movement, lateral motion, locking up, difficulty chewing
- Treated with exercises, bite plates, injections, muscle relaxants, relaxation
- Causes: Trauma, Arthritis, Disk erosion, Inflammatory conditions



Some Other Secondary Headaches

- Subdural hematoma
- Ischemic stroke
- Transient ischemic attack
- Cervicocephalic arterial dissection
- Cerebral venous thrombosis
- Unruptured arteriovenous malformation
- Postcarotid endarterectomy

- Bell's palsy associated with retroauricular pain
- Brain tumors and abscesses
- Dental abscesses
- Sinusitis
- Trigeminal neuralgia
- Low-CSF pressure headache
- Acute glaucoma
- Arterial hypertension

