



Experts on Call: Case Example

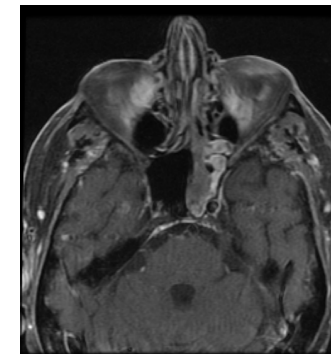
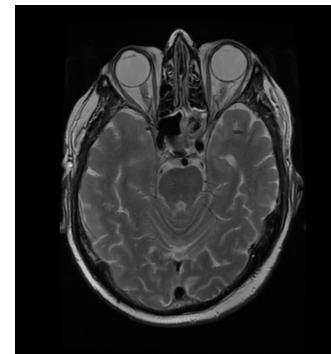
David Dodick, MD, FAAN

Professor Emeritus, Department of Neurology, Mayo Clinic; Chief Science Officer, Atria Academy of Science and Medicine; Adjunct Professor, University of Copenhagen, Norwegian University of Science and Technology USA



79-year-old man referred with headache

- 1 month history of severe left temporal headache associated with scalp sensitivity, and transient partial loss of vision in left eye 1 week ago. Headache worse when lying flat
- Follicular Non-Hodgkin Lymphoma (completed course of chemotherapy 6 weeks ago), hypertension, dyslipidemia, OSAS, prostate cancer
- Examination: VA OD 20/25; OS 20/40; IOP 17/16; 'Macular hole' (Ophthalmology). Neurological examination normal.
- Laboratory results
 - ESR 61; HGB 12.3; WBC 6.2; platelets 252; CRP 4.6
 - Bilateral temporal artery biopsies negative
 - 1 month later, blind left eye

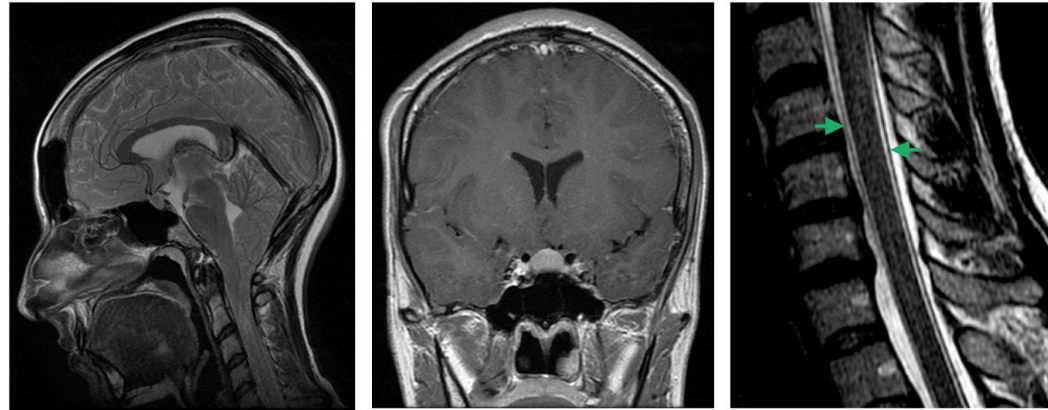
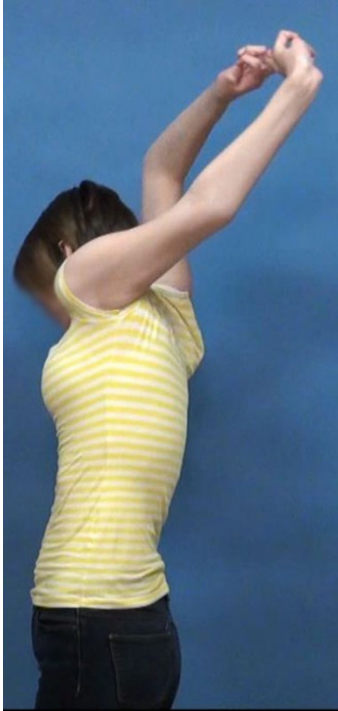


Clinical Pearls

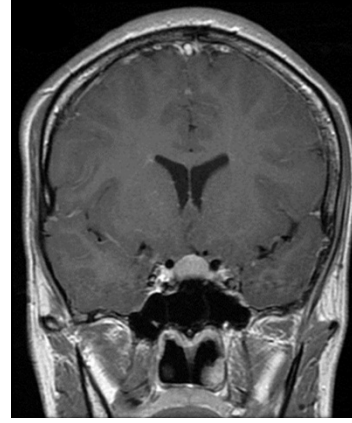
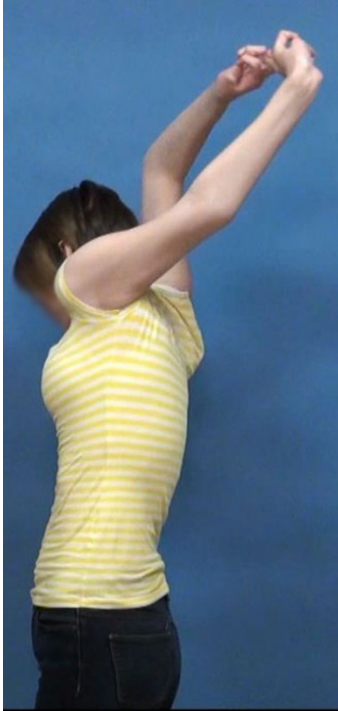


- 1. Look at the sinuses on MRI brain**
- 2. Sphenoid sinusitis is a medical emergency**
- 3. Requires index of suspicion, especially in immunocompromised, headache worsening with Valsalva/supine position**
- 4. There are causes other than GCA of new onset headache in the elderly with elevated ESR**

28 year old woman with persistent headache for 2 months



28 year old woman with persistent headache for 2 months



Spontaneous Intracranial Hypotension Imaging Features

- **SEEPS & COPS**

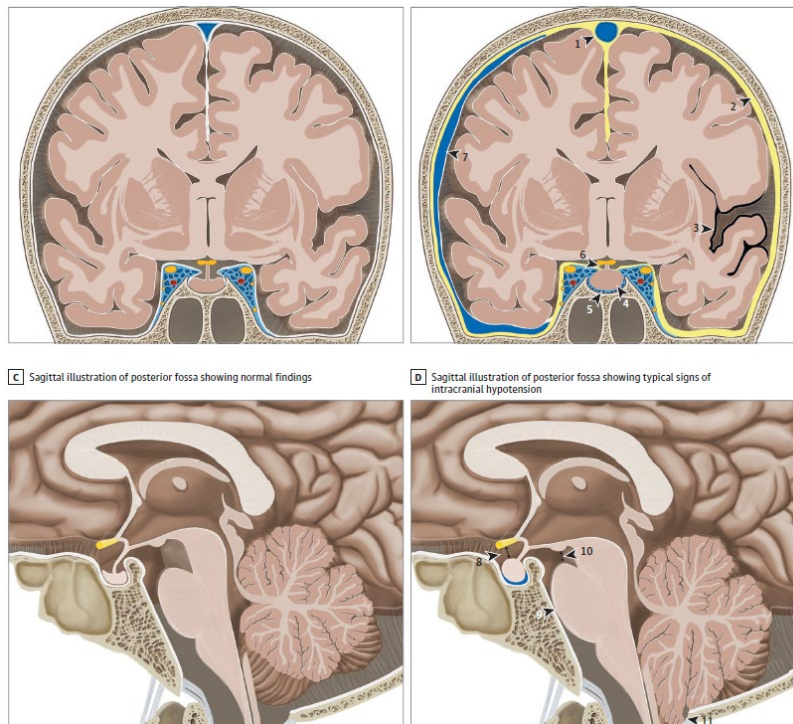
- **S**ubdural fluid collection (36-50%)
- **E**nhancement of pachymeninges (56-83%)
- **E**ngorgement of venous sinuses (48-93%)
- **P**ituitary enlargement/hyperemia (5-63%)
- **S**agging of brain (18-61%)
 - **C**hiari (26-61%)
 - **O**ptic chiasm
 - **P**ontine flattening *
 - **S**inking brain **



in patients with headache and one of these imaging features,
always consider SIH, regardless of whether headache is orthostatic

Six Imaging Signs that are most discriminative for SIH

The score is based on three qualitative and three quantitative signs and identifies a patient with a **high** (score ≥ 5), **intermediate** (score 3 to 4), or **low** (score ≤ 2) probability of having a CSF leak.



Imaging Characteristic	Score point
Engorgement venous sinus	2
Pachymeningeal enhancement	2
Subdural fluid collection	1
Suprasellar cistern ($\leq 4\text{mm}$)	2
Prepontine cistern ($\leq 5\text{mm}$)	1
Mamillipontine distance ($\leq 6.5\text{mm}$)	1

56 year old male referred emergently after angiogram for TIA

- History of MO, <40% R/LICA stenosis, type II diabetes, hypertension, dyslipidemia, 7 MIs, coronary stents
- 2 month history of unilateral (1 bilateral) stereotyped episodes of paresthesias involving fingers, hand, arm, face, and tongue
- Jacksonian march over 30-90 mins; disappear slowly and backward
- All episodes associated with shimmering halo in both eyes, bifrontal and retro-orbital throbbing headache, +/- expressive aphasia

Which diagnostic study is most likely to yield a diagnosis?

- A. MRI brain**
- B. CACNA1A mutation analysis**
- C. ATP1A2 mutation analysis**
- D. Perfusion-weighted brain MRI**
- E. Electroencephalogram (EEG)**
- F. Lumbar puncture**

- **CSF exam revealed 29 WBC (98% lymphocytes); protein 89mg/dl**
- **Diagnosis?**

HaNDL's Syndrome (Syndrome of transient headache and neurological deficits with CSF lymphocytosis)

- **>20 discrete episodes including sensory symptoms (78%), aphasia (66%) and motor deficits (56%). Migraine-aura-like visual symptoms are relatively uncommon (18%).**
- **Some individuals report a “march” of symptoms similar to that reported in typical migraine aura.**

HaNDL's Syndrome (Syndrome of transient headache and neurological deficits with CSF lymphocytosis)

- CSF lymphocytosis (10-760 cells/ul), elevations of CSF total protein (20-250 mg/dl) in >90% of cases, CSF opening pressure (100-400 mm H₂O) in >50% of cases. Papilledema is occasionally present.
- Routine CT and MRI scans (with or without intravenous contrast) and angiography are virtually always normal. Microbiological studies have been uniformly normal.
- EEG and SPECT scans may show focally abnormal areas consistent with the focal neurological deficits.

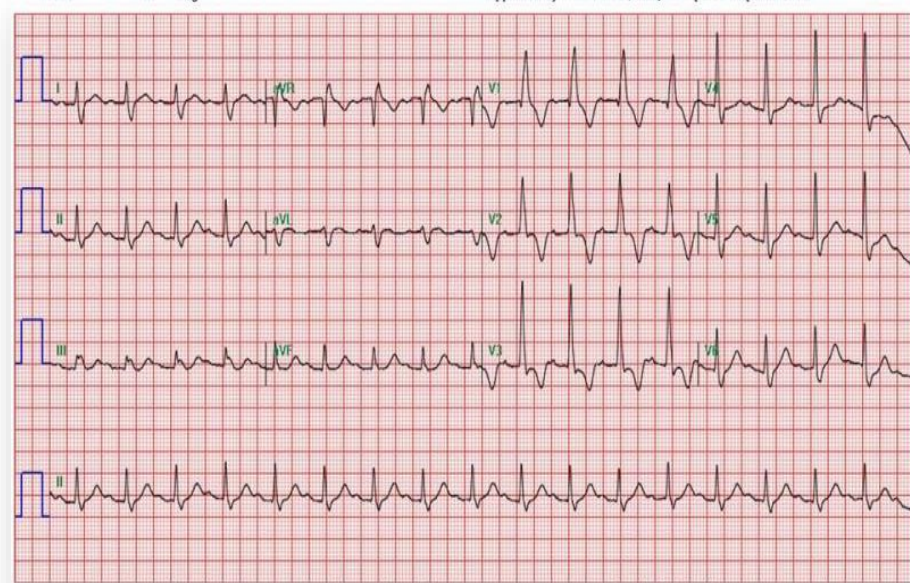
Clinical Pearl



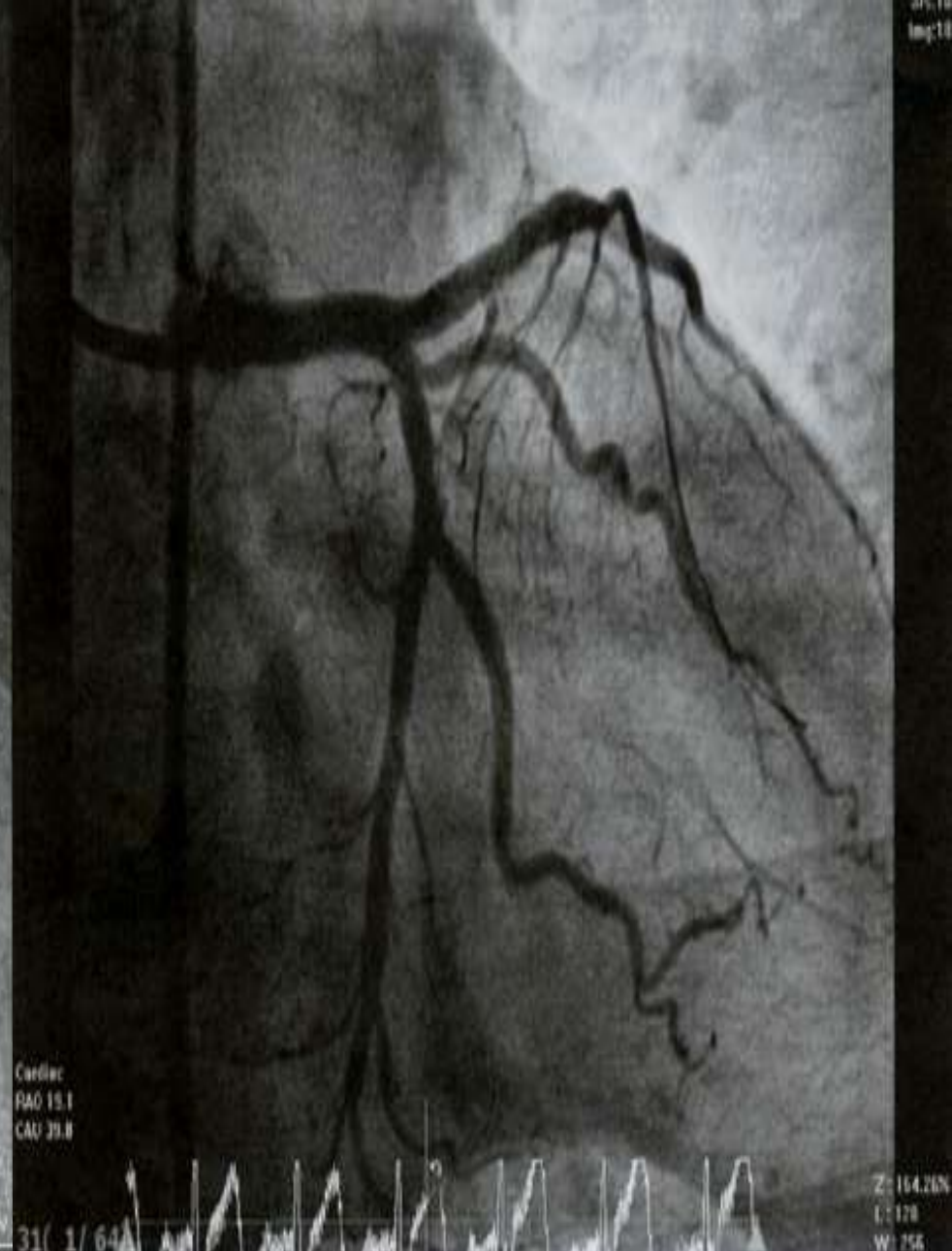
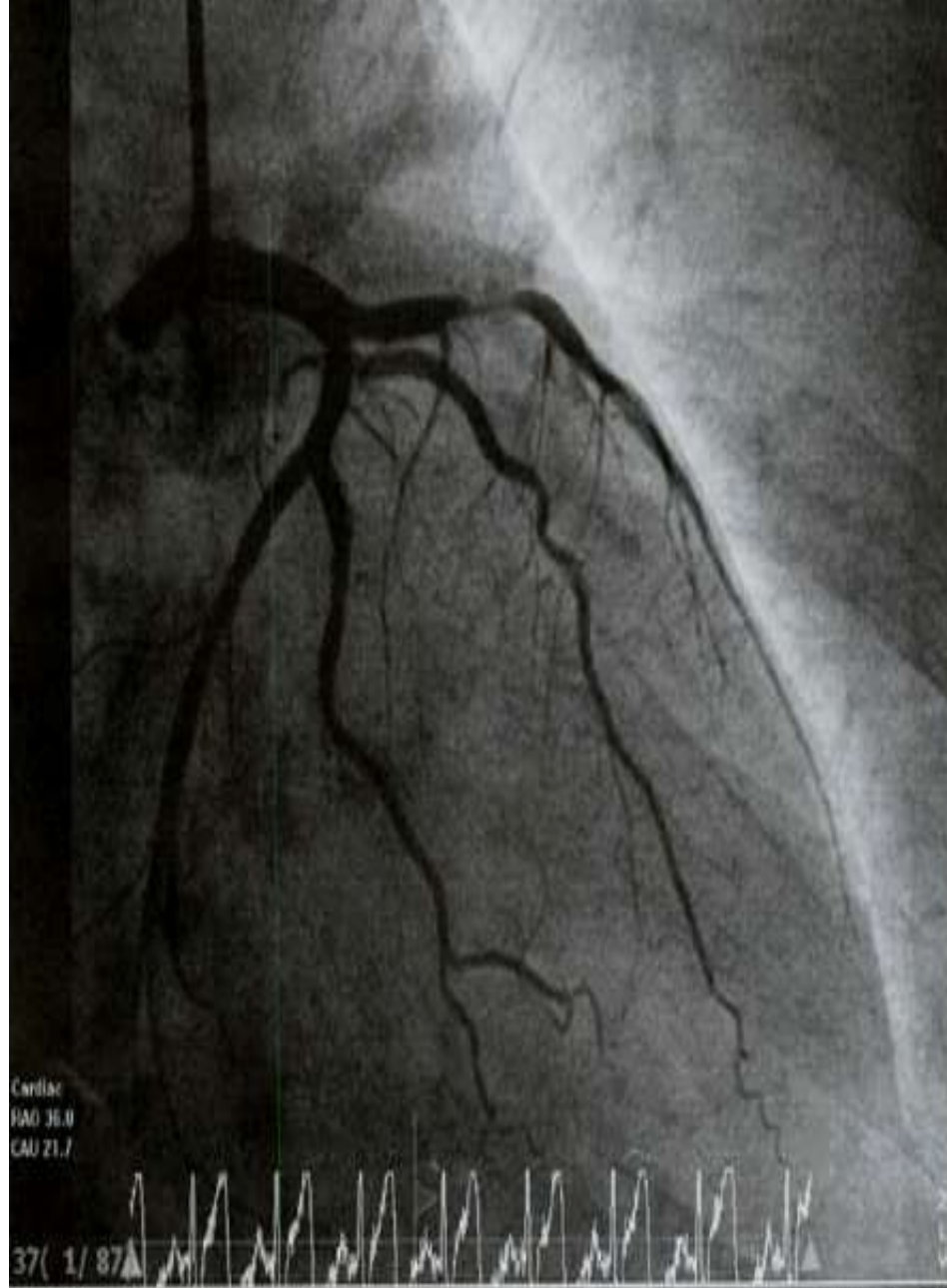
- Think HaNDL's syndrome and CSF exam in patients with frequent, recurrent, stereotyped, aura-like symptoms associated with migraine headache occurring over weeks to months

- **52 year old male presents with 6 week history of very severe left side-locked headache involving jaw, frontal, temporal and occipital regions. No systemic symptoms, history of trauma, new medications.**
- **Past medical history: hyperlipidemia (3 years on Simvastatin), tobacco use (20 pack years), moderate alcohol consumption.**
- **General physical and neurological examination normal. Moderate tenderness over left occipital/suboccipital region.**
- **At least 3 episodes per day, each lasting 5-10 mins, and rapid escalation of pain**
- **Brought on by exertion**

- CBC, ESR, CRP normal
- MRI brain and MRA head and neck normal
- EKG: RBBB
- Exercise stress test normal
- Prescribed aspirin 81mg and atorvastatin 40mg



- Presents 2 weeks later with very frequent episodes of severe headache and chest pain
- EKG normal, but elevated cardiac enzymes
- Diagnosis: NSTEMI



Pearls



- Headache is primary pain location in 3.5% of patients with angina/myocardial infarction
- Exertional headache may be a manifestation of cardiac disease, even with negative treadmill exercise ECG testing
- Cardiac cephalalgia should be considered in patients who present with new onset headache, especially when headache is exertional, patient is over 50 years, or has cardiac risk factors.

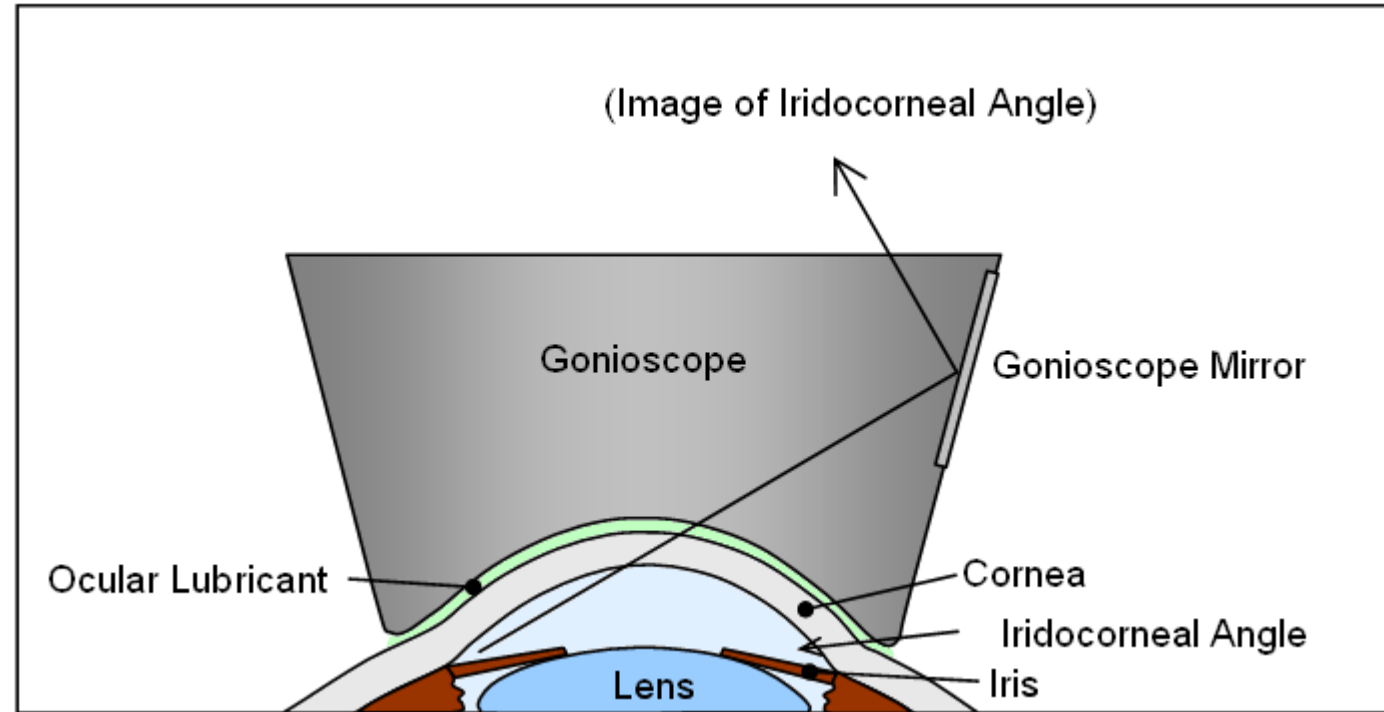
62 year-old woman

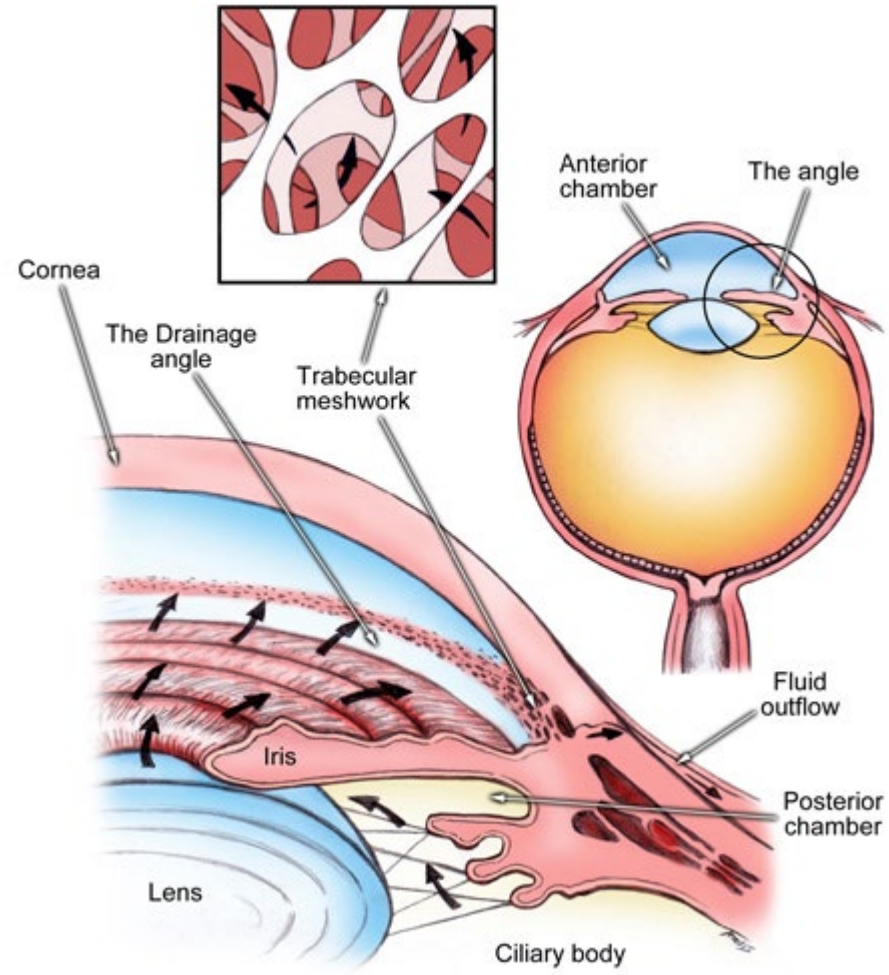
- **9 month history of chronic daily headache ('Chronic tension headache')**
 - **Frontal-temporal, moderate (6-7/10) aching**
 - **Nausea, brief visual blurring**
 - **Several times each week, usually at night, but before going to sleep, 30min-1hour**
 - **Physical and neurological examination are normal.**

Tests?

- CBC normal
- ESR (10)
- CRP (< 0.8)
- Brain MRI – normal
- Va 20/40 OD; 20/25 OS
- IOP
 - OD 15 mm Hg
 - OS 22 mm Hg
- Gonioscopy
 - Narrowed irido-corneal angles; enlarged cup:disc ratios (0.5/0.6); peripheral anterior synechiae OS
- Pilocarpine trial resolved headaches
- Headache free after laser iridotomies

Gonioscopy





Subacute Angle Closure Glaucoma

- > 50 years
- Short duration (< 1 hour)
- Visual symptoms 1/3
- Hyperopia $\leq 50\%$
- Recurrent angle scarring (peripheral anterior synechiae)
- Glaucomatous optic cupping
- Loss of peripheral vision

Diagnosis Requires Gonioscopic Examination



**THANK YOU
QUESTIONS?**

