Optometric Education Consultants

Neural Pearls

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DISCLOSURE:

Dr. Joseph Sowka is/ has been in the past 24 months a consultant or member of the advisory or speaker boards for Zeiss, Visus, and B&L. All relevant relationships have been mitigated. Dr. Sowka has no direct financial interest in any of the diseases, products or instrumentation mentioned in this presentation.

He is a co-owner of Optometric Education Consultants. www.optometricedu.com

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Know what can kill, maim, and blind immediately.

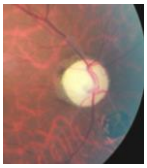
Once you have ruled out bad stuff, you have time to figure things out.

Acute painful ANYTHING is a neuro-ophthalmic emergency

Urgency of evaluation is dictated by duration of condition

46 YOM

- Reports waking up 3 months ago not being able to see OD
- LP OD, 20/20 OS
- Disc pallor OD- no other concurrent findings
- Last medical exam unknown- no medical hx
- Resident gets nervous- sends to ER immediately
- How long do we have to get this worked up?



Neuro-ophthalmic Urgencies and Emergencies

- GCA
  - Any sudden vision loss in the elderly
- Pituitary apoplexy
  - Headache, field loss, diplopia
- Aneurysm
  - Pupils
- Papilledema
  - Clinical suspicion
- Carotid dissection
  - Horner syndrome

66 YOF

ESR = 96

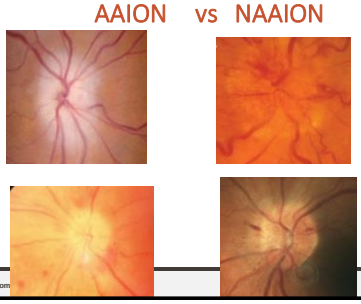
- New onset sudden vision loss
  - VA: 20/400 (longstanding macular scar)
  - Noticed inferior vision loss x 1 day
  - Inferior arcuate scotoma
- OD disc edema- mild pallor, no hemorrhages or teliangectasia
- OS disc- small, crowded disc at risk; C/D < 0.2
- Mild headache- relieved by OTC
- Malaise and loss of appetite- lost 7 lbs over 4 weeks
- No jaw claudication or temporal head pain
- What to do?

ANY acute vision loss in the elderly is GCA until proven otherwise

Anterior ISCHEMIC OPTIC NEUROPATHY

- Hypoperfusion of the posterior ciliary arterial supply to the anterior optic nerve head.
- May be arteritic (AAION) or non-arteritic (NAAION)
- Mechanical factors and atherosclerotic disease play a role in the non-arteritic form while vasculitis contributes in the arteritic form.
- Unilateral presentation but high incidence of subsequent contralateral involvement
  - AAION

AAION vs NAAION

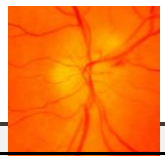


NAAION

- Risk factors:
  - Hypertension, diabetes, atherosclerotic disease, small optic nerves
- Inferior field defects
- Hyperemic swollen nerve- disc at risk
- Progressive moderate vision loss with potential recovery
- Late 30s/ early 40s and beyond
- Painless

AAION

- Pallid optic nerve swelling with flame hemorrhages, arteriole attenuation and NFL infarcts
- Pain (of some sort)
- Severe optic nerve dysfunction
- Visual field defects
- Giant cell arteritis/ PMR- risk factors
- Typically 70s, uncommon under 60
  - Any patient over 50 is at risk
- High risk bilateral involvement



Diagnosis

- Careful history: Must directly ask about nonvisual symptoms
  - Headache (present in over 90%), scalp tenderness, jaw claudication (almost diagnostic), ear pain, arthralgias, temple pain and/or tenderness, malaise, intermittent fevers
- Examination
- Laboratory studies
  - Erythrocyte sedimentation rate
    - Lowered by statins and NSAIDS
  - C-reactive protein
    - Not affected by statins and NSAIDS
  - Elevated platelet count

Initial symptoms in GCA

A normal exam

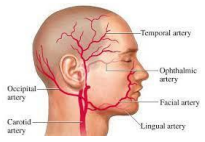
- Headache
- PMR
  - Chair and stair
- Fever
- Visual symptoms without vision loss
  - TIA, diplopia
- Weakness, malaise, fatigue
- What do all of these things have in common?

Vision Loss and Ocular Findings in GCA

- AION
- CRAO
- PION
- TIA
- Transient diplopia

Headache and pain in GCA

- Temporal
- Occipital
- Neck
- Ear
- Jaw
- Scalp



AAION

Diagnosis

- Prodrome, GCA symptoms
- Elevated ESR/CRP (combination of the two gives high specificity [97%])
- Elevated platelet count (acute phase reactant)
- Ophthalmoscopy
- Fluorescein angiography
- Temporal artery Biopsy
  - Negative biopsy: Read the report- "No giant cells, no active arteritis"
  - Focal interruption of the internal elastic lamina= healed arteritis

Treatment

- Prompt steroids and hydration
- Consider IV when vision loss present
  - Very effective in prevention of second eye
  - Occasionally restores vision
  - Best done through ER
  - 250 mg solumedrol QID x 3 days followed by orals

AAION versus NAAION

- Think AAION>>NAAION
  - Systemic symptoms of GCA
  - TVOs/TIAs
  - AION + cilioretinal artery occlusion
  - Evidence of posterior ciliary artery occlusion on FA
  - Multiple unexplained CWS
  - Early massive vision loss
  - Bilateral simultaneous vision loss
  - Chalky white optic disc edema
  - Elevated
    - ESR/CRP
    - Platelets

Hayreh SS, Podhajsky PA, Zimmerman B. Occult giant cell arteritis: ocular manifestations. Am J Ophthalmol. 1998 Apr;125(4):521-6.

NAAION is diagnosed in the negative. Get the tests done promptly with AION

Remember the E's in GCA:
Elderly, ESR is Elevated, only sees the Big E on the Eye chart, and its an Emergency

Neuro-ophthalmic Urgencies and Emergencies

- GCA
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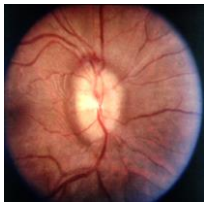
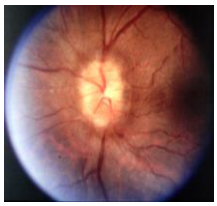
- Pituitary apoplexy
  - Headache, field loss, diplopia
- Aneurysm
  - Pupils
- Papilledema
  - Clinical suspicion
- Carotid dissection
  - Horner syndrome

28 YOF

- Presents with intermittent blurred vision & visual "grey-outs", intermittent horizontal diplopia, and chronic headache steadily worsening X 2 weeks
- MHx: "white coat hypertension", shoulder injury X 6 mos
- Meds: Flexeril® 10 mg BID PRN
- Height / weight: 5'3", 220 lbs.
- VA: OD 20/20, OS 20/20
- Pupils & motility: normal

What questions do you want to ask?
What tests do you want to perform?

28 YOF

- Additional hx: Dull "ringing" in ears
- BP: 142/100
- SLE: unremarkable
- T_A: OU 16 mm Hg
- VF: blind spot enlargement & nasal step defect OU
- Serology Normal
- Imaging: small ventricles, otherwise normal
- LP: O.P. = 510 mm H_2O; all CSF studies normal
- DX: Pseudotumor cerebri (PTC)

PAPILLEDEMA: Signs & Symptoms

- Signs:
  - **bilateral disc edema**
    - **superior & inferior aspects of discs affected FIRST**
    - **obliteration of optic cup**
    - **hemorrhages common**
    - **absence of SVP**
    - **Paton's folds**
  - **highly variable VF defects**
    - **enlarged blind spot (early)**
    - **arcuate defects and constricted (late)**
  - **NO RAPD typically**
  - **VA near normal**
- Symptoms:
  - **Visual:**
    - **transient visual obscurations**
    - **intermittent horizontal diplopia**
  - **General:**
    - **headache common**
    - **nausea & vomiting**
    - **dizziness**
    - **tinnitus**

Papilledema Types:

- Acute
  - **Hemorrhages, exudates, hyperemia, RNFL edema**
- Chronic
  - **Minimal hemorrhage/exudate. Collateral vessels may be present**
- Atrophic
  - **Eventually occurs if papilledema remains chronic. Optic disc pallor**

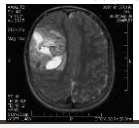
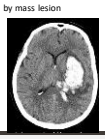
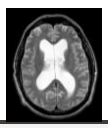


PAPILLEDEMA Pathophysiology

- Disc edema results from axoplasmic stasis
  - intracellular fluids, metabolic by-products accumulate and are regurgitated at the level of the optic nerve head
  - in papilledema, cerebral edema is effectively transmitted along the common meningeal sheaths of the brain and optic nerve producing an engorged, swollen disc.

PAPILLEDEMA Pathophysiology

- Associated with intracranial abnormalities:
  - increased brain volume (intracranial mass lesion)
  - increased intracranial blood volume
    - Intracranial hemorrhage
  - increased CSF volume
    - Hydrocephalus
    - Ventricular blockage by mass lesion

PAPILLEDEMA Management

- Rule out "swollen disc masqueraders"
  - ultrasonography can be invaluable in differentiating ONHD
  - also consider color, margins, SVP, vasculature, etc.
- Acute papilledema constitutes a medical emergency
  - Immediate neuro-imaging to rule out an intracranial mass.
  - If imaging is normal, lumbar puncture to measure CSF pressure and exclude meningitis or other disease processes is necessary.
- Atrophic papilledema with significant vision/field loss:
  - urgent measures must be undertaken to prevent blindness
- Papilledema accompanied by any neurologic abnormalities, fever or stiff neck:
  - Possible serious underlying neurologic abnormality, intracranial infection or bleed requiring immediate medical attention.

PTC vs. IIH

- Pseudotumor Cerebri (PTC)
  - Increased intracranial pressure in the absence of an intracranial mass lesion
  - Many causative agents have been identified
- Idiopathic Intracranial Hypertension (IIH)
  - Increased intracranial pressure without an identifiable cause
  - Young, obese females are at risk
- Primary PTC
  - IIH
- Poor CFS drainage

PSEUDOTUMOR CEREBRI DIAGNOSIS

- Si/SX: consistent with increased ICP
- Papilledema
- Normal neurological examination
  - except for cranial nerve 6 abnormalities
- Neuro-imaging: Normal without evidence of hydrocephalus, mass, or structural lesion, thrombosis
- Normal CSF composition

Elevated LP opening pressure
- Adults: > 250 mm CSF
- Children: > 280 mm CSF
> 250 mm CSF if not sedated/obese

PSEUDOTUMOR CEREBRI DIAGNOSIS

- LP may be deferred if:
- MRI/MRV shows no additional abnormalities and has characteristic findings of flattened globe and empty sella.
- No evidence of fever or acute infection
- Typical profile

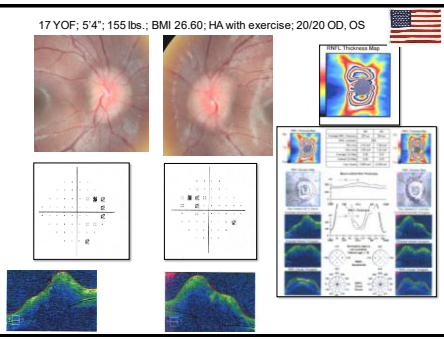
PSEUDOTUMOR CEREBRI MANAGEMENT

- No visual loss
  - Symptomatic headache therapy
  - Acetazolamide 500 mg tid
  - Weight reduction
- Mild visual loss
  - Acetazolamide 500 mg tid
    - Furosemide, Topiramate, Zonisamide
  - Weight reduction

PSEUDOTUMOR CEREBRI MANAGEMENT

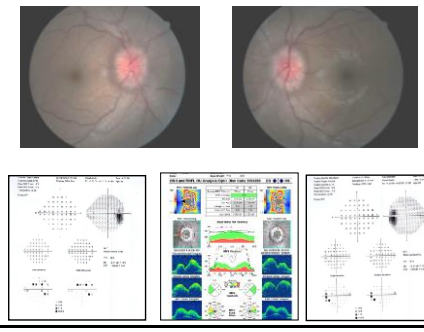
- No/ Mild visual loss
  - Prognosis
    - Excellent (all signs and symptoms, visual loss)
    - 6-9 months
  - Follow-up and visual fields
- Role of weight loss
  - Treat the primary problem
    - 10% weight loss
  - Prevent recurrence
    - Keep the weight down

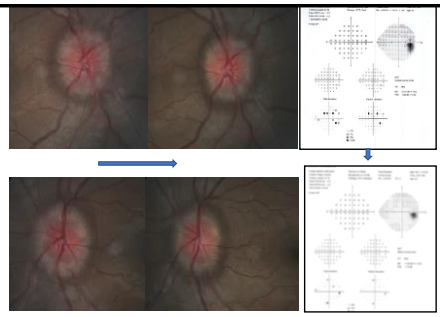
17 YOF; 5'4"; 155 lbs.; BMI 26.60; HA with exercise; 20/20 OD, OS



33 YOF

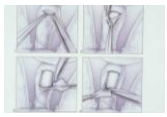
- Horizontal diplopia
- Headache
- TVOs 20/day
- Denies OCP, tetracyclines, vitamin A
- Lost 10 lbs- headaches improved
- 118/72
- 5'5"; 160lbs; BMI 26.62

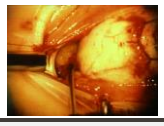




PSEUDOTUMOR CEREBRI MANAGEMENT

- Severe, or progression of visual loss
  - Optic nerve sheath decompression (ONSD)
  - High-dose IV steroids and acetazolamide
  - Lumboperitoneal shunt
    - Failed ONSD
    - Declined ONSD
    - Intractable headache





IIH is a slowly progressive condition…until its not

Fulminant IIH

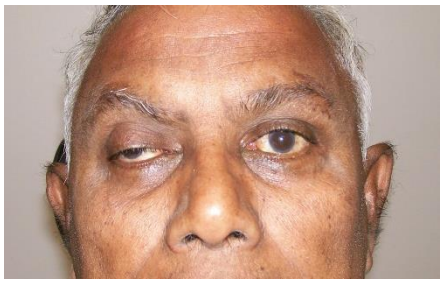
- Same diagnostic criteria for IIH/ PTC
- Less than 4 weeks between symptoms and loss of field/ acuity
- Vision worsening rapidly over several days
- Typically needs CSF diversion surgery and/or ONS fenestration

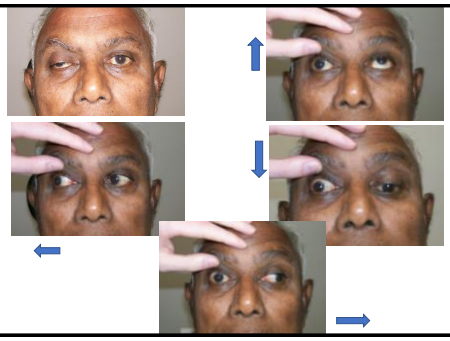
Neuro-ophthalmic Urgencies and Emergencies

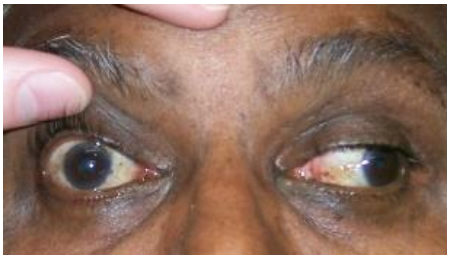
- GCA
  - Any sudden vision loss in the elderly
- Pituitary apoplexy
  - Headache, field loss, diplopia
- Aneurysm
  - Pupils
- Papilledema
  - Clinical suspicion
- Carotid dissection
  - Horner syndrome

63 YOIM

- Long standing glaucoma patient
- Sudden onset of orbital pain x 3 days
- + DM; +HTN
- On coumadin
- Pacemaker
- No vision change
- Presents as walk-in emergency glaucoma eval







5 mm
unresponsive

2 mm
responsive

63 YOIM

- Pupil involved CN III palsy
- 3 days duration at least
- Most likely cause: intracranial aneurysm
- Sent to ED with detailed notes and recommendations
- Endovascular therapy with coils
- Hospitalized 23 days

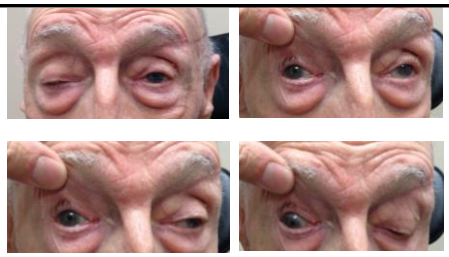
Secondary aberrant regeneration

CN III Palsy Clinical Picture

- An eye that is down and out with a ptosis
- Adduction, elevation, depression deficits
- Isocoric or anisocoric

83 YOM

- Diabetic; LBS in 300s;
- aiC around 11
- Pupils normal MRI ordered through PCP
- Indication for imaging: *Brain ischemia*
- What 2 errors were made here?



The world's best neuroradiologists cannot help you if you don't order the scan, order the correct scan, and them what to look for.

Neuroimaging for the primary care OD

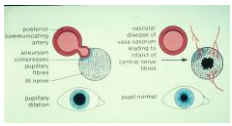
- Disclosure: I do not read MRIs (There are ODs that do- I'm not one of them)
  - What you don't know can hurt you a whole lot
  - That's the reason for residencies in radiology and subspecialties in neuroradiology
    - Thinking that I am as good is irresponsible (e.g. neuroradiologist identifying ciliary body on MRI)
- Rules for ECP: order the correct scan and read the report to ensure that the right thing was done
- If you have questions, doubts, or concerns, reach out to the radiologist
- Form a relationship with an imaging center- find out about the practice
  - Some have better results with MRA and others with CTA

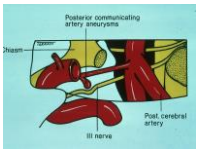
What to order, how, and why

- Disc edema/ suspect papilledema: Brain MRI with and without contrast looking for mass lesion, hydrocephalus, hemorrhage, flattened globe, empty sella; MRV looking for cerebral venous sinus thrombosis.
- Optic nerve/chiasmal disease: MRI orbits and chiasm with and without contrast with fat suppression
  - Snowball in a snowstorm
- Optic neuritis/suspect MS: MRI orbits and chiasm with and without contrast with fat suppression; MRI brain with and without contrast.
- Horner Syndrome: Brain MRI with and without contrast; CTA (or MRA) head and neck looking for cerebral artery dissection; MRI chest with lung apex and brachial plexus
  - Horner protocol or sympathetic plexus
- Suspected aneurysm (CN 3 palsy): CTA/CT and MRA/MRI with concentration to Circle of Willis
  - If high risk aneurysm-send to ER and tell them what to do.
- Don't just send to the ER without helping them. They won't get it right.

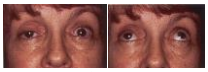
CN III Anatomy

- Vulnerable to compression from aneurysm in subarachnoid space
  - Posterior communicating artery (PCOM)
  - Junction PCOM and ICA
  - Tip of basilar artery





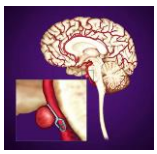
Still More Clues

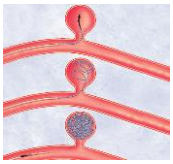


- Pupil involved CN III palsy is PCOM aneurysm until proven otherwise
- Incomplete palsy is PCOM aneurysm until proven otherwise
  - Regardless of pupil
- 30% of CN III palsy are caused by aneurysm
- Pain is pain
  - Only helpful when not present
- Vasculopathic CN III will resolve in time
- Life threatening posterior communicating aneurysm will rupture in time

Still More Clues

- CN III palsy caused by aneurysm
  - 20% die within 48 hrs from rupture
  - 50% overall die
  - Average time from onset to rupture – 29 days
    - 80% rupture w/i 29 days
  - Many never make it to hospital
- Ruptured aneurysms
  - 5% surgical mortality
  - 60% functional impairment post-op
- Unruptured aneurysms
  - No mortality; 75% with normal outcomes; 50% with CN III recovery

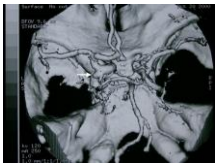


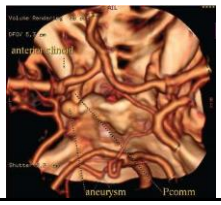


Never dilate a patient with cranial nerve III palsy

Rules for CN III palsy imaging

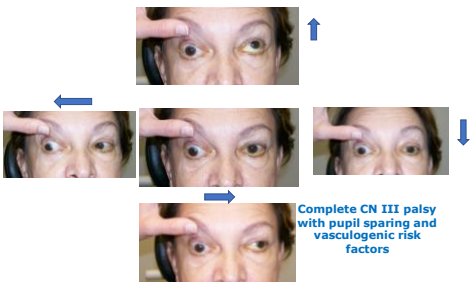
- High suspicion of aneurysm: DSA (gold standard)
- CT/CTA is preferred non-invasive imaging for CN III palsy
  - CT for SAH
- CTA requires contrast- renal impairment prefers MRI/MRA
- CTA superior to MRI when patient can't have MRI
  - Pacemaker, claustrophobia
- MRI superior for non-aneurysmal causes (tumor)
  - MRA adds very little time to scan
- Recent study shows majority of CN 3 palsy patients do not get the appropriate urgent imaging.





A Different patient and Prognosis

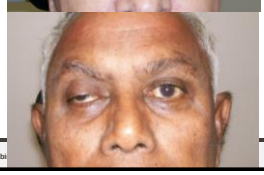
- 63 YOF
- Diabetes and HTN
- Sudden onset retro-orbital pain



Complete CN III palsy with pupil sparing and vasculogenic risk factors

Which is better? One or two?

Resolves over several weeks



Hospitalized 23 days with 2 neurosurgical procedures

Suspect the worst

- Optometrist sees patient with CN III palsy
- Referred to ophthalmologist next day
- Pt dies from SAH before consult

Does presence of vasculopathic risk factors help?

- Arteriosclerotic risk factors in elderly favors microvascular etiology but does not rule out aneurysm
- HTN, DM, atherosclerosis, hyercholesterol all common and don't protect against aneurysm
- Answer: no, but makes me very nervous when NOT present

Does acuteness of presentation help?

- Ans: Yes and No
- Aneurysm expansion usually produces acute manifestations, but chronic and evolving cases well known
- Acute is more worrisome
- Chronic and improving less worrisome but does not rule out aneurysm
- Resolved without recurrence reassuring

Aneurysm Risk Assessment: Isolated CN 3 palsy

• Isolated dilated pupil	none
• Complete CN3-normal pupil	low
• Partial CN3 – normal pupil	high
• Pupil involved CN3	emergency

Never out of the woods

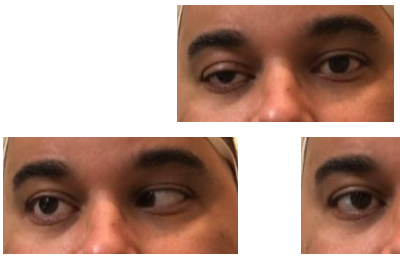
- Pt develops CN III palsy from aneurysm
- Successfully treated with aneurysm clip
  - All coils are inert and MRI safe; not all clips are MRI safe
- Radiologic tech doesn't verify type of clip
- Pt undergoes F/U MRI with non-MRI safe clip in major medical center
- Clip displaces during MRI
- Patient has fatal hemorrhage during procedure
- Patient survived disease…killed by follow up

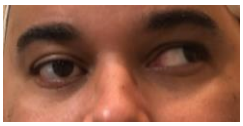
Neuro-ophthalmic Urgencies and Emergencies

- GCA
  - Any sudden vision loss in the elderly
- Pituitary apoplexy
  - Headache, field loss, diplopia
- Aneurysm
  - Pupils
- Papilledema
  - Clinical suspicion
- Carotid dissection
  - Horner syndrome

39 YOM

- Previous history of migraine developed a new and worsening headache.
- He presented to a hospital emergency room where he underwent a non-contrast enhanced computed tomography (CT) and magnetic resonance imaging (MRI) which were subsequently interpreted as normal.
  - His headache was attributed to migraine, and he was medicated as such and discharged.
- Three days later, he developed horizontal and vertical diplopia





39 YOM

- His visual acuity and visual fields were normal.
- He manifested a right pupil-sparing, external partial cranial nerve three palsy and concurrent right sixth nerve palsy. He also complained of worsening headache and lethargy.
- Where is the lesion?
- Let's contact the radiologist for a second reading…

39 YOM

- He was immediately sent for repeat imaging to include contrast-enhanced MRI of the parasellar area and MRA to rule out intracavernous aneurysm and pituitary apoplexy.
- Imaging revealed a pituitary macroadenoma with intratumor hemorrhage consistent with pituitary apoplexy.
- Lateral spread into the right cavernous sinus and possible spread into the left cavernous sinus as well.
- No mass effect on the optic chiasm or prechiasmal intracranial portion of the optic nerve.
  - Hence normal acuity and fields
- The patient was immediately admitted for endocrinological and neurosurgical evaluation

Pituitary apoplexy

- Pituitary apoplexy is a severe and potentially fatal medical condition complicating 2-12% of pituitary adenomas and characterized by the variable association of headache, vomiting, visual impairment, ophthalmoplegia, altered mental state and consciousness, lethargy, and panhypopituitarism.
- Hemodynamic instability may be result from adrenocorticotrophic hormone deficiency, which can be fatal.
- •Occurs due to a rapid expansion, mainly caused by hemorrhage or infarction of a preexisting (known or unknown) adenoma

Pituitary apoplexy

- Most common presenting symptom occurring in 90 % of patients is sudden onset of severe headache
  - Commonly described as frontal or retro-orbital.
    - Pituitary apoplexy is often overlooked as a possible cause of "thunderclap headache" where diagnostic evaluations tend to direct to more common causes of this presentation including subarachnoid hemorrhage, cerebral venous sinus thrombosis, and cervical artery dissection.
- Approximately 50% have visual abnormalities
  - •Blurred vision
- Cranial nerve palsy (CN III) or palsies
  - Cranial nerve VI most common, followed by CN III
- Visual field defects
  - Bitemporal hemianopsia
- Facial weakness

Pituitary apoplexy

- Most symptomatic patients undergo CT scanning in an emergency setting due to the clinical suspicion of acute intracranial hemorrhage
- Acute hemorrhagic infarct may be seen on CT
  - Non-hemorrhagic infarcts will usually show no abnormalities without intravenous contrast
- MRI with contrast is the most effective imaging in cases of suspected pituitary apoplexy
  - MRI is superior to CT

Pituitary apoplexy

- Positive outcome in most cases
  - Conservative medical treatment
  - Stabilize and replace diminished pituitary hormones
- Surgical decompression
  - Trans-sphenoidal or subfrontal transcranial approach
  - Patients with visual impairment and neuro-ophthalmic dysfunction will be selected for surgery.
- Patient was medically stabilized, and surgery delayed due to COVID lock down
- Ultimately underwent successful surgical decompression

Neuro-ophthalmic Urgencies and Emergencies

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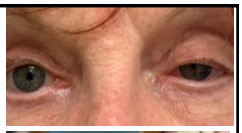
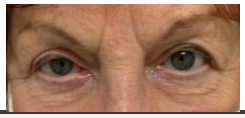
- Papilledema
  - Clinical suspicion
- Carotid dissection
  - Horner syndrome

78 YOF

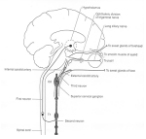
- Sudden onset of ptosis OS
- Immediately following parathyroid surgery
- Headache and eye pain
- Dilation lag and positive lopidine test





What is Horner's Syndrome?

A triad of clinical signs arising from disruption of sympathetic innervation to the eye and ipsilateral face that causes *miosis*, upper lid *ptosis*, mild elevation of the lower lid, and *anhydrosis* of the facial skin.

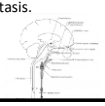


What is the most likely cause?

- Lung cancer
- Carotid dissection
- Direct surgical trauma to the nerve
- Migraine

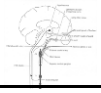
Horner's Syndrome: Etiologies

- First-order neuron disorder: Stroke (e.g., vertebrobasilar artery insufficiency or infarct); tumor; multiple sclerosis (MS), and, rarely, severe osteoarthritis of the neck with bony spurs.
- Second-order neuron disorder: Tumor (e.g., lung carcinoma, metastasis, thyroid adenoma, neurofibroma). Patients with pain in the arm or scapular region should be suspected of having a Pancoast tumor. In children, consider neuroblastoma, lymphoma, or metastasis.



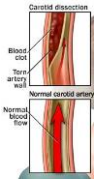
Horner's Syndrome: Etiologies

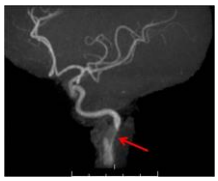
- Third-order neuron disorder: Headache syndrome (e.g., cluster, migraine, Raeder paratrigeminal syndrome), internal carotid dissection, herpes zoster virus, otitis media, Tolosa–Hunt syndrome, neck trauma/tumor/inflammation, prolactinoma.
- Congenital Horner syndrome: Trauma (e.g., during delivery).
  - Heterochromia
- Other rare causes: Cervical paraganglioma, ectopic cervical thymus



Carotid Dissection

- *A 3rd-order Horner's and ipsilateral head, eye, or neck pain of acute onset should be considered diagnostic of internal carotid dissection unless proven otherwise.*





Carotid Dissection

- Carotid artery dissection presents with the sudden or gradual onset of ipsilateral neck or hemicranial pain, including eye or face pain
- Often associated with other neurologic findings including an ipsilateral Horner's syndrome, TIA, stroke, anterior ischemic optic neuropathy, subarachnoid hemorrhage, or lower cranial nerve palsies
  - 52% with ocular or hemispheric stroke with 6 days
    - 67% within first week; 89% within 2 weeks; none after 31 days
- Horner's from suspected carotid dissection should go to ER

Neuro-ophthalmic Urgencies and Emergencies

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- Pituitary apoplexy
  - Headache, field loss, diplopia
- Aneurysm
  - Pupils
- Papilledema
  - Clinical suspicion
- Carotid dissection
  - Horner syndrome

If you listen to patients, they will tell you the diagnosis

73 YOWF

- CC: swollen left eyelid x 3 months
- Not happy with previous doctor
  - *"They aren't listening to me"*
- Highly allergic person- had pain and ear blockage on right side of face while gardening- thinks something got into her eye
- Rx Zylet, Azasite, oral antihistamines, hot and cold compresses- no improvement
- PCP tested for GCA- negative
- Presumed allergic reaction
  - No itching, persistent and unilateral
- Hypothyroid, smoker

Pre-Iopidine Post-Iopidine

Case

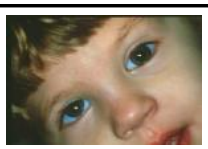
- A 25-year-old woman was involved in a minor automobile accident where she was hit by another driver. The accident was reportedly minor, with no initial injury to either driver, and both cars were able to be driven away. She felt that she experienced only a mild-to-moderate bump during the accident with no head trauma or loss of consciousness. However, immediately upon waking the next morning, though she had no physical pain, she experienced profound double vision.

What is the likely cause?

- A subarachnoid hemorrhage
- A third nerve palsy
- Orbital fracture
- Fourth nerve palsy
- Sixth nerve palsy

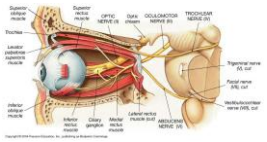
Case

- She described the diplopia as vertical and worse at near. She had a distinct right hyper deviation which, on alternate cover test, worsened in left gaze and right head tilt. This was a signature motility of a cranial IV (trochlear) palsy.



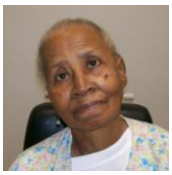
CN IV Anatomy

- Exits the midbrain posteriorly and decussates
- Longest course
- Travels around tentorium, through cavernous sinus, through SOF
- Most prone to trauma



CN IV Palsy

- Longstanding CN IV palsy may present with diplopia from decompensation
  - Observe old photos for head tilt (*Facebook Tomography*)
- Rule of 40-30-20-10
  - 40% traumatic
  - 30% idiopathic
  - 20% vascular
  - 10% CNS lesion



CN IV Management

- Isolated, non-traumatic:
  - Evaluate for ischemic diseases
  - Non-ischemic causes of non-traumatic, isolated CN IV palsy rare
  - Look for longstanding decompensation
    - Increased vertical vergences
    - Old photos

73 YOM

- New onset vertical diplopia
  - Left 4th nerve palsy
  - Relieved by 2 PD BD
- *"Doc, I also noticed that my gripper is off"*
  - Mild left-handed weakness
- Medical history: Treated lung cancer
  - Currently on maintenance chemotherapy
- Approach and outcome?

Never diagnose idiopathic (or ischemic) anything in a patient with a history of cancer

77 YOM

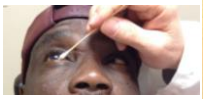
- Pt and sister presents insistent on cataract surgery.
- BVA 20/40 OD, 20/70 OS with commensurate NS
- Chronic horizontal diplopia that has recently gotten worse (2 weeks)
  - Left abduction deficit ~ 40%
- Medical history: Inoperable chondrosarcoma with lysis of clivus extending to left petrous apex and occipital condyle with sphenoidal, ethmoidal and temporal bone involvement. Compression of jugular vein. Has undergone ~ 50 radiation treatments.
  - Vocal paralysis
  - Cranial nerve IX, X, XII palsies
- Imaging obtained: CT with contrast (pacemaker precludes MRI)-new soft mass in left nasopharynx
  - Likely squamous cell carcinoma

CN VI Palsy

- Hallmark sign is horizontal diplopia, greater at distance, with an abduction deficit

CN VI Palsy

- Check motilities at distance
- Forced duction testing



CN VI Management

- Each case of CN VI palsy should be classified as traumatic or non-traumatic.
- Non-traumatic cases should be subdivided as neurologically isolated (just CN VI palsy) or non-neurologically isolated (something else).
- Additionally, patients should be ascribed to one of 3 groups: children, young adults, and older adults

CN VI Demographic Groups

- Older adults (usually not bad)
  - Vascular disease common- resolves-3mos
    - Consider GCA over 60 yrs
- Children (may be bad)
  - Presumed viral illness, trauma, malignancy (50%)
- Young adults (usually bad)
  - Vascular disease (4%) and idiopathic (13%) uncommon
  - Usually complicated CN VI palsy (hemiparesis, Horner syndrome, facial paresis)
    - Cerebrovascular accidents involving the pons, aneurysm (typically within the cavernous sinus) or neoplasm (33%-cavernous sinus, pons), MS (24%).

CN VI Palsy in Older Adults

- In cases of isolated CN VI palsy in older adults with a history of diabetes or hypertension, neuroimaging and other extensive evaluation can be deferred, unless the palsy progresses, fails to improve over 3 months, or other neurologic complications develop.
- Ischemic vascular palsies typically progress over several days, but progression over two weeks warrants neuroimaging.

When looking for mass cause of CN VI palsy, the base of the pons and cavernous sinus are two common hiding spots

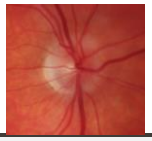
If you are watching a presumptive ischemic CN VI palsy and you are wrong, you likely have not hurt the patient.

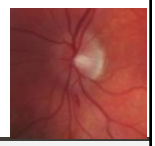
33 YOWM

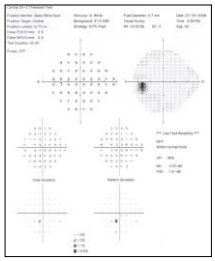
- Occipital HA x 4 mos
  - Visual aura with HA
- Worsens when standing after sitting
- Relieved by sleep
- Denies vision loss, nausea, diplopia, pain on eye movement, behavioral changes

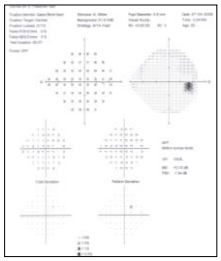
33 YOWM

- 20/20 OD, OS with myopic correction
- Pupils, EOMs, conf fields normal OU
- Biomicroscopy normal OU
- IOP 12 mm Hg OU
- Nasally obliquely inserted nerves









33 YOWM

- Co-manage with PCP- internist
- Complete blood work blood work up including FTA-ABS/RPR ; Lyme titer; CBC w/differential
- MRI w and w/o contrast of brain and orbits
  - Pt had MRI done and mass was identified in fronto/parietal region more toward right side
  - Outcome?

A normal visual field does not mean that there isn't anything wrong

78 YOM

- Acute onset diplopia, blurred vision, and dilated pupils
  - Went to ED-worked up for stroke
    - CT/CTA MRI/MRI all normal
    - Reviewed reports-everything in order
- Referred by colleague after exam
  - Vision improving, pupils less dilated, endpoint nystagmus, non-specific horizontal diplopia
- Pt on anti-muscarinic for bladder
- Mydriasis and blurred vision = ??
- What about that history?
- Outcome?

Take care with *VESI*care

- 56 YOF
- Treated glaucoma
- IOP suddenly in mid 30s with meds
- Bilateral blurred vision and dilated pupils

Case of the curbside consult

- Conference focusing on neurological issues
- 48 YOF-wife of society president
- Complaints of memory loss and loss of smell and taste (Pre-COVID)
  - Fearful of neurodegenerative condition.
- Hx of tonsillectomy two months earlier
- Facial and persistent jaw pain
- Seen by multiple specialists (neuro, ENT, PCP)
- Put on multiple medications including at least 2 oral antibiotics
- *Personal experience with Mucinex*
- Outcome?

Its not always a brain tumor. Think about medication toxicity

Toxicities

- Blurred vision and dilated pupils
- Ethambutol
  - Toxic optic neuropathy
- Amiodarone/ Pacerone
  - Toxic optic neuropathy
- Vigabatrin
  - Anticonvulsant for refractory focal epilepsy in children 2 years of age or older
  - May cause permanent, concentric peripheral visual field loss, thought to be secondary to drug-induced injury to both the retinal photoreceptors and the retinal ganglion cells and their axons.

Immediately referring to the Emergency Department is acceptable management…if you are willing to help

Best handled in the ED*

- Suspected GCA
- Suspected aneurysm
- Suspected papilledema
- Suspected pituitary apoplexy
- Suspected carotid dissection
- CRAO/ BRAO/ TIA

*As long as you are willing to help

Stay Safe Everyone