

MISTAKES YOU CAN'T MAKE IN NEURO-OPHTHALMIC DISEASE

Joseph Sowka, OD, FAAO,
Diplomate

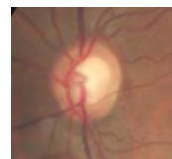
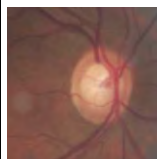
MISTAKES YOU CAN'T MAKE

- Not recognizing a neurologic field
- Thinking glaucoma causes optic disc pallor
- Diagnosing Horner's syndrome...and stopping there
- Not properly assessing a diplopic patient for an aneurysm
- Not properly assessing a patient for giant cell arteritis
- Continuing to treat the chronically red eye

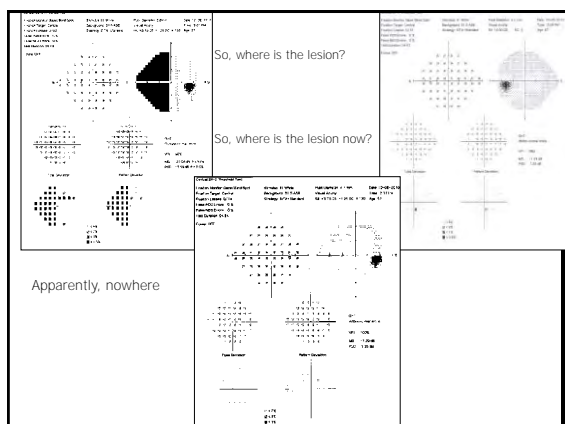
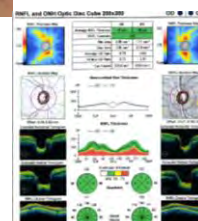
MISTAKE NOT TO MAKE

- Not recognizing a neurologic field

57 YOF- LOW RISK OHTN OU

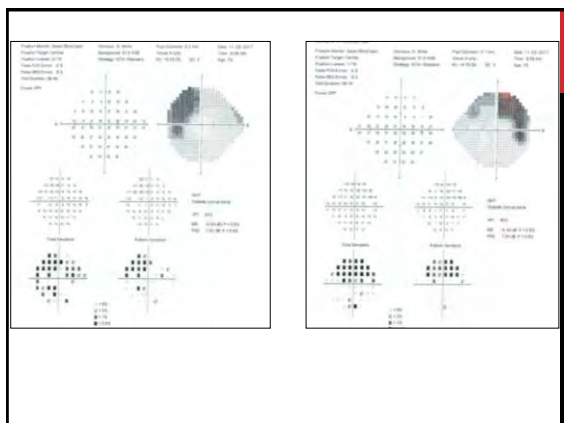
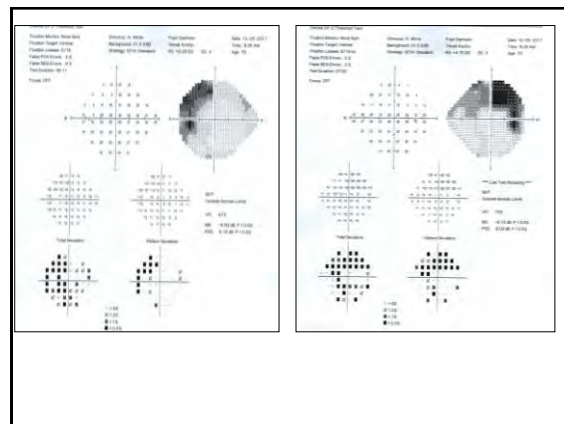
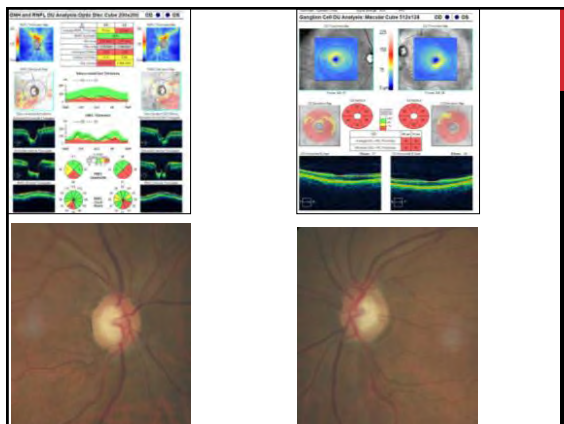


- 6/6/OD, OS
- PERRL(-)RAPD
- Color vision normal OD, OD



74 YOF

- Diagnosed with glaucoma in Jamaica
- Ran out of meds: IOP 20 mm OU
- 6/15 OD, 6/12 OS
- NS 2+
- PERRL(-)RAPD



FINDINGS: There is a large T1 hypointense and T2 iso- to hyperintense lesion extending between the sella into the suprasellar region showing heterogeneous enhancement on the post-contrast images measuring 2.7 cm craniocaudal x 2.1 cm AP x 2 cm transverse. Findings are compatible with a pituitary macroadenoma. It is resulting in compression of the optic chiasm and slightly compressing upon the hippocampus. There is preservation of the signal void of the cavernous carotids. There is possible extension into the cavernous sinus medially. There is slanting of the floor of the sella.

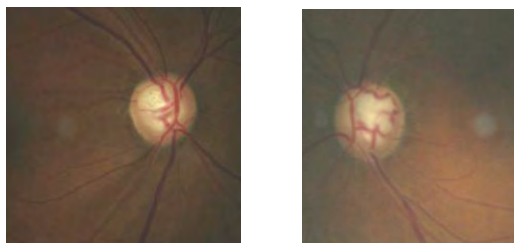
The ventricles are in midline. There are multiple bilateral periventricular and subcortical T2 hyperintensities most commonly representing chronic small vessel ischemia in this age group.

The globes are symmetric. There is no lens dislocation. The post-septal soft tissues are preserved with no definite intra- or extraconal mass. The optic nerves are symmetric at the orbital level showing no abnormal enhancement.

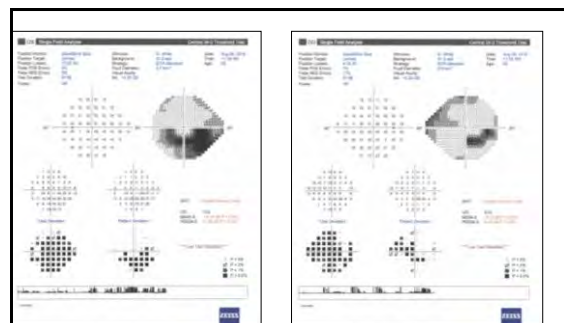
IMPRESSION:

1. Large heterogeneous enhancing sella/suprasellar mass resulting in compression of the optic chiasm compatible with a pituitary macroadenoma.
2. Bilateral periventricular and subcortical T2 hyperintensities compatible with chronic small vessel ischemia.

65 YOF- POAG OU; 20/40 OU



Peak IOP unknown; s/p SLT OU and on latanoprost at first visit.



Oh, by the way, she remembered waking up 10 years ago unable to speak for several hours.

MISTAKE NOT TO MAKE

- Thinking glaucoma causes optic disc pallor

RULE

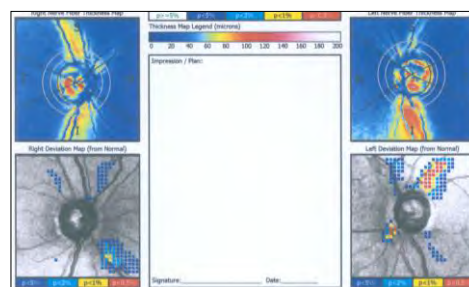
Pallor in excess of cupping indicates something other than, or in addition to, glaucoma

RULE

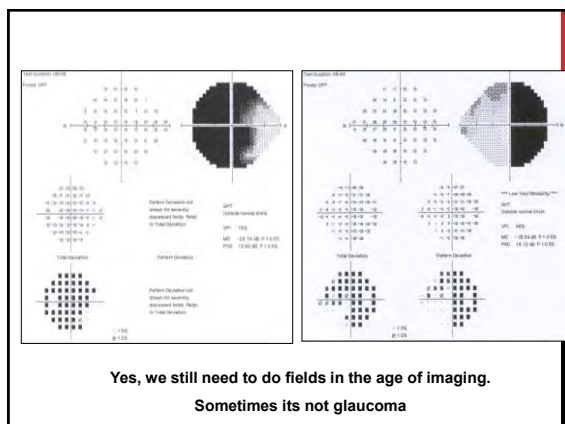
Nothing notches a nerve like glaucoma

IN THE AGE OF IMAGING, DO WE REALLY NEED FIELDS?

- 54 YO Nigerian man
- Referred for glaucoma management
- Told he had glaucoma 6 years earlier- no Tx
- 20/30 OD; HM OS
 - Vision loss from glaucoma- not coming back
- 30 mm Hg OD; 23 mm Hg OS
 - Lumigan- 17 mm Hg OD, 15 mm Hg OS



Do we really need fields in this case?



ODE TO A CUPPED DISC

Oh, to have a cupped disc pink.
That my friend hath a glaucomatous stink.
But to have a cupped disc pale,
Call this glaucoma and you shall fail.
Disc and field damage that is one-sided
Simply cannot be abided.
It might be trauma, infarct or meningioma.
But if the rim is cut always remember,
Nothing notches a nerve like glaucoma

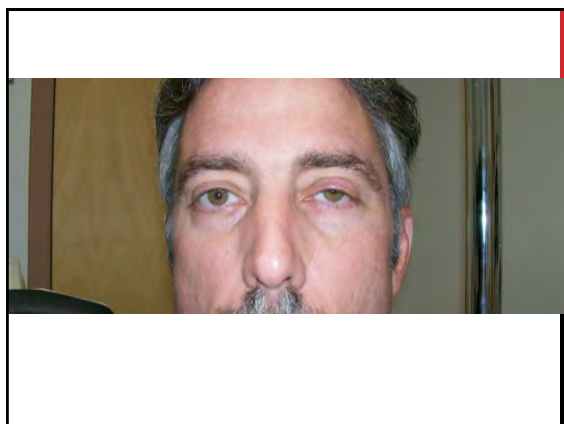
Joseph Sowka, OD

MISTAKE NOT TO MAKE

- Diagnosing Horner's syndrome...and stopping there

46 YOM

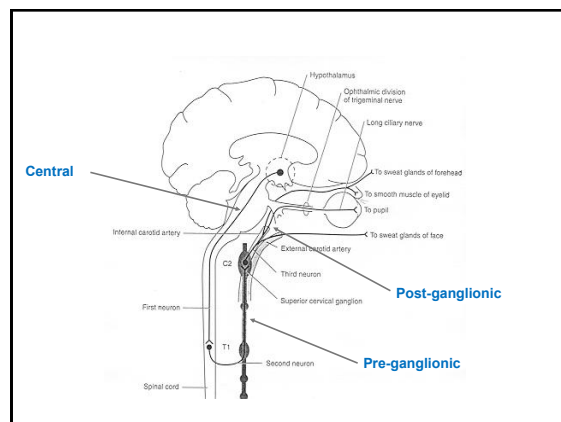
- Patient reports a "droopy left eye" which began about 6 weeks ago. Headache and numbness ipsilateral; hives
 - ER diagnosed with "stye"- treated with topical antibiotic. Patient was referred in by a local optometrist.
- **Observation:** LUL ptosis, Left miosis; anisocoria greater in dark than light
- **All other findings normal**
- **Presumptive Horner syndrome**
 - Confirm with apraclonidine (lopidine)



DISCUSSION

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 *anhidrosis* of the facial skin.



HORNER'S SYNDROME: ETIOLOGIES

Central neuron disorder: Stroke (e.g., vertebrobasilar artery insufficiency or infarct); tumor; multiple sclerosis (MS), and, rarely, severe osteoarthritis of the neck with bony spurs.

Pre-ganglionic 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

- **Post-ganglionic 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).
- **Other rare causes:** Cervical paraganglioma, ectopic cervical thymus
- Remember: Bad stuff happens everywhere

MANAGEMENT

- Neck and facial pain- carotid dissection
- **Necessary Work Up:**
 - MRI of brain, orbits and chiasm with and without contrast, attention to middle cranial fossa.
 - MRA of head and neck-rule out carotid dissection
 - MRI of neck and cervical spine, include lung apex and brachial plexus
 - Horner's syndrome patient needs to be imaged from chest to head- 3 scans
 - Horner's protocol

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 ED

RULE

Diagnosing Horner's syndrome is insufficient. You must try to ascertain a cause and never assume that it is benign.

ODE TO HORNER'S SYNDROME

When the lid is low and the pupil small,
Check to see the sweat don't fall.
Cocaine is no longer universal,
Iopidine will cause reversal.
You have to scan head to chest,
And remember that MRA is best.
Pain in association, will surely cause commotion.
Send to the ER without correction,
Remember, it might be carotid dissection.

Joseph Sowka, OD

MISTAKE NOT TO MAKE

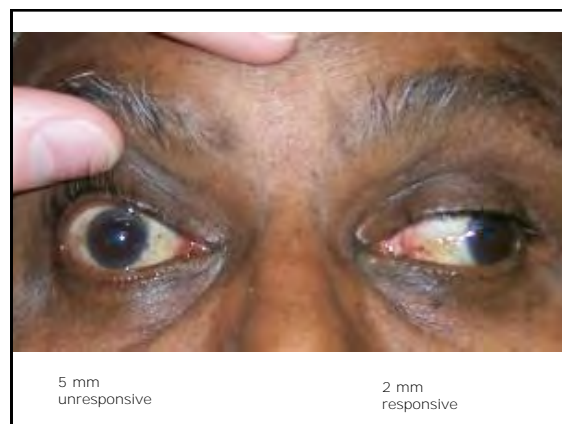
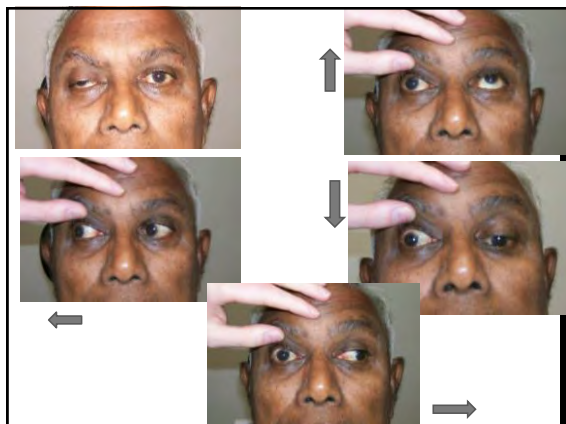
- Not properly assessing a diplopic patient for an aneurysm



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



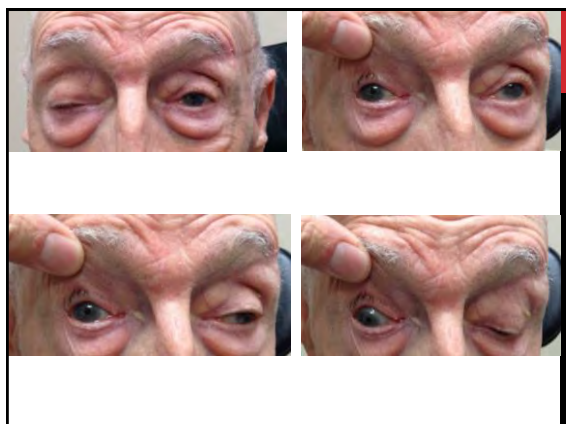
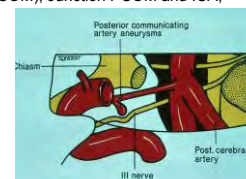
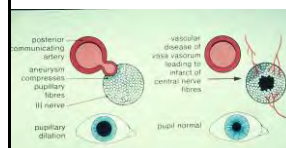


63 YOIM

- 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
- He did survive, however

CN III PALSY CLINICAL PICTURE

- An eye that is down and out with a ptosis
- Adduction, elevation, depression deficits
- Isocoric or anisocoric
- 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- likely ballooning aneurysm developing
- **30% of CN III palsy are caused by aneurysm**
- Pain is pain
 - Only helpful when not present
 - Aneurysms always painful
 - Ischemic palsies painful 90% of the time



STILL MORE CLUES

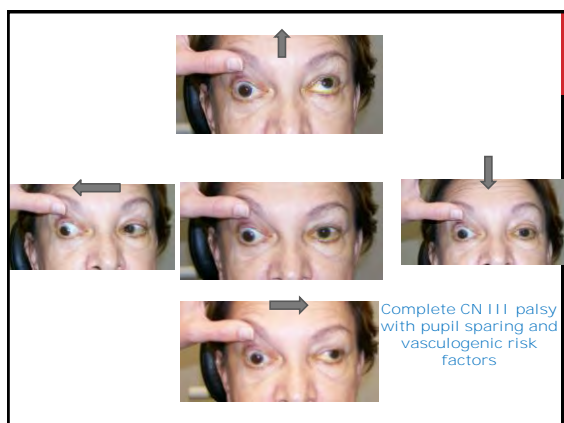
■ CN III palsy caused by aneurysm

- 20% die within 48 hrs from rupture
- 50% overall die
- Average time from onset to rupture – 29 days
 - 80% rupture w/in 29 days
- Many never make it to hospital



A DIFFERENT PATIENT AND PROGNOSIS

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



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, hypercholesterol 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

ODE TO A THIRD NERVE

When the eye is down and out with ptosis,
You better hope for miosis.
If the palsy is total with pupil sparing,
In an Oldie it's vascular and not too daring.
A partial palsy calls for double duty,
Because it's probably an aneurysm going through puberty.
But if the pupil is dilated,
An aneurysm has violated.
No time for deferral and no time for referral.
Send to the ER without debate.
Remember, twenty percent will die within the first forty-eight

Joseph Sowka, OD

MISTAKE NOT TO MAKE

- Not properly assessing a patient for giant cell arteritis

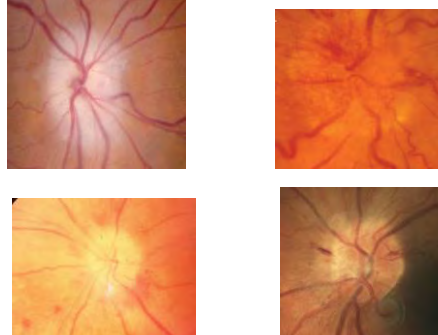
74 YOM

- Presents with 'worst headache of his life'
 - Sees: PA, ED physician; cardiologist; NP;
 - 3 week period
 - Histories: Eye ache; jaw pain, scalp pain, facial pain, somnolence; malaise; jaw claudication
 - Diagnoses: TMJ; Lyme disease
 - "vasculitis such as temporal arteritis highly unlikely", "Not GCA"
 - However, ESR and CRP ordered and elevated- never reviewed
 - Ultimately OD makes diagnosis
 - End result?
 - Bilateral blindness

ANTERIOR ISCHEMIC OPTIC NEUROPATHY

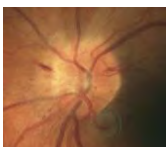
- Hypoperfusion of the posterior ciliary arterial supply to the anterior optic nerve head.
- May be arteritic (AAION) or non-arteritic (NAAION)
- Mechanical factors and atherosclerotic disease play a role in the non-arteritic form while vasculitis contributes in the arteritic form.
- Unilateral presentation 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
- 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
 - Erythrocyte sedimentation rate
 - Lowered by statins and NSAIDS
 - C-reactive protein
 - Not affected by statins and NSAIDS
 - Elevated platelet count
- 65% incidence of bilateral progression; avg 10 days
- Emergency- must get high dose steroid immediately

ODE TO AN ISCHEMIC NERVE

When your patient's optic nerve is ischemic
 You better hope the disc is hyperemic.
 In Non-arteritic no treatment is needed
 And life will rarely be impeded.
 But if the disc is swollen and pale,
 And vision is an epic fail
 If the patient is sixties, seventies or eighties
 You will feel heat like in Hades
 ESR and CRP are required
 And steroids must be acquired
 Remember, when you see a choked disc
 Always assess the giant cell risk

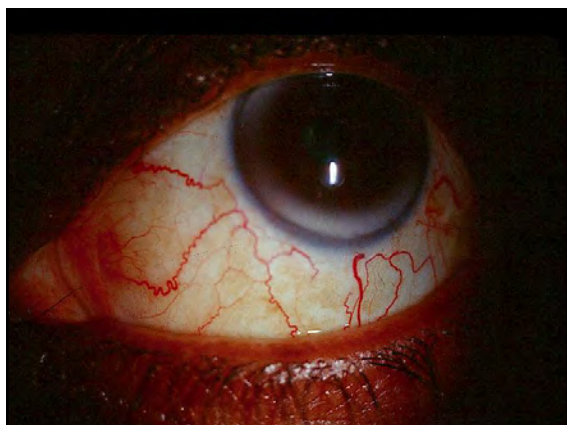
Joseph Sowka, OD

MISTAKE NOT TO MAKE

- Continuing to treat the chronically red eye

47 YOF

- CC:** Horizontal double vision in far left gaze
- BVA:** 6/6 OD, OS
- Medical Hx:** diabetes; chronic red eye
 - Brings in 2 antibiotics, 3 antihistamines, 2 steroids, 2 antibiotic/steroid combos, 4 ATs
- Left abduction deficit in far left gaze**
 - Negative forced duction test
 - Presumptive vascular CN VI palsy?



47 YEAR OLD BLACK FEMALE

CT scan:

*What do
you think
NOW?*



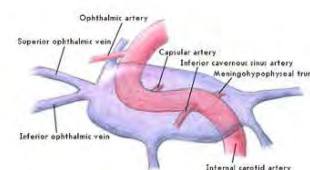
CAROTID CAVERNOUS SINUS FISTULA

Cavernous sinus. . .

- Trabeculated venous cavern
- Houses CN III, IV, VI, V1, oculosympathetics, and ICA
- Drains eye and Adnexa via inferior and superior ophthalmic veins to petrosal sinuses and jugular vein

Fistula. . .

- Rupture of ICA or meningeal branches within sinus
 - Meningohypophyseal, McConnell's Capsular, Inferior Cavernous
- Mixing of arterial blood in venous system



CAROTID CAVERNOUS SINUS FISTULA

Hemodynamic

- High flow vs low flow

Angiographic

- ICA vs meningeal branches

Etiology

- spontaneous vs traumatic



RULE: BEWARE THE CHRONIC RED EYE

- Dilated & tortuous episcleral vessels that go to the limbus and back (omega loops) Ω
- Intervening "clear conjunctiva"
- Red eye that doesn't respond to any topical treatments
 - Bag-o-Meds
- Other non-red eye findings: Chemosis, IOP elevation, proptosis, ophthalmoplegia, ptosis, lid edema

ODE TO A FISTULA

Beware the chronic red eye
 It isn't infected, inflamed, or dry.
 When corkscrew vessels makes the eye red
 And the patient has bag-o-med.
 The problem is deep
 And arterial blood has begun to seep.
 Your first fistula you will always miss
 But on your second case you will never be remiss

Joseph Sowka, OD