Finding the wolf in your visual field testing

Andrew G. Lee, MD
Houston, Texas

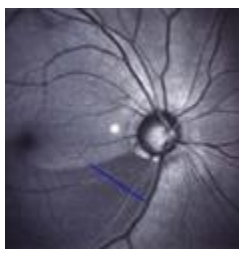
The wolf is hiding in sheep's clothing

Five questions to find the wolf

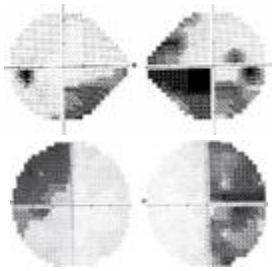
1. Respecting vertical or horizontal meridian?
2. Worse in the temporal or nasal field?
3. Junctional, bitemporal, or homonymous?
4. Lateral geniculate body (wedge sectoranopia)?
5. Optic tract: homonymous, RAPD + band atrophy?

The topography

Nerve fiber layer defects

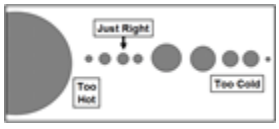


Who has a tumor and who has glaucoma?

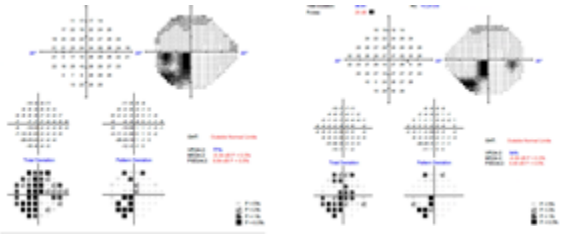


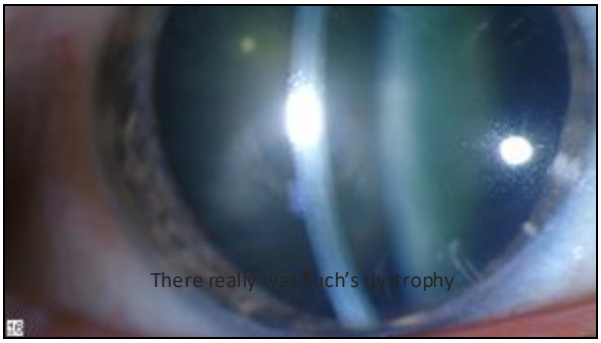
Even my friend Bob, the security guard at Houston Methodist Hospital knows that this is not glaucoma

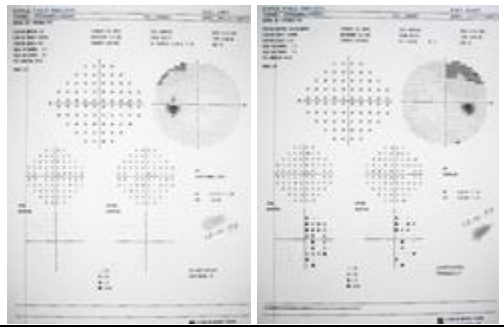
Remember
Goldilock's principle

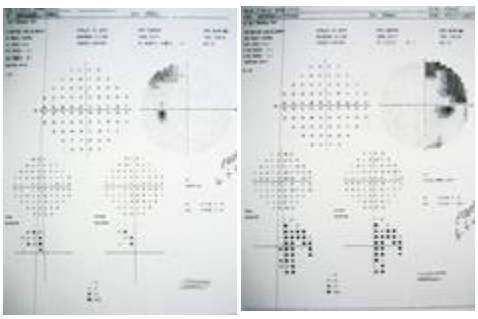


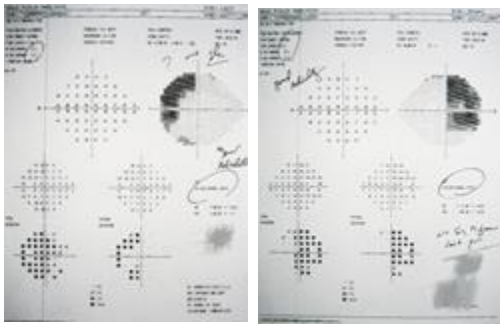
Could this be glaucoma? Too vertical




There really was Fuch's dystrophy
18

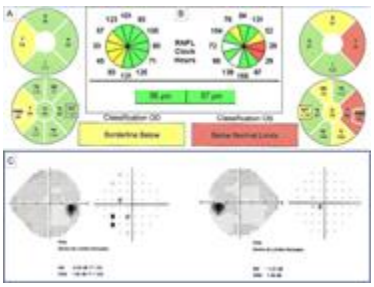




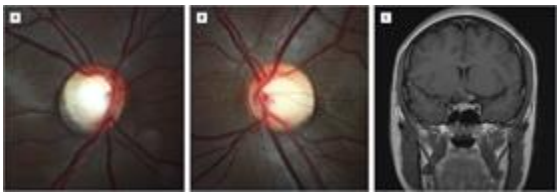


Too vertical
Too fast
Too bad

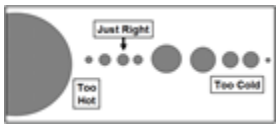
RNFL loss: Too horizontal (papillomacular bundle)

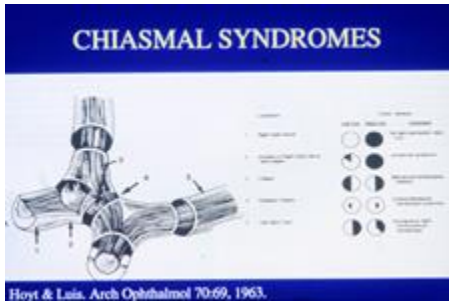


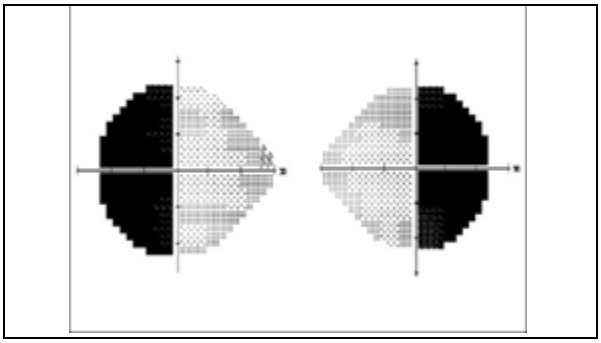
Band cupping, rim pallor

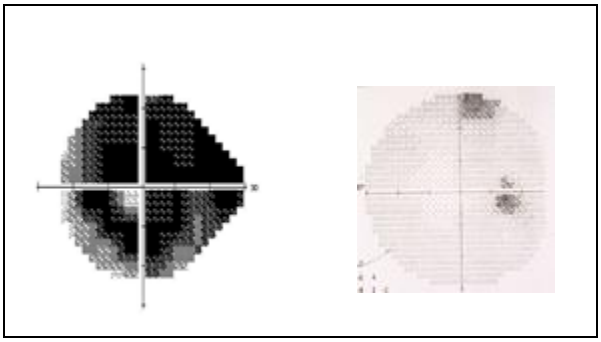


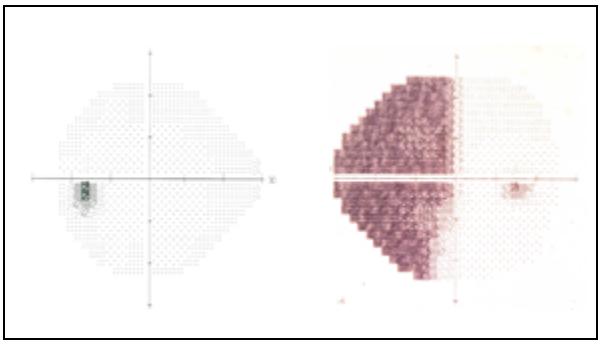
Remember
Goldilock's principle

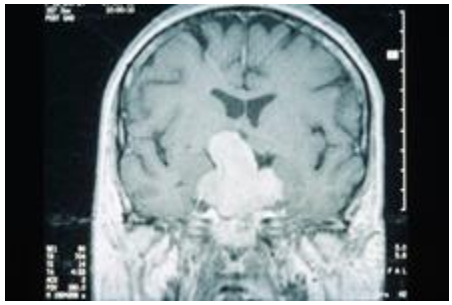


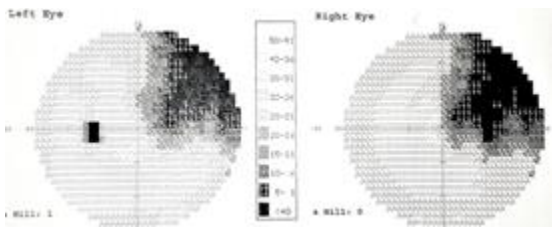


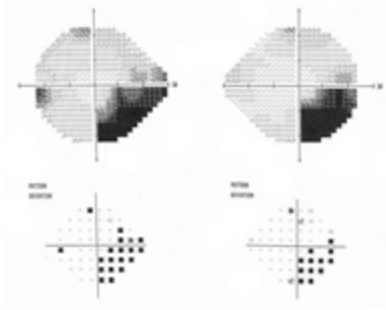


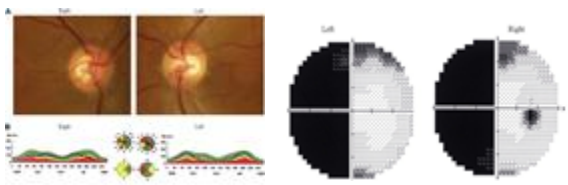


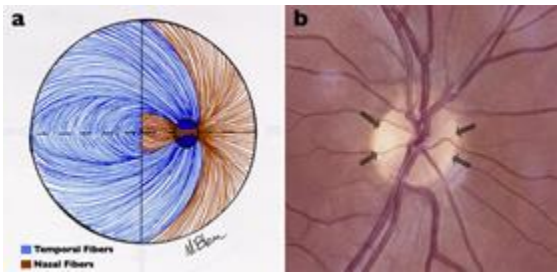




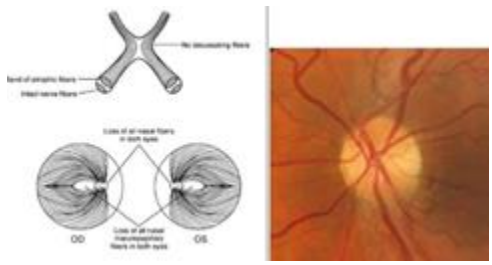

Left Eye
Right Eye

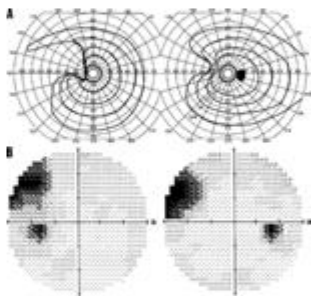


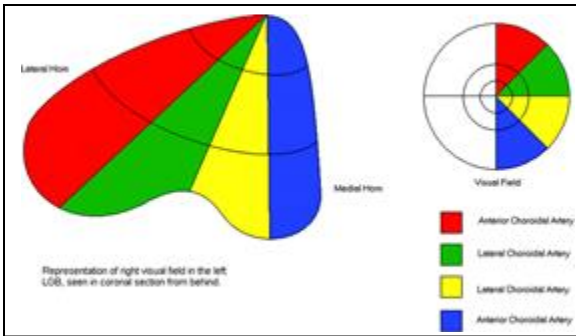


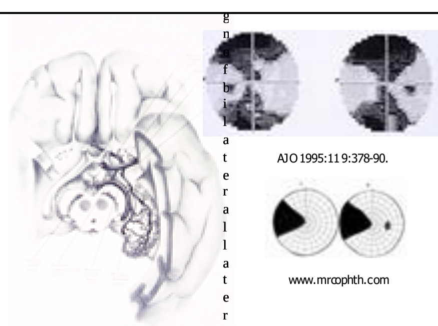


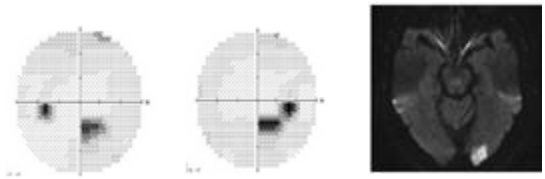
Band atrophy of optic nerve

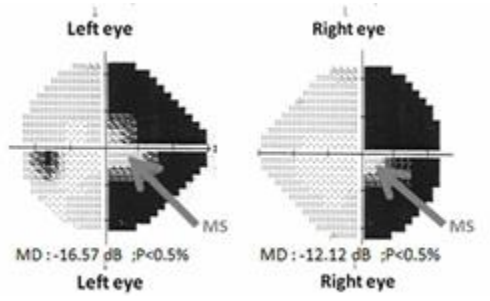


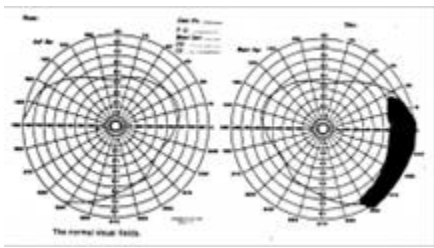

A
B

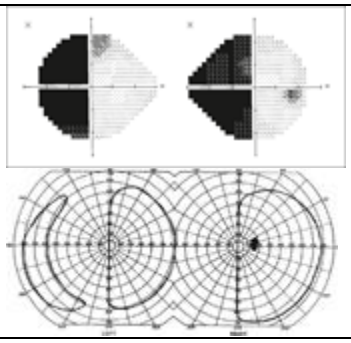

Lateral Horn
Medial Horn
Visual Field
Anterior Choroidal Artery
Lateral Choroidal Artery
Lateral Choroidal Artery
Anterior Choroidal Artery
Representation of right visual field in the left LGB, seen in coronal section from behind.


AJO 1995:119:378-90.
www.mrcophth.com

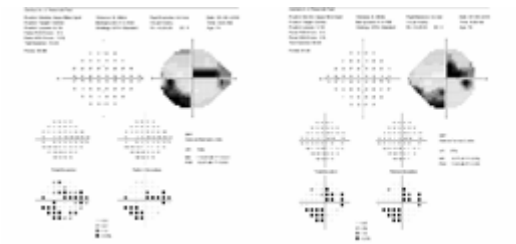


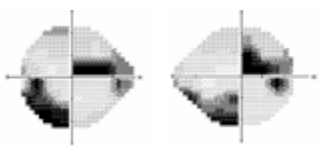

Left eye
Right eye
MS
MS
MD : -16.57 dB ;P<0.5%
MD : -12.12 dB ;P<0.5%
Left eye
Right eye





Beware juxtaposed homonymous VF loss





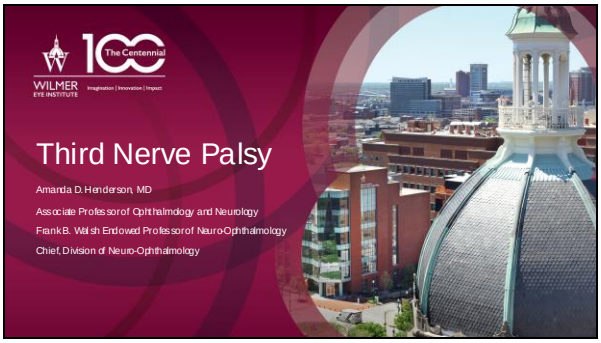
Summary: To find the wolf in VF testing

1. Is is vertical (wolf!) or horizontal meridian?
2. Worse in the temporal (wolfie) or nasal field?
3. Junctional, bitemporal, or homonymous (all wolves)?
4. Lateral geniculate body (wedge sectoranopia): wolfman?
5. Optic tract: homonymous, RAPD + band atrophy? (werewolf)

Thanks for your time & attention

Andrew G. Lee, MD
Houston, Texas

Houston Methodist Texas Medical Center · utmb Health · BCM Baylor College of Medicine · Texas A&M Health Science Center College of Medicine · Weill Cornell Medical College · MD Anderson Cancer Center · University of Iowa Health Care



Disclosures

Horizon Therapeutics/Amgen (Advisory Boards)

Catalyst Pharmaceuticals (Advisory Board)

Argenx (Clinical Trial Site)

Objectives

By the end of this presentation, participants will be able to:

a. Recognize variable presentations of third nerve palsy

b. Identify need for urgent neuroimaging in patients with third nerve palsy

Case 1: 67-year-old man with diplopia

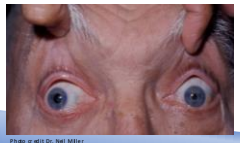


Photo credit Dr. Neil Miller

Past medical history

- +hypertension, controlled on meds
- No diabetes, no cancers, no recent trauma

Approach to Diplopia

History

- Binocular vs monocular?
  - Binocular
- Sudden vs insidious onset?
  - Sudden diplopia yesterday
- Associated symptoms?
  - Lid drooped overnight

Examination

- Motility
  - See next slide
- Check of cranial nerves II-VII
  - Visual acuity OD 20/20, OS 20/20
  - No RAPD
- Other examination findings
  - Pupils
    - OD 6mm → 5.5mm
    - OS 4mm → 2mm

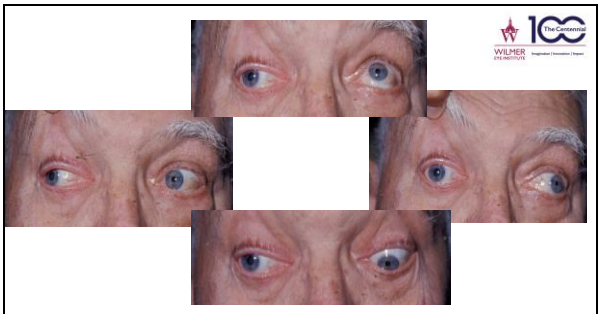
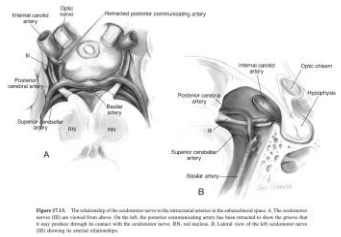


Photo credit Dr. Neil Miller

Localization

- Extraocular muscle(s)
- Neuromuscular junction
- Cranial nerve
  - Orbital apex, cavernous sinus, cisternal space
- Internuclear
- Cranial nerve nucleus

Third nerve anatomy



Walsh & Hoyt's Clinical Neuro-Ophthalmology, 6th Ed. https://collections.lib.utah.edu/ark:/87278/s6rj4hsw/190051

Imaging in Third Nerve Palsy

- Emergent angiography (CTA, MRA) is required in ALL cases of:
  - Pupil-involving third nerve palsy
  - Incomplete third nerve palsy
  - Aberrant regeneration is a red flag for compression!
  - Angiography should be strongly considered in pupil-sparing complete third nerve palsy
- MRI with and without contrast is also indicated in these cases and can be performed concomitantly with the angiography in many cases (ie, MRI/MRA)

Case 1 follow up

- CTA demonstrated right posterior communicating artery aneurysm
- He was admitted, and aneurysm was treated by coiling

Case 2:
55-year-old woman with 3.5 months of droopy left eyelid and diplopia

Past Medical History: Diabetes, Hypertension

MRI/A at Outside ED: negative by report

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Approach to Diplopia

History

- Binocular vs monocular?
  - Binocular
- Sudden vs insidious onset?
  - Sudden onset 3.5 months prior
- Associated symptoms?
  - Left sided headache/discomfort

Examination

- Motility
  - Limitation of adduction, elevation, and depression of OD
  - Elevation of left eyelid with attempted downgaze and attempted adduction
- Check of cranial nerves II-VII
  - Acuity 20/20 OD, 20/60 OS
  - No RAPD
  - Other cranial nerves intact
- Other pertinent examination findings
  - Pupils
    - OD 4mm → 3mm, brisk reaction to light
    - OS 5mm, fixed, no direct or consensual reaction

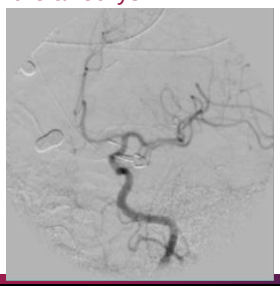
Localization

- Extraocular muscle(s)
- Neuromuscular junction
- Cranial nerve
  - Orbital apex, cavernous sinus, cisternal space
- Internuclear
- Cranial nerve nucleus

Third nerve palsy with aberrant regeneration = **COMPRESSIVE LESION**

Urgent CTA

Clipping of the aneurysm



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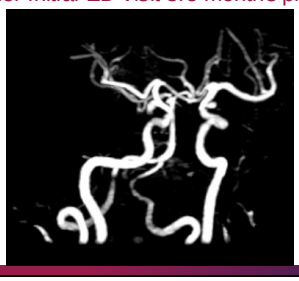
MRA (from her initial ED visit 3.5 months prior)



R

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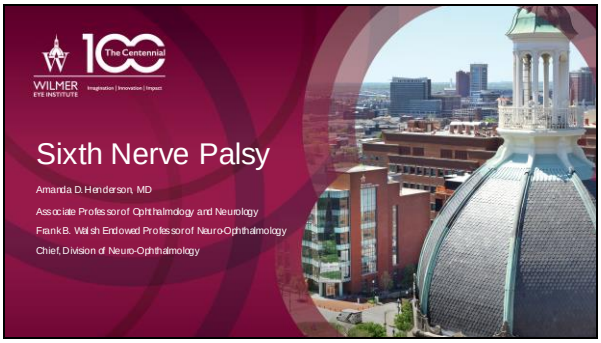
Take Home Points

- Third nerve palsies require urgent neuroimaging, due to concern for potentially life-threatening cause (aneurysm!)
- CTA or MRA can be used to evaluate for aneurysm
- Providing clinical history to your neuroradiologist is key, to prevent "missed" aneurysms



Disclosures

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Catalyst Pharmaceuticals (Advisory Board)

Argenx (Clinical Trial Site)

Objectives

By the end of this talk, participants will be able to:

a. Develop an appropriate evaluation plan for patients presenting with sixth nerve palsy

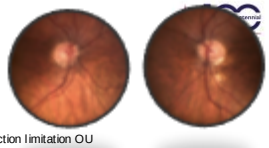
b. Gauge need for and urgency of neuroimaging in patients with sixth nerve palsy

Case 1: 64-year-old man presents with one week of binocular, horizontal diplopia

Past medical history: HIV, CD4 247; Stage 4 adenocarcinoma of the lung with metastases to spine and acetabulum, s/p resection and radiation, currently on Osimertinib; PE, on rivaroxaban; Diabetes, type 2; Hypertension; Hyperlipidemia

History

- Binocular vs monocular?
  - Binocular
- Sudden vs insidious onset?
  - Sudden onset 1 week
- Associated symptoms?
  - None, feels very well



Examination

- Motility
  - Marked abduction limitation OU
- Check of cranial nerves II-VII
  - Acuity 20/20 OD, 20/30 OS
  - No RAPD
  - Other cranial nerves intact
- Other pertinent examination findings
  - 1+ NS OU
  - No papilledema

Case 2

A 52-year-old woman with no significant past medical history presents with two weeks of binocular diplopia. One week prior to the onset of her symptoms, she was kicked in the face by her eight-month-old granddaughter. She had a CT head without contrast that was read as normal two days after the onset of her symptoms.

Exam

Visual acuity 20/20 OD and OS

- Pupils brisk, no RAPD
- Motility shows limitation of abduction of her right eye
  - 6△ esotropia in primary gaze worsening to 18△ in right gaze
- Ophthalmologic and cranial nerve examinations otherwise unremarkable

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Management Questions

- Is neuroimaging required?
- If so:
  - What neuroimaging is required?
  - What is the urgency of neuroimaging?
- Is any additional work up indicated?

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Is Neuroimaging Required?

Whether all patients with acute, isolated sixth nerve palsies require neuroimaging is controversial

- Some recommend imaging all acute, isolated sixth nerve palsies

  (Bendszus, et al., *Neuroradiology* 2001; Chou, et al., *J Neurol Sci* 2004)
- Others argue that non-traumatic, isolated sixth nerve palsy without any "red flags" may be amenable to a more limited work up and close monitoring

  (Richards, et al., *Am J Ophthalmol* 1992; Patel, et al., *Ophthalmol* 2004; Nair, et al., *Indian J Ophthalmol* 2014;
  Miller, et al., *Med Decis Making* 1999)

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Recommended Indications for Neuroimaging

- Age younger than 50 years
- No known microvascular risk factors
- Any history of cancer, whether active or remote
- History of pituitary adenoma
- Recent history of trauma
- Prior history of cranial nerve palsy
- Non-isolated sixth nerve palsy
- Lack of resolution within 3 months

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MRI of the brain and orbits with gadolinium

- Fat saturation allows for improved visualization of orbital pathology
  (Sima, et al., *Indian J Ophthalmol* 2012)
- High-resolution 3D skull base imaging improve sixth nerve visualization
  - Cisternal segment visualized in 98% vs 13% with conventional MRI (Yousry, et al., *Eur Radiol* 2000)
  - Cavernous segment visualized in 95% vs 65% with conventional MRI (Yagi, et al., *Am J Neuroradiol* 2005)

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Urgency?

- Typically, outpatient neuroimaging is appropriate
- Certain clinical scenarios may require emergent MRI
  - Concern for increased ICP, pituitary apoplexy, traumatic hemorrhage, meningitis, etc

Further Evaluation

- Serum
  - GCA rule out
    - ESR, CRP, CBC +/- TAB
  - Mimicker rule out
    - TSI (Graves), AChR abs (MG)
  - Infectious studies
    - Lyme, syphilis, TB, viral titers
  - Inflammatory studies
    - ANA, ANCA, IgG4
- CSF studies
  - Glucose, protein, cell count/diff
  - Consider cultures, viral, and Lyme titers
  - Consider flow cytometry, cytopathology
- Additional imaging
  - CT chest +/- PET/CT if sarcoid suspected

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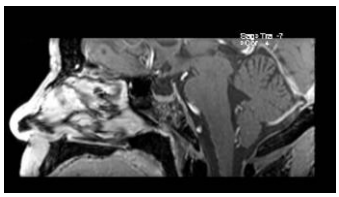
Patient 1 Follow Up

Indications for neuroimaging:

- Age younger than 50 years
- No known microvascular risk factors
- **Any history of cancer, whether active or remote**
- History of pituitary adenoma
- Recent history of trauma
- Prior history of cranial nerve palsy
- **Non-isolated sixth nerve palsy**
- Lack of resolution within 3 months

MRI skull base



MRV

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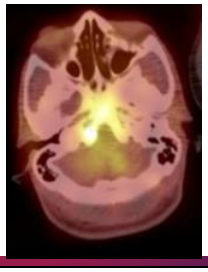
Patient 1- PET



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Patient I

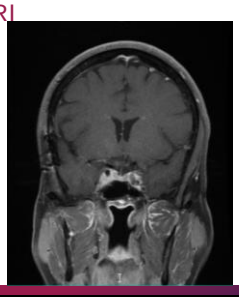
- Lumbar puncture- normal opening pressure
- CSF studies, including cytopathology (x2), unremarkable
- He underwent external beam radiation to the clivus, as well as systemic chemotherapy

Patient 2 Follow Up

Indications for neuroimaging:

- Age younger than 50 years
- **No known microvascular risk factors**
- Any history of cancer, whether active or remote
- History of pituitary adenoma
- **Recent history of trauma (minor, likely red herring in this case)**
- Prior history of cranial nerve palsy
- Non-isolated sixth nerve palsy
- Lack of resolution within 3 months

Patient 2- MRI



Take Home Points

- History and examination can help tailor evaluation plan for patients presenting with sixth nerve palsy
- Neuroimaging is always reasonable in the setting of sixth nerve palsy and is necessary for any patient with:
  - Age younger than 50 years
  - No known microvascular risk factors
  - Any history of cancer, whether active or remote
  - History of pituitary adenoma
  - Recent history of trauma
  - Prior history of cranial nerve palsy
  - Non-isolated sixth nerve palsy
  - Lack of resolution within 3 months

Disclosures

Wilmer Eye Institute
Johns Hopkins Medicine

Horizon Therapeutics/Amgen (Advisory Boards)

Catalyst Pharmaceuticals (Advisory Board)

Argenx (Clinical Trial Site)

Objectives

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By the end of this talk, participants will be able to:

a. Identify red flag signs and symptoms in a patient presenting with anterior ischemic optic neuropathy
b. Develop appropriate treatment plans for patients with giant cell arteritis

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80 yo woman with acute visual loss OD

Visual acuity OD CF @ 1', OS 20/25

+RAPD OD

IOP 12/12

EOM full OU

Anterior segment exam unremarkable aside from PCIOL OU

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Differential Diagnosis

- Anterior ischemic optic neuropathy
  - AAION (GCA)
  - NAION
- Optic neuritis

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What additional information may be helpful?

- History of Present Illness
  - Pain
    - Headaches
    - Scalp tenderness
    - Jaw pain with chewing
    - Polymyalgia rheumatica
    - Pain with eye movements
  - Constitutional symptoms
    - Fever
    - Weight loss
  - Other visual symptoms
    - Preceding episodes of transient vision loss, diplopia
- Past Medical History
  - Hypertension
  - Diabetes
  - Obstructive sleep apnea (or symptoms to suggest, eg, STOP-BANG)
  - Medication history
  - Cancer history

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Patient History

- History of Present Illness
  - Pain
    - Headaches
    - Scalp tenderness
    - Jaw pain with chewing
    - ~~Pain with eye movements~~
    - Polymyalgia rheumatica
  - Constitutional symptoms
    - Fever
    - Weight loss
  - Other visual symptoms
    - Preceding episodes of transient vision loss, diplopia
- Past Medical History
  - Hypertension
  - ~~Diabetes~~
  - ~~Obstructive sleep apnea (or symptoms to suggest, eg, STOP-BANG)~~
  - ~~Medication history~~
  - Cancer history
    - Basal cell on the face

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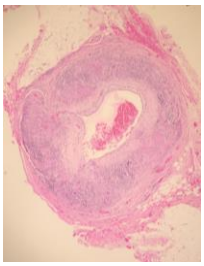
Work Up, Next Steps

- ESR: 105
- CRP: 4.50 (ULN 0.5)
- Platelets: 526
- Started on steroid immediately (high dose IV preferred)

Calculating Risk

Ing EB, Lahaie Luna G, Toren A, Ing R, Chen JJ, Arora N, Torun N, Jakpor OA, Fraser JA, Tyndel B, Sundaram AN, Liu X, Lam CT, Patel V, Weis E, Jordan D, Gilberg S, Pagnoux C, Ten Hove M. Multivariable prediction model for suspected giant cell arteritis: development and validation. Clin Ophthalmol 2017 Nov 22;11:2031-2042.

Temporal artery biopsy: + for GCA



What other testing could be helpful, if case less clear cut?

- Temporal artery ultrasound ??
  - Pros: noninvasive, ability to examine large segment/multiple branches of TA, can include ultrasound of other vessels to improve sensitivity
  - Weighted same as +TAB in 2022 updated of ACR/EULAR Classification Criteria for GCA
  - Recommended as first-line diagnostic test by EULAR, when it is readily available and performed with high quality
  - Con: *Operator dependent*

Schmidt WA, Seifert A, Gromnica-Ihle E, Krause A, Natusch A. Ultrasound of proximal upper extremity arteries to increase the diagnostic yield in large-vessel giant cell arteritis. Rheumatology (Oxford). 2008 Jan;47(1):96-101.
Ponte C, Grayson PC, Robson JC, Suppiah R, Gribbons KB, Judge A, Craven A, Khalid S, Hutchings A, Watts RA, Merkel PA, Luqmani RA; DCVAS Study Group. 2022 American College of Rheumatology/EULAR Classification Criteria for Giant Cell Arteritis. Arthritis Rheumatol. 2022 Dec;74(12):1881-1889.
Dejaco C, Ramiro S, Duftner C, Besson FL, Bley TA, Blockmans D, et al. EULAR recommendations for the use of imaging in large vessel vasculitis in clinical practice. *Ann Rheum Dis*. 2018;77:636–43.

Temporal Artery Ultrasound

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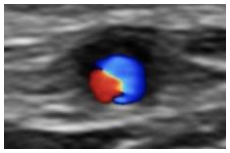
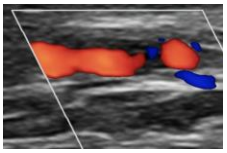
Halo Sign (transverse view)



(longitudinal view)



Compression Sign

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What other testing could be helpful, if case less clear cut?

- Temporal artery ultrasound ??
- MRI ??
  - Vessel wall imaging (cranial, orbital)

MRI for GCA- Orbital Enhancement

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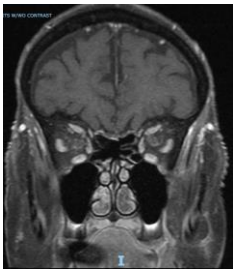

MRI for GCA

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- Enhancement of the vessel walls of the cranial vasculature (i.e., superficial temporal artery)

RheeRL, Rebello R, Tanhankar MA, BanerjeeS, Iiu E, Cao Q, Kurtz R, Baker JE, FanZ, Bhatt V, Amudala N, Chou S, Liang R, Sanchez M, Burke M, Desiderio L, Loewner LA, Morris J S, MerkelPA, SongJW. Combined Orbitaland Cranial VesselWall Magnetic Resonance Imaging for the Assessment of Disease Activity in Giant Cell Arteritis. ACR Open Rheumatol. 2021 Apr;3(4):189-200.

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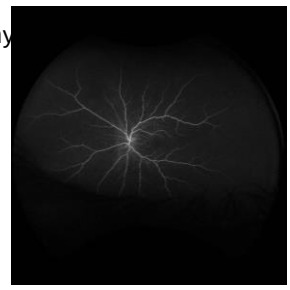
What other testing could be helpful, if case less clear cut?

- Temporal artery ultrasound ??
- MRI ??
  - Vessel wall imaging (cranial, orbital)
- Fluorescein angiography

Fluorescein angiography 81 yo man with GCA



18 seconds

Fluorescein angiography

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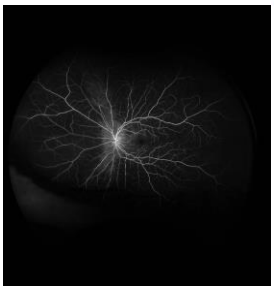

25 seconds

Fluorescein angiography

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60 seconds

Fluorescein angiography

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83 seconds

GCA Management

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- Urgent high-dose steroids
- Rheumatology evaluation
  - Find a co-managing rheumatologist
    - Systemic management
    - Management of steroid side effects
    - +/- Steroid sparing immunosuppression
      - Tocilizumab (anti-IL6R)
        - Reduces steroid requirement, as well as rate of disease flare, when compared with steroid alone
        - Lower steroid doses > decreased risk for steroid complications

Stone JH, Tuckwell K, Dimonaco S, Klearman M, Aringer M, Blockmans D, Brouwer E, Cid MC, Dasgupta B, Rechi J, Salvarani C, Schett G, Schulze-Koops H, Spera R, Unizony SH, Collinson N. Trial of Tocilizumab in Giant-Cell Arteritis. N Engl J Med. 2017 Jul 27;377(4):317-328.

GCA- Take Home Points

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- Giant cell arteritis is a large-vessel vasculitis, occurring almost exclusively in patients over age 50, with a dreaded complication of bilateral blindness
- A diagnosis of GCA should be considered in any case presenting as an AION in a patient over age 50
  - Associated symptoms (temporal headaches/tenderness, jaw claudication, fever, weight loss, PMR, prodrome with transient vision loss and/or diplopia), elevated inflammatory markers, poor presenting acuity, pale disc appearance, lack of disc-at-risk in fellow eye, and patchy choroidal filling on FA increase suspicion
  - GCA Risk Calculator (edse.lng.com/giant-cell-arteritis-nomogram) may help calculate risk, thus avoiding needed for steroids/temporal artery biopsy is very low risk patients
- While AAION is the most common ocular presentation of GCA, patients can also present with CRAO, cilioretinal artery occlusion, posterior ischemic optic neuropathy, PAMM, cranial neuropathy, transient monocular vision loss, extraocular muscle ischemia, vision changes due to choroidal ischemia, or combinations of these- keep a high suspicion for GCA

GCA- Take Home Points

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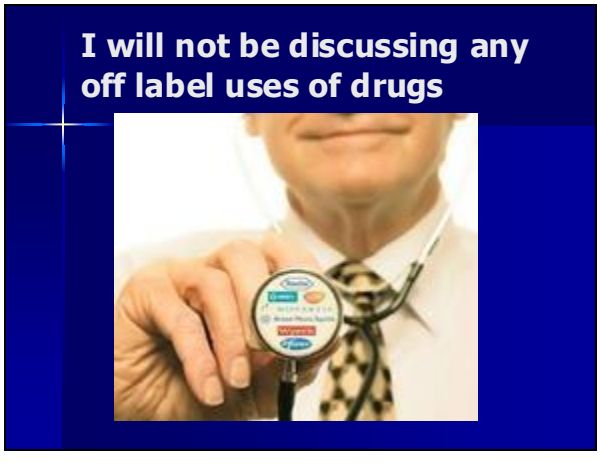
- If GCA is suspected, immediate coverage with steroids is imperative to reduce the risk of further vision loss
- Tocilizumab (anti-IL6R) may reduce steroid requirement, as well as rate of disease flare, when compared with steroid alone


NFL
NASA
•Dr. Lee (Houston Methodist Hospital) works as a consultant for the United States Department of Justice (DOJ), the National Aeronautics and Space Administration (NASA), and the National Football League (NFL) but the views expressed here are his own and do not represent those of these organizations or the United States government.
Other consultant disclosures: Amgen, Viridian, Alexion, AstraZeneca, Bristol Myers Squibb,Catalyst, Stoke
These potential COI have been mitigated per CME rules


I have no financial interest in the contents of this talk


I will not be discussing any off label uses of drugs

•Dr. Lee (Houston Methodist Hospital) works as a consultant for the **United States Department of Justice (DOJ),** the **National Aeronautics and Space Administration (NASA),** and the **National Football League (NFL)** but the views expressed here are his own and do not represent those of these organizations or the United States government.

Other consultant disclosures: Horizon therapeutics, AstroZeneca, Bristol Myers Squibb

These potential COI have been mitigated per CME rules

It turns out that the Jedi superpower: The force is real

Our ability to look at people's eyes and see and deflect the grim reaper is a real superpower (it is the Force)

Overview

- List five potentially life threatening diagnosis in neuro-op
- Define "rule of the pupil"
- Define best imaging study for the 5 dx
- Show key clinical or radiographic features for the above 5 dx

Five dx you cannot afford to miss

1. Arteritis (Giant cell)
2. Apoplexy (Pituitary)
3. Abscess (Mucor)
4. Aneurysm (pupil involved third nerve palsy)
5. Arterial (carotid or vertebral) dissection

Initial symptoms in GCA (n = 100)

Symptom or complaint	Presenting symptom	Finding at diagnosis
Headache	32	68
Polymyalgia rheumatica	25	39
Fever	15	42
Visual symptoms without loss of vision	7	30
Weakness, malaise, fatigue	5	46
Tenderness over arteries	5	27
Myalgias	4	30
Weight loss, anorexia	2	50
Jaw claudication	2	45
Permanent loss of vision	1	14
Tongue claudication	1	6
Sore throat	1	9
Vasculitis on angiogram	1	NA
Stiffness of hands and wrists	1	NA
Decreased temporal artery pulse	NA	46
Erythematous, nodular, swollen arteries	NA	23
Central nervous system abnormalities	NA	15
Synovitis	NA	NA
Dysphagia	NA	15
Limb claudication	NA	NA

Adapted with permission from Hunder GG. Temporal arteritis and polymyalgia rheumatica. In: Kelley WN, et al. Textbook of rheumatology. 4th ed. Philadelphia: Saunders; 1993:1111-22.

OMIC claims: "Whereas…."

1. INDEMNITY PAYMENTS MADE TO SETTLE GCA CASES

	GCA Claims	All OMIC Claims
Closed with a payment	44%	21%
Mean (average) payment	$203,250	$165,282
Median (middle) payment	$335,000	$81,875
Highest payment	$450,000	$3,375,000

https://www.omic.com/giant-cell-arteritis/

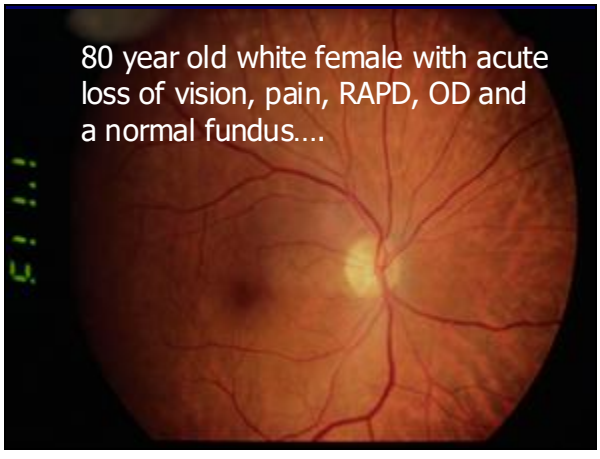
Beware "pallid edema"

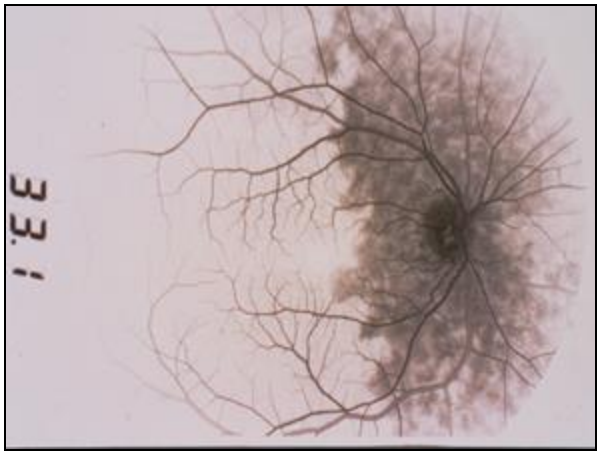
http://webeye.ophth.uiowa.edu/dept/aion/13-AION-A-AION.htm

And the MRI of head was normal…..

- WHY?


20 year old white female with acute loss of vision, pain, RAPD, OD and a normal fundus….


80 year old white female with acute loss of vision, pain, RAPD, OD and a normal fundus….



Beware "optic neuritis" in elderly….likely GCA

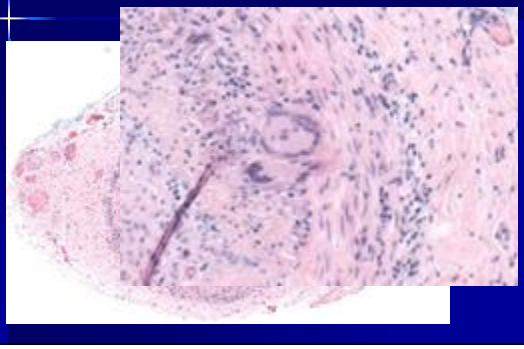
- Wicked good pearl: retrobulbar optic neuritis in elderly might be PION due to GCA….Pallid edema sometimes looks like no edema (dead nerve cant swell)

Five easy mistakes to avoid in GCA = don't call these NAION

- Severe visual loss (e.g. LP or NLP)
- Bilateral simultaneous visual loss
- Transient visual loss (not seen in non-arteritic form of ischemic optic neuropathy)
- PMR with visual symptoms

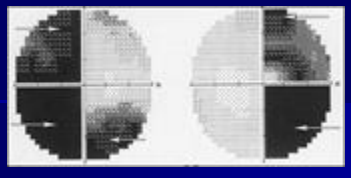
Neurologic Clinics 2016 Aug;34(3):611-29. doi:10.1016/j.ncl.2016.04.005.

Biopsy proven giant cell arteritis



There are five things to remember about acute visual loss in the elderly

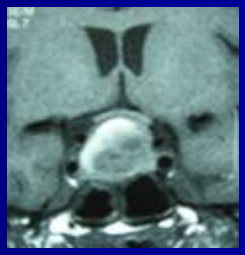
- One is GIANT CELL ARTERITIS….
- And the other four are Giant Cell Arteritis



Life threatening diagnosis?

Pituitary apoplexy

- Acute onset
- Usually severe headache
- Bitemporal hemianopsia
- Apoplexy can kill (8%)
- Hypopituitarism (cortisol)
- Emergent scan



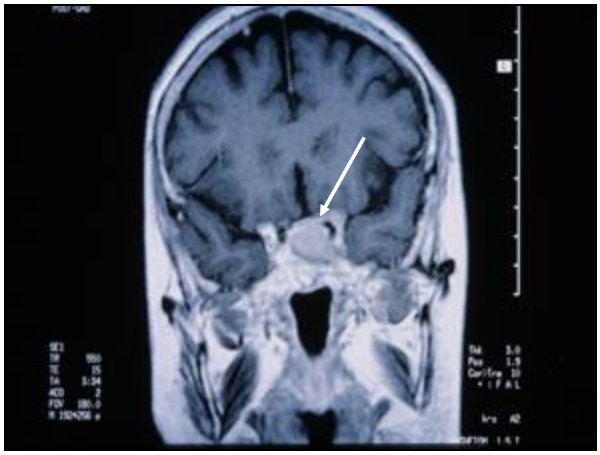
Neurologic Clinics 2016 Aug;34(3):611-29. doi: 10.1016/j.ncl.2016.04.005.

biocomp.stanford.edu

Acute ophthalmoplegia in a diabetic

- 35 y/o WM with diabetes
- History of diabetic ketoacidosis
- Complete left ptosis
- Acute onset almost complete left sided ophthalmoplegia
- What should be the evaluation?

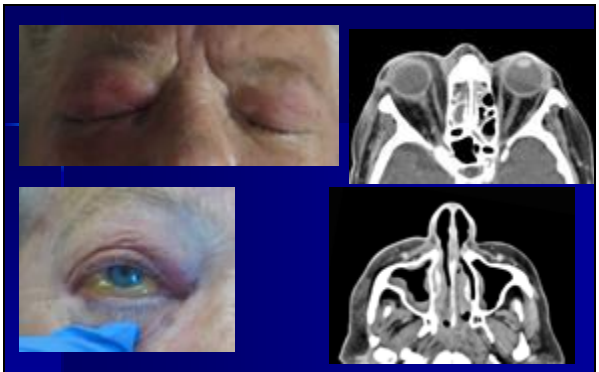
Life threatening diagnosis?



Case from Iowa

- 76-year-old woman with with acute myelogenous leukemia (AML)
- Induction chemotherapy (day 13)
- Two day history of worsening right-sided periorbital swelling & erythema

http://webeye.ophth.uiowa.edu/eyeforum/cases/108-Orbitorhinocerebral-Mucormycosis.htm

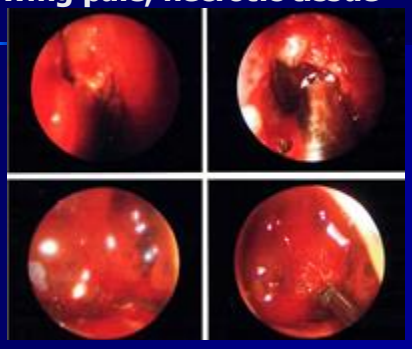


http://webeye.ophth.uiowa.edu/eyeforum/cases/108-Orbitorhinocerebral-Mucormycosis.htm

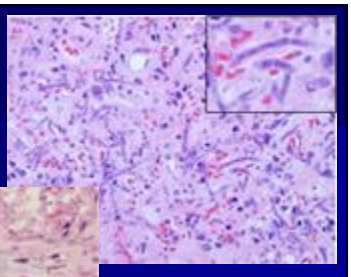
Is this orbital inflammatory pseudotumor? Tolosa Hunt?

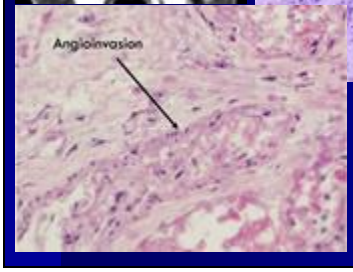
- Wicked good pearl: Don't give patients who are immunosuppressed the diagnosis of autoimmune disease!

Intraoperative endoscopic photos showing pale, necrotic tissue



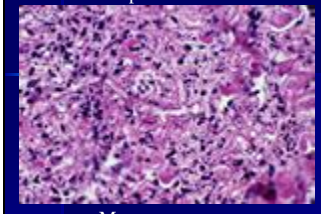
http://webeye.ophth.uiowa.edu/eyeforum/cases/108-Orbitorhinocerebral-Mucormycosis.htm





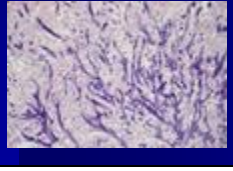
http://webeye.ophth.uiowa.edu/eyeforum/cases/108-Orbitorhinocerebral-Mucormycosis.htm

From: http://medic.med.uth.tmc.edu/edprog/Path/InfDis.htm



Mucor

Does not have to show black eschar

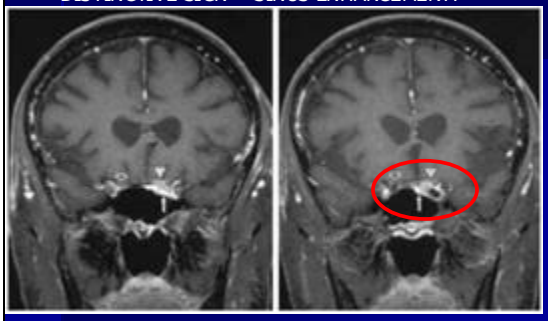


Can be Aspergillus too!

And the MRI of head was normal…..

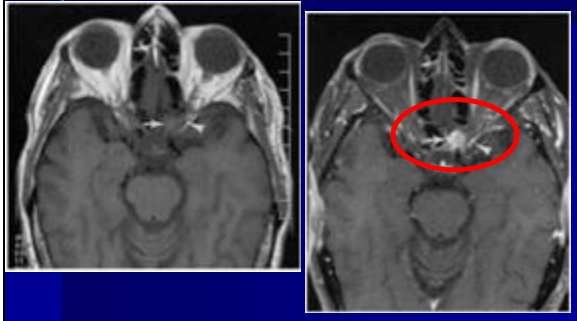
- WHY?

YOU NEED CONTRAST.
DISTINCTIVE SIGN = SINUS ENHANCEMENT!



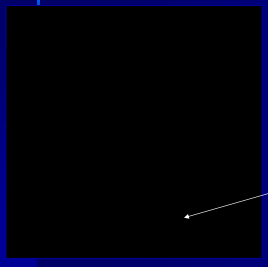
http://www.mayoclinicproceedings.com/inside.asp?AID=230&UID=

Aspergillosis of orbital apex



http://www.mayoclinicproceedings.com/inside.asp?AID=230&UID=

What happens if you don't give contrast?....



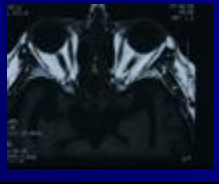
My house at NIGHT!!!

Why didn't they give MRI contrast?

- Diabetic
- Diabetic nephropathy
- Poor renal function (GFR)
- They wont give the gadolinium
- Fear nephrogenic systemic dermatopathy

What is Fat suppression ("fat-sat")? technique

- T1 weighted signal
- Increase contrast (light and dark) between structures
- Fat is "too bright" on T1

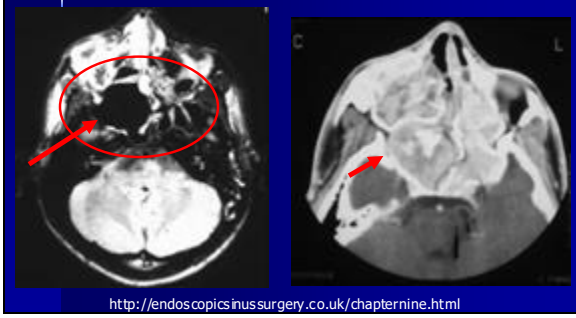


And the MRI of head was normal.....

- WHY?

Polar bear in a snowstorm

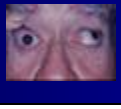
Complementary roles for CT & MR in fungal orbital apex disease (T2 dark)



http://endoscopicsinussurgery.co.uk/chapternine.html

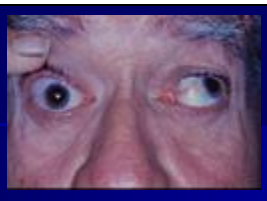
What's wrong with this picture?

- 60 y/o diabetic man
- New onset ptosis right
- Right adduction, elevation, & depression deficit
- 45 exotropia (XT)
- Diagnosis: "Ischemic third nerve palsy"
- Plan: "Return 6 weeks"



Tell your technicians….

- If the patient's complaint is diplopia or ptosis or….
- If you have to lift a ptotic lid to put in the dilating drops then….
- STOP, come get the doctor before dilating



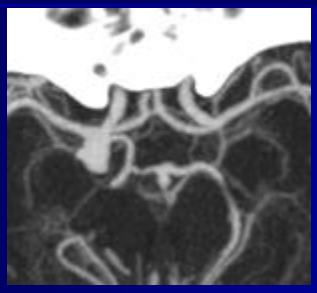
Acute pupil involved third n. palsy
Life threatening diagnosis?

Rule of the pupil

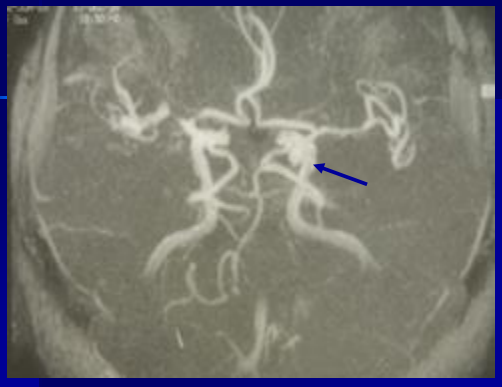
- A pupil involved third nerve palsy
- Aneurysm of posterior communicating artery until proven otherwise

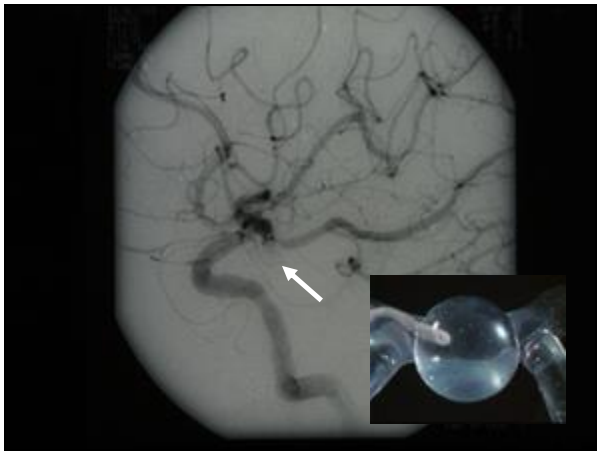
Neurologic Clinics 2016 Aug;34(3):611-29. doi: 10.1016/j.ncl.2016.04.005.

CTA: R posterior communicating a. aneursym



http://www.cedars-sinai.edu





5 myths that even some NOPs believe in CN III palsy

1. You can observe CN III palsy in diabetics
2. MRI is sufficient for CN III (You need an A to find that A)
3. MRI is better than CT (CT/CTA first to look for SAH/aneurysm)
4. If CT/CTA negative you are done: You need MRI/MRA first to look for non-aneurysmal etiologies or do MRI second if CTA negative
5. Catheter angiography is no longer needed in post MRI/MRA era: If MRI/MRA and CTA not of sufficient quality or insufficient confidence level to rule out aneurysm

Neurologic Clinics 2016 Aug;34(3):611-29. doi:10.1016/j.ncl.2016.04.005.

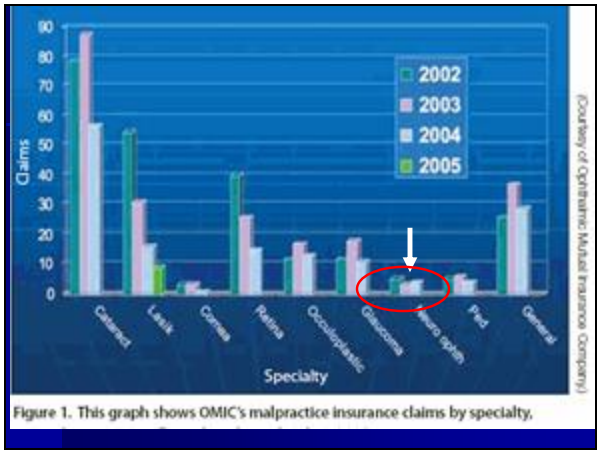


Figure 1. This graph shows OMIC's malpractice insurance claims by specialty.

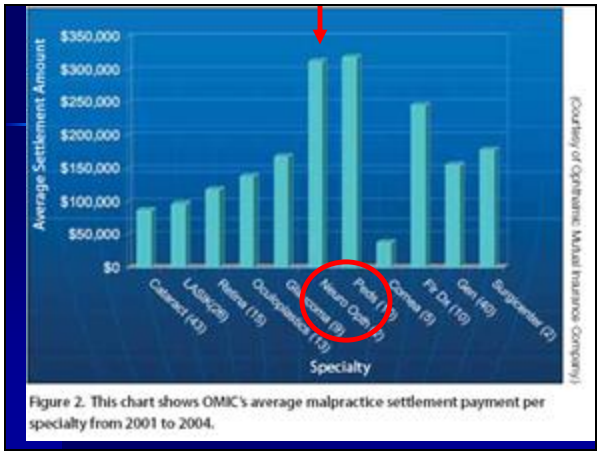


Figure 2. This chart shows OMIC's average malpractice settlement payment per specialty from 2001 to 2004.

Acute painful anisocoria after car accident

Life threatening diagnosis?

Horner syndrome

- Wicked good pearl: In acute setting just image sympathetic axis for Horner syndrome

As if death weren't enough….

And the MRI of head was normal…..

- WHY?

Summary

- List five potentially life threatening diagnosis in neuro-op
- Define "rule of the pupil"
- Define best imaging study for the 5 dx
- Show key clinical or radiographic features for the above 5 dx

Summary: Lee's "A"s: The five chances to save the life of your next neuro-ophthalmology patient

1. Arteritis (Giant cell)
2. Apoplexy (Pituitary)
3. Abscess (Mucor)
4. Aneurysm (pupil involved third nerve palsy)
5. Arterial (carotid or vertebral) dissection

But really my teaching point is that using our super power to detect & deflect the reaper is our best defense against physician burnout

Bottom line: Its your job

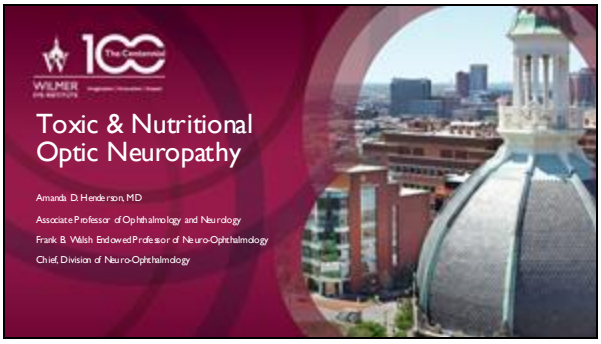
There is only one thing to say to the god of death…

Not today, not on my watch



Thanks for your time & attention

- Andrew G. Lee, MD
- Chair Ophthalmology, Houston Methodist Hospital, Professor of Ophthalmology, Neurology, & Neurosurgery, Weill Cornell Medical College; Adjunct Professor: Baylor College of Medicine, U. Iowa & Clinical Professor, UTMB Galveston, UT MD Anderson Cancer Center, U. Buffalo, SUNY



Toxic & Nutritional Optic Neuropathy

Amanda D. Henderson, MD
Associate Professor of Ophthalmology and Neurology
Frank B. Walsh Endowed Professor of Neuro-Ophthalmology
Chief, Division of Neuro-Ophthalmology

Disclosures

Wilmer Eye Institute

Horizon Therapeutics/Amgen (Advisory Boards)

Catalyst Pharmaceuticals (Advisory Board)

Argenx (Clinical Trial Site)

Objectives

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By the end of this talk, participants will be able to:

a. Identify typical presentations of toxic and nutritional optic neuropathies
b. Perform an appropriate work up for patients with toxic and nutritional optic neuropathies

Toxic Causes

- Methanol
- Ethyleneglycol
- Lead
- Organic solvents
- Tobacco
- Ethanol
- Ethambutol
- Amiodarone
- Linezolid
- Disulfiram
- Cyanide

Nutritional Causes

Deficiencies of:

- B12
- Folate
- Thiamine
- Vitamin A (usually a retinopathy)
- Zinc

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78-year-old man presents with gradually progressive peripheral visual loss for several months

POH
Remote history of ocular trauma, with no remaining visual effects
No prior ocular surgeries

PMH
Hypertension
Mycobacterium avium intracellulare
Depression

FH unremarkable

SH
Moved from the Ukraine to the US 25 years ago
Retired shoemaker
Nonsmoker

Meds
Metoprolol succinate
Amlodipine/valsartan
Lorazepam
Azithromycin
Ethambutol 1200mg daily

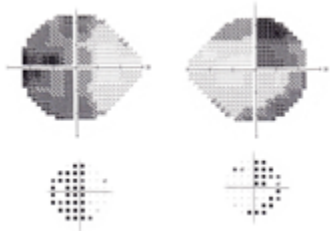
Exam

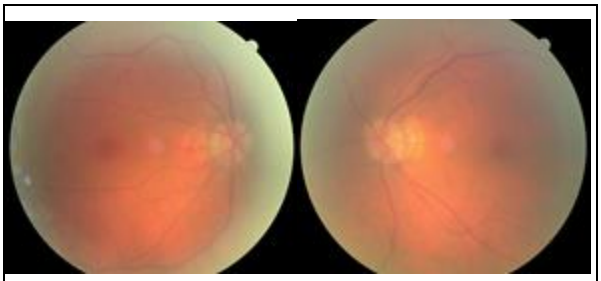
VA OD 20/70
OS 20/400
No RAPD

Color plates
Right eye 4/10
Left eye 4/10

Anterior segment unremarkable

HVF 24-2

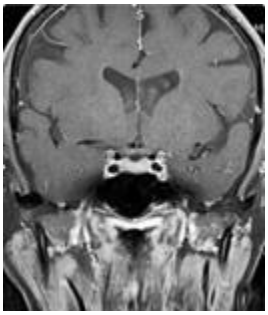




HVF 24-2

4 mos prior to presentation

At presentation



Serum labs
CBC wnl
eGFR slightly reduced at 57 (LLN 59)
Vitamin B12 922 (normal range 211-946)
Folate >19.9

Diagnosis?

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Ethambutol toxicity affecting the optic chiasm

Ethambutol

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- Bacteriostatic agent used to treat mycobacterial infections
- Optic neuropathy is a well-known side effect of ethambutol treatment
- Ethambutol preferentially affects the papillomacular bundle, causing central or cecocentral scotomas, and has also been reported to affect the optic chiasm, causing a bitemporal hemianopia

Ethambutol Toxicity

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- Dose-related
  - No "safe" dose
- Sometimes reversible with discontinuation of medication
- Incidence
  - 15% with doses of ≥35 mg/kg/day
  - 5-6% with dose of 25 mg/kg/day
  - 1% with dose of 15 mg/kg/day
- Mean time of onset is 7 months after initiation of therapy
  - Rarely occurs earlier than 1.5 months after initiation of therapy

Ethambutol Toxicity: Risk Factors

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- Renal insufficiency
- Diabetes
- Hepatic disease
- High dose of ethambutol
- Older age

Clinical course

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- Ethambutol therapy stopped
- Ciprofloxacin 500mg bid substituted for ethambutol
- Vision improved to 20/40 OD and 20/100 OS
- Bitemporal hemianopia resolved gradually over ~4 months

18-year-old woman from Kuwait presents with 8 months of progressively blurred vision

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Previously treated in Kuwait with steroid and with IVIg with no improvement.

Otherwise, PMH unremarkable

ROS: + paresthesias in lower extremities, +distal > proximal weakness in extremities

Exam

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Acuity 20/100 OD and OS

Sluggish pupillary reactions with no RAPD

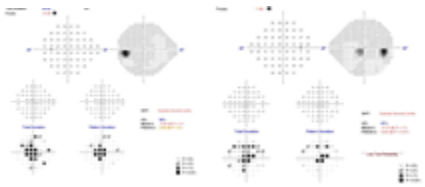
Color plates 11/13 OD and OS

Temporal pallor of optic discs OU

Decreased vibration and proprioception, distal > proximal extremity weakness (Neurology)

HVF

Wilmer Eye Institute
Johns Hopkins Medicine



Work up

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- Unremarkable MRI
- **Vitamin B12, vitamin A, vitamin D all low**
- Unremarkable: Aqp4-IgG, thiamine, anti-Gq1b, MOG-IgG, sensory-motor neuropathy ab panel, SPEP
- LP initially had high protein in Kuwait, but repeat at Hopkins was unremarkable
- Striational ab positive at low titer, otherwise paraneoplastic panel neg

Further history

- Patient had been vegan for 2 years
- Family reported that she had been "eating very little" for the last couple of years, in an attempt to lose weight

Vitamin B12 deficiency

- May cause bilateral, slowly progressive optic neuropathy, with central/cecocentral scotomas
- Optic neuropathy may precede anemia and other neurologic signs/symptoms in patients with deficiency, though usually patients will have signs/symptoms of sensorimotor polyneuropathy
- Usually related to impaired absorption (though not always), in the setting of pernicious anemia or a history of bariatric surgery

Clinical course

- Supplementation was started, initially with B12 injections, then switched to oral
- She had excellent improvement in serum vitamin levels with the supplementation
- Acuity improved to 20/60 OD and 20/40 OS
- Some improvement in mean deviation on the fields, but still with central scotomata

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Summary

- When evaluating bilateral, gradually progressive optic neuropathy with central/cecocentral scotomas, the 3 main differential diagnostic considerations are **toxic**, **nutritional**, and **hereditary** optic neuropathies
- Careful review of patient medications may identify potentially offending agents
- Historical characteristics may raise suspicion for nutritional deficiencies, and serum lab evaluation can include B12, thiamine, RBC folate, vitamin A, and zinc
- When evaluating a bitemporal hemianopia, it is almost always a compressive lesion. When there is no compressive lesion, think ethambutol.