# What Went Wrong? Neuro-Op M&M

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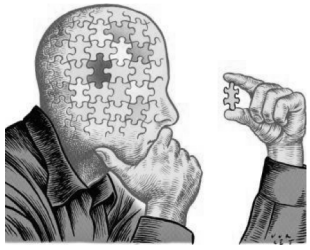
## Disclosures

- No relevant disclosures
- Consultant for GenSight Biologics

## Case 1



## 20-year-old woman

- Recurrent episodes of headaches
  - "Migraine"
  - Over the counter medications
- 2 weeks prior, headaches worsened:
  - More severe
  - More frequent (daily)

- PCP recommends Neurology
- Neurology not available
- Goes to "an" ER:
  - Normal examination – "sinus infection"
  - Narcotics, sent home
- Comes back to "another" ER:
  - "Cranial nerves intact"
  - Normal head CT no contrast; normal CSF
  - Narcotics, sent home

- Returns to same ER (one week of severe headaches):
  - "Normal ophthalmic examination"
  - Normal head CT no contrast
  - Neurology consultation:
    - "Normal examination"
    - "Cranial nerves 1-12 normal"
    - => MRI/MRA normal
  - Sent home

- Goes to another ER two days later:
  - Complains of horizontal diplopia
  - Neurologic consultation:

PHYSICAL EXAMINATION: The patient is sitting in a dark room with some sensitivity to light. She has her head covered with either a towel or a pillow.
HEENT: Her pupils equal, round, reactive to light. Extraocular muscles are intact. She stated she did have some diplopia on extreme gaze previously.
NEUROLOGIC: There is no evidence of cranial nerve 6 involvement. Funduscopic examination, while limited secondary to photosensitive, does not reveal any papilledema. The face is symmetric. V1 through V3 are intact to light touch and temperature. The tongue is midline. The palate and the uvula elevate

  - Sent home…

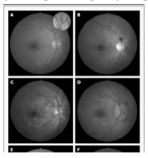
## Two days later

- Diplopia worsens
- Headaches worsen
- Develops visual loss
- =>Another ER (ours…)

## FOTO-ED Study


The NEW ENGLAND JOURNAL of MEDICINE

**Nonmydriatic Ocular Fundus Photography in the Emergency Department**



N Engl J Med 2011;364:387-9.


## Bilateral papilledema


## In the ER (Saturday)

- Ophthalmology:
  - VA 20/30 OU
  - Left RAPD
  - Bilateral optic nerve edema
- Neurology:
  - Normal examination
  - MRI/MRV: no venous thrombosis
  - LP: CSF OP 50cm; normal contents

## Stays until Sunday

- Headaches improved
- Acetazolamide 500 mg bid

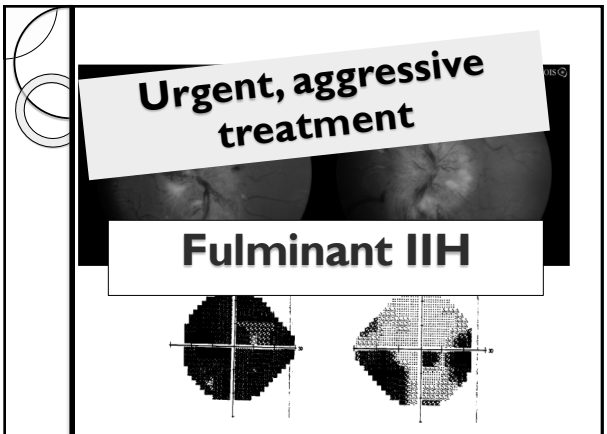
- Send home
- "Follow-up with Neuro-op"

## At home

- Headaches worse
- Diplopia
- Vision worse

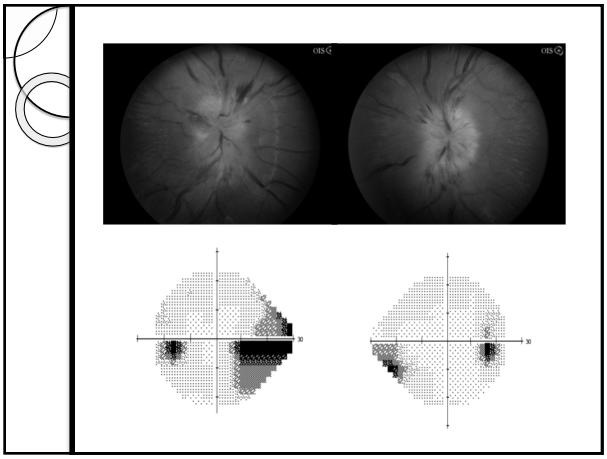
## Neuro-op on Tuesday

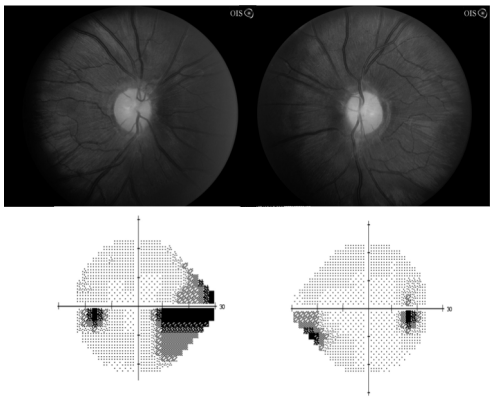
- Severe headaches
- VA 20/70 OU
- Large left RAPD
- Esotropia




## From Neuro-Op

- Immediate LP
- Admission with lumbar drain
- Optic nerve sheath fenestration OS NOW!
- CSF shunting procedure two days later



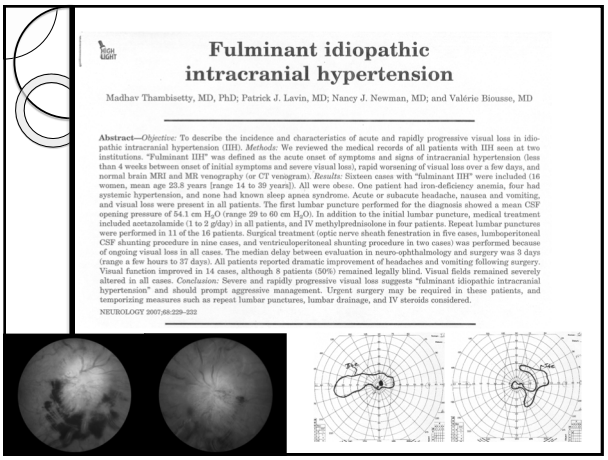


# What Went Wrong?

- No funduscopic examination in headache patient
- Inadequate funduscopic examination
  - =>Learn to do it
  - Or, buy a non-mydriatic fundus camera

# What Else Went Wrong?

- Correct diagnosis but inappropriate evaluation of severity:
- Discharge happened too quickly !
  - Fulminant IIH requires immediate and aggressive treatment



**Fulminant idiopathic intracranial hypertension**

Madhav Thambisetty, MD, PhD; Patrick J. Lavin, MD; Nancy J. Newman, MD; and Valérie Biousse, MD

**Abstract**—*Objective:* To describe the incidence and characteristics of acute and rapidly progressive visual loss in idiopathic intracranial hypertension (IIH). *Methods:* We reviewed the medical records of all patients with IIH seen at two institutions. "Fulminant IIH" was defined as the acute onset of symptoms and signs of intracranial hypertension (less than 4 weeks between onset of initial symptoms and severe visual loss), rapid worsening of visual loss over a few days, and normal brain MRI and MR venography (or CT venogram). *Results:* Sixteen cases with "fulminant IIH" were included (16 women, mean age 23.8 years [range 14 to 39 years]). All were obese. One patient had iron-deficiency anemia, four had systemic hypertension, and none had known sleep apnea syndrome. Acute or subacute headache, nausea and vomiting, and visual loss were present in all patients. The first lumbar puncture performed for the diagnosis showed a mean CSF opening pressure of 54.1 cm $H_2O$ (range 29 to 60 cm $H_2O$). In addition to the initial lumbar puncture, medical treatment included acetazolamide (1 to 2 g/day) in all patients, and IV methylprednisolone in four patients. Repeat lumbar punctures were performed in 11 of the 16 patients. Surgical treatment (optic nerve sheath fenestration in five cases, lumboperitoneal CSF shunting procedure in nine cases, and ventriculoperitoneal shunting procedure in two cases) was performed because of ongoing visual loss in all cases. The median delay between evaluation in neuro-ophthalmology and surgery was 3 days (range a few hours to 37 days). All patients reported dramatic improvement of headaches and vomiting following surgery. Visual function improved in 14 cases, although 8 patients (50%) remained legally blind. Visual fields remained severely altered in all cases. *Conclusion:* Severe and rapidly progressive visual loss suggests "fulminant idiopathic intracranial hypertension" and should prompt aggressive management. Urgent surgery may be required in these patients, and temporizing measures such as repeat lumbar punctures, lumbar drainage, and IV steroids considered.

NEUROLOGY 2007;68:229–232

# Case 2

"Off hand, I'd say you're suffering from an arrow through your head, but just to play it safe, I'm ordering a bunch of tests."

# 87 yo W woman

- Binocular diplopia
- PMHx:
  - Atrial fibrillation on Coumadin
  - 3 episodes of transient hemiparesis (last 3-4 y prior)
  - Recurrent falls – fell twice over last month and hit her head (bruises on her face)
  - s/p cataract extraction/PCIOL OU

- Day 1: pulled by her dog and fell
  - Presumed head trauma with loss of conciousness (no CT)
  - Left wrist fracture
- Day 4:
  - Binocular vertical diplopia
  - No headache or other neurologic sign

- Day 6 (Ophthalmology):
  - VA: 20/25 OU
  - Extraocular movements full
  - Right flick hypertropia and esotropia
- Same day (Strabismus):
  - Same
  - Given prism (3 D base down OD)

- Day 7 (Neuro-oph)
  - VA: 20/25 OU
  - Extraocular movements full
  - 2 prism D ET and right HT
  - Fundus: mild temporal pallor OU (had cataract surgery: "pseudophakic")

## "Decompensated latent strabismus", but… let's be careful (fall)

- INR: 2.69. CBC, platelet: normal
- Head CT (same day):
  - No bleed
  - Remote left MCA infarction

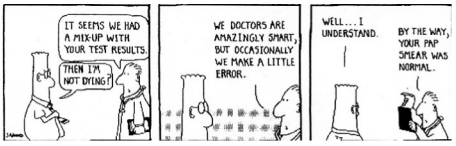
## However…

- Day 20:
  - Vision loss OD > OS
  - Elevated ESR and CRP
  - Temporal artery biopsy: florid inflammation

# What Went Wrong?

- Just **think** about GCA in any patient:
  - Older than 50
  - Visual symptoms
    - Visual loss (nerve, retina, choroid)
    - Unexplained visual loss
    - Transient visual loss
    - Diplopia
    - Transient diplopia
    - Cranial nerve palsy
  - Headaches

## Case 3




37 year old white woman with visual loss right eye

Past Medical History: Unremarkable

Medications: None

Family History: Migraines (mother)

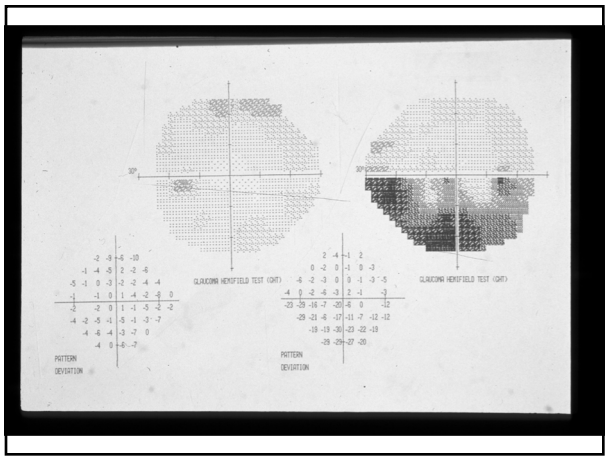
5 weeks prior:
- Irritation/itching right eye
- Better with artificial tears

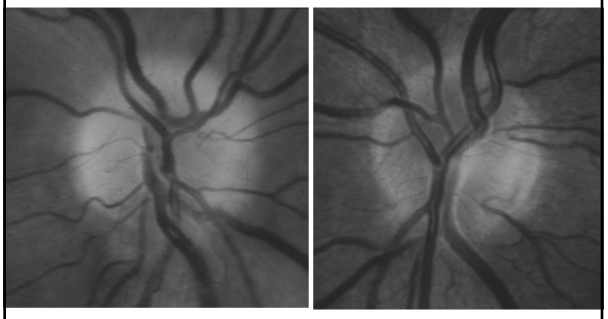
5 days later:
- Decreasing vision in right eye lower VF
- Worsened over 4-5 days
- No headache, pain, pain on eye movement

No previous visual loss or neurologic symptoms
No change in vision over next 3-4 weeks

Ophthalmologist examination:

| | | |
|---|---|---|
| Vision: | 20/20 | 20/20 |
| Color: | 14/14 | 14/14 |
| | 20% red desat | |
| Orbits: | Normal | Normal |
| SLE: | Normal | Normal |
| IOPs: | 14 | 14 |
| Pupils: | + RAPD | |
| EOMs: | Full | Full |



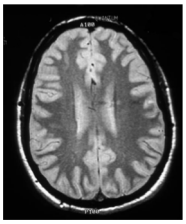


## Optic Neuropathy

- Compression/Infiltration
  - Neoplastic vs non-neoplastic
- Inflammation
  - Infectious vs non-infectious
- Ischemia
- Toxic/Metabolic
- Hereditary
- Trauma
- Mechanical
  - Elevated intraocular pressure
  - Elevated intracranial pressure

Sent to a neurologist:
Otherwise normal neurologic examination
Orders brain MRI without contrast

Neurologist plans:
LP
IV steroids
Considers multiple sclerosis



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## AION vs. ON

Why Do We Care?

- Prognosis for visual recovery
- Recognition of giant cell arteritis
- Prognosis and treatment for multiple sclerosis

## AION vs. ON

- Rate of visual loss same as for optic neuritis
  - *Rizzo JF, Lessell S. Arch Ophthalmol 1991*
- Range of acuities same as for optic neuritis
  - *Rizzo JF, Lessell S. Arch Ophthalmol 1991*
- NAION in pts < 50 yrs old
  - 169/727 (23%) pts
  - Final VA 20/40 or better in 64%
  - VF defects persist
  - 2nd eye involved in 41%
    - *Preechawat P, Bruce BB, Newman NJ, Biousse V. AJO 2007*

## AION vs ON

| | | |
|---|---|---|
| Age | Older (>50) | Younger |
| Gender | M = F | F > M |
| Visual loss | Acute | Rapidly progressive |
| Pain | Infrequent | Frequent with EOM |
| Color Vision | May be normal | Commonly abnormal |
| Visual Field | Altitudinal defect | Central defects |
| Optic Disc | Acute: edema<br>Small c/d | Normal or edema |
| | Late: seg pallor | Temporal pallor |
| MRI | Nl optic nerve | Abnl optic nerve |
| Visual prognosis | Poor | Good |
| Systemic disease | HTN, DM, r/o GCA | Subsequent MS |

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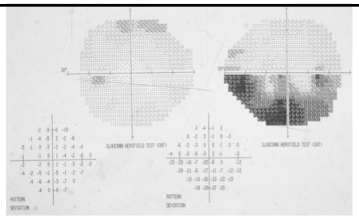
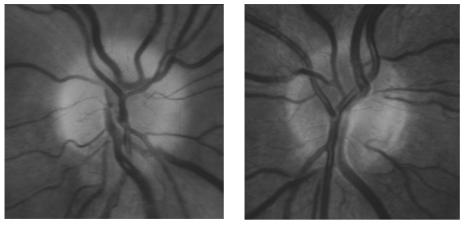




JAMA Ophthalmology | Original Investigation

### Incidence and Causes of Overdiagnosis of Optic Neuritis

Leanne Stunkel, MD; Nathan H. Kung, MD; Bradley Wilson, MA; Collin M. McClelland, MD; Gregory P. Van Stavern, MD

JAMA Ophthalmology Dec 2017

**IMPORTANCE** Diagnostic error is an important source of medical error. Overdiagnosis of optic neuritis may prompt unnecessary and costly diagnostic tests, procedures, and treatments.

**OBJECTIVE** To assess the incidence of and characterize factors contributing to overdiagnosis of acute optic neuritis.

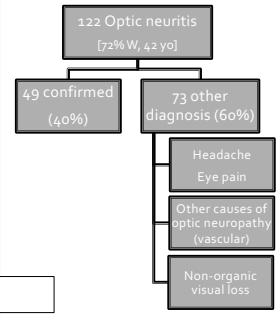
**DESIGN, SETTING, AND PARTICIPANTS** In this retrospective clinic-based cross-sectional study of new patient encounters, 122 patients referred for acute optic neuritis at a university-based Midwestern neuro-ophthalmology clinic between January 2014 and October 2016 were studied. Data were analyzed from September 2016 to July 2017.

**INTERVENTIONS** Definite diagnosis was determined by neuro-ophthalmologists. For patients with alterative diagnoses, the Diagnosis Error Evaluation and Research taxonomy tool was applied to categorize the type of diagnostic error.

**MAIN OUTCOMES AND MEASURES** The primary outcome was the primary type of diagnostic error in patients erroneously diagnosed as having optic neuritis. Secondary outcomes included final diagnosis and interventions undergone prior to referral.

**RESULTS** A total of 122 patients were referred with acute optic neuritis during the study period; 88 (72.1%) were female, and the mean (SD) age was 42.6 (14.0) years. Of these, 49 patients (40.2%; 95% CI, 31.4-49.4) were confirmed to have optic neuritis, and 73 (59.8%; 95% CI, 50.6-68.6) had an alternative diagnosis. The most common alternative diagnoses were headache-and-eye pain, functional visual loss, and other optic neuropathies, particularly nonarteritic anterior ischemic optic neuropathy. The most common diagnostic error was eliciting or interpreting critical elements of history, which occurred in 24 of 73 patients (33%) with alternative diagnoses. Other common errors included errors weighing or considering alternative diagnoses (23 patients [32%]), errors weighing or interpreting physical examination findings (15 patients [21%]), and misinterpreting diagnostic test results (11 patients [15%]). In patients with alterative diagnoses, 12 (16%) had normal magnetic resonance imaging findings preceding the referral, 12 (16%) had received a lumbar puncture, and 8 (11%) had received unnecessary treatment with intravenous steroids.

**CONCLUSIONS AND RELEVANCE** These data suggest that nearly 60% (95% CI, 50.6-68.6) of patients referred for optic neuritis have an alternative diagnosis, with the most common errors being overreliance on a single item of history and failure to consider alternative diagnoses. Understanding pitfalls leading to overdiagnosis of optic neuritis may improve clinicians' diagnostic process.



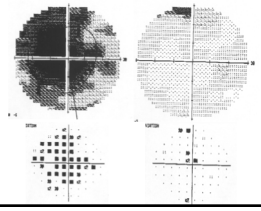

Author Affiliations: Department of Neurology, Washington University School of Medicine, St Louis

## Case 4

- 20-yo WM with visual loss in both eyes
- PMHx
  - Unremarkable
- Fam Hx:
  - Unremarkable
- College student – no ETOH or drugs

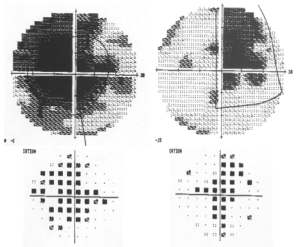
- Age 8: told he had « swelling OU » during routine examination
  - Asymptomatic
  - Observed yearly, without change

- Age 20 (8 months prior seeing us)
  - Sudden, painless visual loss OS
    - VA: 20/20 OD; 20/200 OS
    - "Swelling" OS



- MRI brain/orbits: normal
- CBC, bartonella, toxo, RPR, FTA: normal or negative
- Told he had optic neuritis

- 6 months later:
  - Visual loss OD
  - VA: 20/100 OD; CF OS

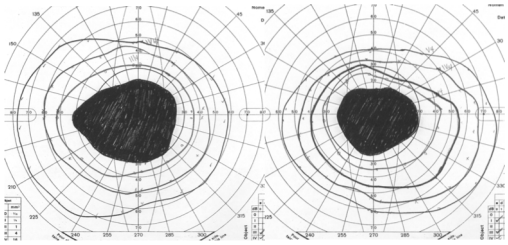


- Repeat MRI: normal
- More blood tests (NMO): all normal
- Lumbar puncture:
  - OP: 16 cm
  - CSF contents: normal

## Examination 2 months later

| | OD | OS |
|---|---|---|
| VA | CF | CF |
| Col Vis | No control | No control |
| Orbit | Normal | Normal |
| Lid | Normal | Normal |
| IOP | 14 | 15 |
| SLE | Normal | Normal |
| Pupils | Normal | 1.2 RAPD |
| EOM | Full | Full |

## Goldmann Visual Fields



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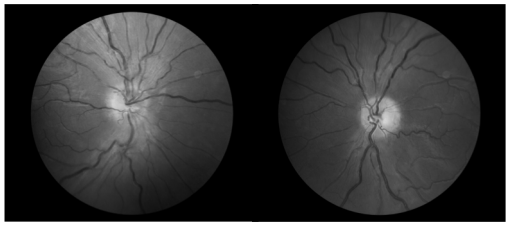
## Fundus



## Work-up

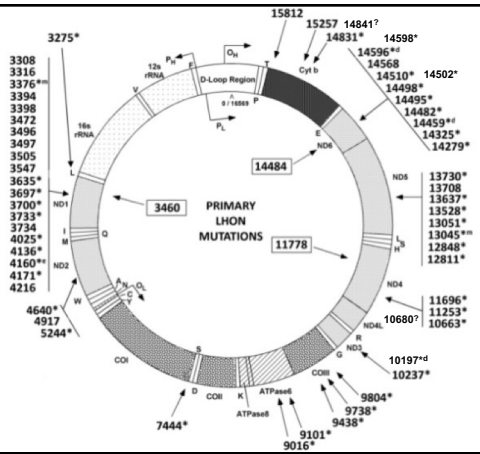
- Leber's hereditary optic neuropathy:
  - 3 primary mutations:
    - mt DNA 11778
    - mt DNA 14484
    - mt DNA 3460
- Negative

-

## Work-up

- Blood sent to lab with expertise in LHON
  - Complete sequencing of mtDNA
    - 10 mutations found
    - Including, novel mtDNA mutation at np 12848 (heteroplasmic)
      - Alters complex 1

-




## Case 5

## Case 6

- 41 yo woman with visual loss in left eye
- PMHx:
  - Hypertension, borderline diabetes
  - Migraine headaches (no aura)
- Medications:
  - Hydrochlorothiazide, aspirin 81, vitamins, ibuprofen prn

## HPI

- Followed by neurologist for episodes of tingling of both legs and occasionally left arm shaking
  - Normal brain MRI
- Saw ophthalmologist for annual visit:
  - Decreased vision left eye
  - Left optic nerve pallor
  - => "Left optic neuritis"

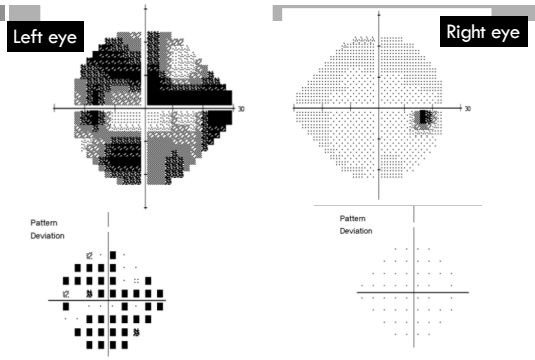
- Neurologist:
  - Repeat brain MRI (normal)
  - Planned LP for possible multiple sclerosis
- Patient:
  - Panicked
  - Refused LP
  - Second opinion

## Neuro-Ophthalmology

| | Right eye | Left eye |
|---|---|---|
| Visual acuity | 20/20 | 20/40- |
| Color vision | 14/14 | 3/14 |
| Slit lamp | Mild cataracts | |
| IOP | 12 | 13 |
| Pupils | Normal | RAPD++ |
| Eye movements | Full | Full |

## Fundus Examination

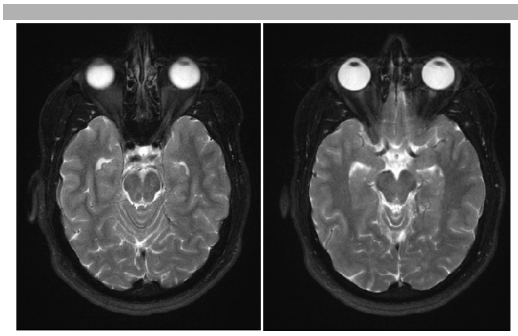

## Humphrey Visual Fields

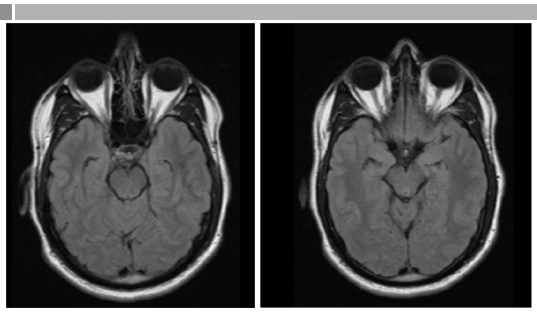


## Diagnosis

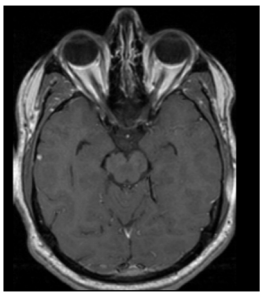
- Left optic neuropathy (chronic)
- Incidentally found
- No pain

## Review MRI

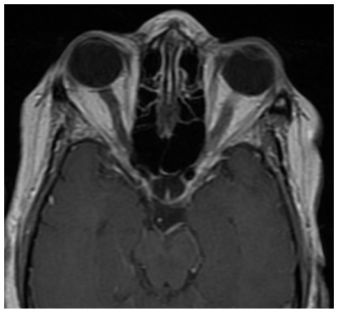




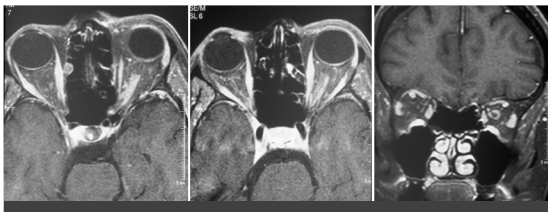
## With contrast



## MRI **Orbits** with Contrast



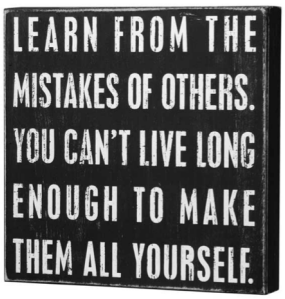
## MRI **Orbits** with Contrast **and Fat Suppression**



Left Optic Nerve Sheath Meningioma


"We'll make a mess of this one, you need to learn how to handle a malpractice complaint."

## Case 6




## 39 yo Chinese woman

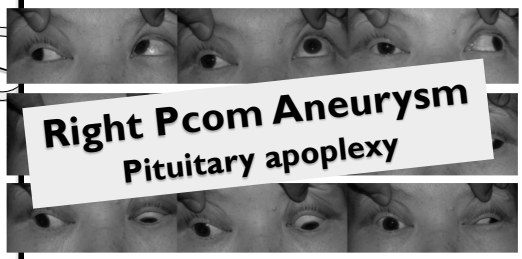
- Systemic lupus erythematosus (2004)
  - Hydroxychloroquine, ASA
- No ocular or neurologic hx

- March 1:
  - Severe right eye and brow pain
  - Lasted 2 days and resolved
- March 5:
  - Diplopia and vomiting
- => Local ER
  - Right third nerve palsy

## Admitted to hospital

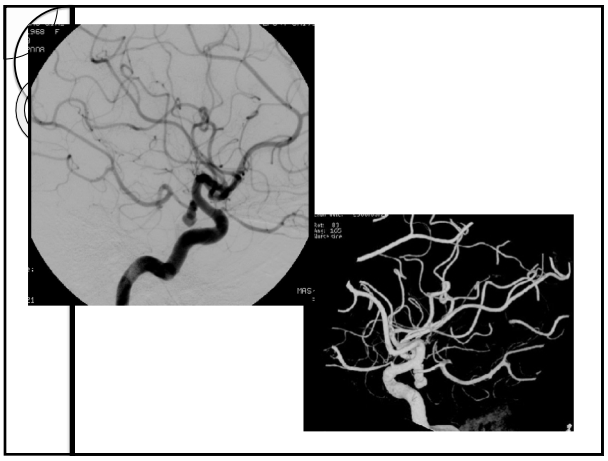
- Normal head CT
- Normal MRI brain (no contrast)
- Normal MRA brain
- LP: 1WC; 39 RBC; gluc 61; protein30 (not checked for xanthochromia)
- Rheumatologist (lupus)
- IV decadron
- Discharged home

- One week later:
  - Eye unchanged
  - => Sees "eye doctor"
- Sent to Neuro-Op



**Right Pcom Aneurysm**
**Pituitary apoplexy**

**Right third nerve palsy complete, with pupil**



# What Went Wrong?

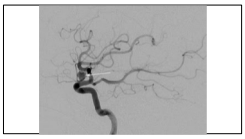
- Know the radiologist
- Talk to the radiologist
- Do not trust a printed report


## Underdiagnosis of Posterior Communicating Artery Aneurysm in Noninvasive Brain Vascular Studies

Valerie I. Elmalem, MD, Patricia A. Hudgins, MD, Beau B. Bruce, MD, Nancy J. Newman, MD, Valérie Biousse, MD

**Background:** Expert interpretation of modern noninvasive neuroimaging such as computed tomographic angiography (CTA) or MRA should detect nearly all aneurysms responsible for an isolated third nerve palsy. Whether a catheter angiogram should still be obtained in cases with negative CTA or MRA remains debated and mostly relies on whether the noninvasive study was correctly performed and interpreted. The aim of our study was to review the diagnostic strategies used to evaluate patients with isolated aneurysmal third nerve palsy at a large academic center.
**Methods:** Retrospective review of all cases with posterior communicating artery (PCom A) aneurysmal third nerve palsies seen at our institution since 2001.
**Results:** We identified 417 cases with third nerve palsy, aneurysm, or subarachnoid hemorrhage, among which 17 presented with an acute isolated painful third nerve palsy related to an ipsilateral PCom A aneurysm (mean age: 52 years; range: 33–83 years). Patients were classified into 3 groups based on the results of the noninvasive imaging obtained at initial presentation. Group I included 4 cases with subarachnoid hemorrhage on initial noncontrast head CT initially obtained in an emergency department for evaluation of their isolated third nerve palsy. Group II included 5 cases with isolated third nerve palsy and normal noncontrast head CT at presentation, immediately correctly diagnosed with a PCom A aneurysm at the referring institution. Group III included the 8 remaining cases who all had aneurysms that were missed on noninvasive studies at outside institutions. Review of these outside studies at our institution showed a PCom A aneurysm, confirming misinterpretation of these tests by the outside radiologists, rather than inadequate technique. Absence of specific training in neuroradiology and inaccurate clinical information provided to the interpreting radiologist were associated with test misinterpretation at the outside institutions. The average size of PCom A aneurysms causing an isolated third nerve palsy across all 3 groups was 7.3 mm and was similar in each group.
**Conclusion:** Our study suggests that aside from an accurate history, the training and experience of the interpreting radiologist is probably the most important factor in determining the reliability of a noninvasive scan in patients with isolated third nerve palsies.

**Journal of Neuro-Ophthalmology 2011;31:103–109**
**doi: 10.1097/WNO.0b013e31818f6985**

## Case 7


**"I already diagnosed myself on the Internet. I'm only here for a second opinion."**

## 84 yo white man with

- Bilateral optic nerve edema
- PMHx:
  - Pace-maker (3rd degree block)
  - Afib/anticoagulated
  - Diabetes/mild NPDR
  - CHF
  - s/p cataract surgery – good outcome

## Routine examination optometrist:

- Bilateral optic nerve head edema
  - No headache
  - No visual loss

## Ophthalmologist:

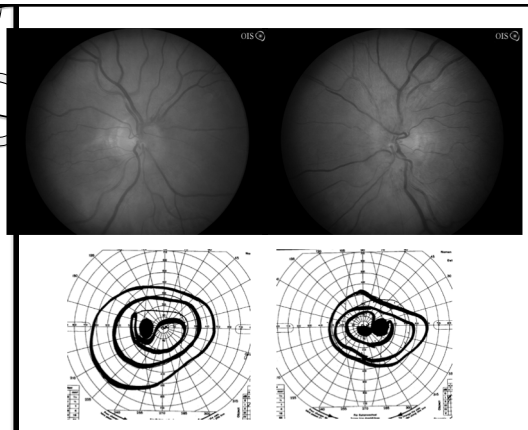
- Bilateral disc edema
- => Head CT normal
- Normal/neg ESR, CRP, CBC, ACE, B12, folate, ANA, Bartonella, RPR

## One month later:

- Mild decreased vision
- Same optic nerve edema
- => Neurologist:
- LP: normal CSF OP, normal contents

## One month later neuro-oph:

| | OD | OS |
|---|---|---|
| • VA | 20/30- | 20/30+ |
| • Color | 12/14 | 13/14 |
| • Pupils | RAPD | |
| • SLE | PCIOL | PCIOL |
| • EOM | Full | Full |



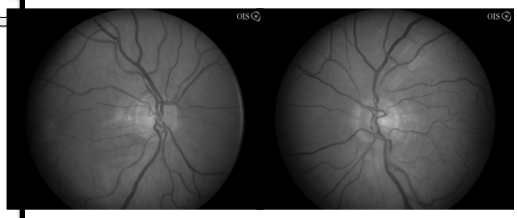
## What are we missing?

- Lipitor
- Coreg
- Synthroid
- Coumadin
- Lisinopril
- Pacerone
- Aspirin
- Nexium
- Metformin

## What are we missing?

- Lipitor
- Coreg
- **Amiodarone**
- Lisinopril
- **Pacerone**
- Aspirin
- Nexium
- Metformin

## One month after d/c amiodarone



## What Went Wrong?

**Mistakes easily repeated when using electronic med record**

- Ophthalmologist's note: medication list copied from ...

**Always double check with patient possible "toxic" drugs**

- ... student
- Neurologist's note: medication ...

**You only find what you are looking for**


- 17 y/o man with anisocoria
- Myelodysplastic syndrome
- S/p bone marrow transplant
- Intubated for respiratory compromise
- Two days after extubation:
  - New onset seizures
  - Anisocoria left pupil larger than right

- **Neurologic consultation:**
  - Dilated left pupil
  - Normal visual acuity and EOMs

- **Ophthalmologic consultation:**
  - Normal visual acuity, intraocular pressures, anterior segment, fundi
  - Normal extraocular movements
  - Normal lids
  - OD pupil normal; OS 8mm and minimally reactive, no RAPD

- **Work-up:**
  - MRI/MRA and LP: all normal
  - Seizures secondary to metabolic derangement

## And now what ?

- **Pupil testing:**
  - **0.1% pilocarpine: no constriction**
  - **1.0% pilocarpine: constriction of OD only**

## Diagnosis

- **Pharmacologic mydriasis**

- Respiratory therapy X 3d:
  - Albuterol sulfate
    - Anticholinergic
  - ipratropium bromide
    - Beta-adrenergic
  - Loose fitting mask – mist escaping to left

## Follow-up

- Respiratory treatments held overnight:
  - Normal pupils next day