Ellerbrock Presents Grand Rounds, Part I

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Headache Eye Pain Vision Loss and a Bloody Nose. What's the Diagnosis?
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Headache, Eye Pain, Vision Loss and a Bloody Nose. What's the diagnosis?

An 87 year old male presents with severe eye pain, headache and epistaxis. Vision measures Hand Motion OU. Mental status quickly declines, and patient begins vomiting blood. Stat head CT scan reveals large intraparenchymal hemorrhage.

I. Case History
   a. Patient demographics- 87 year old white male presents 6-3-2010
   b. Chief complaint-Patient sent from ER physician for urgent consult for severe eye pain OS x 3 days, headache and sudden vision loss.
   c. Medical history
      i. Prostate cancer
      ii. Hyperlipidemia
      iii. Hip arthralgia
      iv. Hypertension
      v. Umbilical Hernia
      vi. Old myocardial infarction
      vii. Hearing loss
      viii. Osteoarthritis
      ix. Occlusion and stenosis of carotid artery
      x. Chronic renal insufficiency
      xi. CVA 7-2008
      xii. Spinal stenosis
      xiii. Cardiac pacemaker
      xiv. Atrial fibrillation
      xv. s/p Cardiac stent
      xvi. Arteriosclerotic Cardiovascular Disease
      xvii. Left heart failure, systolic
      xviii. Abdominal aortic aneurysm
   d. Ocular history
      i. Non-arteritic Anterior Ischemic Optic Neuropathy OD 8-2005

ii. Dry macular degeneration OU
iii. Carotid occlusive disease:
    1. Hollenhorst plaque OS
    2. Peripheral retinal hemorrhages OD
iv. Choroid nevus OD
v. Cataract OD and pseudophake with PCIOL OS

e. Medications
i. Acetaminophen 325 mg tablets. Take 4 tablets po bid
ii. Clopidogrel bisulfate 75 mg tablet qd po
iii. Finasteride 5mg tablet qd po for prostate
iv. Furosemide 20mg tablet qd po
v. Guaifenesin 600mg tablet bid po for mucous
vi. Lisinopril 2.5 mg tablet po qd
vii. Metoprolol 100mg tablet qd po
viii. Nitroglycerin 0.4mg tablet prn for chest pain
ix. Simvastatin 80mg tablet qd po
x. Aspirin 325 mg tablet qd po
xi. Coumadin 1.25mg Sun, 2.5mg 6x/week

f. Other salient information
i. Coumadin prescribed because patient has cardiac stent
ii. INR goal: 2.0-3.0

II. Pertinent findings
a. Clinical
i. Patient presented to eye clinic on a gurney
ii. He denied head trauma. He had a nose bleed since noon that day.
iii. Psychological status: anxiety and severe pain
iv. Neurological status deteriorated during exam. Unable to state day of week or name the US president
v. Vision measured Hand Motion in each eye
vi. Patient developed nausea and began to vomit blood

b. Physical
i. Blood pressure 139/77 at 4pm in ER
ii. PERRL +APD OD-longstanding afferent pupil defect OD
iii. EOM and Confrontation visual field: unable to test
iv. Adnexa/Orbits: normal OU
v. Anterior segment exam with direct ophthalmoscope
    1. Unable to perform slit lamp
    2. Lids/lashes: squinting from pain, mild erythema
    3. Conjunctivae: white, quiet OU
    4. Cornea: clear, no stain OU

5. Irides: grossly intact OU
6. Anterior chambers formed OU: shadow test OD grade 2 and OS grade 4
7. Perkins tonometry @6:00pm
    a. OD 14
    b. OS 15
8. Dilated with 1% tropicamide and 2.5% phenylephrine
9. Examined with BIO and direct ophthalmoscope
10. Limited views OU, poor cooperation, expedited exam
11. Lenses: OD nuclear sclerosis and OS PCIOL
12. Vitreous: grossly normal OU
13. Maculae: grossly normal OU
14. Optic nerve
    a. OD 0.3 temporal pallor trace
    b. OS 0.2 good color
15. Peripheral retinae: flat, intact 360 degrees with no holes/tears OU

c. Laboratory studies

Test name	Result	units	Ref. range	Site Code
PT	>100 H*	SEC	10.0 - 13.8	[405]
INR	>12.5 H*	RATIO	0.8 - 1.2	[405]
PTT	51.2 H	SEC	19 - 33	[405]

Test name	Result	units	Ref. range	Site Code
WBC	7.1	K/cmm	4.5 - 11.0	[405]
RBC	4.25 L	M/cmm	4.7 - 6.1	[405]
HGB	13.0 L	g/dl	14 - 18	[405]
HEMATOCRIT	39.2 L	%	40 - 52	[405]
MCV	92.3	fl	80 - 98	[405]
MCH	30.7	pg	27 - 34	[405]
MCHC	33.3	g/dl	32 - 37	[405]
RDW	14.4	%	11.5 - 14.5	[405]
PLT	238	K/cmm	130 - 450	[405]
MPV	8.3	fl	7.4 - 10.4	[405]
LYMPH %	7.4 L	%	10 - 55	[405]
MONO %	6.7	%	2 - 12	[405]
NEUT %	82.9 H	%	40 - 75	[405]
EOS %	2.6	%	0 - 7.0	[405]
BASO %	0.4	%	0 - 2.0	[405]

Test name	Result	units	Ref. range	Site Code
GLUCOSE	123 H	mg/dL	65 - 100	[405]
Eval: Random and 2HR post-prandial 70-120 mg/dL.				
UREA NITROGEN	25	mg/dL	7 - 25	[405]

CREATININE 1.59 H mg/dl 0.5 - 1.5 [405]
Eval: ***Revised creatinine method/IDMS traceable; started 3/16/2010***
eGFR 41 L mL/min Ref: >=60 [405]
Eval: eGFR= estimated Glomerular Filtration Rate eGFR < 60 ml/min indicates the
Eval: presence of chronic kidney disease. Lower doses of some drugs may be
Eval: needed in patients with eGFR < 60. eGFR<60: associated with increased
Eval: risk of cardiovascular disease. Patients with eGFR <30 should be referred
Eval: for renal consultation.
SODIUM 140 mmol/L 135 - 145 [405]
POTASSIUM 4.3 mmol/L 3.5 - 5.0 [405]
CHLORIDE 109 mmol/L 100 - 110 [405]
CARBON DIOXIDE 21 mmol/L 20 - 30 [405]
CALCIUM 9.2 mg/dL 8.5 - 10.5 [405]
PHOSPHORUS 2.4 L mg/dL 2.5 - 5.0 [405]
Eval: WIDE VARIATION OCCURS WITH MEALS.
MAGNESIUM 2.3 mg/dL 1.8 - 2.4 [405]

d. Radiology studies
    i. Stat Head CT scan

Impression:

1. A 9.3 x 5.2 cm new acute intraparenchymal hemorrhage within the left parietal, left posterior temporal, and left occipital lobes with extension into the left lateral and third ventricles, causing enlargement of the left lateral ventricle, a 14 mm left-to-right midline shift, and significant mass effect in the left hemisphere.

2. Old infarcts in the left frontal and left parietal lobes, also seen on the prior study.

3. Interval development of chronic infarct in the right frontal lobe, not seen on the prior study.

4. No fracture.

e. Other

III. Differential diagnoses

a. Primary/leading-Hemorrhagic CVA-intraparenchymal cerebral hemorrhage
b. Others
    i. Subarachnoid hemorrhage
        1. Terson's syndrome was negative
        2. No intraocular hemorrhage
    ii. Increased intracranial pressure
    iii. Meningitis

iv. Pituitary apoplexy
v. Substance abuse
vi. Malignant hypertension
vii. Migraine
viii. Giant cell arteritis
ix. Acute angle closure glaucoma
x. Ocular ischemic syndrome
xi. Tumor, aneurysm, AV malformation
xii. Epidural or subdural hematoma
xiii. Cluster headache

IV. Diagnosis and discussion
a. Elaborate on the condition-Intracerebral hemorrhage
i. Hemorrhagic CVA accounts for 10-15% of strokes and has a higher mortality rate than ischemic CVA
ii. Headache, altered mental status, seizures, nausea, vomiting, marked hypertension are common features
iii. Brain infarction or hemorrhage can cause pain from pressure on the meninges or increase intracranial pressure
iv. Intense periocular pain can be caused by ipsilateral stroke to occipital lobe
v. Ischemia to cranial nerves III through VI can also cause severe periocular pain
vi. Hemorrhagic CVA will cause visual field loss based upon the location and extent of the stroke
b. Expound on unique features
i. Patient presented with alarming complex of problems
1. Sudden vision loss
2. Severe eye pain and headache
3. Persistent epistaxis
4. Nausea and vomiting of blood
5. Declining mental status
ii. Supratherapeutic INR >12.5

V. Treatment, management
a. Treatment and response to treatment
i. Contacted ER physician and ordered stat head CT scan
ii. Send patient back to ER for imaging
iii. Declining mental status and worsening somnolence noted in radiology and ER
iv. Patient was given IV mannitol in the ER
v. FFP-fresh frozen plasma not available in time before transfer
vi. Vitamin K not given

vii. No Cushings response was noted
    1. Cushing response is increased blood pressure and
    2. Bradycardia due to increased intracranial pressure
viii. Transferred to Dartmouth Hitchcock Medical Center for neurosurgery consult
ix. Notes from Dartmouth indicate that the patient became non-responsive to pain. He was removed from life support and died shortly thereafter on 6-11-2010

b. Refer to research when appropriate
c. Bibliography, literature review

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vi. Hemorrhagic Stroke in Emergency Medicine. Liebeskind DS. http://emedicine.medscape.com/article/1916662-overview Updated 8-17-2011.

VI. Conclusion

a. Clinical pearls : Ocular signs and symptoms may be among the first indicators of an acute intracranial vascular problem that can significantly impact morbidity and mortality. Any patient who presents with acute vision loss, severe headache and worsening mental status requires immediate head CT scan and co-management with an ER physician or neuro-surgeon based upon the findings.

I. Case History

A. 67-year-old Caucasian male presents with a chief complaint of diplopia

1. He noticed the diplopia after a recent VP shunt revision

2. The diplopia is oblique, constant, and present at distance and near

3. When shutting either eye the diplopia disappears

4. He also complains of oscillopsia (things appear to move) especially in upgaze

5. He presents in a chin up position, tipping his head backwards at all times

B. Pertinent medical history

1. Ventricular peritoneal shunt in 2008 for normal pressure hydrocephalus

2. Two subsequent shunt revisions were performed

3. His most recent shunt revision resulted in the symptom of diplopia

C. Prior medical history

1. Mitral valve repair

2. Hypertension

3. Hyperlipidemia

4. Thyroid disease

D. Prior ocular history

1. Last eye exam 10 years ago

2. He wears reading glasses

3. No ocular injury, surgery, or disease either himself self or his family

II. Pertinent findings

- **CLINICAL**

A. Alignment

1. 5 prism diopters of left hyper with eight prism diopters EXO diplopia in primary gaze

2. It is non-concomitant eye turn

a. The left hyper gets larger in left gaze

b. The EXO gets larger both in left gaze and in right gaze

B. Ocular motility

1. Upgaze is absent and eyes are deviated downward called the “setting sun” sign

2. Horizontal gaze shows bilateral adduction deficits and abducting nystagmus, INO

3. Optokinetic nystagmus is absent vertically and slow horizontally

4. Vertical bobbing nystagmus is apparent in both right and left end gaze

5. Convergence is absent

C. Pupils

1. 3 mm and nonreactive to light

2. Pupils react to near better than they do to light

3. This is a light near dissociation

3. No afferent defect is present

D. Eyelids

1. In down gaze the eyelids don't follow the globe

2. This is Collier’s “tucked lid” sign

- **PHYSICAL**

A. His balance is off he is seated in a wheelchair

B. Ataxia

C. Sluggish speech

D. Weakness of lower extremities

- **DIAGNOSTIC**

A. CSF pressures noted as “normal”

B. MRI confirms enlarged ventricles

III. Differential diagnosis

A. Trauma is always a potential source of neurologic injury

B. Infection

C. Inflammation

D. Demyelinating disease

E. Vascular compromise, Stroke

F. Auto-immune

IV. Diagnosis and Discussion

(1) A. Primary findings in Dorsal Midbrain Syndrome

- Vertical gaze disturbance
- Convergence retraction nystagmus
- Light near dissociation of the pupils
- Lid retraction (Collier’s sign)

(2) B. Secondary findings

- Spasm/paresis of convergence
- Spasm/paresis of accommodation
- Pseudoabducens palsy (thalamic esotropia)
- Associated Ocular Motility Deficits
- Skew deviation
- Third nerve palsy
- Internuclear ophthalmoplegia
- See-saw nystagmus

C. Other symptoms may include:

1. Horner's syndrome

2. Motor weakness

3. Loss of balance from vestibular pathway involvement

4. Ataxia

V. Treatment and Management

A. Medical treatment of the source

B. Physical therapy

C. Occupational Therapy

D. Vision

1. Diplopia is addressed with corrective prism and vision rehabilitation

2. Ocular motor therapies are aimed at increasing range of motion

3. Doll's head eye movements can assist with upgaze paralysis in some patients

4. Visual adaptations can be made in one's environment

VI. Conclusion

A. This case highlights the importance of observation of eye movements in neuro-ophthalmic disease diagnosis.

B. It also exhibits associated lid and pupil abnormalities

C. Ocular findings do accompany motor anomalies

1. Ataxia and gait abnormalities are present

2. Localizes to brainstem and pathways

D. Understanding neuro-anatomy is the key to putting together the clinical diagnosis in dorsal midbrain syndrome

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Papilledema Secondary to Sleep Apnea
Jay Rofsky O.D., FAAO

Papilledema Secondary to Sleep Apnea

Jay Rofsky, OD, FAAO

Course Outline

I. Case History
   a. A 44 y.o. Hispanic woman presents with complaints of headaches, dizziness, and transient vision loss with bending over or coughing.
   b. Medical History: Type 2 DM, HTN, Obesity, CHF, sleep apnea; two separate hospitalizations for pneumonia, and H1N1; smokes.
   c. Medications: Glipizide, enalapril, furosemide.

II. Pertinent Findings

a. Exam shows VA 20/20 OD, and 20/25 OS; pupils, EOMs, cover testing, and confrontation fields are normal OU; IOPs are 21 OD, and 22OS; anterior segment exam is unremarkable OU. Blood pressure is 140/88.
b. Posterior segment exam (Images available) shows marked bilateral disc edema, venous tortuosity, and superficial hemorrhage. Vitreous is clear, and retinal exam is otherwise normal OU.
c. Emergent CT is negative. Lumbar puncture reveals normal CSF, as well as opening pressure.
d. While in E.R., patient experiences acute, hypercapnic respiratory failure, and is treated with BiPAP in the ICU, leading to improvement in her condition. She is discharged with CPAP machine.

III. Differential Diagnosis of Papilledema
a. Space-occupying lesion (tumor, hematoma) (6)
b. Meningitis, encephalitis, cerebral edema(6)
c. Cerebral venous sinus thrombosis (3,7)
d. Medications (tetracyclines, vitamin A, anabolic steroids, etc) (7)
e. Sleep apnea (2)
f. "Idiopathic intracranial hypertension" (2)
g. Malignant hypertension (6)
h. Diabetic papillopathy(6)
i. Hyaline bodies (disc drusen) (6)

IV. Diagnosis and Discussion
a. "Sleep apnea" or "Sleep disordered breathing" - Defined as episodic cessation of breathing for at least 10 seconds, or a decrease in airflow with a drop in Hb saturation of at least 4%. (1)
b. Proposed cause of elevated intracranial pressure in sleep apnea:
    i. Nocturnal hypoxemia and hypercarbia causes chronic cerebral vasodilation, which leads to increased intracranial pressure. (2)
    ii. Increased intracranial pressure could collapse of the transverse sinuses, leading to obstructed venous outflow, resulting in continued elevated pressure during the day. (3)

V. Treatment/Management
a. Treatment options for sleep apnea:
    i. Nighttime ventilatory support (CPAP or BiPAP). (1)
    ii. Surgical intervention to open airway. (1)
    iii. Oral appliances. (4)
    iv. Weight loss. Improves mechanical/anatomical factors in airway function.(5,7)

b. Our patient receives CPAP. Since starting, her symptoms disappear. At follow-up exam 6 months later, her exam is normal, and her papilledema is resolved (Images available).
c. References

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VI. Conclusion
a. Sleep apnea is an uncommon cause of papilledema; however it should be included on the list of differential diagnoses.
b. Patients with papilledema should be questioned about any known sleeping disturbances and snoring, and the co-managing physician should be informed.
c. Proper treatment of sleep apnea can be life-changing, in terms of quality of life and reduced risk of morbidity, for the patient.(4)

I Am Looking Up!
Erin Draper O.D., FAAO

AAO 2012 Grand Rounds Outline **Erin M. Draper, O.D., F.A.A.O.**

"I am Looking Up!"

Patient presents with rare transient dorsal midbrain syndrome(DMS). Normal neuro-imaging and serum testing warrants CSF analysis. The presence of oligoclonal bands and MRI spinal cord lesions reveals demyelinating disease. MS seldom presents initially with DMS.

CASE REPORT:

II. Case History

Patient Demographics: - 33 year-old, 5 ft 11 in, 250 lb right-handed man

Chief Complaint:

- Vertical diplopia 5 days ago which resolved within 3 days
- Other symptoms - Eyes now feel constantly strained at distance and near and he is having difficulty focusing to read.

Ocular / Medical History:

- Bell's Palsy 4 months ago affecting the lower left side of his face which resolved within 2 months

Medications:

Took oral prednisolone 4 months ago when diagnosed with Bell's Palsy

Other Salient Information:

At the onset of the Bell's Palsy the patient's primary care doctor ran laboratory testing in the form of a CBC, ESR, Lyme titer, Epstein-Barr virus antibody, and CMV antibody. The results of these labs were remarkable for an elevated Epstein-barr virus IgG antibody (4.68) and an elevated Epstein-Barr virus nuclear antigen antibody (IgG) (2.18). The report indicated that this was consistent with past infection. The patient saw a neurologist two months after the onset of the Bell's Palsy, at which time his symptoms were mostly resolved. No further work-up was ordered and the patient was not asked to return for follow-up.

B. Pertinent Findings:

a. INITIAL PRESENTATION

i. *Clinical:* (photos included in presentation where appropriate)

- VA 20/20 OD and 20/20 OS.
- Ishihara color plates: 14/14 OD and 14/14 OS,
- Pupils equal with no RAPD, but poorly reactive to light. Pupils have a greater reaction to a near accommodative stimulus consistent with light-near dissociated pupils.
- Confrontation fields full bilaterally
- No ptosis or proptosis
- Supraduction deficit with 0% normal supraduction capacity OD and OS. Definite eyelid retraction with convergence retraction nystagmus in attempted upgaze. Ductions full in all other gazes.
- Cover testing at distance and in primary gaze demonstrates an orthophoric horizontal and vertical deviation. On left gaze, there is 2 left right hyper. On right gaze, there is 3 right hyper. Convergence retraction nystagmus is noted on attempted upgaze. On downgaze, there is no deviation noted.
- Slit lamp examination remarkable for a few endothelial deposits OU and mild conjunctiva injection OS. No cells or flare noted in either eye. IOP: normal OU / Blood pressure: normal
- Dilated fundus examination:
  a. Optic discs: distinct margins with no evidence of edema or pallor. 0.4 x 0.4 cupping OD and OS
  b. Macula flat and clear OD and OS
  c. Vitreal opacities OD and OS consistent with old intermediate uveitis and no active inflammation noted.

ii. *Physical:*

- Neurologic examination: cranial nerves V, VII - XII intact
- Some mild left upper and lower extremity weakness noted, but otherwise motor, sensory, and coordination testing unremarkable.

iii. *Laboratory Studies: (INITIAL WORK_UP)*

- Serum
    a. CBC with differential, platelet count, ESR, C-reactive Protein, Lyme Titer, ANA with reflex titer, RPR, FTA-ABS, ACE, toxoplasma serology panel, quantiferon TB Gold In-tube test. All results within normal reference ranges.
    b. Epstien Barr virus antibodies elevated as before, consistent with prior infection

iv. *Radiology Studies:*

- MRI of the brain and orbits with and without contrast – normal
- CT scan of the chest with and without contrast - normal

b. FOLLOW-UP (3 days later)

i. *Clinical:* (photos included in presentation where appropriate)

- No new visual or neurological symptoms since last visit. No episodes of diplopia
- VA 20/20 OD and 20/20 OS
- All afferent system testing same as prior visit, except pupils react somewhat better to light than last visit, but light-near dissociation of the pupils is still apparent. The right pupil is not round, but has area of flattening around the pupillary ruff and has mild sectoral paralysis from 12 o'clock to 3 o'clock.
- Ishihara color plates: 14/14 OD and 14/14 OS,
- Pupils equal with no RAPD, but poorly reactive to light. Pupils have a greater reaction to a near accommodative stimulus consistent with light-near dissociated pupils.
- Confrontation fields full bilaterally; as well as Humprey visual field testing
- Ocular motility testing shows definite improvement with normal ductions, versions, and saccades. No supraduction deficit or eyelid retraction and convergence retraction nystagmus in attempted upgaze.
- Slit lamp examination remains unchanged
- IOP: normal OU / Blood pressure: normal
- Dilated fundus examination remains unchanged with normal optic discs, clear and flat macula and evidence of an old intermediate uveitis with no active inflammation noted.

ii. *Additional Laboratory Studies:*

- Cerebrospinal Fluid
    a. Elevated WBC count at 12-14
    b. Elevated IgG
    c. Greater than 5 oligoclonal bands
    d. NMO antibody testing negative
- Neuro-imaging
    a. MRI of the spine shows multiple enhancing lesions in the cervical spinal cord

III. Differential Diagnoses:

- *Light Near Dissociation of Pupils:*

a. **Dorsal Midbrain Syndrome** – associated with upgaze paresis, convergence retraction nystagmus and eyelid retraction.
    i. Causes: compressive or ischemic injury to dorsal midbrain
        1. Tumor
        2. **Demyelinating Disease**
        3. Stroke
b. Tonic Pupil
c. Argyll Robertson Pupils – associated with miotic pupils; almost pathoneumonic for syphilis
d. Aberrant regeneration of CN III
e. Amaurotic pupil – blind eye

- *Vertical Gaze Palsy*
    a. **Degenerative conditions - MS**, Progressive Supranuclear Palsy
    b. Brainstem injury – compressive, stroke, or hemorrhage
    c. Infectious – Prion disease, Whipple's Disease
    d. Metabolic Disease – Niemann-pick disease
    e. Myasthenia Gravis
    f. Grave's Disease

C. Diagnosis and Discussion

a. Dorsal Midbrain Syndrome Secondary to CNS Demyelinating Disease

i. Ocular manifestations are frequently the first sign of demyelinating disease. Optic neuritis is most commonly associated, but ocular misalignment, pathologic nystagmus, impaired saccades, saccadic intrusions, impaired pursuits, and uveitis are also possible presentations of the disease.
ii. In this case, the remarkable resolution of the dorsal midbrain syndrome signs is characteristic of many early manifestations of MS. Multiple sclerosis is characterized by exacberations and remissions of neurological signs and symptoms separated by time and location.
iii. Dorsal midbrain syndrome as a manifestation of MS is a rare, but should be considered in young patients without an alternative cause.
iv. Pars planitis and periphlebitis occur with a higher frequency in MS patients than in the general population.
v. An MRI of the brain with and without contrast is necessary in the setting of dorsal midbrain syndrome to rule out any compressive, ischemic or hemorrhagic etiologies.
vi. In the setting of normal serum laboratory testing and brain MRI, additional testing of the CSF is necessary to further determine the etiology of the gaze paresis.
vii. Oligoclonal bands IgG are present in the CSF of 75% of clinically definite MS patients.
viii. The absence of white matter lesions in the brain in the diagnosis of demyelinating disease requires additional testing to rule out Neuro-myelitis Optica(NMO). It is important to differentiate between NMO and MS because the treatment and prognosis differs.

D. Treatment / Management:

a. *Treatment / Response:*
- Start Copaxone treatment
- Educated patient on potential visual and ocular symptoms related to multiple sclerosis.

b. *Bibliography / Literature Review*

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IV. Conclusion:

a. Pupils that poorly react to light should be checked for light near dissociation. The presence of LND will help aid in narrowing the differential diagnosis.
b. Even though the patient's symptoms resolved within a few days and all initial work-up was negative, further investigation is necessary.
c. Multiple sclerosis is the cause of a variety of ocular motor disorders in additional to optic neuritis and uveitis. Thus, MS must remain in the differential for many of these cases particularly if the motor disorders are transient in nature and occur in a younger patient.
d. The absence of white matter lesions in the brain in the diagnosis of demyelinating disease requires additional testing to rule out Neuro-myelitis Optica (NMO).

The Case of the Dubious Mathematician
Mika Moy O.D., FAAO

The Case of the Dubious Mathematician
Asymptomatic optic tract glioma caught on routine automated visual field screening. Close involvement with optic tract made removal impossible. Radiation therapy is utilized with serial visual fields thereafter to monitor for progression.

I. Case History
   a. 38 year old white male presents for routine care
II. Pertinent findings
   a. Screening visual field shows OD: infero-nasal non-repeatable defect, OS: repeatable infero-temporal defect
   b. Threshold visual field confirms OD: small infero-nasal defect, OS: larger infero-temporal defect, however, patient is not good at taking visual fields which confounds the diagnosis. Of note: patient states that visual fields are not mathematically sound and have little diagnostic benefit.
   c. Visual field interpretation is non-congruous (left much more than right) homonymous left hemianopsia which can be caused by a lesion in the optic tract.
   d. All other exam findings within normal limits
III. Differential diagnosis
   a. Intracranial space occupying lesion
   b. Glaucoma
IV. Diagnosis and discussion
   a. MRI revealed a partially peripherally enhancing lesion posterior to the pituitary stalk very near the optic chiasm, basilar artery, and hypothalamus.
   b. Biopsy identified the mass as a Juvenile Pilocytic Astrocytoma (JPA) which is a type of glioma that is generally benign with good prognosis.
   c. JPA is one of the most common types of optic tract tumors that tend to form cysts.
V. Treatment and management
   a. Intensity Modulated Radiation Therapy (IMRT) was used as the tumor was deemed too risky to remove due to its proximity to the optic tract.
   b. IMRT uses radiation from many different directions to minimize the amount of radiation on normal tissue while maximizing dosage at the tumor site.
   c. Of interest is that neurology is requesting that we follow the tumor for progression with sequential visual fields rather than sequential MRI due to location of lesion and difficulty in determining growth by MRI alone.
VI. Conclusion
   a. The patient was treated successfully with a six-week course of IMRT.
   b. Visual fields worsened significantly due to radiation therapy, but have remained stable since that time.
   c. Screening visual fields are a useful tool in detecting asymptomatic intracranial masses.

VII. References

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Surprising Complications of Tumor Removal
Stuart Podell O.D., FAAO

Case Report Submission- Academy Meeting 2012

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Abstract: A 50 year old female presents with a chief complaint of diplopia. Extensive examination and radiologic studies MRI studies reveal an intraconal cavernous hemangioma. Surgical removal of the lesion results in complete ptosis. The surgical correction of the ptosis results in inability to close the eye.

I. Case History

- 50 year old female
- Chief complaint of diplopia
- No significant ocular history other than chief complaint
- No significant medical history

II. Pertinent findings

- Clinical
  - Unilateral proptosis
  - Unilateral EOM restriction of gaze
  - Diplopia
  - Unilateral Increased hyperopia
  - Unilateral blurred optic disc margins
- Physical
  - None
- Radiology studies
  - MRI reveals unilateral intraconal cavernous hemangioma
- Differential diagnosis
  - Lesion behind eye
  - Thyroid
  - Diabetes – 3rd nerve involvement
- Diagnosis and discussion
  - MRI studies unquestionably reveal an intraconal cavernous hemangioma impinging on the medial rectus muscle. This impingement is the cause of the restriction of gaze. The lesion is also pressing on the posterior of the globe

causing reduced hyperopia. The lesion is also pressing on the optic nerve causing a blurred disc margin. The space occupying lesion is also the cause of the proptosis.

- The unique features of this growth is that it is causing all of the above clinical findings. The true uniqueness of this case is what occurs after surgical removal of the lesion.

- Treatment management and response
  - The treatment is surgical removal of the tumor. There are two approaches; a fontal approach or a lateral orbitotomy. The surgeon chose the frontal approach. Unfortunately the levator was severed and the patient had no ability to open her eye and was left very disfigured. Pictures of the patient are provided in the presentation. The surgeon attempted a second lid repair to correct the complete ptosis and this resulted in the patient's inability to close the eye. This resulted in exposure keratitis, severe discomfort and further cosmetic disfigurement. The patient sought a second option and this surgeon performed a tutoplast fascia latta sling which allowed the eye to open and close using the frontalis muscle rather than the obicularis. Pictures of the patient at all stages of this process are included in the presentation. As the scar tissue healed the patient has been able to regain limited control of her eyelid, however cosmesis is still an issue. The exposure keratitis has been almost completely resolved and the patient remains on ocular lubricant.
  - Literature review of preferred method of tumor removal is discussed
- Conclusion
  - Unilateral proptosis and diplopia must be investigated carefully
  - Treatment options and risks must be carefully explained to the patient