A Day of Cases

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Financial Disclosure

I have no financial interests.

Opinions in this presentation do not represent the United States Department of Veterans Affairs nor the US Government.
What is this next hour about?

- Cases that I have seen since joining the VA
- Cases that usually made me stop and think
- I’ll share “What I learned”….My takeaways from each case
- We will NOT be talking about Diabetes, Glaucoma, Cataracts, Macular Degeneration or Dry Eye
8:00 Patient

- 69yo White Male complaining of **vertical diplopia** for the past few days
- Med Hx: Diabetes (diet controlled), hypertension, PTSD, Hypothyroidism
- Medications: Metoprolol, Levothyroxine
- Ocular History: Cataracts, No DR (last exam was 5 months previous at another VA)
- VA corrects to 20/25 OD, 20/30 OS
- 1+NS/CS OU
- Nerves, Retina are fine

8:00 Patient – I HATE DIPLOPIA

**FIRST RULE OF DIPLOPIA**

**TALK THEM OUT OF IT**
8:00 pt – I HATE DIPLOPIA

• Why I hate diplopia
  • Kills your groove

• Takes more time, patients are going to wait longer

• Possible huge downside for not getting things right.

4 Categories of causes

- Brain (Cerebral Cortex and Brainstem)
- Cranial Nerves
- Neuromuscular Junction
- Orbit
Diplopia Examples due to Brain Issues

- **Gaze Palsy** - inability to look to one side
- **Internuclear Ophthalmoplegia** - adduction deficit on the side of lesion in the MLF along with a compensating nystagmus in the Abducting eye.
- **One and a Half Syndrome** - Ipsilateral gaze palsy and INO. Only horizontal movement seen is Abduction of the eye contralateral to the lesion.
- **Skew Deviation** - vertical misalignment. Hyper eye is intorted and lower is extorted. Usually due to damage at the brainstem.

**Cranial Nerves**

- **Subarachnoid Space** - CNs are subject to trauma, especially CN IV
- **Cavernous Sinus** - CNs III, IV, and VI converge in the cavernous sinus.
  - If multiple CNs affected, consider orbital apex or cavernous sinus lesion such as neoplasm, carotid artery aneurysm, inflammation, fistula or thrombosis.
  - May complain of pain if CN V is affected as well.
Diplopia 2/2 Cranial Nerves

- Isolated CN palsies in patients over 50 are often due to microvascular ischemia
  - Diabetes, hypertension
  - Resolve in three to six months
- Imaging?
  - "A recent study found that overall 16.5% of adult patients presenting with acute ocular motor mononeuropathy had structural lesions on MRI scan and 4.6% with fourth and sixth nerve palsies and no risk factors were found to have positive MRI scans."
- Lab work up
  - ESR, CRP, CBC

Diplopia 2/2 Neuromuscular Junction Issues

- Myasthenia Gravis
  - Acetylcholine can't fuse to its receptors
  - Patients may present with ptosis and ocular misalignment
  - In-office tests
    - Orbicularis weakness
    - Ice-Pack
    - Eyelid fatigue
Diplopia 2/2 Orbit

- Exophthalmometry and forced duction testing
- Orbital Fractures
- Mucocele
  - When there is scarring and obstruction of the sinus ostium, a mucocele can develop
- Thyroid Eye Disease

8:00 Patient – BACK TO THE CASE

- What is your in-office diplopia work-up?
  - Ductions
  - Cover test in all directions of gaze
    - Comitant or non-comitant
  - EOMs
  - Vertical Diplopia
    - Park’s 3 Step: [https://www.eyedock.com/parks-3-step](https://www.eyedock.com/parks-3-step)
The Diplopia Evaluation

• Monocular vs Binocular
• Measure Ductions and Saccades
  • Check ductions monocularly and binocularly
• Is it Comitant vs non-comitant
  • Comitant is usually a non-paralytic cause
  • Non-comintancy of greater than 5pd can indicate paralytic or restrictive cause
• Phoria vs. Tropia
  • Cover test, Maddox Rod, Red Lens

8:00 Patient – THE EXAM

• EOMS: Full and smooth, OD, OS, and OU
• Alignment: Red Lens Test: 6pd Base Down OD
• Parks Three Step: Left Inferior Oblique Palsy
• https://www.eyedock.com/parks-3-step
8:00 Patient – I HATE DIPLOPIA

Orders: ESR, CRP, MRI, MRA, A1C
Results: Posterior Circulation Infarct (Ischemic Stroke)
Treatment: Prism and observation

Final Diagnosis: Left Inferior Oblique Palsy secondary to microvascular ischemia

What I learned:

• It is important to have a consistent, step-wise approach
• Observation and case history can point the way
8:30 Patient

47yo White Male

Med Hx: Lupus, Hyperlipidemia, Sleep Apnea, DVT, Sensorimotor Dysfunction

Medications: Zolpidem, Tramadol, Lacosamide, Atorvastatin, Warfarin, Topiramate,

Ocular History: Myopia

8:30 Patient – NOT AGAIN

CC: Diplopia OU x 2mos. Horizontal Diplopia. Episodes are brief but occur several times a day.

VA corrects to 20/20 OD, OS c -2.00 sph

Cover test is ortho

Distance Phoria testing 1BI horizontal, ortho vertical

Internal and external ocular health are unremarkable
8:30
Patient – NOT AGAIN

• What could the cause be?
  • Not ocular alignment
  • Not Lenticular or Retinal

• Looking back at medications
  • Medications: Zolpidem, Tramadol, Lacosamide, Atorvastatin, Warfarin, Topiramate

Anti-Epileptic Drugs (AEDs)

• How do they work?
  ○ By Affecting Ion Channels in the cell membrane
    ■ Sodium
    ■ Potassium
    ■ Calcium
    ■ Chloride
  ○ By Altering neurotransmitters in the Synapses
    ■ GABA (Gamma-Aminobutyric Acid)
    ■ Glutamate
Anti-Epileptic Drugs (AEDs)

- Visual disturbances are a common side-effect of many antiepileptic drugs.
- Retino- or Neurotoxic visual abnormalities such diplopia, blurred vision, and nystagmus
- Over dosage or prolonged use
- Some anticonvulsants are associated with specific visual problems that may be related to the mechanistic properties of the drug, and occur even when the drugs are administered within the recommended daily dose
AED Examples

- Acetazolamide
- Brivaracetam
- Cannabidiol
- Carbamazepine
- Clobazam
- Clonazepam
- Eslicarbazepine acetate
- Ethosuximide
- Everolimus
- Gabapentin
- Lacosamide
- Lamotrigine
- Levetiracetam
- Oxcarbazepine
- Perampanel
- Phenobarbital
- Phenytoin
- Piracetam
- Pregabalin
- Primidone
- Rufinamide
- Sodium valproate
- Stiripentol
- Tiagabine
- Topiramate
- Valproic acid
- Vigabatrin
- Zonisamide

Vimpat® (Lacosamide)

- AED given either orally or by IV to reduce the frequency of seizures
- Common Side Effects include diplopia, dizziness, headache, and nausea.
8:30 Patient – NOT AGAIN

- Upon questioning, diplopia symptoms correlated with patient starting Lacosamide.
- Refer

What I learned:

- If EOMS and alignment are good, look at the patient’s medications to find possible causes of diplopia
What I learned: Medications that cause diplopia

<table>
<thead>
<tr>
<th>Very Common</th>
<th>Common</th>
<th>Uncommon</th>
</tr>
</thead>
<tbody>
<tr>
<td>Lacosamide</td>
<td>Botulism toxin</td>
<td>Amlodipine</td>
</tr>
<tr>
<td>Zonisamide</td>
<td>Pregabalin</td>
<td>Pravastatin</td>
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<td></td>
<td>Sildenafil</td>
<td>Lamotrigine</td>
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<td></td>
<td>Gabapentin</td>
<td>Sertraline</td>
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<td></td>
<td>Topiramate</td>
<td>Ciprofloxacine</td>
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9:00 patient

- 70-year-old, white male
- CC: decreased VAsc and cc OU at near and far
- Med Hx: Hyperlipidemia, Supranuclear Palsy, Hypothyroid
- OC Hx: unremarkable
9:00
patient – You have what?

- VA corrects to 20/40 OD, 20/50 OS
- EOMS: restricted on down gaze
- Cover Test: unable to accurately obtain
- Significant ocular findings: 2+ Nuclear Sclerotic Cataracts

What is Progressive Supranuclear Palsy (PSP)???

- Progressive supranuclear palsy, also called Steele-Richardson-Olszewski syndrome, is an uncommon brain disorder that causes serious problems with walking, balance and eye movements. The disorder results from deterioration of cells in areas of your brain that control body movement and thinking.
Progressive Supranuclear Palsy

- Supranuclear disorders result from lesions above the level of the ocular motor nerve nuclei. If oculocephalic maneuvers move the eyes appropriately, the lesion causing the gaze palsy is supranuclear.
- Supranuclear disorders account for almost 10% of all patients with disorders of eye movements.
Progressive Supranuclear Palsy

- Eye movement abnormalities of supranuclear origin are characterized by gaze palsies, tonic gaze deviation, saccadic and smooth pursuit disorders, vergence abnormalities, nystagmus, and ocular oscillations.
- Apraxia of lid opening, blepharospasm, abnormal fixation, decreased saccadic function, reduced reading speed, vertical supranuclear gaze palsy.

OD’s role in treatment of PSP

- Proper referrals, such as neurology and/or physiotherapy due to fall risks.
- Best refraction, consider prism for tasks such as reading and eating.
  - Try to bring their world up!
- Single Vision only.
- Ocular surface disease secondary to decreased blink rate.
9: 30 – Eye Pain

• 59yo white male
• CC: Eye Pain OD x 1-2 days, +photophobia, +water discharge, +decreased vision
• Oc History: Dry Eye
• Ocular Meds: Art Tears PRN
• Med Hx: Diabetes, Hyperlipidemia
• A1C: 9.6 one month earlier

9:30 – Eye Pain

• Vasc 20/250-, NIPH OD
• OD: LL edema, ectropion. 1+mucopurolent discharge
• OD: 4+ conj chemosis inferiorly
• OD: 3+ corneal edema
• AC: 2mm hypopyon
9: 30 – Eye Pain – Day 1 treatment

- Durezol q2h
- atropine bid,
- moxifloxacin q1h
- Keflex 500mg po bid

9: 30 – Eye Pain, 2 Day follow-up

- Vasc 20/300, NIPH  OD
- Using all drops except Atropine
- Pt feels like it looks better
- Pain is improved
9: 30 – Eye Pain, 2 Day follow-up

- OD: LL edema, ectropion. 1+ mucopurulent discharge
- OD: 3+ conj chemosis inferiorly
- OD: 5mm corneal abrasion and stromal folds
- AC: 1mm hypopyon
9: 30 – Eye Pain, 2 Day follow-up. TREATMENT

- Inserted BCL Air Optix Night and Day
- Change Durezol to q4h OD
- Atropine bid OD, if able to tolerate
- Moxifloxacin q4h OD
- Keflex 500mg bid po x 10d
- Art tears q1h/prn

9: 30 – Eye Pain, Summary

- Day 5 – hypopyon was gone
- Day 14 – abrasion 75% healed
- 1month – SPK only
9: 30 – Eye Pain; What I learned

• Reasons for Hypopyon
  - endophthalmitis, infectious corneal ulcer, severe iridocyclitis, retained intraocular foreign body, intraocular tumor necrosis, recurrent corneal erosion, drugs (eg, rifampin), leukemia and can be seen post cataract surgery – sterile or infected due to device contaminant

• Bell’s Palsy is associated with Diabetes Mellitus
  - Approximately 11% of Bell’s Palsy Pts have DM
  - Studies have shown recovery and facial movement score is slowed in diabetics

10:00 pt

• 90yo African-American, male, NEW patient.
• CC: decreased nva OU
• Med Hx: Hypertension, Emphysema
• Oc Hx: Pseudophakia OD, Cataract OS
10:00 pt – Where did it go?

- MRx: +0.5 sph OD 20/40+, NI CF@4ft OS
- IOP: 17/19
- Anterior Segment: wnl
- Posterior Segment…..

RIGHT EYE
LEFT EYE

10:00 pt - B: Scan

- Diagnosis for this patient
  - Decentered IOL OD
  - Luxated Lens OS
OD: IOL dislocation Categories

- Decentered within an intact capsular bag
- Partially luxed: one haptic in the bag, one out
- Lens in the ciliary sulcus
- Lens in the bag, but both bag and lens are subluxated and decentered

OD: IOL dislocation Causes/ Risk Factors

- Trauma
- Pseudoexfoliation Syndrome
- Previous Vitreoretinal surgery
- Increased Axial Length
- Retinitis Pigmentosa
- Uvieits
OD: IOL dislocation Treatment

- Observation
- Spectacle rx change
- Lens Exchange
  - New PCIOL
  - Iris Sutured
  - Scleral Fixated
  - ACIOL

OS: Luxated Lens

- Ectopia lentis is a displacement or malposition of the eye's crystalline lens from its normal location. A partial dislocation of a lens is termed lens subluxation or subluxated lens; a complete dislocation of a lens is termed lens luxation or luxated lens.
OS: Luxated Lens Causes

- Trauma
- Glaucoma
- Uveitis
- Tumors
- Cataracts
- Marfan syndrome
- Homocystinuria
- Ehlers-Danlos syndrome
- Hyperlysinemia
- Pseudoexfoliation Syndrome

OS: Luxated Lens Complications

- Increased Intraocular Pressure
  - Vitreal Prolapse into the anterior chamber
- Uveitis
- Retinal Breaks
- Hypermature lens
OS: Luxated Lens Treatment

- Optical Correction
- Treatment of underlying condition
- Surgical
  - Lensectomy/vitrectomy with aphakic contact lens or spectacles
  - Iris-fixated intraocular lens
  - Scleral-sutured posterior intraocular lens
  - Implantation of in-the-bag intraocular lens with a capsular tension ring

10:00- Where did it Go?

What I learned

- Observation is an option
- Multiple surgical techniques to reposition or replace a dislocated IOL.
59yo WF presents with a painful left eye
- Pain is a 9/10, starts from left brow and goes through her temple and down the back of her neck.
- Can't open her eye

Med Hx: substance abuse, ADHD, recently hospitalized with pneumonia
OC Hx: Chronic blepharitis
Allergies: tetracycline, erythromycin, neomycin, bacitracin, formaldehyde, lidocaine, tacrolimus, benzoyl peroxide
VA: 20/30 OD, LP OS
No APD by reverse. Unable to see OS
Bullous Keratopathy OS
Treated with Durezol qid, NaCl 5% qid, atropine bid
Prompt referral
• Assessment
  • 1. Bullous Keratopathy
  • 2. Previous corneal perforation; gave BCL, con't meds. Rheum referral. Pt referred to Duke Eye

• Plan:
  • 1. BCL OS, Rheumatology Referral
  • 2. Continue gtt.
  • 3. Send to Duke Eye

• Rheum work up came up empty.
• Pt started on oral acyclovir
1030pt – Corneal Perforations

- Causes: Trauma, Herpetic Infection, Inflammatory conditions, Dry Eye
- Treatment: BCL for perforations less than 3mm, Surgery, Human Fibrin Glue
- Adherent Leukoma may develop
  - corneal scar which has fibrous tissue adherent to its deeper surface. It always indicate a perforation unless an adherent leukoma of congenital origin.

11:00 Pt

- 70yo AA male
- 1st present to clinic in 2018 with “long-standing decreased VA OD” of unknown etiology
- Med Hx: DM, HTN, OSA, Obesity
- BVA: 20/80 OD, 20/25 OS
- Pupils: 1+ APD OD
- CF: UTO due to poor fixation
Optic Atrophy Diff Dx

- **Compressive** – secondary to papilledema, tumor, bony growth (fibrous dysplasia, osteoporosis), thyroid eye disease, chiasmal (pituitary etc), optic nerve sheath meningioma, disc drusen, increased intraocular pressure (glaucoma)
- **Vascular** – arteritic and non-arteritic ischemic optic neuropathy, diabetes
- **Inflammatory** – sarcoid, systemic lupus, Behcet’s, demyelination (MS), etc.
- **Infectious** – viral, bacterial, fungal infections - herpes, TB, bartonella, etc.
- **Toxic & nutritional** – many medications such as ethambutol, amiodarone, methanol, vitamin deficiency etc.
- **Metabolic** – diabetes
- **Neoplastic** – lymphoma, leukemia, tumor, glioma
- **Genetic** – Autosomal dominant optic atrophy (OPA1), Leber's hereditary optic neuropathy, Leber's hereditary optic neuropathy, as a late complication of retinal degeneration.
- **Radiation optic neuropathy**
- **Traumatic optic neuropathy**
• What to do?
  • Long-standing….so do I need to do anything?
  • Blood work?
    • Glucose, ESR/CRP, B12, CBC
  • Chest X-ray
    • If respiratory symptoms, think Sarcoid
  • Imaging
    • MRI of brain and orbits

• What I did
  • DFE, OCT, Fields, Photos
  • Reviewed labs
    • A1C = 6.5
    • CBC = normal
    • HDL = High
    • Folate = High
  • Observation only is what I chose
    • Monitor as a glaucoma suspect. Large CDs and monocular patient
What I did

- Pt returned 2019
- VA OD: 20/100, OS: 20/20
- IOP: 13/14
- DFE/OCT: unchanged
- Ordered MRI of Brain and orbits

“Constellation of findings suggesting a diagnosis of multiple sclerosis with multiple bilateral periventricular/callosal periventricular T2 and FLAIR hyperintensities with additional focus of hyperintensity within the left middle cerebellar peduncle. There is segmental T2 and FLAIR hyperintensity of the right optic nerve could represent sequelae of prior optic neuritis. There is no abnormal enhancement to suggest active demyelination. Consider further imaging of the spinal cord if clinical concern dictates.”
If he does have demyelinating disease, it is mild and inactive. We discussed that optic neuropathy has many causes and does not always result in a diagnosis of MS. At this time, I recommend clinical surveillance as well as repeat MRI in 1 year. If the imaging is stable at that time, he will not need to follow up long term in this clinic.

11:00 Pt – What I learned

- Rule out potential serious causes of Optic Atrophy, even if long standing
- Life is messy, optometry can be VERY messy
  - Labs and positive findings may not be conclusive
1300 PT

- 43yo, white, female
- h/o optic neuritis secondary to MS

1300 PT - History

- April 2012 complains of “objects swimming across field” OD
  - Dx w/ Floaters
- May 2012 VF Defect OD – thought to be retrobulbar optic neuritis
- MRI scans showed concern for progression to MS, but nothing definitive
  - Followed annually by Neurology with MRI
1300 PT - History

- 2018
  - “Normal MRI of the orbits. 2. Small nonspecific, nonenhancing foci of T2 hyperintensity in the right frontal lobe white matter. The differential is broad including sequelae of old inflammatory/infectious etiologies, migraine-related white matter change, and possibly demyelination. Consider follow-up MRI of the brain without and with IV gadolinium in one year for further evaluation.”
1300 PT

- VA: 20/20 OD, OS
- Pupils: ERRL (-) APD
- CF: OD – sup-temp restriction, OS: full
- IOP: 11/13
- CD: 0.6round OU without pallor
1300 pt – ONH vs Retina

- Does this case make sense?
  - ONH and rNFL look healthy
  - Visual field is stable in the OD
  - Could this be retinal??
1300 PT – What is AZOOR?

• Acute zonal occult outer retinopathy (AZOOR) is a presumed inflammatory disorder with outer retinal dysfunction
• Acute and unilateral
• Symptoms include photopsias and nasal field loss; scotoma is usually contiguous with the optic nerve.
• Later, the other eye is involved in nearly three fourths of patients

• A diagnosis of AZOOR should be suspected in cases of loss of one or more zones of visual field, particularly when associated with photopsia, absence of discomfort with eye movement, relatively spared visual acuity and without funduscopic explanation for visual loss
• Electrophysiology should be carried out to differentiate AZOOR from optic neuritis and other lesions affecting the posterior visual pathways
• Patients with AZOOR showed a pattern of visual dysfunction that was photoreceptor in origin.
What is AZOOR?

- “Fundus: May be normal in the beginning but may show a grayish-white line at the border of normal and involved retina, usually in peripapillary area. This line disappears within weeks and is replaced with an orange zone. With time, retinal vessels attenuate and a large zone of retinal pigment epithelium (RPE) depigmentation appears, sort of a sector retinitis pigmentosa (RP) or unilateral or asymmetric RP.”


May be part of the spectrum of “White-Dot Syndrome”

- Acute idiopathic blind spot enlargement (AIBSE) syndrome
- multiple evanescent white dot syndrome (MEWDS)
- Multifocal choroiditis
- punctate inner choroidopathy
- acute posterior multifocal placoid pigment epitheliopathy (APMPPE)
- serpiginous choroiditis
- Birdshot retinochoroidopathy (HLA-A29 associated in 95% of cases)
- Presumed ocular histoplasmosis syndrome (POHS)
13:00 Pt – What I learned

- Pretty much everything I just talked about regarding AZOOR
- AZOOR is associated with demyelinating white mater lesion and multiple sclerosis
- Relatively new disease; first reported in 1992
  - Not fully understood.