Case history

- M.C., Caucasian female, 46 y.o.
- Computer worker
- Referred for presumed corneal warpage 2nd to continuous wear of Focus N&D™

Subjective

- Had ceased to wear c.l. x 3 wks
  - After her OD's recommendation
- Chief complaint with c.l.:
  - visual fluctuation at far
  - night driving
  - occurrence: several months ago
  - increased over time

Non compliance history

- Admit to overwear
  - 2 months / pair, even more sometimes
- Fitted in 2000 with Focus N&D™
  - BC 8.4 ; od -2.25 OS -2.00
  - presumed monovision; OS dominant at far
- Has no glasses for back-up

Subjective (2)

- General health: good
- Medication: Venlafaxine (Effexor™)
  - antidepressant and anxiolytic medications
  - 75 mg once daily
  - abnormal vision as possible side effects
- Family Backgrounds:
  - Ocular and systemic : nothing to report
Objective

- UCVA: 20/200 OD, OS, OU
- Preliminary and pupil tests: WNL
- BCVA
  - OD -2.50 -1.00 x 180 (20/20)
  - OS -1.50 -1.25 x 175
- No previous findings available except c.l. (-2.25/-2.00 ; SE??)

Topo map – OD

- Corneal asymmetry
  - Ks: 44.1 x 44.6
  - E value: 0.61@127 ; 0.82 @37

Alien profile

- More severe corneal asymmetry
  - Ks: 43.2 x 44.6
  - E value: 0.66@85 ; 0.89 @175

Skull head profile

- SLE: unremarkable
- No microcysts, nor neovascularization
- Grade 1 papillae upper palpebral conj. OU
- Fluoresceine: negative – no SPK
- Dilated fundus: normal
- IOP: 12 mm Hg. O.u. (compensated for 560 um corneal thickness)

Assessment

- Corneal warpage secondary to contact lens wear
- Forme frustre keratoconus (FFK)
- Congenital corneal asymmetry
- Side effects of medication

DDX

- Data collected 3 wks after lens withdrawal
  - Are they the « real picture »?
  - In case of warpage, corneal changes occur mostly from 3 wks up to 6 months
- No previous data available (congenital)
- Symptoms vary (night/day):
  - not compatible with medication side-effects
- Warpage vs FFK: only time will tell us
Plan

- Already fitted with highest DK
- Items to improve
  - Modulus
  - Continuous wear
  - Non compliance
  - Glasses for back-up (not at this time)

Plan 2

- Temporary fit: Oasys for astigmatism OU
- Daily wear, 2 wks disposable, with Clear Care
- BCVA of 20/20-2 OD and 20/25 +2 o.s.
- Position and movement : optimal

Follow-ups

- It took 4 months to stabilize the cornea
- Final refraction
  - OD -2.50 -0.75 x 10 (20/20)
  - OS -2.25 -0.75 x 5 (20/20)
- AOA prescribed, 2wks disposable, DW
  - OD -1.75 -0.75 x 10
  - OS -2.25 -0.75 x 180

Final topo maps

- More symetrical
- Regular toric pattern

- K values
  - OD 43.25 x 43.87
  - OS 42.5 x 43.25
- Definitively not FFK

Discussion

- Visual fluctuation can come from
  - Surface irregularity
  - Ocular dryness
  - Uncorrected astigmatism
  - Media opacities
  - Systemic /drug induced

Discussion

- Corneal warpage can occur from
  - Chronic Hyposia
  - Bad contact lens fit (RGP, high modulus, distorted lens, etc)
  - Eye rubbing
  - Ocular surgery
Conclusion

- DK/t is not the end of the story
- Corneal warpage can occur in soft and High DK Si-Hy lenses
  - Lower modulus lenses can help
- Management of these cases can take up to 6 months
  - Should be addressed for immediate (BCVA, discomfort) and future needs (refractive surgery, cataracts)

A Visual Field Defect In a Patient With Small, Tilted Optic Discs and Disc Drusen: Which One is Responsible For the Defect?

Benjamin P. Casella, O.D.

Disclosure Statement

- Nothing to disclose

The Game Plan

- Case presentation
- Some differentials
- What do you think it is?
- What do I think it is?
- Conclusion

Vita

Private Practice (Augusta, GA) 2008 – Present
SUNY State College of Optometry (Residency) 2008
UAB School Of Optometry (O.D.) 2007
University of Georgia (B.A.) 2003

26yo Asian Female

- Presents for routine follow-up
- Self-referred with existing diagnosis of retinal hemangioma OD
- Lasered by retinal surgeon (4 months out)

26yo Asian Female

OHx / MHx
- Hemangioma is well-lasered and stable
- Pt states had w/u for von Hippel-Lindau
- Pt diagnosed with disc drusen at time of laser
- Meds: Ortho Tri-Cyclen
- Allergies: Benzoyl Peroxide
- PMH: unremarkable
- FMH: unremarkable
Exam
- VA: 20/20 OD/OS
- Pupils normal
- IOP: 12 OD/OS
- Anterior segments unremarkable
- Fibrosis and exudate within area lasered OD
- Retinas intact and stable: rt x 4 months or pm

Disc Photography

B-Scan
- OD: Low Gain
- OS: Normal Gain

Baseline Visual field
GDx

Back To the Field Defect

Inferotemporal OS (Causes of a VF Defect)
- Glaucoma?
- Disc Drusen?
- Optic Pit?
- Tilted Disc?
- Optic Nerve Hypoplasia?
- Neurological?

Glaucoma?
- Nature and location of the visual field defect
- Nature and location of the RNFL defect
- ONH appearance
- Possible?
- Probable?

Optic Disc Drusen?
- Nature and location of the visual field defect
- Disc appearance
- B-scan
- Buried vs Exposed
- Possible?
- Probable?
- Risks (CNVM)

Optic Pit?
- Nature and location of the visual field defect
- Disc appearance
- Possible?
- Probable?
- Risks (serous detachment)

Tilted Disc?
- Nature and location of the visual field defect
- Disc appearance
- Bilaterality of defects?
- Possible?
- Probable?
Optic Nerve Hypoplasia?
- Nature and location of the visual field defect
- Disc appearance
- Visual acuity?
- Double ring sign?

Optic Nerve Hypoplasia
- ~1.3 : 1000
- Variable presentation
- 20/20 to severely visually impaired
- Visual field defects common
- Bilaterality common
- Males = Females
- Measuring considerations

Optic Nerve Hypoplasia
- Can be related to midline abnormalities in the brain (S-O-D)
  - Absent septum pellucidum, pituitary dwarfism, corpus callosum may be absent or thinned.
- Fetal Alcohol Syndrome
- Can be associated with hormone imbalances
- Can be sectoral
- Nature of the VF defect
  - Depends on each optic nerve, itself

Summary
- Abnormal Discs
- Small Discs
- Look for structure / function abnormalities
- This patient, like others with optic nerve head anomalies, deserves a baseline visual field and ONH analysis
Summary

In the age of ONH imaging, visual fields are still:

- Necessary
- Challenging
- Fun (?)

I’m not a Fellow in the American Academy of Optometry…

…I did stay at a Holiday Inn Express last night.

Questions?

Neuromyelitis Optica Diagnosis via Serum Autoantibody Marker

Tina R. Porzukowiak OD, FAAO

Disclosure Statement
- Nothing to disclose

HPI

- 55 y.o. Mexican female
- ER consult
- CC: gradual onset, constant, throbbing retrobulbar pain OS (5/10) w/ eye movement associated w/ a sudden onset of central blurred vision OS; OD asymptomatic.
**OHx**
- No h/o previous ocular examination
- Presbyopia OU (+2.75 OTC readers)
- Negative for trauma, surgery, or eye disease
- FOHx: negative

**MHx**
- HTN
- Primary hypothyroidism
- Diverticulitis, s/p partial Lt colectomy
- s/p partial hysterectomy
- Herpes Zoster
  - Dx 1.5 wks prior to ER presentation @ outside medical facility
  - Rash high Lt thoracic dermatome spread to Lt upper back, shoulder, & neck

**Meds**
- Self-d/c HTN med
- Levoxyl (intermittent use)
- Acyclovir
- Prednisone
- Endocet prn
- Allergies: Sulfa (unknown rxn)

**SHx**
- Tobacco: negative
- ETOH: occasional beer
- Drugs: negative
- Orientation: P/P/T
- Mood: normal

**Vision Exam**
- **BCVA:**
  - OD: 20/20
  - OS: 10/600—NI (Feinbloom DFV Number Chart)
- **Refraction:**
  - OD: pl sph 20/20
  - OS: pl sph 10/600
  - Add: +2.75 20/20
- **P: ERL, 3+ L APD**
- **EOMS: FROM OU, +pain all gazes OS**
- **CF: FTFC OD, central blur OS**
- **CT: ortho**

- **Ishihara CV:**
  - OD: 12/12
  - OS: 0/12
- **Red cap: 95% desaturation OS**
- **Amsler grid:**
  - OD: NL
  - OS: central blur w/o metamorphopia
Vision Exam

- Physical:
  - Negative for lymphadenopathy
  - Negative tenderness on palpation of temporal arteries with strong pulsation bilaterally

- CN / Neuro Exam:
  - III-XII intact
  - Dysesthesia Lt V₁ & V₂ dermatomes
  - Unsteady tandem gait

Vision Exam

- SLEx:
  - L/L: NL OD, OS
  - Conj: W/Q OD, OS
  - K: clear OD, OS
  - Iris: flat, brown OD, OS
  - A/C: D/Q OD, OS
  - TA: 21 mmHg OD, OS @ 1:19 pm

Vision Exam

- DFE: 1% M, 2.5% P OD, OS; 78D/20D/BIO
  - Lens: tr NS OD, OS
  - ONH: 0.30 w/ healthy rims OD, OS
  - Mac: NL OD, OS
  - Vit: clear OD, OS
  - Ves: NL OD, OS
  - Peri: flat w/o breaks OD, OS

GVF

DDx

- RBON, unknown etiology
- Posterior scleritis, unknown etiology
- Retrobulbar mass

Plan

- MRI brain & orbits w/ & w/o contrast STAT
- Neuro consult
**MRI brain & orbits w/ & w/o contrast**

**Neuro consult results**
- III-XII intact
- Dysesthesia Lt V₁ & V₂ dermatomes
- Motor, sensory, reflex testing: NL
- +RUE tremor
- Unsteady tandem gait
- Romberg sign: neg
- Physical: no evidence of rash; 2 red, papules behind the hairline along the Lt posterior skull

**Neuro Plan**
- Hospital admittance
- ID Consult
- Meds:
  - Metoprolol 50 mg bid
  - Levoxyl 0.1 mg qd
  - CPM x HZ
- Lab w/u:
  - CBC w/diff
  - Serum chem
  - PT
  - INR
  - ESR
  - CRP
  - ACE
  - ANA
  - c-ANCA
  - p-ANCA
  - RI
  - CXR
  - MRA

**DDx**
- Giant cell arteritis
- Neoplastic dz
- Vasculitis dz
- Neurosarcoidosis
- Collagen vascular dz
- Wegener’s granulomatosis
- Carotid artery dissection
- Disseminated herpes zoster
- Multiple sclerosis

**Neuro Plan**
- LP w/ opening pressure
- Glucose
- Protein
- Cytology
- RBC
- IgG
- PCR w/ infectious screen x viral, bacterial, and fungal etiology
- OCB
- MBP
- Lab w/u
  - Lyme titer
  - TFT
  - HIV
  - HbA1c (↑ 6.6%)
  - RPR
  - Dermatology consult
  - Meds:
    - IV acyclovir
    - IV methylprednisolone
    - Insulin

**DDx**
- Disseminated HZ
- MS
- Meningitis
- Lupus
- HBV
- HSV
- HIV
- Syphilis
- Lyme dz
- Cryptococcosis
- Coccidioidomycosis
- Mycoplasma pneumonia
- Aspergillosis
- Sarcoidosis
- Cerebral vasculitis
- Neoplastic dz
ID consult results
- 1 mm papules on Lt upper shoulder w/o rash
- Working dx: Presumed disseminated HZ sine herpete w/ associated LL RBON
- Meds: IV acyclovir 10 mg/kg/d x 14d

Dermatology consult results
- Linear hyperpigmentation on the Lt upper posterior shoulder thought to represent post-inflammatory hyperpigmentation associated w/ HZ.

Neuro-ophthalmology Grand Rounds
- 6 days s/p ER presentation
- CC: gradual worsening of VA OS, but less pain (1/10) with eye movement; asymptomatic OD.
- BCVA:
  - OD: 20/20
  - OS: HM@1'
- Ocular examination unchanged

Neuro-ophthalmology Plan
- Repeat MRI orbits
- PPD
- Serial DFE's

MRI orbits w/ & w/o contrast

Neuro F/U & Plan
- Weakness Lt arm & leg
- MRI cervical spine
  - No focal cord lesions
  - DDD C5-6, C6-7
ID Plan

- Meds: Oral Acyclovir 800 mg 5x/d x 7d
- Hospital Discharge: 16 days post presentation

Vision Exam – Out-patient

- 19 days s/p ER presentation
- Unchanged examination
- Persistent LUE weakness

Vision Exam – Out-patient

- 26 days s/p ER presentation
- BCVA:
  - OD: 20/20
  - OS: 5/180
- Unchanged examination

Vision Exam – Out-patient

- 43 days s/p ER presentation
- BCVA:
  - OD: 20/20
  - OS: 20/160
- 1+ diffuse ONH pallor OS
- Improved GVF

Fundus photos

GVF
Emergency Room, Part II

- s/p 46 days from initial ER presentation
- CC: low grade fever, chills, mild HA, & diffuse myalgia across chest & back
- W/U:
  - CBC w/ diff
  - Chemistries
  - Glucose (↑ 182 mg/dl)
- Dx: viral syndrome suspect URI
- Tx: Tylenol prn, ↑ fluid intake, CPM
- Gen med consult x diabetic control

Emergency Room, Part III

- s/p 49 days from initial ER presentation
- CC: 3-day h/o N/V & hiccups; no BM x 4 days
- W/U:
  - CBC w/ diff
  - Abdominal X-ray
  - Abdominal CT
  - CAR
  - UA
- Hospital admittance x dehydration
- Tx:
  - Promethazine HCl 12.5 mg po q6h x N/V
  - NaCl 0.9% injections x electrolyte stability
  - Fleet’s enema x constipation
  - Gastroenterology consult

Gastroenterology consult results

- Sx: intractable hiccups, fever, gastroesophageal reflux-like symptoms
- Examination was essentially NL w/o clear diagnosis for constitutional sx.
- Meds:
  - Chlorpromazine 25 mg t.i.d. x hiccups
  - Omeprazole 20 mg bid

Vision consultation

- s/p 52 days from initial ER presentation
- CC: sudden, painful onset of central vision loss OD assoc. w/ pain on eye movement (6/10 OD, 1/10 OS) & weakness of all 4 extremities including complete paraparesis of the legs
- Examination was essentially NL w/o clear diagnosis for constitutional sx.
- Meds:
  - Chlorpromazine 25 mg t.i.d. x hiccups
  - Omeprazole 20 mg bid

Vision Exam

- BCVA:
  - OD: HM@5’ superiority→NI (confabulation)
  - OS: 20/160→NI
- P: ERRL, 3+ R APD
- EOMS: FROM OU, ±pain all gazes OU
- CF: inferior altitudinal defect OD, central blur OS
- CT: ortho
- Ishihara CV: 0/12 OD, OS
- TA: 17 mmHg OD, 20 mmHg OS
- SLEX / DFE: unchanged

Vision Plan

- Dx: Presumed RBON OD
- Psych consult
  - Medication-induced hallucinations sec to chlorpromazine & promethazine
- ID follow-up
- Consult Rheum
- Neuro-oph grand rounds

- W/U
  - MRI brain, orbits, spine w/ & w/o contrast
  - LP
    - CMV
    - EBV
    - Cryptococcus
    - WNV
    - VZV
    - HTLV-1
ID Follow-up

- Meds:
  - IV methylprednisolone 1g/d
  - IV acyclovir

- After review of all lab results and past hx, no evidence of serum or CSF infection was found. Consider demyelinating vs. vasculitic etiology.

Neuro-oph Grand Rounds

- s/p 55 days from initial ER presentation
- Confined to cardiac chair w/ spastic quadriparesis
- BCVA:
  - OD: bare LP
  - OS: 20/160 – NI
- P: elliptical OD which rounded w/ light stimulation, round OS. +direct OD, OS; 3+ R APD
- EOMS: FROM OU, +pain at gazes OU
- CF: amplitud OD, central blur OS
- TPN: 17 @ 5% mmHg OD, 17 @ 5% mmHg OS
- 20D w/ penlight ant seg / DFE: unchanged

Neuro-oph Grand Rounds

- DDx
  - Neuromyelitis optica (NMO / Devic’s Disease)
  - West Nile Virus myelopathy
  - Disseminated CNS viral Dz
  - Neoplastic Dz
  - Arteritic Dz
- W/U
  - NMO Ab serology via Mayo Clinic, Rochester, MN

MRI orbits w/ & w/o contrast

MRI spine

Rheumatology consult results

- No evidence of connective tissue dz based on physical examination & previous laboratory w/u.
Vision Exam

- s/p 60 days from initial ER presentation
- BCVA:
  - OD: NLP
  - OS: 20/160 → NI
- P: ERE; RPG APD
- EOMS: FROM OU, no pain
- CF: unable OD, central blur OS
- TPN: 19 @ 5% mmHg OD, 22 @ 5% mmHg OS
- 20D w/ penlight ant seg / DFE: unchanged

Neurology follow-up

- Physical condition worsens
- ARF sec to acyclovir
- Meds:
  - ↓ 450 mg IV q8h
  - Prednisone taper
- W/U
  - CSF & serum forwarded to Neurovirologist, D. Gilden, M.D. @ Univ. of Colorado
  - Plasmapheresis considered

Vision exam

- s/p 75 days from initial ER presentation
- BCVA:
  - OD: LP
  - OS: 20/80
- Tolerating plasmapheresis

Neurology follow-up

- No evidence of VZV Ab in CSF or serum.
- Dx: Presumed NMO
- Meds:
  - d/c acyclovir
  - Taper prednisone
  - Continue plasmapheresis

Vision Exam

- s/p 82 days from initial ER presentation
- BCVA:
  - OD: HM@6" inf-temp quad
  - OS: 20/80
- DFE: 2+ diffuse pallor ONH OU

NMO Ab Serology Results

- Positive
- 1:480 titer (norm < 1:120)
- Hospital discharge
- Rehabilitation Institute of Chicago
Vision Exam / Outcome

- s/p 132 days s/p initial ER presentation
- BCVA:
  - OD: HM@6” inf-temp quad
  - OS: 20/40
- Paraplegic
- UE weakness
- Severe pain (gabapentin)

NMO (Devic’s Disease)

- Inflammatory demyelinating disorder of the central nervous system with a predilection for (but not restricted to) the optic nerve and spinal cord.

Diagnostic Criteria – revised 2006

- Optic neuritis
- Acute myelitis
- 2 of 3 characteristics
  - Disease-onset brain MRI non-diagnostic x MS
  - MRI spinal cord lesions extending over 3+ vertebral segments
  - NMO-IgG seropositive

Diagnostic Testing

- Neurologic exam
- Ophthalmic exam
- MRI brain, orbits, spine
- LP
- NMO-IgG serology
  - Mayo Clinic, Rochester, MN
  - 4 mL serum cool or frozen
  - 73% sensitivity
  - 90% specificity for NMO
  - Test is not + in MS

Disease Classifications

- Monophasic (~10%)
  - Unilateral or b/l optic neuritis
  - 1 episode myelitis
  - Episodes occur w/in ~1 month of each other
  - Do not recur
- Relapsing (~90%)
  - Recurrent exacerbation of optic neuritis and/or myelitis after initial NMO diagnostic criteria are met.
  - Recurrence w/in 5 yrs x 90% of pts
  - Cumulative impairment

Epidemiology

- Age of onset
  - Infancy-9th decade
  - Mean: late 30’s
- Sex ratio
  - 0.9 : 1.0 (monophasic)
  - 4.5 : 1.0 (relapsing)
- Incidence / Prevalence
  - Unknown

Race
- AA
- Hispanics
- Japanese
- Pacific Islanders
Symptoms
- Viral prodrome: 30-50%
  - HA
  - Fatigue
  - Myalgia
  - Pyrexia
- Respiratory complaints
- GI complaints
  - Nausea
  - Vomiting
  - Hematemesis
  - Sphincter dysfunction
- Neurologic dysfunction
  - Weakness
  - Lower extremity paresthesias
  - Paraplegia / Quadriplegia
  - Paroxysmal tonic spasms
- Respiratory complaints
- GI complaints
  - Nausea
  - Vomiting
  - Hematemesis
- Sphincter dysfunction
- Hemiparesis
- Diplopia
- Vertigo
- Nystagmus
- Postural tremor

Visual Course
- Acute, severe attack
  - RBON w/ or w/o pain
- VF defects
  - Central
  - Paracentral
  - Altitudinal
  - Chiasmal
- Monophasic
  - Majority recover
  - >20/30
  - 22% remain <20/200
  - in one eye
- Relapsing
  - 40% eyes NLP at nadir during 1st episode
  - 80% of these improve to 20/200 over 6 mo.

Systemic Course
- Stepwise deterioration in motor, sensory, visual, and bowel / bladder function as a result of cumulative attack-related neurologic injury.

Physical Outcome
- 5-yr survival rate
  - 90% monophasic
  - 68% relapsing (respiratory failure sec to myelitis)
- 1/3 pts die from respiratory failure

Etiology
- Unknown
- Autoimmune associations
  - Hypothyroidism (30%)
  - Sjögren’s syndrome
  - Lupus
  - Connective tissue d/o
- Infectious associations
  - 25% preceded w/ viral infection
  - Only 2 cases of NMO followed by bilateral ARN sec to varicella zoster virus described in patients w/ AIDS
  - Past q/o MS-associations
- Immunizations

NMO Spectrum Disorders
- Longitudinally Extensive Transverse Myelitis (LETM)
- Recurrent isolated optic neuritis
- Japanese optic-spinal MS
- High risk for NMO conversion
- Order NMO-IgG
Treatment

- 1000 mg methylprednisolone / day x 5d
- Plasmapheresis
- IV Rituximab
- Azathioprine (Imuran) & prednisone
- Cyclophosphamide (Cytoxan)
- Methotrexate (Rheumatrex)
- β-interferon
- IV mitoxantrone
- IV IgG

Summary

- Case exhibits challenges of Devic’s dx
- Rarely does disseminated HVZ affect immunocompetent pts
- Well known in pts with HIV/AIDS, hematologic malignancy, or chemotherapy
- Serial DFE’s / ARN & PORN
- Timely NMO-Ab serology acquisition

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- James Goodwin MD
- Charles W. Kinnaird OD, FAAO
- Michelle M. Marciniak OD, FAAO
- Victoria M. Butcko OD, FAAO
- Danielle Weiler OD, FAAO

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- Michelle M. Marciniak OD, FAAO
- Victoria M. Butcko OD, FAAO
- Danielle Weiler OD, FAAO

A Case for Scleral Lenses: Atopic Dermatitis and Keratoconus

Muriel Schornack, OD, FAAO
Department of Ophthalmology, Division of Optometry
Mayo Clinic, Rochester, MN

American Academy of Optometry
Disclosure Statement

- Nothing to disclose

Case History
62-year-old female

- Medical History
  - Atopic dermatitis
  - Asthma
  - Polyarthralgia

- Ocular History
  - History of iritis
  - History of HZO, OS
  - Cataracts OU (OD>OS)
  - Chronic allergic conjunctivitis, resultant scarring of the palpebral conjunctiva
  - Extensive atopic dermatitis of the eyelids, resultant disruption of normal lid architecture

At Presentation

Right Eye

- Keratoconus
  - Spectacle Rx:
    - -6.00-5.00 X 180 OD (20/80+)
    - -3.25-2.25 X 058 OS (20/80+2)
  - Keratometry:
    - 56.50 @ 180, 58.87 X 090, distorted mires OD
    - 52.00 @ 180, 50.50 @ 090, distorted mires OS
  - Topography not available

Left Eye

Oh, by the way...

- Keratoconus
  - Spectacle Rx:
    - -6.00-5.00 X 180 OD (20/80+)
    - -3.25-2.25 X 058 OS (20/80+2)
  - Keratometry:
    - 56.50 @ 180, 58.87 X 090, distorted mires OD
    - 52.00 @ 180, 50.50 @ 090, distorted mires OS
  - Topography not available
Current Medications

- **Topical medications**
  - Restasis 0.05%, BID OU
  - Livostin 0.05%, TID OU
  - Cromolyn sodium 4%, TID OU

- **Oral medications**
  - Singulair 10 mg PO once daily
  - Theophylline SR 400 mg PO once daily
  - Zantac 300 mg PO TID
  - Allegra 180 mg PO once daily
  - Claritin 1 tablet PO PRN
  - Albuterol inhaler, 1-2 puffs q 4 hours PRN

Our Challenge

- **Vision**
  - Inadequate visual acuity with spectacles
  - Inadequate vision acuity with hydrogel torics
  - Unable to wear corneal RGP’s due to disruption of lid architecture

- **Comfort**
  - Severe ocular itching
  - Moderate dry eye symptoms

Scleral Lens Fit, OD

- Diagnostic lens parameters: 6.47 (52.25)/8.2, 6.87/2.0, 9.0/1.0, 12.25/1.5, 14.5/0.5
- Initial corneal clearance adequate, but lens settled back within 15 minutes of wear
- Parameters ordered: 6.47/8.2, 6.87/2.0, 8.8/1.0, 12.25/1.5, 14.5/0.5
- With initial lens ordered, patient achieved 20/70 acuity, and reported significantly increased comfort compared to no lens wear

Scleral Lens Fit, OS

- Diagnostic lens parameters: 6.75 (50.00)/8.2, 7.05/2.0, 9.0/1.0, 12.25/1.5, 14.5/0.5
- Scleral alignment appeared adequate, but bubble developed beneath lens within 15 minutes of wear
- Parameters ordered: 6.89 (49.00)/8.2, 7.29/2.0, 9.0/1.0, 12.00/1.5, 14.0/0.5
- With initial lens ordered, patient achieved 20/25 acuity, and reported significantly increased comfort compared to no lens wear

Right Eye, with ScL

Left Eye, with ScL
Here's the Rub...

What Happened?

- At 6-month follow-up:
  - Slow uptake of NaFl into post-lens fluid reservoir
  - Feather touch noted over apex of cone, without epitheliopathy

- At 12-month follow-up:
  - Minimal uptake of NaFl into post-lens fluid reservoir
  - Apical bearing with epitheliopathy
  - 20/20 acuity!

Current Plan

- Replace left lens
  - 6.75 (50.00 D)/8.2, 7.12/2.0, 8.8/1.1, 12.0/1.4, 14.0/0.5
  - Follow-up pending

Atopy and Keratoconus

- Conditions are frequently coexistent

- Eye rubbing may be the common link between the two conditions

- Elevated IgE levels are noted in both conditions

Ocular Complications of Atopic Dermatitis

- Blepharitis
- Keratoconjunctivitis
- Keratoconus
- Uveitis
- Subcapsular cataract
- Ocular herpes simplex


Scleral Lenses: Calming the “Perfect Storm”

- Visual benefits
- Ocular surface protection
- Improved comfort

Venous Macroaneurysm Associated with Hemi-Retinal Vein Occlusion

Brooke Smith, O.D.
Hampton VA Medical Center
Hampton, Virginia

American Academy of Optometry

References


Patient History

- 88 year-old White Male
- Medical History
  - Hypertension
  - Hyperlipidemia
  - CVA
  - Hyperkalemia
  - Anemia
  - Diverticulitis
- Ocular History
  - Advanced POAG
  - Non-ischemic HRVO
- Medications:
  - Topical:
    - Travatan QHS OU
    - Timolol BID OD
  - Systemic:
    - Nifedipine 30mg
    - HCTZ / Lisinopril 20mg
    - Simvastatin 40mg
    - Ferrous Iron
- Allergies: None

Clinical Presentation: Arterial Macroaneurysm

- Types
  - Fusiform
  - Saccular
- Location
  - Arteriolar bifurcation or A/V crossing
  - Supratemporal Artery
- Findings
  - Hemorrhage
  - Exudate
Clinical Presentation: Arterial Macroaneurysm

- Symptoms
  - Asymptomatic
- Epidemiology
  - Hypertension
  - Arteriosclerosis
  - Hyperlipidemia
- Etiology
  - Age: Sixth-Seventh Decade
  - Gender predilection: Women
- Ocular Associations
  - Eales, Leben, Gaes, BRVO

Retina Hemodynamics

- Vascularature
  - Retinal
  - Choroidal
- End Artery System
  - Extraretinal
  - Intraretinal
- Circulation
  - Autoregulation
  - F=AP/R
  - Perfusion Pressure
  - Resistance
- Intrinsic Factors
- Physiologic Adaptation
  - HTN

Differential Diagnosis

**Microaneurysm**
- Diabetic Retinopathy
- Retina Telangiectasis
- Vein Occlusion
  - BRVO
  - HRVO
  - CRVO

**Macroaneurysm**
- Arterial
  - Age
  - Atherosclerosis
  - HTN
- Capillary
- Collateral

Causes and Effect

- Vein Occlusion
  - Atherosclerosis
  - F=AP/R
  - Capillary Non-Perfusion
  - Neovascularization
  - Collaterals

- Venous Macroaneurysm
  - Increased Intraluminal Pressure
  - Retina Ischemia
  - Degeneration of inner BRB
  - Collaterals

Clinical Course

- Natural History
  - Cystoid Macula Edema
  - Venous Hemorrhage
  - Macula Holes
- Treatment
  - Photocoagulation
    - Direct
    - Indirect
  - CME
  - Focal Laser
  - Grid Laser
- Management
  - Monitor q3 months

3+pallor
0.7 H x 0.85 V
Exudates vs. Drusen

Intra-retinal hemorrhages
NVE vs. Collaterals
**Pseudo-Retinitis Pigmentosa**

Dennis J. Light, OD, FAAO
Dayton, Ohio VAMC Eye Clinic

**References**

**CASE HISTORY**
- 52 year old black male
- Complains of difficulty with vision at all distances over the last several years; unable to wear bifocals from last eye exam b/c “they didn’t help me to see any better with or without them.”

**OCULAR HISTORY**
- Dx with “retinitis pigmentosa” while serving in military in 1976 (age 20)
- Describes vision loss at this time as sudden onset:
  “It happened quickly. One day I just couldn’t see very well out of either eye. They took me to medical for a couple of weeks. A few weeks later they discharged me right out of the service and said I would eventually be blind.” (military records unavailable)

**FAMILIAL OCULAR HISTORY**
- ? Hx. of retinitis pigmentosa in one nephew
MEDICAL HISTORY

- HIV (+) x 1986
- Erectile dysfunction vs. anorgasmia
- Hyperlipidemia
- Depression, PTSD
- Hx of gonorrhea while in military
- Hx of being raped by father at age 8 and by same sex duty officer while in military service
- Multiple homosexual relations
- Denies past/present IV drug use
- ~0.5 ppd smoker x 30 years

MEDICATIONS

- efavirenz 600mg (HIV)
- lamivudine 300mg (HIV)
- paroxetine HCL 20mg (depression)
- buspirone HCL 30mg (anxiety)

Allergies: penicillin, codeine

ADDITIONAL OCULAR HISTORY

Record review of last 2 eye exams:

4 years ago:
- BCVA OD 20/50; OS 20/40
- Confrontation visual fields:?
  - full OD; restricted all quadrants OS
- Dx: Retinitis pigmentosa; HIV without ocular manifestations; myopic astigmatism w/presbyopia

2 years ago:
- BCVA OD 20/50; OS 20/40
- Goldmann visual fields (results will be shown later)
- Dx: Retinitis pigmentosa; HIV without ocular manifestations; myopic astigmatism w/presbyopia

CLINICAL FINDINGS

- Uncorrected vision
  - OD 20/200 ph 20/70+
  - OS 20/80 ph 20/50+
- Best corrected vision
  - OD 20/50+
  - OS 20/40+
- EOM: full
- Cover testing with manifest
  - Ortho distance; 4 exophoria near
- Pupils: ERRL (-) APD
- Visual fields (will show later)

BIOMICROSCOPY (OU)

Lids: mild MGD
 Conj: mild diffuse benign melanosis
 KC: mild areas; clear central without edema
 AC: deep/clear; no cell or flare
 I: brown; unremarkable
 Lens: +1+NS

Tonometry (applanation) @ 11:30
 16mmHg OD, OS

FUNDUS EXAMINATION OS
FUNDUS EXAMINATION OS
ONH: C/D ~ 0.85 V/H mod/deep cupping with waxy appearance
MAC: mottled
FUND: dense, thickened bone spicule chorioretinal degeneration mid-periphery/peripheral
VESS: marked attenuation
VIT: syneresis without cells

VISUAL FIELDS OS
Goldmann visual fields stable.

DIAGNOSIS- classic retinitis pigmentosa
- Previous diagnosis of RP
- Mid-peripheral/peripheral bone spicules and RPE atrophy
- Waxy pallor of the optic nerve
- Retinal arteriolar attenuation
- Heavily constricted visual field

Seems pretty straight forward, right? Except for the fact that the title slide mentions pseudo-retinitis pigmentosa!
FUNDUS EXAMINATION OD

ONH: C/D ~ 0.85 V/H mod/deep cupping without waxy appearance
MAC: dense hyperpigmented RPE scarring/mottling
FUND: dense pigmented chorioretinal scarring contained mostly within arcades
VES: trace attenuation
VIT: syneresis without cells

“Waxy” disc? (not this time)
Attenuation? (not so much)

VISUAL FIELDS OD

Goldmann visual fields stable.
Now, we have a mystery…
is this really retinitis pigmentosa?

RETINITIS PIGMENTOSA
- Group of inherited retinal disorders characterized by progressive degeneration of photoreceptors and RPE
- Affects ~ 1 in 3,000 to 1 in 5,000; ~1.5 million people worldwide.
- Age of onset may vary, typically between adolescence and early adulthood.
- Has been classified by retinal location, age of onset, mode of inheritance, predominant photoreceptor involvement, and most commonly as Primary (confined to eye alone) or Syndromic.
- Syndromic associations include:
  - Usher’s syndrome
  - Bassen-Kornzweig syndrome
  - Refsum’s disease
  - Bardet-Biedl syndrome

RETINITIS PIGMENTOSA
- Earliest symptoms typically manifest as night blindness with progressive peripheral VF loss that can lead to central vision loss.
- VF defects typically present by teenage years.
- VF loss slow but relentless. Estimated by one study to be ~4.6% per year.
- Central vision is typically preserved until later stages when cone degeneration occurs.

RETINITIS PIGMENTOSA
- Fundus findings are typically symmetric.
- Classic ophthalmic triad includes mid-peripheral bone spicules, attenuated vessels, and waxy pallor of the optic nerve.
- Cataract is the most common anterior segment complication from RP; up to 50% of adult patients with RP will have PSC.

RP- what doesn’t fit our patient?
- Asymmetric fundus findings
- Acute onset of central vision loss as initial presentation
- Stability of VF loss

What else could it be?

DIFFERENTIAL DIAGNOSES
- TORCH (infectious)
  - Toxoplasmosis
  - Other (syphilis, hepatitis B, HZV, HIV, parvovirus 19)
  - Rubella/congenital rubella
  - Cytomegalovirus
  - Herpes simplex
- Toxicity
  - Thoridazine, chloroquine
- Trauma

That’s a lot of Ts. I pity the fool who don’t look at labs closely.
CONSIDER RECENT LABS

- HIV viral load - 10,602 copies/mm³ (high)
- CD4 – 283 cells/mm³ (low)
- CBC w/diff – low new WBC
- Toxoplasma (IGG) – negative
- CMV (IGG) - negative
- Rapid plasma reagin (RPR) - negative
  - also (-) 05/2003; 07/2003; 12/2003; 06/2006

NEW LABS ORDERED

- Repeat RPR
  - Negative
- Fluorescent treponemal antibody absorption (FTA-ABS)
  - Reactive

NEW DIAGNOSIS:
pseudo-retinitis pigmentosa secondary to SYPHILIS

SYPHILIS

- Term came from the epic poem written in 1530 by Veronese physician Girolamo Fracastoro entitled “Sphylis or the French Disease.”
  - “Sphylis” was the main character in the poem; a shepherd who became the first man stricken with the disease. It was given to him by the mythological god Apollo as punishment for defiance.

- Infection by the spirochete Treponema pallidum.
- Humans are only host
- Transmitted primarily through sexual contact via breaks in the skin or by penetration through intact mucous membranes; also perinatally, blood transfusion (rare), or with needle sharing

SYPHILIS- demographics

- CDC reported 40,920 cases (all stages) in US in 2007.
- Rates of 1 and 2 cases have increased annually since 2000
- Occurrence higher in southern and southwestern US, and in large urban populations
- Incidence is greater in males and among nonwhites
- Frequency greatest in young adults during years of peak sexual activity
- 2004 CDC estimated 64% of all primary and secondary cases were among homosexual males. Also have high rate of HIV co-infection.

SYPHILIS- clinical course

- 4 stages to classic clinical course:
  - Primary
  - Secondary
  - Latent
    - Early latent
    - Late latent
  - Tertiary
- Stages tend to overlap, often best to describe as:
  - Early syphilis
    - Primary, secondary, early latency
  - Late syphilis
    - Late latency, tertiary
CLINICAL COURSE - primary

- Begins as nonpainful chancre at site of sexual contact
- Initial incubation period is 9-90 days; avg 3 weeks
- Lesions will heal treated or untreated within 6-8 weeks without scarring, but organisms spread through lymphatics and bloodstream
- If untreated, virtually all 1st cases will proceed to secondary syphilis

CLINICAL COURSE - secondary

- Concentration of systemic spirochete greatest at this stage
- Characterized by generalized rash involving entire trunk and extremities (most prominent on palms and soles of feet)
- Occurs ~4-10 weeks after 1st infection
- Constitutional symptoms may include:
  - Fever, nausea, malaise, headache, joint pain
- Other organs/systems may be involved including:
  - Liver, GI tract, kidneys, CNS, skeletal
  - 10% will have ocular manifestations
- Untreated 2nd syphilis persists 4-8 weeks before patient becomes symptom and lesion free
- 30% of untreated 2nd cases will proceed to 3rd syphilis

CLINICAL COURSE - latent

- Early latency
  - CDC defines as < 1 year after 1st infection
  - Still considered infectious
  - May relapse back into 2nd
- Late latency
  - > 1 year after 1st infection
  - Considered noninfectious
  - Relapse rare

CLINICAL COURSE - tertiary

- Benign late syphilis
  - Gummatous (granuloma) lesions of skin/mucous membranes
- Cardiovascular syphilis
  - May involve coronary arteries and aorta
- Late neurosyphilis
  - Collection of syndromes found in all stages, but most devastating effects are found in 3rd stage
  - May present as tabes dorsalis (slow degeneration of sensory neurons in spinal chord) or general paresis
  - CNS affected via vascular pathway or direct involvement of parenchyma

SYPHILIS - ocular manifestations

- Rarely seen earlier than 6 months following the 1st chancre
- Frequently found in 2nd and 3rd syphilis
- Ocular involvement occurs in ~5% of untreated cases of 2nd
- Disease progresses to neurosyphilis, ~10% will have ocular involvement

SYPHILIS - ocular manifestations

Anterior Segment

- Conjunctivitis
- Episcleritis/scleritis
- Interstitial keratitis (90% congenital)
- Secondary glaucoma (mainly congenital)
- Cataract
- Anterior uveitis - most common 2nd complication
  - 56% unilateral; 50% granulomatous KP
  - acute or chronic; recurrent
SYMPHILIS - ocular manifestations

Posterior Segment
- Posterior uveitis also common
  - Multifocal chorioretinitis (including pseudo-RP)
  - Acute syphilitic posterior placoid chorioretinitis (ASPPC)
- Retina/Vitreous
  - CMV, neuroretinitis, vasculitis, serous RD, vitritis
- Neuro-ophthalmic
  - Papillitis, optic neuritis, optic atrophy
- EOM (with neurosyphilis)
  - Cranial nerve palsies
  - Pupil abnormalities (with neurosyphilis)
  - Argyll Robertson (unequal, miotic, with light-near dissociation)
  - Tonic pupil

SYMPHILIS - laboratory testing

Nonreponemal serological testing
- Detects nonspecific cardiolipin-lecithin cholesterol antibodies (found in immune response to T. pallidum infection)
- Minimum of 1-3 weeks post infection to turn sero (+)
- If treated within 6 months, usually sero (-) by 12 months
- May be used to monitor and quantify treatment effectiveness.
- Nonspecific so can give false positive results
  - Other spirochete infections (Lyme), auto-immune disease, HIV, other acute or chronic infections

RPR (Rapid Plasma Reagin)
VDRL (Venereal Disease Research Laboratory)
* INDICATES ACTIVE SYMPHILIS (confirmed only when coupled with positive specific treponemal testing)

SYMPHILIS - laboratory testing

Treponemal serological testing
- Detects specific antibodies to T. Pallidum; once (+) will remain so for life
- FTA-ABS 98% sensitive even in late syphilis
- May have false (+) with SLE, biliary cirrhosis, and rheumatoid arthritis

FTA-ABS (fluorescent treponemal antibody absorption)
TP-PA (T. Pallidum particle agglutination)
MHA-TP (micro-hemagglutination T. Pallidum)
* INDICATES LIFETIME EXPOSURE TO SYMPHILIS (does not indicate if active)

SYMPHILIS & HIV

Co-infection with HIV can accelerate natural course of syphilis
Greater frequency of ocular involvement and neurosyphilis
Higher rates of treatment failure and relapse
May give false (+)/(-) serological testing
ALL patients with HIV and syphilis should have cerebrospinal fluid (CSF) evaluation because of increased frequency of neurosyphilitic complications

SYMPHILIS- treatment

Medication
- CDC recommends neurosyphilis regimen for ocular syphilis:
  Aqueous crystalline penicillin G, 2.4 million U IV q4h x 10-14 days
  Or alternatively:
  Procaine penicillin IM daily & probenecid 500mg PO x 10-14 days
  *Many recommend following either regimen with:
  Benzathine penicillin G, 2.4 million U IM x 3 weeks
*PCN allergy: skin tested, desensitized, and managed by expert
SYMPHILIS - treatment

- Consultation with infectious disease specialist recommended
- Sometimes there is an “eye” in TEAM

SYMPHILIS - our patient

- EYE CLINIC
  - Patient informed that vision loss was not from RP, but as a result of a previous syphilis infection
  - Advised him that his vision loss was stable and is not expected to follow clinical course of retinitis pigmentosa (progression to blindness)
  - Glasses and possible low vision aids to help with vision
  - Monitor annually
  - Referred to Infectious Disease Clinic

- INFECTIOUS DISEASE
  - Recommend lumbar puncture. Patient declined.
  - Consider PCN treatment as unable to establish clear history of previous treatment. Patient declined.
  - ID Clinic agreed to monitor given:
    - Longstanding and stable history
    - Lack of other symptoms
    - Multiple negative RPR
    - Allergy to PCN
    - Patient’s wishes

CONCLUSIONS

- Important to look at previous diagnoses critically
  - May impact treatment and management
  - May impact psychology of patient

- Syphilis is a great imitator and should be considered in unusual presentations

- Laboratory testing is useful, but you should know what the results say and do not say
  - () RPR does not mean that syphilis should not be considered
  - Recommend FTA-ABS be ordered in addition to RPR when considering syphilis
  - Remember false negatives/positives do occur

- Syphilis/HIV are a dangerous combination
  - If one is present, test for the other
  - Manage with Infectious Disease

REFERENCES


REFERENCES


Questions?

Thank you!