Oh yeah, I have Ankylosaurus Spondywhatsis?

Emily M Thompson, OD, FAAO
AFOS Annual Meeting
Nov 1, 2021
• I have no financial disclosures
• None of the photos are of the actual patient
Initial Presentation: Day 1 Apr 30

- 36 y/o Caucasian male aviator
- CC: Sore & bruised feeling OS, also noticed OS pupil won’t dilate
- Onset 10 days ago
- Vision worse when he leans forward & looks down
- Had been to ER before coming to Optometry, was given Tobradex QID OS
- Oh yeah, this happened a few years ago OD. Possible history of trauma - hit in face with football

- DVA sc:
  - OD: 20/15       OS: 20/20
- IOP NCT:
  - OD: 10       OS: 08
- Pupils:
  - OS miotic but reactive equally in dim & bright light
  - (-) APD
- Slit lamp:
  - Conj: tr-1 chemosis OS
  - Cornea: tr diffuse edema OS
  - A/C: 2+ cells, tr flare OS
- DFE: (-) post seg inflammation OD, OS
Initial Presentation: Day 1  Apr 30

• Treatment Plan
  • Stop Tobradex
  • Begin Durezol q 2 hrs OS
  • Declined cycloplegic
  • Made note of 2nd iritis within a few years, previous episode seemed to be related to trauma
  • RTC 1 day
Follow Up #1  May 1

CC:
- Using Durezol q 2 hrs OS.
- Eye still feels irritated and vision is slightly foggy.
- No achy or sore sensation.

DVA sc:
- OD: 20/20    OS: 20/15-1

IOP NCT:
- OD: 09    OS: 09

Pupils:
- OS miotic but reactive equally in dim & bright light.
- (-) APD

Slit lamp:
- Cornea: 1+ edema esp inf OS
- A/C: Tr cells, 0.5 flare OS
Follow Up #1  May 1

- Treatment Plan
  - Improved comfort
  - Reduced inflammation
  - Cont Durezol q 2 hrs OS
  - Declined cycloplegic
  - RTC 2 days

SUN Grading system for AC cell and flare

- Cells per high-power field in 1x1 mm slit beam.
  - 0 = < 1 cell/hpf
  - 0.5+ = 1 - 5 cells
  - 1+ = 6 - 15
  - 2+ = 16 - 25
  - 3+ = 26 - 50
  - 4+ = > 50

- Flare
  - 0 = none
  - 1+ = faint
  - 2+ = moderate, (iris/lens details clear)
  - 3+ = marked (iris/lens hazy)
  - 4+ = intense (fibrin or plastic aqueous)
Follow Up #2  May 3

• CC:
  • Using Durezol q 2 hrs OS
  • Vision not as foggy
  • Eye sometimes feels achy or scratchy

• DVA sc:
  • OD: 20/20  OS: 20/15

• IOP NCT:
  • OD: 12  OS: 09

• Pupils:
  • OD=OS
  • (-) APD

• Slit lamp:
  • Cornea: 1+ edema esp inf OS
  • A/C: Tr cells, 0.5 flare OS
Follow Up #2  May 3

• Treatment Plan
  • Begin Durezol taper to QID OS
  • Add Muro 128 gtts QID OS to speed corneal edema resolution
  • RTC 5 days
  • Edu will likely be on steroid gtts for several weeks
Follow Up #3  May 8

• CC:
  • Durezol QID OS
  • Muro 128 gtts QID OS
  • Vision still a little foggy

• DVA sc:
  • OD: 20/15-2  OS: 20/15-2

• IOP NCT:
  • OD: 11  OS: 12

• Pupils:
  • OD = OS
  • (-) APD

• Slit lamp:
  • Cornea: tr edema esp endothelial with some PEE superficially OS
  • A/C: D & Q (-) cells (-) flare
Follow Up #3  May 8

• Treatment Plan
  • Cont Durezol taper to BID OS
  • Muro 128 gtts BID OS
  • RTC 1 week
Follow Up #4  May 14

• CC:
  • Durezol BID OS
  • Muro 128 gtts QID OS
  • Vision still a little foggy
  • Wife pointed out yesterday that pupil OS is smaller again

• DVA sc:
  • OD: 20/15-1  OS: 20/20

• IOP NCT:
  • OD: 10  OS: 10

• Pupils:
  • OD = OS
  • (-) APD

• Slit lamp:
  • Cornea: Rough endothelium inferior sector w/tr pinball NaFL staining
  • A/C: 1 single cell (-) flare
Follow Up #4  May 14

- Treatment Plan
  - Cont Durezol taper to once daily OS
  - Muro 128 gtts BID OS
  - RTC 5-7 days
Follow Up #5  May 17

• CC:
  • Durezol QD OS
  • Muro 128 gtts BID OS
  • Feels like left eye is “focusing harder” since iritis started

• Manifest:
  • OD: +0.25-0.50*085....20/15
  • OS: PL-1.00*080......20/15

• DVA sc:
  • OD: 20/15  OS: 20/20

• IOP NCT:
  • OD: 09  OS: 07

• Pupils:
  • OD = OS
  • (-) APD

• Slit lamp:
  • Cornea: Tr Rough endothelium, no NaFL staining
  • A/C: (-) cell (-) flare
Follow Up #5  May 17

- Treatment Plan
  - Stop Durezol
  - Muro 128 gtts BID OS
  - RTC 1 week, consider repeat MR and specs at that time if pt still bothered by vision
Pt walked into clinic saying he had a flare up of the iritis over the weekend and went to ER

Referred patient to off base ophthalmology at 1245 same day

That ophthalmologist referred him to retinal specialist for possible panuveitis....He has an appointment the next day with him
May 23
Pt was diagnosed with posterior uveitis
Started oral and topical steroids
Recommended auto-immune workup
Retinal Specialist Reports

- June 4
- Pt was found to be HLA B27+
- Planned to be on oral steroids for 6 weeks minimum
- Rheumatology consult ordered to rule out ankylosing spondylitis

- Typical ankylosing spondylitis workup:
  - HLA B27
  - SI joint films
  - Rheumatology consult
Retinal Specialist Reports

• Sept 17
• Reported he felt flare up coming on, started steroids again
• Developed macular edema OS at some point that was not resolving
• Now has panuveitis with extensive debris inferiorly
• Nongranulomatous
• HLA B27 (+) HLA B51(-)
Jan 31
Last one, promise
Tapered off both oral and topical steroids
VA OD, OS 20/20
Toxo and quantiferon negative
F/U in 1 year

Rheumatologist started him on Humira April 9 the following year
UVEITIS

- Anterior
  - Noninfectious
    - Nongranulomatous
  - Infectious
    - Granulomatous

- Intermediate

- Posterior

Classified by
- Pathology
- Location
- Course
- Etiology
Anterior Uveitis: Iritis, Iridocyclitis

- Inflammation of iris and ciliary body
- Breakdown of blood-aqueous barrier = leaking of WBC into anterior chamber
Signs & Symptoms

- Pain, redness, photophobia, tearing, reduced vision
- Photophobia, usually dramatic
- Cells
- Flare
- Ciliary flush
- Keratic precipitates
  - Fine: nongranulomatous
  - Mutton fat: granulomatous
- Iris nodules
- Increased or decreased IOP
- PAS
- Posterior synechiae
- Hypopeon (esp suggestive of HLA B27 associated or Behcet’s)
- Most commonly idiopathic or autoimmune
Cells & Flare

**Standardized Grading Scales for Uveitis**

**SUN Grading Scheme for Anterior Chamber Cells**

<table>
<thead>
<tr>
<th>Grade</th>
<th>Cells in Field</th>
</tr>
</thead>
<tbody>
<tr>
<td>0</td>
<td>&lt; 1</td>
</tr>
<tr>
<td>0.5+</td>
<td>1 – 5</td>
</tr>
<tr>
<td>1+</td>
<td>6 – 15</td>
</tr>
<tr>
<td>2+</td>
<td>16 – 25</td>
</tr>
<tr>
<td>3+</td>
<td>26 – 50</td>
</tr>
<tr>
<td>4+</td>
<td>50+</td>
</tr>
</tbody>
</table>

(using 1mm slit beam)

**SUN Grading Scheme for Anterior Chamber Flare**

<table>
<thead>
<tr>
<th>Grade</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>0</td>
<td>None</td>
</tr>
<tr>
<td>1+</td>
<td>Faint</td>
</tr>
<tr>
<td>2+</td>
<td>Moderate (iris/lens details clear)</td>
</tr>
<tr>
<td>3+</td>
<td>Marked (iris/lens details hazy)</td>
</tr>
<tr>
<td>4+</td>
<td>Intense (fibrin/plastic aqueous)</td>
</tr>
</tbody>
</table>

Image from “Anterior Uveitis – Pearls and Pitfalls, 2017”
Keratic Precipitates

- Nongranulomatous
  - Lymphocytes
  - Plasma cell infiltrates

- Granulomatous “mutton fat”
  - Epithelioid infiltrates
  - Giant cell infiltrates
Intermediate Uveitis / Pars Planitis

- Affects vitreous, pars plana, ciliary body
- Usually children & young adults
- Typically bilateral
- Decreased VA, floaters, usually no pain, redness or photophobia
- Associated with MS, sarcoidosis

- Snowballs, snow banking, vitreous cells
Posterior Uveitis

- Cystoid Macular Edema
- Retinal detachment
- Vasculitis
- Optic neuritis
- Neovascularization
- Hypotony
- Phthisis

- Usually no pain, redness, or photophobia unless anterior uveitis also present
Panuveitis

• Inflammation of entire uveal tract
• “A pattern of severe, diffuse inflammation of both anterior and posterior segments” according to Wills Eye Manual
### Panuveitis Causes: Most to least common

- **Sarcoidosis**
  - Candle-wax drippings
- **Birdshot retinochoroidopathy**
- **Behcet disease**
  - Recurrent oral & genital ulcerations, hypopeon
- **Vogt-Koyanagi-Harada VKH syndrome**
  - Asian, Afro-Caribbean, Japanese
  - Bilateral
  - Exudative retinal detachment
- **Sympathetic ophthalmitis**
  - Rare, bilateral, following penetrating trauma
- **Tuberculosis choroiditis**
  - Uncommon
- **Acquired syphilis**
  - Rare
  - Large yellow placoid lesions in post pole
Visual Prognosis

• Depends on:
  • Etiology
  • Ability to control inflammation
  • Sequelae
Recommended Tests: Nongranulomatous

- Negative past iritis history
  - CBC
  - ESR erythrocyte sedimentation rate
  - Rapid plasma regain or VDRL
  - FTA-ABS or MHA-TP (syphilis)
  - HLA B27
Recommended Tests: Granulomatous

- ANA
- Rheumatoid Factor
- Angiotensin Converting Enzyme
- Purified Protein Derivative
- Herpes titers
- ELISA
- HIV antibody
- Chest radiograph
- Chest CT
- Sacroiliac radiograph
- Knee radiograph
- Gallium scan
- Urinalysis
- Urethral cultures

- Special Diagnostics
  - HLA typing
  - ANCA
  - Raji cell & C1q binding assays
  - Complement proteins
  - Soluble interleukin-2 receptor

- Rheumatology consult!
Ankylosing Spondylitis

- New bone formation in spine
- Arthritis affecting spine and hips
  - Particularly sacroiliac joint
- Can limit breathing if affecting ribs
- Various methods to test joint pain by PCM
- [https://spondylitis.org/](https://spondylitis.org/)

- Gaenslen Test
Ankylosing Spondylitis

- Typically Caucasian male in 30s
- Diagnostic testing can include:
  - HLA B27
  - C Reactive protein
  - X-Ray, MRI
  - Urinalysis
- Positive X-ray/MRI AND one of following to diagnose:
  - Limited chest expansion for demographic
  - Limited range of motion of lower back
  - Back pain x3 months, improves w/exercise, worsens w/rest
- Treatments
  - Exercise
  - Physiotherapy
  - Medications
    - NSAIDS, steroids, biologics
- Prognosis generally good
Uveitis Treatments

- Topical steroids
- Topical cycloplegic
- May need to use IOP lowering drops
- Oral steroids
- Ranitidine (Zyrtec) with oral steroids
- Sub-tenon steroid injection or implantable sustained release steroid devices

- NSAIDS
- Immunosuppressive chemotherapy
  - Antimetabolites (methotrexate)
  - Inhibitors of leukocyte signaling (cyclosporine)
  - Alkylating agents (chlorambucil)
  - Biologics (Enbrel, Humira)
  - Others (dapsone, colchicine)
HLA B27 on Chromosome 6

• Protein on surface of WBC that’s a marker for certain autoimmune conditions

PAIR:
• Psoriatic arthritis
• Ankylosing spondylitis
• Inflammatory bowel disease
• Reiter syndrome aka Reactive arthritis
  • Can’t see, can’t pee, can’t climb a tree
Resources

• Ophthalmology: Clinical Signs and Differential Diagnosis
  • Kanski, 1999

• The Massachusetts Eye and Ear Infirmary
  • Friedman & Kaiser

• The Wills Eye Manual

• Google image search
• How to diagnose and manage uveitis - EyeGuru
• https://spondylitis.org/
H35.9999 Unspecified Retinal Goomba

Michael V. Hall OD

PRIMARY EYE CARE RESIDENT, WOMACK ARMY MEDICAL CENTER/NSUOCO
Introduction

Case Overview
Learning Points
Update on current status

The views expressed in this presentation are those of the author and do not reflect any official policy or position of the Department of the Army, Department of Defense or the US Government.

No financial disclosures
CC: Newly Diagnosed with Diabetes

30 yo Hispanic Male

Happy with current Rx: OD -3.25-1.75x177 20/20
OS -3.25-2.50x176 20/20

No visual complaints

1\textsuperscript{st} diagnosed with Type II Diabetes Aug 2021, no current treatment other than lifestyle changes
Entrance and Anterior Segment

• Pupils: PERRL (-) APD
• EOM: Smooth and Full Both Eyes
• IOP: 11 Right Eye, 12 Left Eye @ 0940
• Anterior Segment: No Abnormalities
What to do?

- Area of temporal retinal hemorrhage right eye only
  - Diabetic Retinopathy?
  - Distal Vein Occlusion?
  - Some other systemic etiology?

Service Member is deploying in 2 months

Plan?
1 Month Later

• Pupils: PERRL (-) APD
• EOM: Smooth and Full Both Eyes
• IOP: 12 Right Eye, 13 Left Eye @ 0945
• Anterior Segment: No Abnormalities
Unspecified Retinal Goomba

• Hemorrhaging following retinal vein
• Scattered exudate
• Amelanotic, lobular goomba, elevated?
• White without pressure, hazy retina temporal to lesion

• Further Testing? Referral?
Refer to Retina Specialist
Differential Diagnosis
Diabetic Retinopathy
Proliferative Sickle Cell Retinopathy

- Peripheral arteriolar occlusion leading to non-perfusion...
- Peripheral AV Anastomoses
- Pre-retinal sea fan neo posterior to non-perfusion
Amelanotic Choroidal Melanoma

- Thickness greater than 2mm
- Fluid present in subretinal space
- Symptoms—flashes, floaters, decreased vision
- Orange pigment overlying the lesion
- Ultrasound hollowness
Coats Disease

- Male (85%) and usually one eye involved (95%)
- Leukocoria
- Typically progressive, 6-8 yo
- Primarily aneurysms and telangiectasia of vessels
- Temporal retina with various degrees of exudation
- OFTEN confused with retinoblastoma (calcific findings on CT or Ultrasound exclude Coats)
Familial Exudative Vitreoretinopathy (FEVR)

• Bilateral failure of the temporal retina to vascularize
• Peripheral Exudation with traction
• Like Retinopathy of Prematurity but normal birthweight and no O² tx
Retinal Cavernous Hemangioma

- Cluster of grapes appearance
- Usually unilateral
- Usually asymptomatic unless involving macula
- Strong autosomal dominant inheritance
Phakomatoses

• Neurofibromatosis type 1
• Neurofibromatosis type 2
• Tuberous Sclerosis
• Sturge Weber Syndrome
• Von Hippel-Lindau
Capillary Hemangioma (Von Hippel-Lindau)

- Characterized by “hemangioblastoma” of the retina and CNS
  - Cerebellum, Medulla, Pons, and Spinal Cord in 20% of patients
  - “Visceral” manifestations are common (renal cell carcinoma, cysts of kidney, pancreas, liver)
Von Hippel-Lindau (VHL) continued

• 2nd-3rd Decade and can occur anywhere in retina
  • Roughly 70% of VHL patients have retinal capillary hemangioma

• Starts no larger than Diabetic microaneurysm located in ST mid periphery

• Feeder vessels and capillary network incompetent
  • “Steal Phenomenon”

• Grows/proliferation of fibro vascular material
# AFOS2021
Resident Grand Rounds

## TABLE 1. CRITERIA FOR THE DIAGNOSIS OF VON HIPPEL-LINDAU SYNDROME

<table>
<thead>
<tr>
<th>If family history is:</th>
<th>Feature</th>
</tr>
</thead>
<tbody>
<tr>
<td>Positive</td>
<td>for any one of the following:</td>
</tr>
<tr>
<td></td>
<td>• retinal hemangioblastoma</td>
</tr>
<tr>
<td></td>
<td>• brain hemangioblastoma</td>
</tr>
<tr>
<td></td>
<td>• visceral lesion&lt;sup&gt;a&lt;/sup&gt;</td>
</tr>
<tr>
<td>Negative</td>
<td>for any one of the following:</td>
</tr>
<tr>
<td></td>
<td>• two or more retinal hemangioblastomas</td>
</tr>
<tr>
<td></td>
<td>• two or more brain hemangioblastomas</td>
</tr>
<tr>
<td></td>
<td>• single retinal or brain hemangioblastoma with a visceral lesion&lt;sup&gt;a&lt;/sup&gt;</td>
</tr>
</tbody>
</table>

<sup>a</sup>Visceral lesions include renal cysts, renal carcinoma, pheochromocytoma, pancreatic cysts, islet cell tumors, epididymal cystadenoma, and endolymphatic sac tumor.
Treatment/Prognosis

• Depends on f/u ability (watch vs treat)
• Progressive Subretinal Exudation leading to total RD
• Small lesions easy/large notoriously difficult
• Photocoagulation for <1DD size
• Trans-Scleral Cryo is viable
• Photodynamic therapy with Verteporfin seems to be preferred due to precision
  • Can combo with anti-VEGF
Vasoproliferative Tumor

- Elevated dome-shaped, Echo Dense, IT region
- MINIMALLY dilated retinal feeding and draining vessels
- Exudation, SRF, and Hemorrhaging
- Related to ROP, RP, Uveitis, FEVR
- Primary vs Secondary
- Treatment
More Information from Specialists
Conclusions

Not textbook presentation
Update on current status
Utilize technology
Medicine is a team sport
Wide list of differentials
References

A RASH, A RED EYE, AND AN ADVERSE REACTION
Chief Complaint

• A 63-year-old Caucasian male was referred to the eye clinic from the urgent care clinic complaining of a one-month history of ocular redness, foreign body sensation, light sensitivity, and blurred vision in left eye.

History of Present Illness

• Ocular symptoms started after he developed a rash on his cheek that spread to his eyelid.
• Left eye was swollen shut for about one week and had resolved two weeks ago.
• He saw an urgent care provider and was prescribed erythromycin ointment QID OS with no improvement.
• He also reports
  • hives all over his body that had started 7 weeks prior and lasted about three weeks
  • a fever that had since resolved
  • Relatively new shortness of breath
  • Joint and back pain when he first broke out with hives that had somewhat improved
• COVID-19 testing was negative in the ED before reporting to the eye clinic
OCULAR HISTORY

Past ocular history

- Pseudophakia (PCIOL in 2008)
- Hyperopic astigmatism OD, simple astigmatism OS, Presbyopia

Ocular Medications

- Erythromycin QID OS
Medical History

- Hypertension
- Hyperlipidemia
- Depression
- Low back pain
- Hearing loss
- Anxiety
- Erectile dysfunction
- Microcystic colitis
- Benign prostatic hypertrophy s/p prostatectomy

Medications

- Diltiazem 180 mg qdaily po
- Losartan 100 mg qdaily po
- Cholestyramine oral powder daily
- Mirtazapine 30 mg QHS
- Duloxetine 60 mg EC BID po
- Aripiprazole 5 mg QHS po
- Gabapentin 300 mg TID po
- Methocarbamol 750 mg QID prn po
- Mesalamine 1200 mg BID po
- Prazosin 2 mg QHS po
- B12 injection 100 mcg IM monthly
### SOCIAL HISTORY

<table>
<thead>
<tr>
<th>Alcohol use</th>
</tr>
</thead>
<tbody>
<tr>
<td>Drug use</td>
</tr>
<tr>
<td>• Marijuana</td>
</tr>
<tr>
<td>• Heavy cocaine use that had decreased recently since moving away from triggers</td>
</tr>
<tr>
<td>Smoker</td>
</tr>
<tr>
<td>• 10 cigarettes a day x 35 years</td>
</tr>
<tr>
<td>Divorced</td>
</tr>
<tr>
<td>Recently relocated from Florida</td>
</tr>
<tr>
<td>Admits to high-risk sexual encounters</td>
</tr>
<tr>
<td>Denies any history of sexually transmitted diseases</td>
</tr>
</tbody>
</table>
OCULAR EXAMINATION – ENTRANCE TESTING

Acuity

- OD: 20/25-2 PH NI
- OS: 20/20-2 PH NI

Entrance Testing

- Pupils: EERL OU, No APD
- Confrontation Fields: Full OU
- Extraocular Motilities: full and smooth OU
- Ocular alignment: ortho by Hirschberg
## OCULAR EXAMINATION – ANTERIOR SEGMENT

<table>
<thead>
<tr>
<th>OD</th>
<th>OS</th>
</tr>
</thead>
<tbody>
<tr>
<td>Normal</td>
<td>Normal</td>
</tr>
<tr>
<td>Fine keratic precipitates</td>
<td>Fine keratic precipitates</td>
</tr>
<tr>
<td>White and quiet</td>
<td>2+ perilimbal injection</td>
</tr>
<tr>
<td>Gr1 cells, trace flare</td>
<td>Gr2-3+ cells, gr1 + flare</td>
</tr>
<tr>
<td>Normal, no nodules</td>
<td>Normal, no nodules</td>
</tr>
<tr>
<td>3/3 n/t</td>
<td>3/3 n/t</td>
</tr>
<tr>
<td>19 mmHg</td>
<td>IOP @ 1327 by Goldmann</td>
</tr>
<tr>
<td></td>
<td>22 mgHg</td>
</tr>
</tbody>
</table>
**OCULAR EXAMINATION: POSTERIOR SEGMENT**

<table>
<thead>
<tr>
<th></th>
<th>OD</th>
<th>OS</th>
</tr>
</thead>
<tbody>
<tr>
<td>PCIOL clear and centered</td>
<td></td>
<td>PCIOL clear and centered</td>
</tr>
<tr>
<td>Syneresis</td>
<td></td>
<td>Vitreous</td>
</tr>
<tr>
<td>0.15v/0.15h</td>
<td>C/D</td>
<td>0.25v/0.25h</td>
</tr>
<tr>
<td>NRRI, pink, no pallor or edema</td>
<td>Disc</td>
<td>NRRI, pink, no pallor or edema</td>
</tr>
<tr>
<td>No pathology</td>
<td>Macula</td>
<td>No pathology</td>
</tr>
<tr>
<td>Blot heme inferior, drusen scattered throughout arcades</td>
<td>Fundus</td>
<td>Indistinct white lesion with pigmented borders superior temporal to optic nerve</td>
</tr>
<tr>
<td>No pathology</td>
<td>Vessels</td>
<td>No pathology</td>
</tr>
<tr>
<td>Flat and intact 360 degrees</td>
<td>Periphery</td>
<td>Flat and intact 360 degrees</td>
</tr>
</tbody>
</table>
OCULAR EXAMINATION – ANCILLARY TESTING
### Working diagnosis

- Bilateral nongranulomatous panuveitis of unknown etiology

### Treatment

- Difluprednate QID OU
- Cyclopentolate 0.5% TID OU
- Order serology
- Referral to retina and uveitis specialists
DIFFERENTIAL DIAGNOSIS

Infectious
- Syphilis
- Tuberculosis
- Lyme’s Disease
- Toxoplasmosis
- Bartonella

Inflammatory
- HLA-B27 associated uveitis
- Sarcoidosis
- Herpes Simplex
- Behcet's Disease
- Lupus
Contact PCP to report hives on back that have spread to torso and legs.
Told it is probably viral and will go away on its own.

Went to urgent care and given steroid injection.
Was prescribed oral prednisone and an antihistamine.

Contacts PCP and reports hives remain and that he now has swollen lymph nodes in his neck and pain in his shoulder and back.
PCP ordered allergy consult.
PCP ordered inflammatory labs due to reported pain.

Allergy Consult - continue loratadine.
Referred to ED due to complaints of dizziness, shortness of breath, increased thirst, low grade fevers, and sores under his tongue.
Vet left ED AMA due to wait time.

ESR: 61 mm/hr
CRP: 5.279 mg/dL
RF: 18.2 I/mL
ANA: neg
PCP ordered rheumatology consult but vet moved from area before appointment.

Urgent Care Visit due to eye "getting so bad".
Prescribed erythromycin ointment.

Reports to Salem VAMC Emergency Department.
Consulted with Infectious Disease and next day admission with lumbar puncture was recommended
## LUMBAR PUNCTURE AND CSF ANALYSIS

<table>
<thead>
<tr>
<th>Test</th>
<th>Result</th>
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<tbody>
<tr>
<td>VDRL, CSF</td>
<td>Reactive 1:1</td>
</tr>
<tr>
<td>Protein</td>
<td>59.1 mg/dL</td>
</tr>
<tr>
<td>Glucose</td>
<td>56 mg/dL</td>
</tr>
<tr>
<td>Turbidity</td>
<td>Clear</td>
</tr>
<tr>
<td>Color</td>
<td>Colorless</td>
</tr>
<tr>
<td>RBC</td>
<td>0 cells/uL</td>
</tr>
<tr>
<td>Nucleated cells</td>
<td>32 cells/uL</td>
</tr>
<tr>
<td>Lymphocyte %</td>
<td>98%</td>
</tr>
<tr>
<td>Monocyte %</td>
<td>2%</td>
</tr>
<tr>
<td>Supernatant appearance</td>
<td>Colorless</td>
</tr>
</tbody>
</table>

*abnormal results are in bold

**Diagnosis:** NEUROSYPHILIS WITH OCULAR SYPHILIS
TREATMENT AND MANAGEMENT COURSE

Treatment

- **Treatment**: 4 million units of IV penicillin q4 hours x 14 days
- At 4 months, treatment was successful with a 4-fold decrease in RPR titer (1:4 from 1:64)
- Repeat lumbar puncture:
  - CSF VDRL: negative
  - Glucose: 61
  - Clear and colorless
  - Cell count not performed
- RPR will be repeated out to 2 years

Management

- Developed acute renal insufficiency from interstitial nephritis 2/2 PCN. Switched to ceftriaxone.
- Developed rash from ceftriaxone and was switched back to penicillin.
- Developed secondary membranous nephropathy confirmed by biopsy due to Syphilis leading to nephrotic syndrome. Treated with oral steroids, furosemide, losartan, and chlorthalidone. He remains in CKD stage 3
- Found to have C. Diff. Treated with oral vancomycin.
- Echocardiogram showed mildly dilated aortic root and mildly enlarged ascending aorta.
### Ocular Outcome

**Day 2**
- Subjective improvement in vision and comfort.
- Tr cell OD; 1+ cell OS
- Stable vitritis and retinal findings OS
- **IOP: 16/36**
- Timolol started bid OU

**Day 15**
- No cell OD; trace cells OS
- Rare vitreous cells
- Stable retinal findings
- IOP controlled with timolol

**7 weeks**
- 20/30 OD, 20/25 OS
- No cells OD, 1-2 cells OS
- **Vitritis resolved**
- **Retinal findings appear inactive**
- IOP controlled with timolol

**2 months**
- Uveitis specialist stopped cyclopentolate and timolol
- Decrease Durezol to once daily x 1 week then discontinue

**4 months**
- Uveitis specialist note indicates that the eyes remain quiet with no recurrence of inflammation off treatment.
IMAGING DAY ONE VS 7 WEEKS – LEFT EYE
**DISCUSSION**

- **Organism:** Treponema Pallidum
  - corkscrew shaped spirochete
- **Mode of Transmission:**
  - Direct contact with chancre
- **Neurosyphilis occurs in 5-10% of untreated cases**
  - 1/3 are asymptomatic
  - 70% spontaneously revert

---

**Table: Disease Stage, Findings, Timing**

<table>
<thead>
<tr>
<th>Disease Stage</th>
<th>Findings</th>
<th>Timing</th>
</tr>
</thead>
<tbody>
<tr>
<td>Primary Syphilis</td>
<td>Painless chancre on the site of inoculation (genitalia, lips, anus, oropharynx, conjunctiva), non-tender regional lymphadenopathy, uveitis.</td>
<td>May present 1 week - 3 months after initial inoculation. Chancres will usually heal on their own in 1 to 3 weeks</td>
</tr>
<tr>
<td>Secondary Syphilis</td>
<td>Fever, rash (many variations but the classic rash is described as a maculopapular rash that involves the upper trunk, palms, and soles), alopecia, pharyngitis, CNS involvement, uveitis, scleritis, renal dysfunction, and hepatitis.</td>
<td>Weeks to months after initial chancre (though may still have the primary chancre in cases with HIV)</td>
</tr>
<tr>
<td>Early Latent</td>
<td>Asymptomatic though positive serological testing</td>
<td>First year after initial infection</td>
</tr>
<tr>
<td>Late Latent</td>
<td>Asymptomatic though positive serological testing</td>
<td>More than 1 year after initial infection or initial infection date is unknown.</td>
</tr>
<tr>
<td>Tertiary Syphilis</td>
<td>Gummatous disease, cardiovascular lesions (aortitis), Argyll-Robertson pupils, optic atrophy, vasculitis, uveitis, scleritis.</td>
<td>1-30 years after initial infection</td>
</tr>
<tr>
<td>Neurosyphilis</td>
<td>Early: meningitis, uveitis, stroke, hearing loss Later: Tabes dorsalis, Argyll-Robertson Pupils, general paresis</td>
<td>May occur during any time throughout the infectious process. Late neurosyphilis usually develops 10 to 25 years after initial infection.</td>
</tr>
</tbody>
</table>
DISCUSSION

### Treatment for Neurosyphilis

- **10–14-day course of systemic antibiotics**
  - Preferred: 18 to 24 units per day of IV Penicillin G
  - Alternative: 2g daily of ceftriaxone
- **Repeat lumbar puncture** 6 months post-treatment. Continue to repeat every 6 months until cell count is normal and CSF-VDRL is nonreactive.
- Re-treat if CSF WBC count does not decrease by 6 months or CSF-VDRL does not decrease by 4-fold or become nonreactive by 1 year post treatment.

<table>
<thead>
<tr>
<th>Treponemal testing results (FTA-ABS, MHA-TP, EIA)</th>
<th>Nontreponemal testing results (VDRL, RPR)</th>
<th>Interpretation</th>
</tr>
</thead>
<tbody>
<tr>
<td>Nonreactive</td>
<td>Nonreactive</td>
<td>No syphilis infection.</td>
</tr>
<tr>
<td>Reactive</td>
<td>Reactive</td>
<td>Confirmed active syphilis if no prior infection. Recurrent infection if nontreponemal tests return four-fold greater than post-treatment titers.</td>
</tr>
<tr>
<td>Reactive</td>
<td>Nonreactive</td>
<td>Latent syphilis or previously treated syphilis infection.</td>
</tr>
<tr>
<td>Nonreactive</td>
<td>Reactive</td>
<td>Likely false-positive nontreponemal testing.</td>
</tr>
</tbody>
</table>
SUMMARY

- 63-year-old Caucasian male presented with blurred vision, redness, and irritation associated with persistent rash, sores, joint/back pain that had not resolved after seeing multiple health care providers.
- Serologic testing returned positive for syphilis, and he was diagnosed with ocular syphilis. Lumbar puncture was performed confirming neurosyphilis. Staging points to secondary syphilis with ocular and neurosyphilis.
- A 14-day treatment of IV Penicillin G was started. The veteran developed acute renal insufficiency as a result of the treatment and was switched to ceftriaxone. Unfortunately, he developed a rash and was switched back to penicillin to finish his treatment course. He was also treated for C. Diff infection.
- He developed membranous nephropathy secondary to syphilis that was treated with furosemide and steroids. He currently has CKD stage 3b.
- He was also found to have a dilated aortic root, which is likely coincidental and not a result of aortitis from syphilis as this typically does not develop until 10-30 years after initial infection during tertiary syphilis.
- Treatment was successful with 4-fold decrease in RPR titer and resolution of ocular symptoms.


ACKNOWLEDGEMENTS

- David A. Martinez, OD – 2020-2021 Optometry Resident
- Dr. Stephanie Nagy-Agren and Dr. Shikha Vasudeva, Salem VAMC Infectious Disease Specialists
Thank you for your attention

Please clap and don't ask difficult questions