

Grand Rounds

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SUNY Downstate Medical Center

Ophthalmology

January 16, 2014

Our Patient

HPI: 41 year old male, first visit here, complaining of blurry vision at night for the past 2-3 weeks.

- Previously seen elsewhere for poor vision 3 years ago, told that “he has fluid in the left eye” but reports vision had been better in the interim.
- Denies trauma, excessive lacrimation, flashes, floaters or curtains.
- Of note, he has a month history of intermittent parotid gland swelling, elevated ACE levels, and hilar adenopathy on CXR. He is scheduled by ENT for parotid gland biopsy.

Core Competency: Patient Care

Our Patient

- **PMH:** HTN
- **PSH:** none
- **POH:** self-reported poor vision OS 3 years ago, no refractive error, surgery or trauma
- **FH:** no blindness or glaucoma
- **SH:** denies smoking, drugs, illicit
- **Meds:** HCTZ
- **Gtts:** none
- **Allergies:** NKDA

Core Competency: Patient Care

Our Patient

Vitals: BP ranges from 130s-150s / 70s-90s

Exam:

DVasc: 20/20 OD, 20/100-2 PH 20/60 (nasal letters only)

Pupils: 4 to 2 OU, no apd

EOM: full OU

CVF: full OU

Tapp: 10, 11 @ 1:20 pm

SLE:

LLA: wnl OU

C/S: w/q OU

K: clear OU

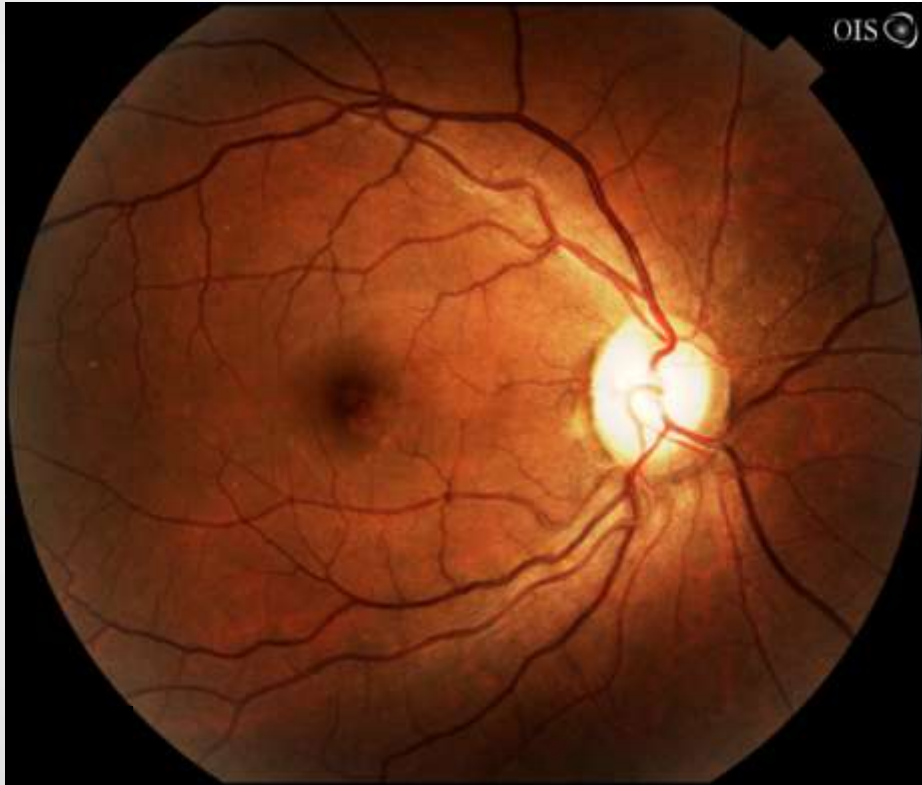
AC: deep and quiet OU

I/P: round and reactive OU

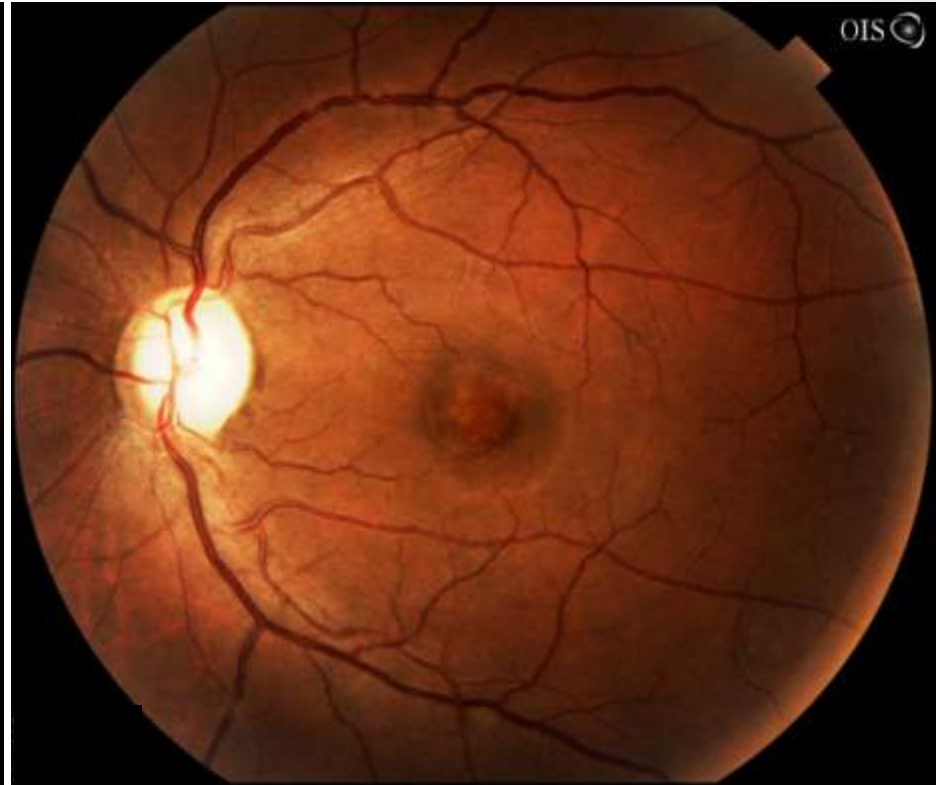
L: clear OU

Core Competency: Patient Care

Our Patient



OD

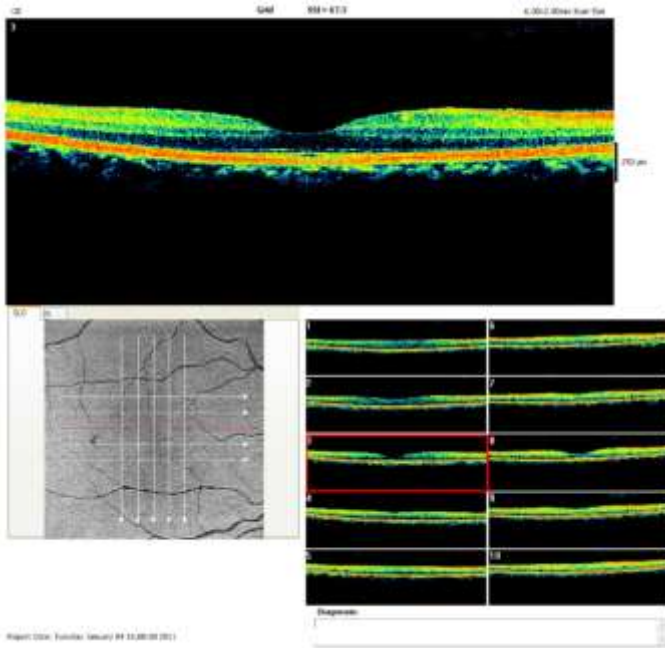


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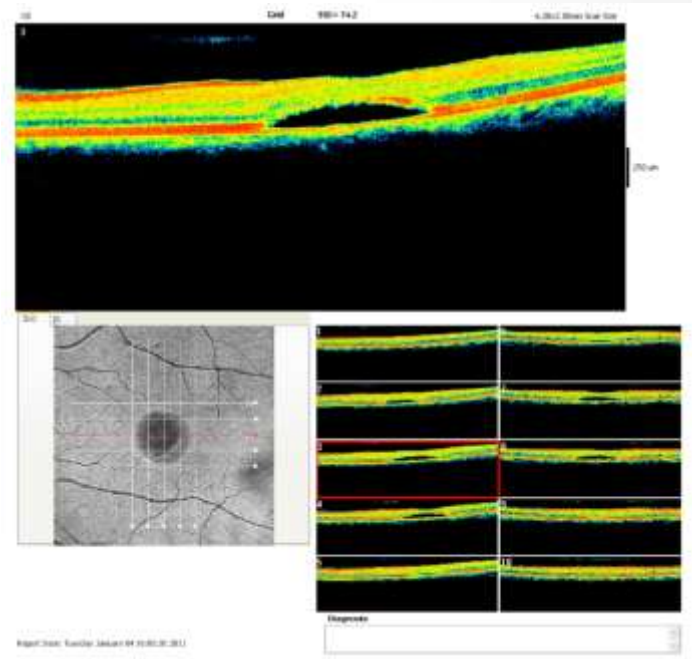
Core Competency: Patient Care

Next Step



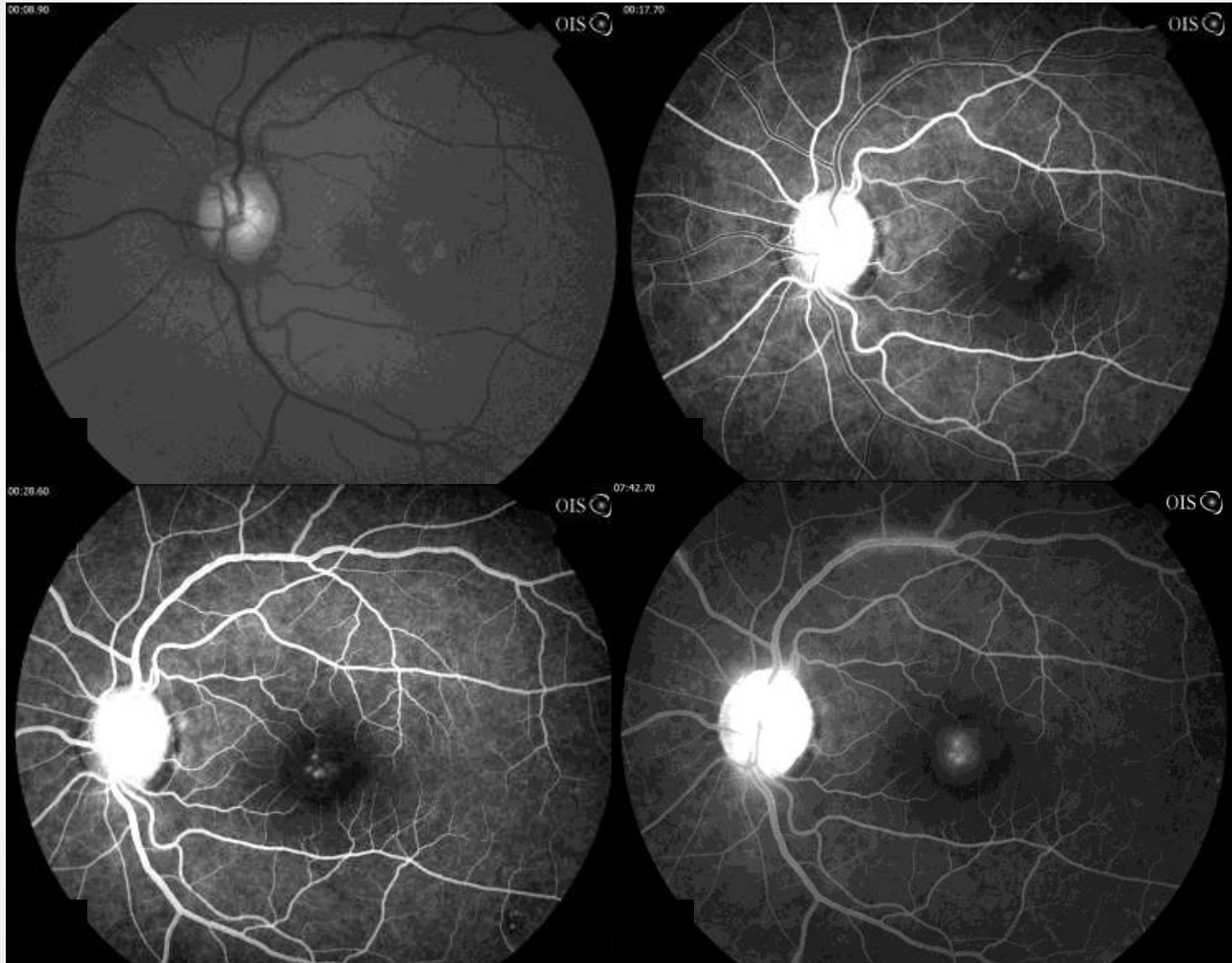


OD

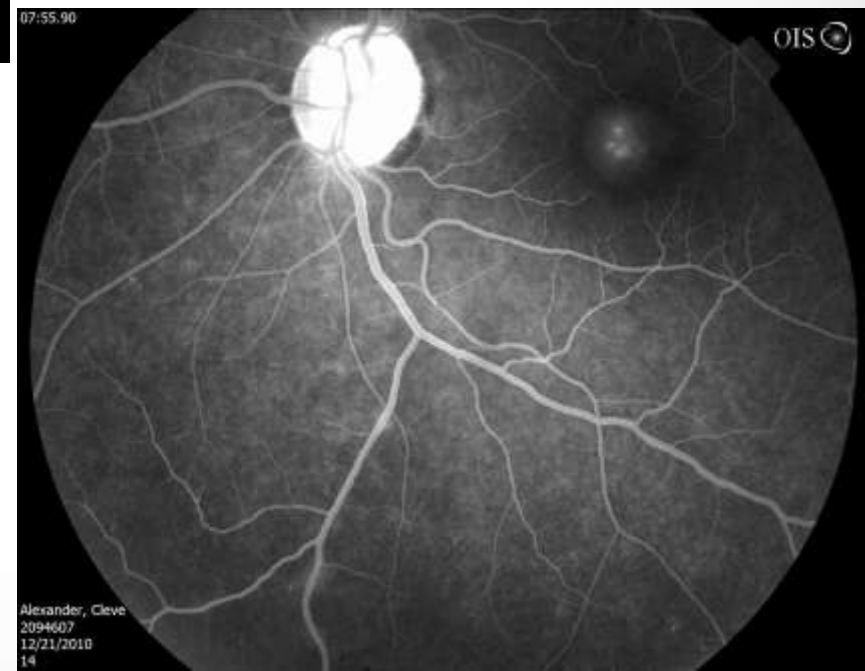


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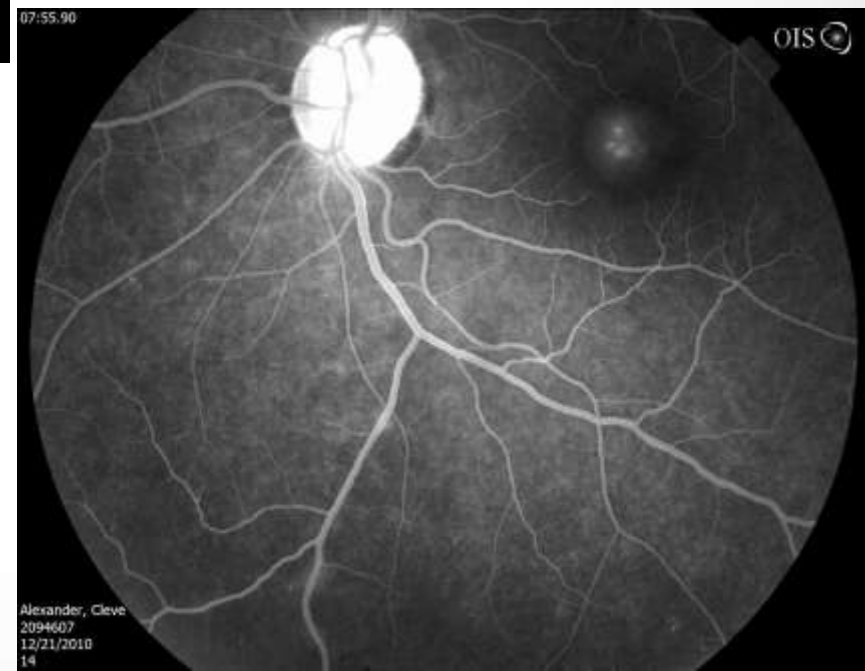
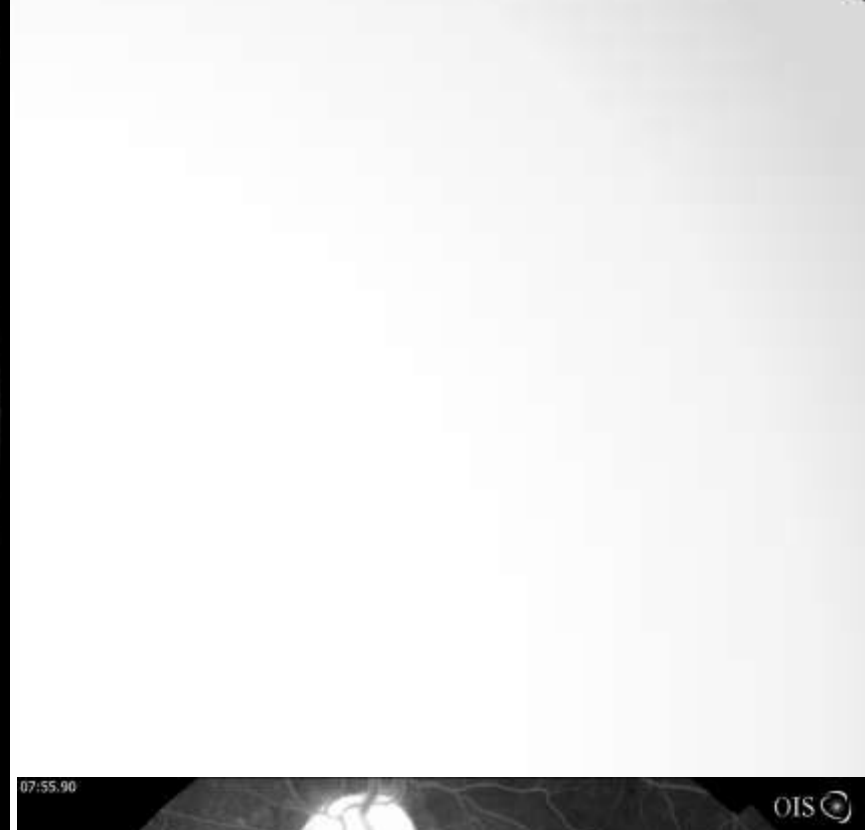
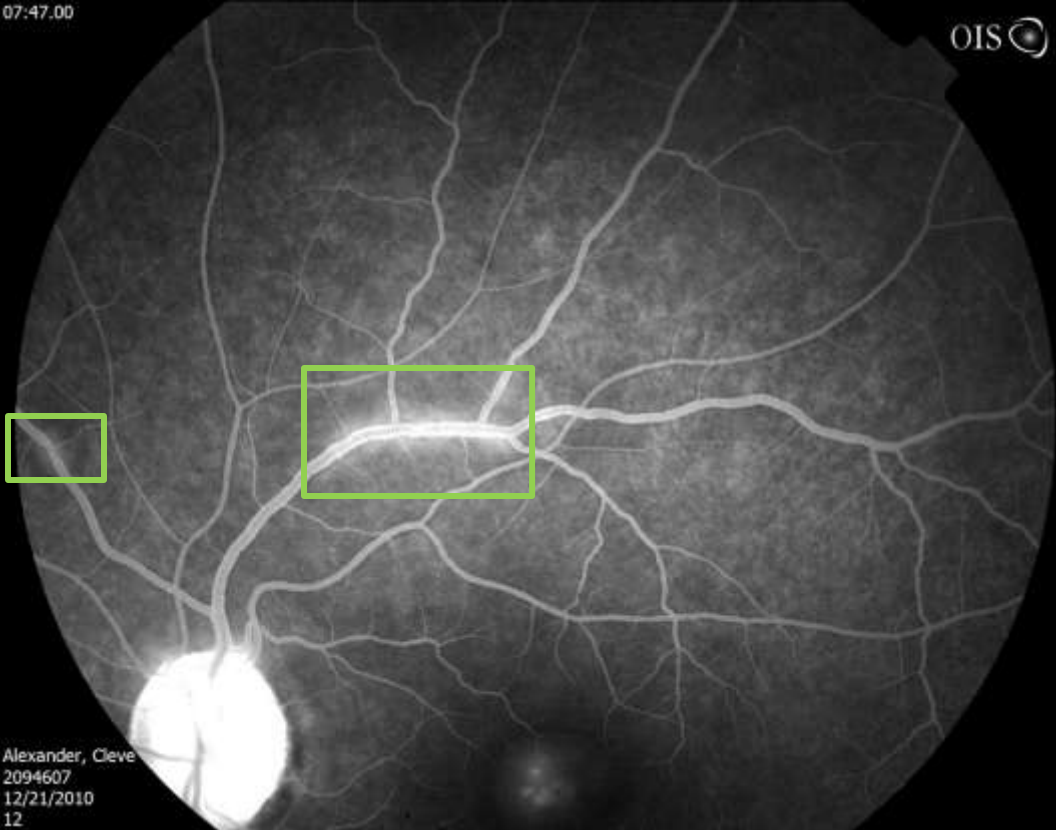
Core Competency: Patient Care



Core Competency: Patient Care



● Core Competency: Patient Care



● Core Competency: Patient Care

Further workup?

Review of Ancillary Tests

- **B-scan**: symmetric choroidal thickness
- **Labs**: ACE 185 (8-52)
- **CT Chest**: mediastinal adenopathy with 1.2cm R paratracheal node and 1.1 cm node anterior to carina
- **Bronchial Biopsy**: RLL: multiple non-necrotizing granulomata
- **Parotid Gland Biopsy**: unable because swelling resolved

Core Competency: Patient Care

Goals

- Periphlebitis
 - Differential diagnosis
- Sarcoidosis: Posterior Segment
 - Presentation
 - Epidemiology
 - Findings
 - Treatment
 - Prognosis

Differential Diagnosis

Periphlebitis

- Panuveitis
 - Sarcoidosis
 - Behcet's
 - Tuberculosis
 - Sympathetic Ophthalmia
 - VKH
- MS
- Infectious
 - ARN, Toxo, CMV
- Leukemia

Unilateral Neurosensory Detachment

- CSCR
- Uveitis
- RD
- CNV

Core Competency: Medical Knowledge

Differential Diagnosis

Periphlebitis

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Unifying Diagnosis?

Core Competency: Medical Knowledge

Differential Diagnosis

Periphlebitis

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Unilateral Neurosensory Detachment

- CSCR
- **Uveitis**
- RD
- CNV

Unifying Diagnosis?

Core Competency: Medical Knowledge

Retinal Vasculitis

- **Starts with**
 - Breakdown of inner blood-retina barrier (endothelial cell junctions)
 - Diapedesis of inflammatory cells
- **Leads to**
 - Continued leak -> edema, or
 - Occlusion -> neovascularization and fibrosis
- **“Healing phase”**
 - Gliosis of vascular walls (venous sheathing)
 - 4 types of peripheral venous sheathing:
 - Congenital sheathing (persistent embryonic connective tissue)
 - Post-inflammatory sheathing (perivascular gliosis)
 - Halo sheathing (venous sclerosis)
 - Periphlebitic sheathing (active periphlebitis)

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FA: diffuse microvascular leakage
found in 100% of cases

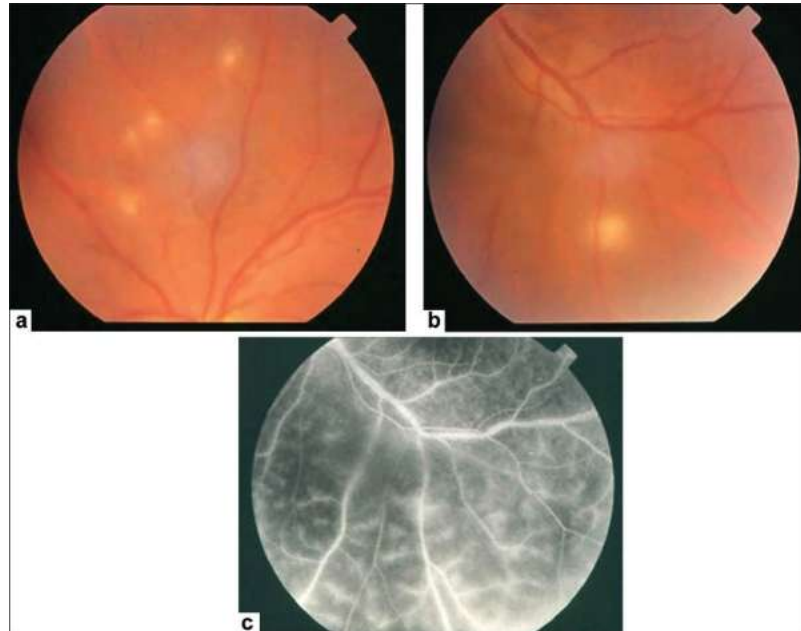


Photo credit: Differential Diagnosis of Retinal Vasculitis

Core Competency: Medical Knowledge

Retinal Vasculitis

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 - **Tuberculosis**
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FA: Most common retinal finding is **periphlebitis**

Usually a presumed diagnosis based on symptoms and CXR findings



Core Competency: Medical Knowledge Photo credit: Differential Diagnosis of Retinal Vasculitis

Retinal Vasculitis

- Panuveitis
 - Sarcoidosis
 - ~~Behcet's~~
 - ~~Tuberculosis~~
 - **Sympathetic**
 - VKH

No history concerning for rupture or previous PPV

- MS
- Infectious
 - ARN, Toxo, CMV
- Leukemia

Core Competency: Medical Knowledge

Retinal Vasculitis

- Panuveitis
 - Sarcoidosis
 - ~~Behcet's~~
 - ~~Tuberculosis~~
 - ~~Sympathetic~~
 - **VKH**

No history consistent with prodromal, acute, convalescent, or recurrent phase

- MS
- Infectious
 - ARN, Toxo, CMV
- Leukemia

Core Competency: Medical Knowledge

Retinal Vasculitis

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- **MS**
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FA: patchy perivascular cuffing indicates activity, wall whitening with no leakage represents chronic sclerotic change

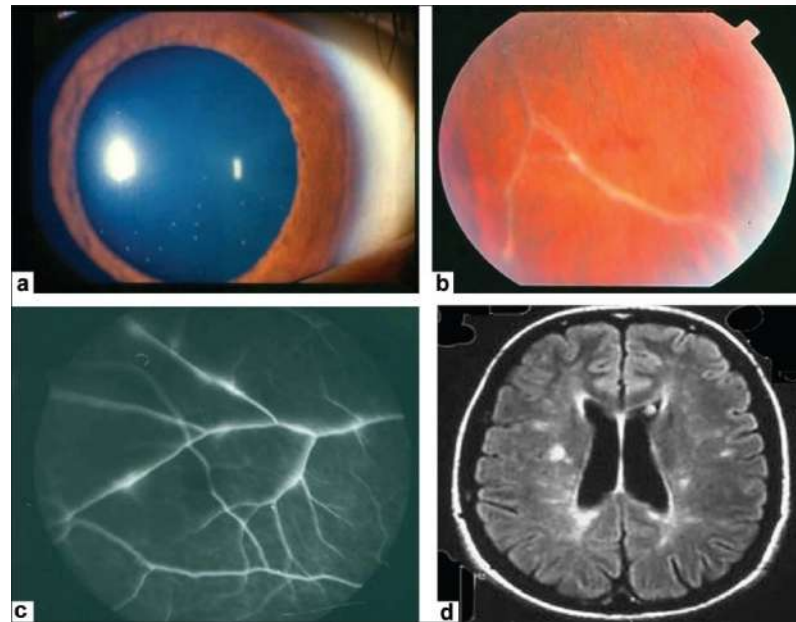


Photo credit: Differential Diagnosis of Retinal Vasculitis

Core Competency: Medical Knowledge

Retinal Vasculitis

- Panuveitis
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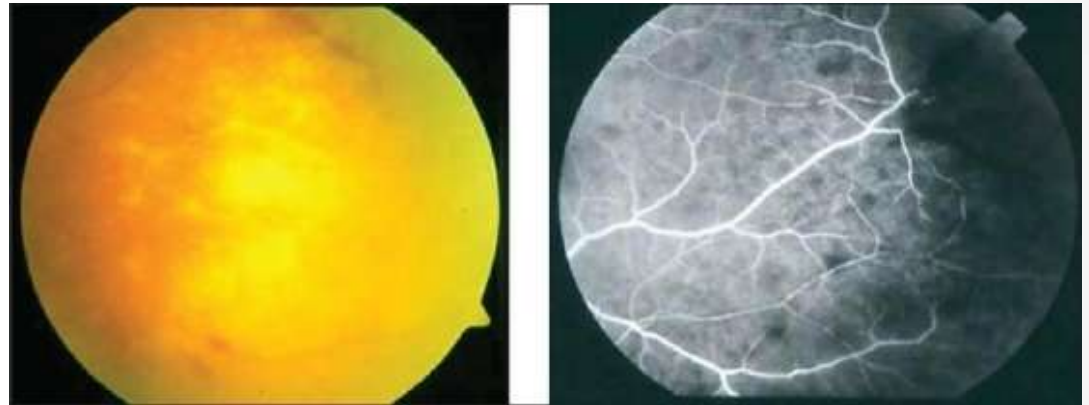


Photo credit: Differential Diagnosis of Retinal Vasculitis

Core Competency: Medical Knowledge

Sarcoidosis

○ Epidemiology

- Affects all ethnic groups, highest prevalence in northern Europeans
- In US, affects AA > white
- ACCESS study: workplace exposure to mold, musty odors, insecticides is associated with increased risk of sarcoidosis
- HLA-DRB1
- Mycobacterial organisms may be etiologic factors

Core Competency: Medical Knowledge

Sarcoidosis

- **Pathology**
 - Noncaseating granuloma
 - Sarcoid tubercle: epithelioid cells, Langhans, rim of lymphocytes
- **Systemic Manifestations**
 - Lofgren syndrome
 - Heerfordt syndrome
 - Acute and chronic forms

Core Competency: Medical Knowledge

Sarcoidosis

○ Ocular Manifestations

- Orbital, lacrimal, conjunctival, corneal, glaucomatous, iris
- Anterior uveitis found in 67% of patients
- Intermediate uveitis
- Posterior uveitis found in 25-30%
 - Macular edema: most common sight-threatening consequence of sarcoid uveitis
 - Neovascularization is most significant factor contributing to poor prognosis [Spalton et al. 1981]
 - Sequelae include vitreous hemorrhage, ischemia, and retinal detachment
- Neuro

Core Competency: Medical Knowledge

Posterior Sarcoid Uveitis

- 1/3 have no anterior findings! [Wertheim 2005]
- Active periphlebitis: fluffy white haziness around vessels
- Candle-wax drippings:
 - segmental venular cuffing, sheathing and perivenous exudation
 - non-necrotising granulomas
 - not pathognomic for sarcoid! (syphilis, TB, Behcet's)
- 15% of posterior uveitis develop neovascularization
- FA: venule wall staining, focal leakage, capillary closure, cystoid macular edema, neovascularization

Sarcoidosis

○ Diagnosis

- Screening:
 - CXR: 90% sensitivity, thin-cut spiral CT is more sensitive
 - ACE, lysozyme are better for tracking active disease (not diagnostic or specific)
 - Positive gallium + ACE elevation + uveitis is highly specific
- Vitreous fluid analysis
 - vitreous tap and immunological analysis
 - CD4/CD8 > 3.5: sensitivity of 100% and a specificity of 96.3%.
- Diagnosis ultimately made with biopsy

Core Competency: Medical Knowledge

Sarcoidosis

○ Treatment

- TB skin test
- Anterior uveitis:
 - topical steroids (Pred Forte 1% > generic > prednisolone phosphate) + cycloplegia
 - Lotemax; Vexol and FML good for maintenance
- Vision threatening posterior uveitis:
 - Systemic corticosteroids (prednisone 40-80 mg/day)
 - Intravitreal fluocinolone acetonide implant
- Immunomodulation therapy
- PRP
- Surgery
 - cataract surgery risks much higher with anterior uveitis
 - Trabeculectomy or glaucoma drainage device
 - Vitrectomy

Core Competency: Medical Knowledge

Vitreotomy for Sarcoid

- Inconclusive sarcoid diagnosis [Matsuoka, Clin Ophthal 2012]
 - Case reports
 - Epithelioid and multinucleated giant cells pathognomic for sarcoid
- Vitreous opacity [Ieki et al, Ocul Immunol Inflamm. 2004]
 - 11 eyes with vitreous opacities and uveitis associated with sarcoidosis resistant to corticosteroid therapy
 - 7 gained 2 or more lines of Snellen VA 6 months postoperatively
- Epiretinal membrane [Kiryu, JPn J Ophthalmol. 2003]
 - 11 eyes with epiretinal membrane and uveitis with sarcoidosis underwent pars plana vitrectomy
 - 9 gained 2 or more lines of Snellen VA 1-12 months after surgery; however 4 of these lost 2 or more lines by final visit 2/2 cataract formation and membrane reformation

Sarcoidosis

- **Prognostic factors**

- Chronic posterior uveitis, glaucoma, delay in presenting to uveitis specialist for > 1 year, presence of intermediate or posterior uveitis associated with visual loss

Returning to our patient

- Multiple visits over the last 3 years
- VA improved from 20/100-2 to 20/60, neuroretinal detachment resolved, no sub-retinal fluid
- Course involved no steroids (minimal inflammation)
- Still following closely

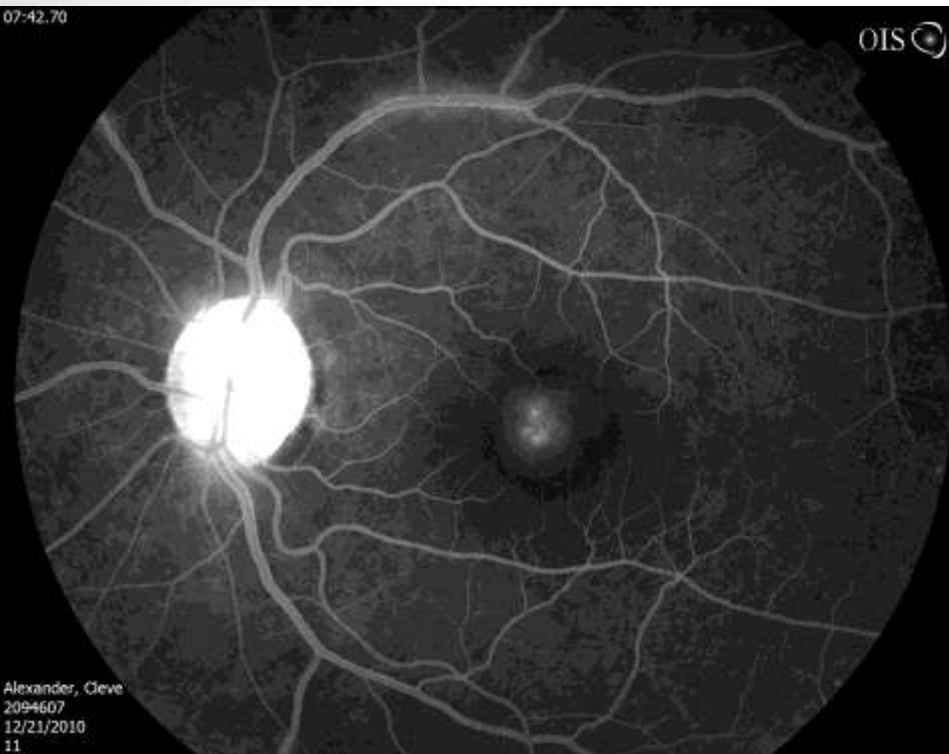
Core Competency: Patient Care

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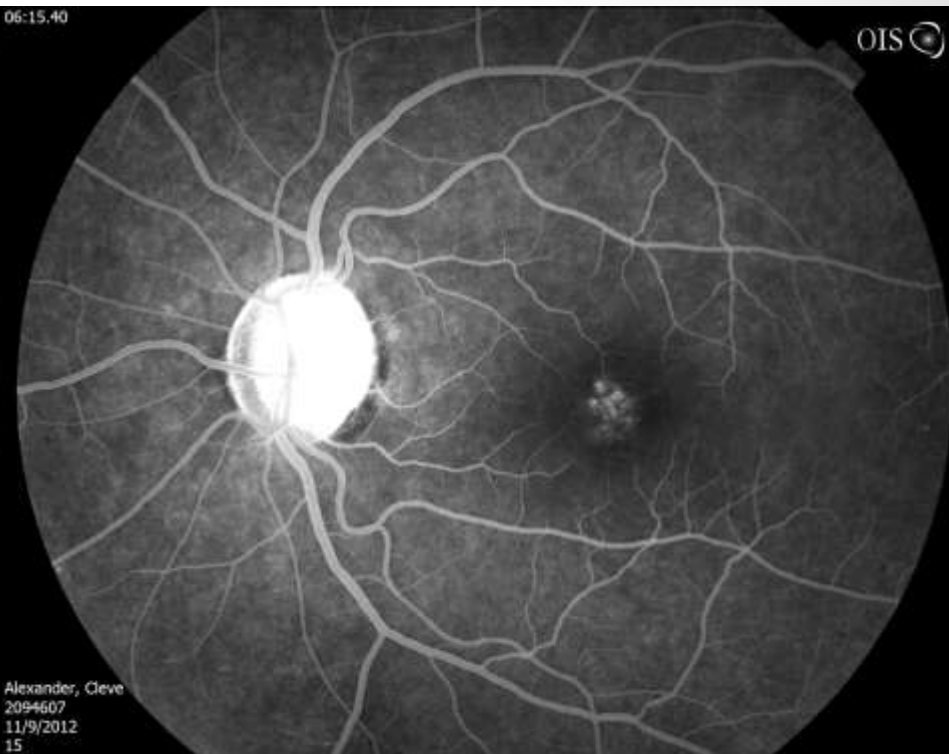
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OIS



Alexander, Cleve
2094607
12/21/2010
11

On presentation



Alexander, Cleve
2094607
11/9/2012
15

2 years later

Reflective Practice

- This case represented application of careful history taking, ophthalmic examination and creation of a complete differential diagnosis to evaluate and treat a complex retinal disorder.

Core Competencies

- Patient Care**: The case involved thorough patient care and careful attention to the patient's past medical history.
- Medical Knowledge**: This presentation allowed me to review the presentation, differential diagnosis, proper evaluation/work up, treatment options for sarcoidosis.
- Practice-Based Learning and Improvement**: This presentation included a literature search of current diagnostic and treatment modalities.
- Interpersonal and Communication Skills**: The patient was made aware of the complexity of the diagnosis and is actively engaged
- Professionalism**: The patient provided information for the grand rounds presentation only after careful exam, explanation of findings and use of information.
- Systems-Based Practice**: The patient's history was obtained through multiple health care deliverer's (PMD, ENT, ophthalmology)

References

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- Kanski
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- Spalton DJ, Sanders MD. Fundus changes in histologically confirmed sarcoidosis. *Br J Ophthalmol.* May 1981;65(5):348-58
- Matsuoka M et al. Two cases of ocular sarcoidosis in which vitreous cytology was useful for supporting the diagnosis. *Clinical Ophthalmol.* 2012; 6:1207-9

Acknowledgements

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- Dr. Joseph Tseng