

Ophthalmology Grand Rounds

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February 16, 2012

Case Presentation

- CC: “seeing a dark spot”
- HPI:
46 yo AA male Hx HTN c/o dark spot in vision OD,
x3-4 weeks, constant
1 month Hx bilateral periorbital fullness OD>OS

Denies eye pain, flashes, floaters

Denies SOB, cough, trauma

Case Presentation

- PMHx: HTN
- Meds: metoprolol, HCTZ
- POHx: denies
- Gtts: none
- FHx: neg glc/blindness
- SHx: +smoking ½ - 1 ppd, social drinking, from North Carolina, was in National Guard
- NKDA

Case Presentation

- Dva sc: 20/30 OD 20/30 OS
- MRx:
+0.25 -0.50 x90 → 20/30+ OD
+0.50 -0.75 x110 → 20/20- OS
- Pupils: 4-2 ou no apd
- CVF: full ou
- Color: 10/10 ou
- Red-desat: +OD (orange)
- Tapp: 18/18 at 2:15pm
- BP 157/107, HR 79, RR 20,
100% on RA, Tc 97.9
- Gross/EOM: photo
- HEENT: CN III-XII intact



Patient Care, Interpersonal & Communication Skills



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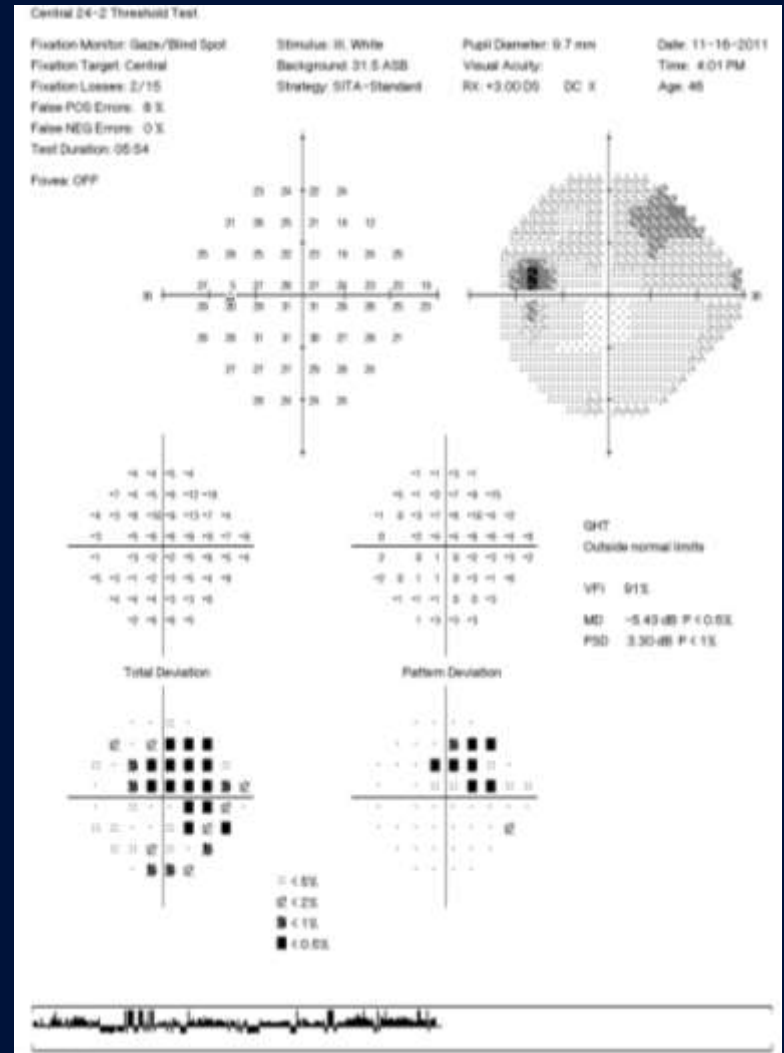
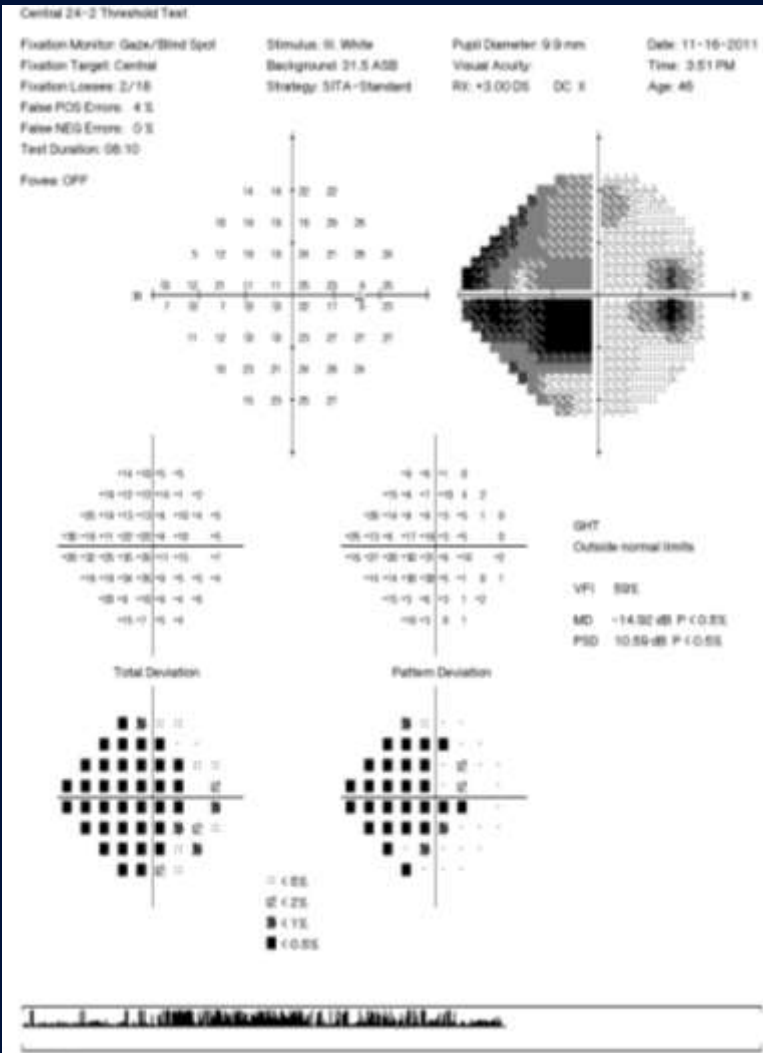
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Case Presentation

- SLE:
LLA: no lid lag
K: clear ou, no evidence exposure keratopathy
AC: d/q ou
IP: r/r ou, no nvi
L: clear ou
- DFE:
Vit: clear ou
ON: 0.2 severe disc edema w blurred margins & peripapillary hemes OD, 0.3 pink with mild disc edema OS
Mac: flat ou
BV/P: wnl ou, no vasculitis, heme, tears, holes



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Differential Diagnosis

- Lymphoproliferative disease
 - Lymphoid hyperplasia
 - Lymphoma
 - Multiple myeloma
- Idiopathic
 - Idiopathic inflammatory pseudotumor
 - Sarcoidosis
- Infectious
 - Tuberculosis

Next Step?

- Labs:
 - ACE, lysozyme
 - PPD
- Imaging
 - CXR
 - Orbits
- Lumbar puncture

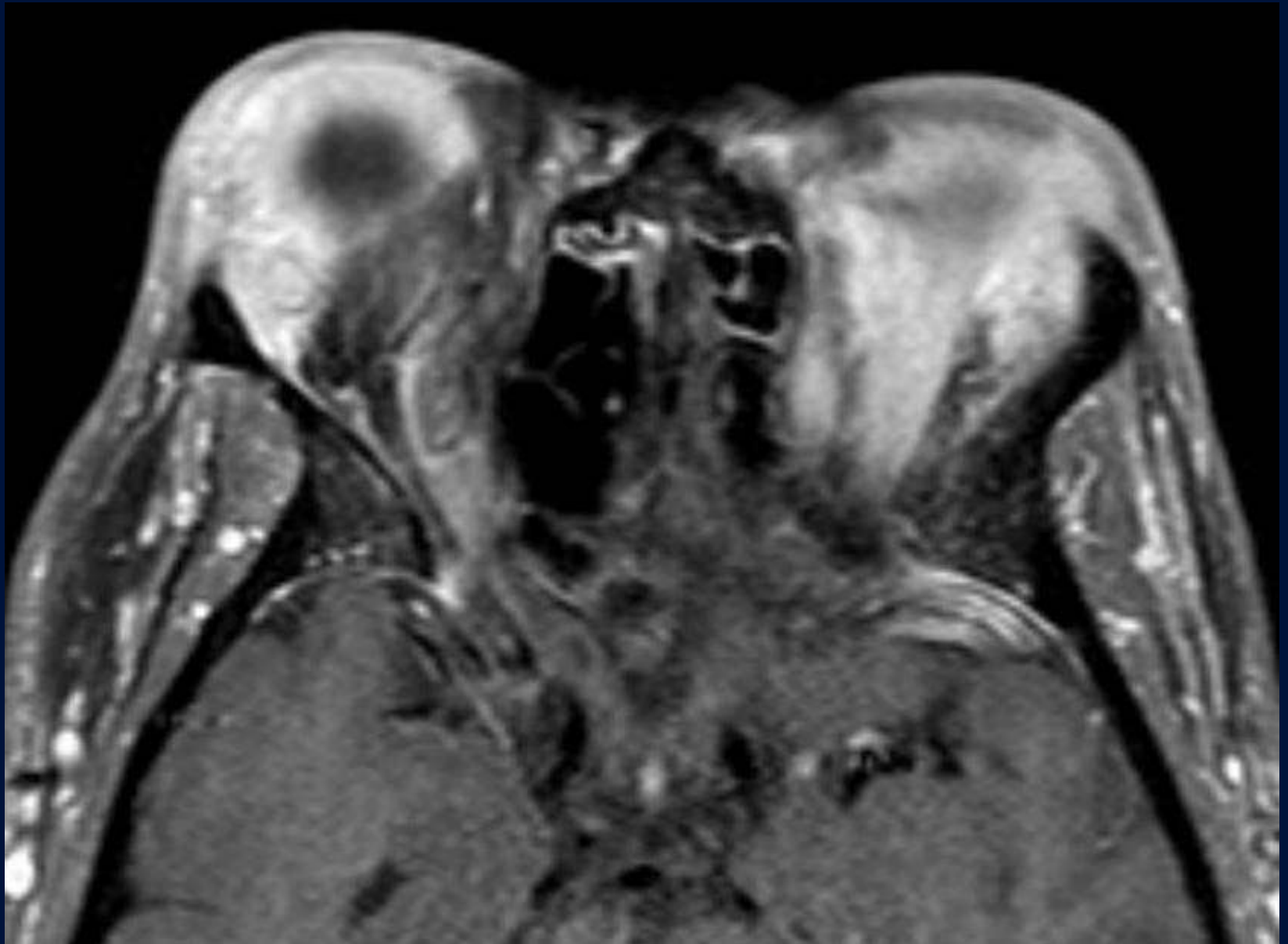
Case Presentation

- Labs:
 - ACE serum: 56 (↑)
 - Lysozyme: 15 (↑)

CBC, BMP, TFTs, CRP, C-ANCA, PPD WNL



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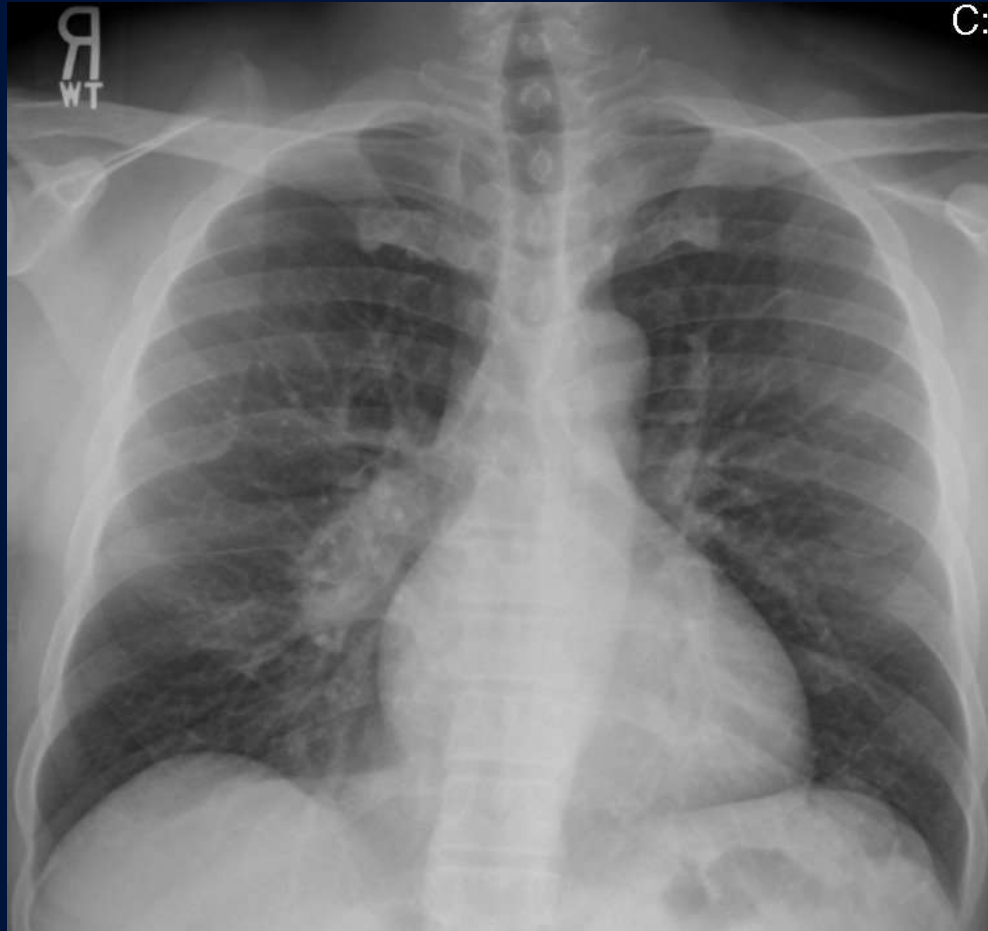
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NSE -3 SENSE

7 DM

Case Presentation



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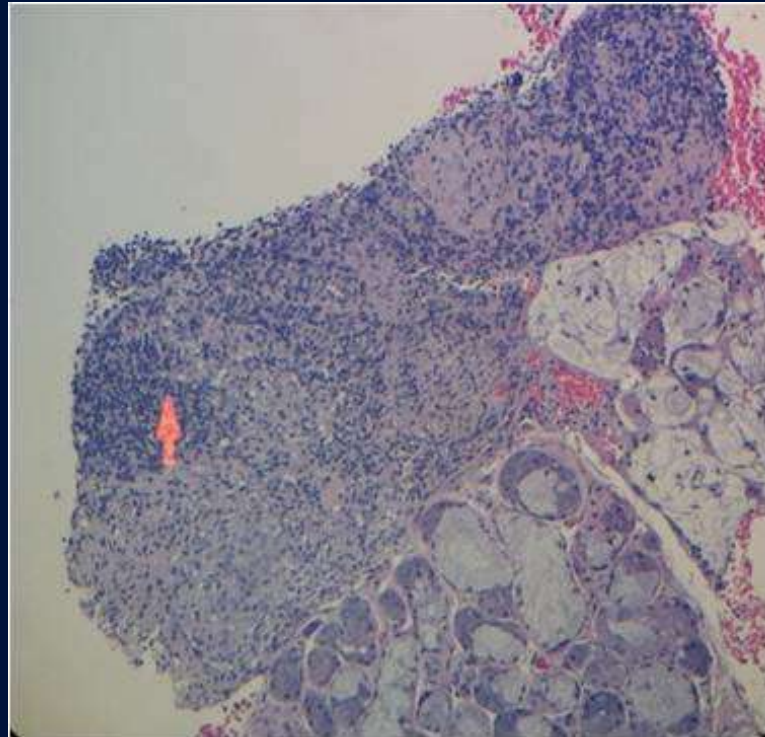
Case Presentation

- LP:
 - Opening Pressure 24 (nml 5-20mmHg)
 - ACE 3
 - Protein 42
 - Glucose 85
 - WBC 10 (nml <5mm³)
 - Cultures neg
 - Cytology neg

Case Presentation

- Transbronchial biopsy

Non-necrotizing granulomas, negative for AFB and fungi
Consistent with sarcoidosis



Patient Care, Interpersonal & Communication Skills

Recap

- 46yo M Hx HTN c/o dark spot in vision OD
- Exam reveals:
 - bilateral lacrimal gland enlargement OD>OS w superior orbital spillover OD
 - hypoglobus OD
 - disc edema OD>OS
 - nasal HVF defects OD>OS

Neurosarcoidosis

Epidemiology and Etiology

- Sarcoidosis estimated 15 per 100,000 people
More common in blacks
- 5-15% develop neurosarcoidosis (NS)
50% patients present with NS at the time
sarcoidosis first diagnosed
- Etiology likely multifactorial (immune, familial,
environmental)

Clinical Manifestations

- Uveitis (25%; ant>int>post)
- Cranial neuropathy--most common presentation
 - Facial--most frequently involved
 - Trigeminal
 - Oculomotor
 - Optic nerve
 - Abducens
 - Vestibular
 - Vagus
 - Others
- Peripheral neuropathy
- Granulomatous infiltration of the CNS
 - Seizure
 - Psychiatric symptoms
 - Space-occupying feature
 - Neuroendocrine dysfunction
- Idiopathic communicating hydrocephalus
- Aseptic meningitis

Diagnostic Labs

- Serum ACE (elevated 5-50%)
- Serum interleukin-2 receptor (IL2R)
 - Measure of T-cell activation, also elevated in lymphoma
 - Preferred for monitoring disease activity

Diagnostic Imaging

- MRI preferred neuro imaging, most sensitive
- High resolution CT preferred for chest
- Whole-body gallium scan or fluorodeoxyglucose (FDG) PET scan can be utilized to find inflammation or neoplasia
 - insensitive for CNS involvement and limited by poor specificity (particularly sarcoidosis vs lymphoma)

Lumbar Puncture and Biopsy

- CSF abnormalities usually non-specific; may have elevations of ACE, IgG, oligoclonal bands, CD4:CD8 lymphocyte ratios, lysozyme, B2-microglobulin levels
 - CSF-ACE may be used to monitor disease activity
- Caution with LP as patients may have elevated ICP
- Conjunctival, transbronchial, skin, lymph node, peripheral nerve biopsy
 - Demonstrating non-caseating granulomas

Spaide RF, Ward DL. Conjunctival biopsy in the diagnosis of sarcoidosis. (MEE 1990)

- Prospective study of 47 sarcoidosis suspects comparing transbronchial lung biopsy (TBBX) and conjunctival biopsy (CBX)--35 AA, 12 Caucasian
- 77.1% TBBX vs 45.7% CBX positive in the AA patients
33.3% TBBX vs 25% CBX positive in Caucasian patients
- TBBX positive in pts with pulmonary infiltrates **80.6%** (vs 37.5% without)
- CBX more likely to be positive in patients with pulmonary infiltrates (51.6% vs 23.1%)
- CBX positive with conjunctival follicles **66.7%** (vs 31.4% without), and **60%** with any ocular abnormality consistent with sarcoidosis (vs 25.9%)

Treatment

- Corticosteroids first line
 - Neuropathy treated with 0.5 mg/kg/day x4 weeks
 - Encephalopathy/vasculopathy with 1.0-1.5 mg/kg/day x4 weeks
 - Then slow taper by 5mg every 2 weeks as tolerated
 - If recurs, double the dose or give at least 10-20mg/day

Treatment

- Immunomodulating agents for refractory disease
- Mycophenolate mofetil effective for CNS
- Cyclosporine dosed at renal transplant levels
- Azathioprine, chlorambucil, cyclophosphamide targeted to lower total WBC count to 3500 or lymphocyte count to 1000 per mm³
- Radiation therapy for refractory disease

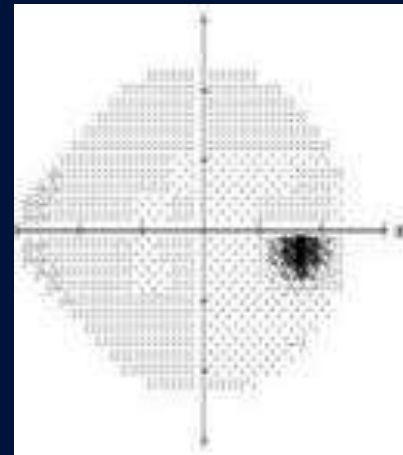
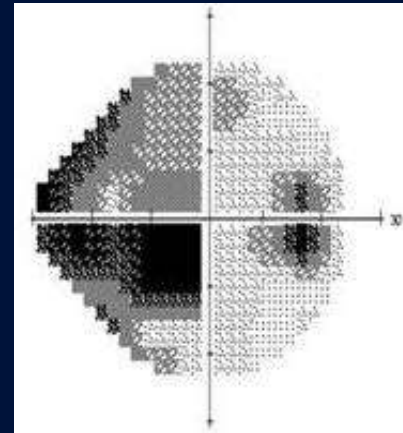
Prognosis

- 2/3rd monophasic illness
- May have relapsing –remitting course or progressive disease
- Approximately 10% die as a result from inflammatory process or its treatment

Patient Update

- Admitted for IV steroids, discharged on prednisone 60mg PO with taper
- Started azathioprine 100mg PO daily with prednisone 20mg PO daily
- Following up with rheumatology this week

Patient Update



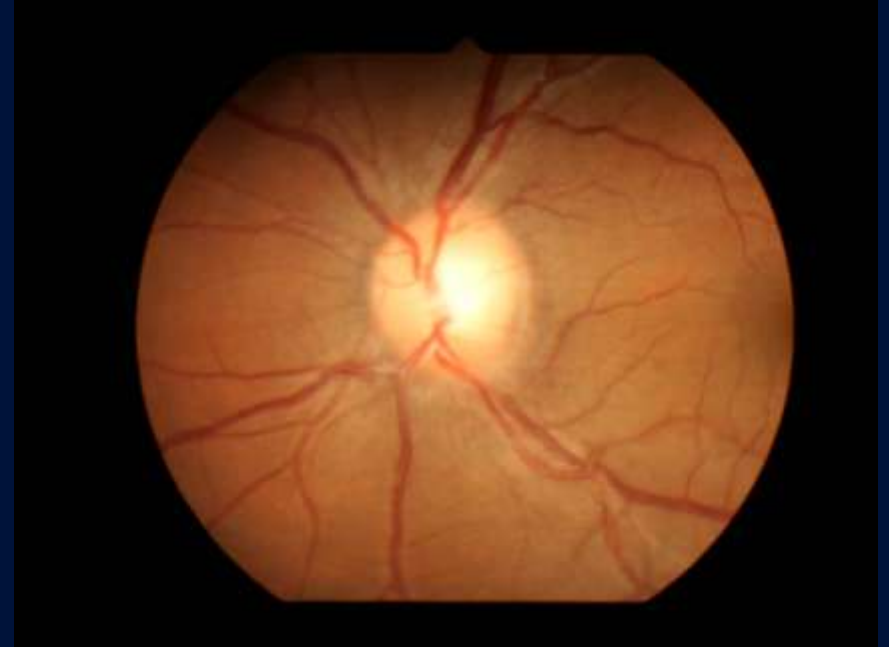
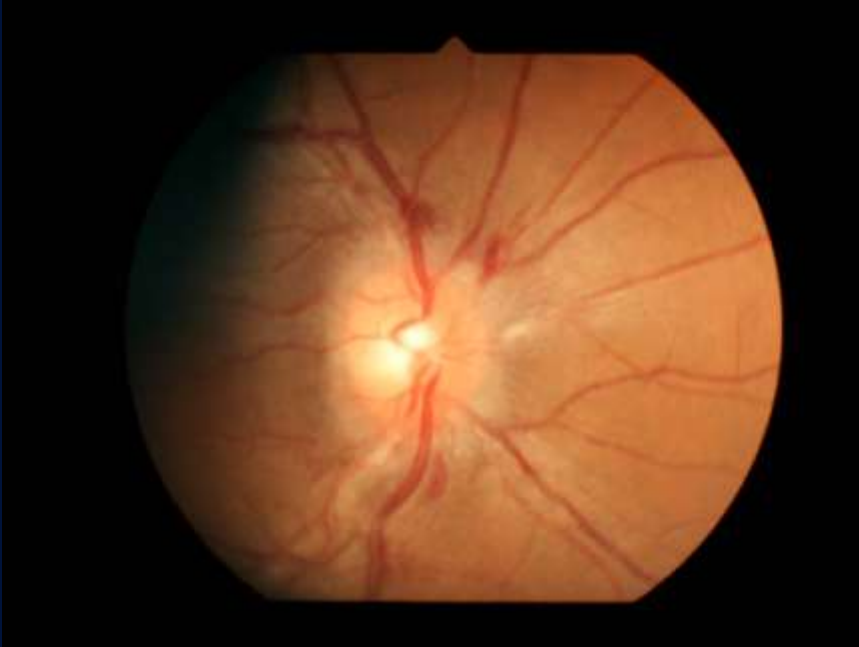
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Patient Update



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Fundus Photos – after inpt steroids



Patient Care, Interpersonal & Communication Skills

Reflective Practice

- **Patient Care:** This patient was appropriately evaluated by physical examination, imaging, and testing. Patient received appropriate counseling and treatment for his ocular diseases.
- **Medical Knowledge:** This presentation provides an overview of neurosarcoidosis, including associated manifestations.
- **Practice-Based Learning and Improvement:** This presentation included information regarding the evaluation, differential diagnoses, and evidence-based management of patients with neurosarcoidosis.
- **Interpersonal and Communication Skills:** The patient was appropriately counseled on the associated disease manifestations of neurosarcoidosis. Treatment options were explained, follow-up was maintained.
- **Professionalism:** Patient was treated appropriately and with due urgency at KCHC over the course of several months.
- **Systems-Based Practice:** Appropriate care was provided via the services of neurology, radiology, pathology, rheumatology, ENT, and ophthalmology.

Thank You

Dr. Shinder

Dr. EC Lazzaro

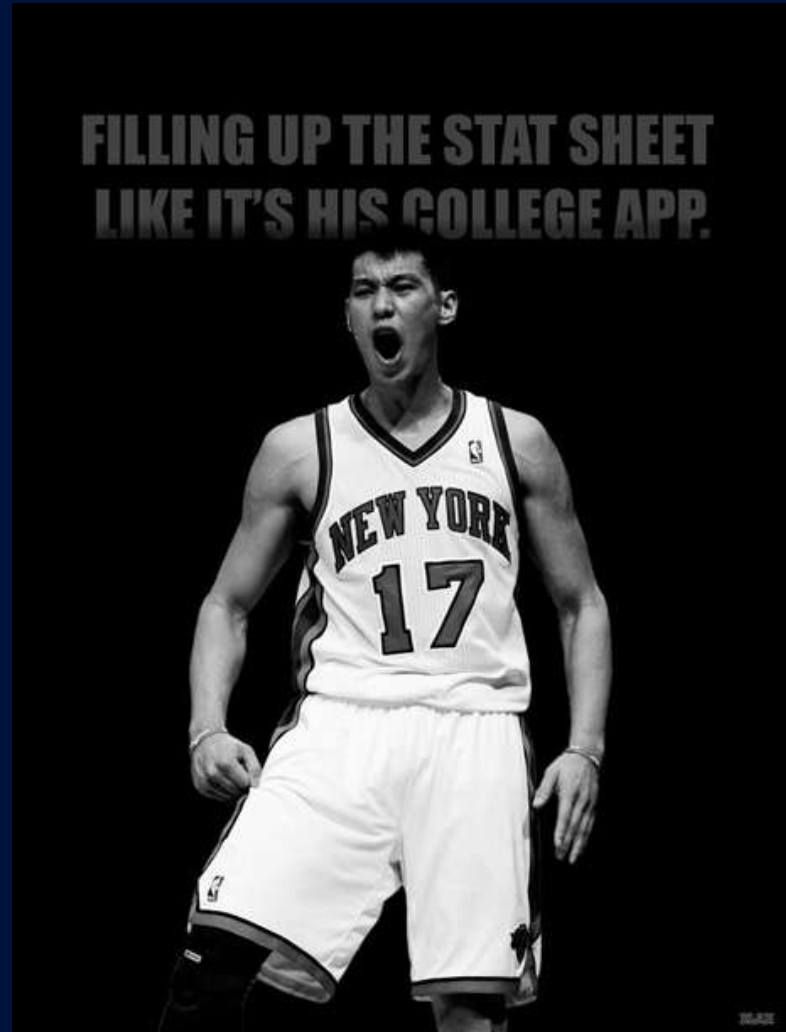
Dr. Scott

Dr. Asoma

Dr. Thode

Dr. Mostafavi

Our patient



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References

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