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 \(\rightarrow\) 20/30+ OD
  +0.50 -0.75 x110 \(\rightarrow\) 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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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
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
Patient Care, Interpersonal & Communication Skills
Case Presentation

Patient Care, Interpersonal & Communication Skills
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
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 x 4 weeks
  – Encephalopathy/vasculopathy with 1.0-1.5 mg/kg/day x 4 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

• 2/3\textsuperscript{rd} 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

Patient Care, Interpersonal & Communication Skills
Patient Update

Patient Care, Interpersonal & Communication Skills
Fundus Photos – after inpt steroids
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
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Dr. Scott
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Dr. Thode
Dr. Mostafavi
Our patient

Patient Care, Interpersonal & Communication Skills
References


