Looking at Lab Work: A Case Report of Isolated Optic Disc Edema Secondary to Ocular Syphilis

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Presentation Format: Case Report, poster first, paper second

General Topic: Ocular Disease

Primary Topic: Posterior Segment

Abstract:
Optic neuropathy has a myriad of etiologies and syphilis may be overlooked. With the recent re-emergence, this case shows the importance of ordering proper lab tests since irreversible visual consequences can result if left undiagnosed.

I. Case History

- Patient Demographics
  - 39 year old Black male

- Chief Complaint
  - Reports visual distortions in both eyes for the last year. Vision seems to be distorted and objects appear tilted especially at distance.
  - No previous history of glasses wear
  - Denies eye pain, flashes, floaters, light sensitivity

- Ocular, medical history
  - Pertinent Ocular History
    - Diagnosed with bilateral disc edema at last eye exam 1 year ago
  - Medical History
    - (+) Herpes Simplex Virus 1 & 2, previously treated with Acyclovir
    - (+) Depression
    - (+) High risk bisexual relationship
    - NKDA

- Medications: Currently not on medications. History of Acyclovir 200mg TID for treatment of genital herpes

II. Pertinent Findings

- Clinical
  - Entering VAs without spectacles:
    - Right eye: 20/20-2
    - Left eye: 20/20-
  - Pupils: PERRL (-) APD
  - Extraocular muscles: full, no restriction both eyes
Confrontation fields: full to finger counting both eyes

Manifest Refraction
  - Right eye: plano
  - Left eye: plano

Physical

Slit Lamp Exam
  - Lids/Lashes: grade 1 capped meibomian glands both eyes
  - Conjunctiva: scattered melanosis both eyes
  - Cornea: clear both eyes
  - Iris: flat, intact (-)NVI both eyes
  - Angles: 4x4 both eyes
  - Anterior chamber: deep and quiet both eyes

Tonometry (one drop proparacaine 0.5% and NaFl strip)
  - Right eye: 16 mmHg
  - Left eye: 17 mmHg

Dilated fundus exam (1 drop 1% Tropicamide, 1 drop 2.5% Phenylephrine)
  - Lens: clear both eyes
  - Vitreous: clear both eyes
  - Optic nerve head
    - Right eye: 0.1, mildly blurred disc margins 360 but no obscurcation of vessels (-)SVP
    - Left eye: 0.1, mildly blurred disc margins 360 but no obscurcation of vessels (-)SVP
  - Macula
    - Right eye: flat, intact
    - Left eye: flat, intact
  - Vessels
    - Right eye: normal caliber, 2/3
    - Left eye: normal caliber, 2/3
  - Periphery: flat, intact, no holes, no tears 360 both eyes

OCT Macula 5 Line Raster: flat, all macula layers intact both eyes

OCT Optic Nerve Head: thickening of nerve fiber layer, vitreal traction of optic disc both eyes

Visual Field 30-2 at visit #2:
  - Bilateral inferior nasal step

Fluorescein Angiography performed at visit #4: early and late fluorescein leakage of optic nerve head in both eyes

Laboratory studies collected after visit #2
  - Syphilis antibody with reflex RPR testing: Positive syphilis antibody with reactive RPR
  - HIV: positive
  - Quantiferon-TB: negative

Radiology studies after visit #2
MRI scan of orbits with and without contrast: Ocular bulbi, extraocular muscle conus, optic nerves, lacrimal glands appear bilaterally symmetrical, unremarkable, and within normal range. No intraorbital signal abnormalities or areas of abnormal enhancement. Optic chiasm and optic tracts appear normal. Pituitary not enlarged. Paranasal air spaces appear clear.

III. Differential Diagnosis
- Primary/leading: Bilateral optic nerve head edema secondary to active syphilis
- Others:
  - Optic disc drusen: Fluorescein angiography did not show focal areas of staining and OCT did not show drusen
  - CRVO: no other retinal findings such as cotton wools spots or hemorrhages
  - Anterior ischemic optic neuropathy: No signs of ischemia on fluorescein angiography
  - Compressive optic neuropathy: MRI of brain and orbit showed no compressive lesion
  - Optic neuritis: no pain on eye movements, usually unilateral, VA was not decreased

IV. Diagnosis and Discussion
- Elaborate on the condition: Ocular syphilis
  - Syphilis is a sexually transmitted disease, caused by the Gram-negative spirochete, Treponema pallidum. Direct contact with an infectious sore is necessary for transmission. Congenital syphilis can be transmitted from an infected pregnant mother. There are three stages of the disease:
    - Stage 1: Presence of a single, painless syphilitic sore (chancre)—the point of entry by the bacterium. Chancres heal within three to six weeks regardless of treatment.
    - Stage 2: Progression of Stage 1 with the presence of skin rashes and mucous membrane lesions (condyloma lata). Other symptoms include fever, malaise, swollen lymph nodes, muscle aches, and sore throat. Again, symptoms can disappear with or without treatment.
    - Stage 3: Progression of Stage 2 to latent syphilis, after primary and secondary signs and symptoms have disappeared without treatment. Late stages of syphilis can affect the central nervous system (neurosyphilis), internal organs, and infiltrate blood vessels. Symptoms include paralysis, blindness, and dementia.
  - Ocular syphilis, a form of neurosyphilis, can occur at any stage
  - According to the CDC, ocular syphilis has reemerged within the last two years, with a majority of cases occurring in the HIV positive population, namely men who have sex with men (1).
  - Known as the “great masquerader,” the clinical manifestations are non-pathognomonic, and can include anterior and posterior uveitis, optic
neuropathy, vitreous precipitates, retinal vasculitis, retinal detachment, chorioretinitis, episcleritis, scleritis, and interstitial keratitis (2).

- Optic nerve head involvement may present as optic disc edema, optic atrophy, or papilledema (3).
  - Fluorescein angiography can show fluorescein leaking or staining at the optic nerve head, vascular leakage, nonperfusion, papillary neovascularization (2), and delayed venous return (4).

- There is an association of syphilis and HIV due to an immunocompromised state. The rate of co-infection has been as high as 70% during outbreaks. However, unlike other associated ocular conditions such as CMV retinitis, initiating HAART therapy is not shown to decrease the incidence of ocular syphilis (3).
  - Since co-infection is common and HIV may be unsuspected, HIV testing in patients with ocular syphilis is recommended (4).
  - There is no correlation between ocular syphilis and the stage of HIV (5).

- Expound on unique features
  - At the patient’s previous eye exam one year ago, patient had the same visual symptoms and optic disc findings, but did not return for the ordered lab work and scans
  - Patient did not have any other active inflammatory ocular findings associated with ocular syphilis, such as anterior or posterior uveitis.

V. Treatment, Management

- Retina consult #1:
  - Following ocular examination and review of lab results, the patient was taken to Adult Medicine for treatment of active syphilis
  - Treatment: Intramuscular injections of 2,400,000 units (4mL) Penicillin G Benzathine for 3 weeks. Patient was advised to contact sexual partners and inform them to seek treatment. Patient advised not to engage in unprotected sex until self and partners have been treated.

- Retina consult #2:
  - Residual bilateral optic disc edema. Medication review stated two injections of Penicillin G Benzathine were completed. Patient was to return in 4-6 weeks for another retinal evaluation. Patient was lost to follow up after this visit.

- The CDC states cases of ocular syphilis should be reported to the state or local health department within 24 hours of diagnosis (1).

- Diagnosis of syphilis with blood tests:
  - Nontreponemal tests: VDRL and RPR. These tests are non-specific for syphilis and are insufficient for diagnosis.
    - RPR has been shown to be more sensitive in the secondary stage, and insensitive during the primary and tertiary stages (4).
    - A positive RPR represents active disease
  - Treponemal tests: FTA-ABS, TP-PA. These tests detect syphilis-specific antibodies that are detectable in a patient for life, despite successful treatment.

- Standard treatment of syphilis:
Primary, secondary, and early late syphilis: single intramuscular injection of Benzathine penicillin G is sufficient to cure (1).

Late latent syphilis or latent syphilis of unknown duration: three doses of Benzathine penicillin G at weekly intervals (1).

Neurosyphilis: requires intravenous benzylpenicillin for up to 17 days (3). If a patient is allergic to penicillin, then doxycycline, tetracycline, or ceftriaxone with close follow up and lab tests are alternatives, but without sufficient research.

Patients with ocular syphilis are recommended to obtain a lumbar puncture to determine if the central nervous system is affected (6).

- An abnormal CSF can present in both HIV-positive and negative patients (6).
- CSF cell counts, protein levels, glucose levels, and VDRL are assessed (4).

**Prognosis:**
- Treatment is sufficient to cure syphilis if detected early
- Syphilis does not recur once successfully treated; however, it does not prevent reinfection. Since re-treatment may be necessary, titers must be closely monitored.
  - Relapse or reinfection is seen in up to 14% of infected patients (4).
- Depending on the site and severity of ocular involvement, vision may improve with appropriate treatment (3).
- Antibiotic treatment can recover or improve visual acuity to up to 20/20 (5).

**Prevention:**
- Protection of infected sores with condoms is recommended
- Abstinence from partners with infected sores

**VI. Conclusion**

**Clinical pearls**
- Syphilis can present in a variety of ocular manifestations, the most common of which are inflammatory conditions such as anterior and posterior uveitis
- It is important to also consider syphilis when presented with unexplained acute or gradual visual disturbances/loss and optic disc edema (unilateral or bilateral).
- Ordering proper lab work is essential to diagnosing syphilis in a timely manner to ensure treatment and prevention of the course of the disease (7).
- Early detection and treatment of ocular syphilis can lead to a better visual prognosis

**References**


