Acute Syphilitic Posterior Placoid Chorioretinitis in an HIV+ Patient

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Abstract:

Acute syphilitic posterior placoid chorioretinitis is diagnosed in a 46 yo HIV+ white male. Differential diagnosis, laboratory testing, and treatment are discussed. Resolution of chorioretinitis is monitored with serial SD OCT and autofluorescence imaging.

I. Case History

- Patient demographics: 46 year old white male
- Chief Complaint: Flashes and a shadow in superior field of vision OS
- Medical history: HIV+, IDDM
- Medications: Abacavir/Lamivudine, Efavirenz, Insulin, Aspirin 81mg, Cyclobenzaprine, Gabapentin, Levothryroxine, Lisinopril, Nortriptyline, Omeprazole, Simvastatin, Sildenafil, Zolpidem
- Patient stopped all HAART medications for a two month period six months prior to presentation. He also had a new male oral sex partner in the past few months.

II. Pertinent Findings

- Clinical:
  - BCVA: 20/20 OD, 20/20-2 OS
  - CF: full OD, mild constriction superior nasal OS
  - PERRL –APD
  - Color: WNL OD and OS
  - Anterior segment: WNL OD; few KPs centrally and trace cells OS
  - IOP: 18/16
  - Posterior segment: Moderate NPDR OU. C/D 0.25 with distinct margins OU. Yellow/white oval lesion along inferior temporal arcade OS with vasculitis and overlying vitritis. RPE stippling surrounding the lesion.
- Physical: chancre on tongue, fatigue, general malaise
- Laboratory studies:
  - CD4: 730
  - Viral Load: detectable
  - HbA1c: 8.9%
  - RPR: reactive, titer 1:32
  - CSF VDRL: non-reactive
  - CSF FTA-ABS: reactive
  - CSF: high nucleated cells
  - T. Pallidum AB: reactive
  - CMV IgG: high
  - CMV IgM: normal
  - HSV 1: negative
  - HSV 2: high
  - ACE: normal
  - Lysozyme: high
  - Serum Lyme AB: detected
  - CSF Lyme: not detected
  - Toxoplasmosis: negative
- Imaging:
  - Fundus Photos: yellow/white placoid lesion along inferior temporal arcade with surrounding RPE stippling
  - SD OCT: area of retinal whitening shows disruption of all retinal layers with overlying vitreous cells. Hyperreflective nodular deposits on RPE throughout inferior arcade and macula. Loss of IS/OS junction starting 1000um inferior to center of fovea and extending through inferior arcade.
  - Fundus Autofluorescence (FAF): two distinct circular areas of hyperautofluorescence. One encompassing ONH and macula, and another brighter area encompassing inferior arcade. Punctate hyperreflective dots surrounding the lesion.
  - FA: early patchy hypofluorescence with late progressive staining of lesion. Punctate hypofluorescent dots surrounding lesion.

III. Differential Diagnosis
- Primary: Syphilitic etiology causing chorioretinitis, posterior placoid chorioretinitis, and/or vascular occlusive disease
- Others: CMV retinitis, HSV retinitis, APMPPE, ARN, PORN, toxoplasmosis

IV. Diagnosis and Discussion
- Our patient was suspected of having syphilis based on his fundus appearance and presence of a chancre on his tongue. There was also a higher index of suspicion given his history of HIV and male sexual partners. Diagnosis was confirmed at day 2 with positive RPR and treatment was initiated immediately.
- CMV and HSV were high on the differential list, but were ultimately ruled out after laboratory results were obtained.
  - There are two antibody tests for CMV: IgG and IgM. If a patient has been previously infected with CMV, IgG antibodies will always be detectable. Active CMV infection is diagnosed with high IgM antibodies or a fourfold increase in IgG antibodies in samples taken 2-4 weeks apart. Our patient did have high IgG antibodies, but IgM was not detectable, so he was not suspected of having active disease.
  - HSV 2 infection is lifelong and patients will always remain seropositive. Active infection is determined by symptoms and presence of lesions. Our patient did have high HSV 2 levels, but no active lesions.
- Ocular syphilis can manifest in any stage of the disease and in many different forms. It can affect almost any ocular structure, but posterior uveitis and panuveitis are the most common.
- Acute syphilitic posterior placoid chorioretinitis (ASPPC) is defined as a yellowish/whitish placoid lesion near the macula, often with a faded center and stipulation of the adjacent RPE. This produces a classic leopard spot appearance.
- The development of a placoid deposit in the outer retina is thought to be from subretinal deposition of RPE-photoreceptor complex material and incomplete phagocytosis of outer segments. It also may be due to the accumulation of lipofuscin in the RPE.
- SD OCT scans show irregular hyperreflectivity with nodular elevations at the junction of the photoreceptors and the RPE. This is associated with segmental loss of the IS/OS band.
- Multiple case reports have shown restoration of the IS/OS junction, remodeling of the outer retina, and improvement of visual function after treatment.
V. Treatment and Management
- Penicillin G 4 million units IV q4hr x 14 days, Pred Forte TID with slow taper
- At day 23:
  - Patient reports shadow and flickering are improved, but still present
  - Rare anterior chamber and vitreous cell, retinitis resolved, placoid lesion resolved with residual pigment mottling
  - OCT: hyperreflective dots on RPE improved, some restoration of IS/OS junction and ELM, reorganization of retinal layers in area of retinitis
  - FAF: circular area of hyperautofluorescence surrounding ONH and macula resolved, circular area around inferior arcade improved, punctate dots improved
  - RPR titer decreased twofold, from 1:32 to 1:16

VI. Conclusion
- ASPPC is a rare form of ocular syphilis with potentially devastating visual consequences. However, treatment of the disease can lead to dramatic remodeling of the outer retina (as seen on SD-OCT) and restoration of visual function.
- Immunocompromised patients are at a higher risk of severe infection from diseases such as syphilis, CMV, and HSV. The ability to properly order and interpret laboratory studies is critically important when differentiating and diagnosing these vision-threatening conditions.
- Although syphilis is not commonly encountered in optometry, the CDC has reported an increase in cases of ocular syphilis over the past year. Awareness of the disease and the multitude of clinical presentations will aid in prompt diagnosis and treatment.

VII. References