CLINICAL VIGNETTE

Hindsight is 20/50 in a Patient with Neuro-Ocular Syphilis

Vindeep Bhandari, DO and Bryan Lopez, MD

Case

A 61-year-old man was referred to the emergency department from Ophthalmology with progressive bilateral vision loss and "shock-like" headaches for two weeks. Past medical history includes hypertension, psoriatic arthritis, latent tuberculosis, and diabetes mellitus complicated by severe non-proliferative diabetic retinopathy (NPDR). His prior visual acuity was 20/50 bilaterally, however he was only able to see objects three feet away, and visual acuity bilateral was reduced to less than 20/400.

Vital signs were normal and physical exam did not reveal meningismus or temporal tenderness. Ophthalmoscopic examination with Optical Coherence Tomography (OCT) revealed outer retinal atrophy inconsistent with non-proliferative diabetic retinopathy, and lack of papilledema or optic disc pallor. It did reveal non-specific marked distortion of retinal architecture as shown in Figure 1. MRI/MRA of the brain and MRI of the orbit were unremarkable (Figure 2). Initial labs included elevated ESR 93 mm/hr. Beside lumbar puncture by the procedural team with opening pressure of 15 cm, clear colorless fluid with 3 rbcs, 9 wbc's and normal glucose and protein. Basic CSF analysis and cultures were negative. Due to sudden onset vision loss in the setting of elevated ESR there was concern for temporal arteritis, and he was started on prednisone 60mg daily. After two days he had notable improvement in visual acuity to OD 20/400 and OS 20/160. However, additional analysis revealed a positive serum RPR at 1:128, reactive serum TP-PA, and minimally reactive CSF TP-Ab. Given positive syphilis serologies and ophthalmologic findings demonstrating insult to the outer layers of the retina, a diagnosis of syphilitic chorioretinitis was established.

Ophthalmology recommended treatment for presumed tertiary neurosyphilis with IV penicillin. Due to a penicillin allergy, he was transferred to the ICU to undergo penicillin desensitization, and subsequently started Penicillin G 4 million units every 4 hours. Although steroids were initially held due to concern for worsening neurosyphilis, given prior improvement in visual acuity after steroids, he was also given a four-day prednisone taper (40mg oral dose reduced by 10mg each day). The patient completed a fourteen-day course of IV penicillin G. On discharge, he demonstrated significant improvement in his visual acuity to OD 20/100 and OS 20/70 and repeat OCT revealed marked improvement in retinal architecture as showed in Figure 1. At his post-discharge Ophthalmology follow-up visit two weeks later, his visual acuity had improved to OD 20/70 and OS 20/50.

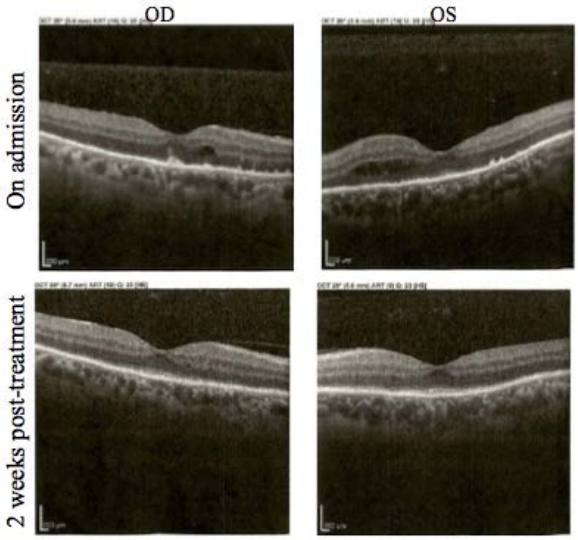

Figure 1: Optical Coherence Tomography (OCT).
Marked distortion of the retinal architecture is noted on admission OCT with reversal of the notably thickened and nodular retinal pigment epithelium seen two weeks post treatment IV Penicillin G.

Figure 2: MRI orbits with contrast.
This study demonstrated no soft tissue masses, normal globes, optic nerves and nerve sheath complexes, and no focal areas of abnormal enhancement.

Discussion

Ocular involvement in syphilis is rare but potentially debilitating and can occur in any stage of the disease. Although uveitis is the most common finding in ocular syphilis, multiple eye structures can be involved causing variable presentations, including chorioretinitis. This is important to recognize given increasing incidence of syphilitic infections. The majority of cases have been among individuals co-infected with HIV. One review reported a relative risk 14.5 times higher than for HIV-uninfected patients.[1] This case presents the phenomenon of syphilitic chorioretinitis in an HIV-uninfected patient without prior manifestations of syphilitic disease. Diagnosis is generally made through a combination of serologic (both non-treponemal screening and treponemal-specific confirmatory tests) and ophthalmologic findings, including focal thickening and increased nodularity of the choroid-retinal pigment epithelium seen on OCT.[2] First line treatment of neuro-ocular syphilis is parenteral penicillin therapy.[3-5] For patients who are penicillin allergic, desensitization is recommended. As shown here, in select cases, adjunctive glucocorticoids – in combination with appropriate antibiotic therapy – may represent a potentially successful therapeutic strategy to enhance vision improvement by decreasing inflammation as well as a prophylactic measure to prevent the Jarish-Herxheimer reaction.[5]

REFERENCES

1. **Amaratunge BC, Camuglia JE, Hall AJ**. Syphilitic uveitis: a review of clinical manifestations and treatment outcomes of syphilitic uveitis in human immunodeficiency virus-positive and negative patients. *Clin Exp Ophthalmol*. 2010 Jan;38(1):68-74. doi: 10.1111/j.1442-9071.2010.02203.x. PMID: 20447104.
2. **Burkholder BM, Leung TG, Ostheimer TA, Butler NJ, Thorne JE, Dunn JP**. Spectral domain optical coherence tomography findings in acute syphilitic posterior placoid chorioretinitis. *J Ophthalmic Inflamm Infect*. 2014 Jan 27;4(1):2. doi: 10.1186/1869-5760-4-2. PMID: 24468306; PMCID: PMC3917537.
3. **Yadlapati A, Chang E, Curcio E, Shah B, Yadlapati R**. Ocular Syphilis in Patients Infected With Human Immunodeficiency Virus: Case Report and Review of the Literature. *Infectious Diseases in Clinical Practice*. 2014 Mar;22(2):68-70. doi: 10.1097/IPC.0b013e3182948d6c.
4. **CDC**. Clinical advisory: ocular syphilis in the United States 2015. Atlanta, GA: US Health and Human Services, CDC; 2016. Available at: http://www.cdc.gov/std/syphilis/clinicaladvisoryos2015.htm
5. **Aldave AJ, King JA, Cunningham ET Jr**. Ocular syphilis. *Curr Opin Ophthalmol*. 2001 Dec;12(6):433-41. doi: 10.1097/00055735-200112000-00008. PMID: 11734683.