Young Woman with Bilateral Vision Loss and Macular Findings

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Presentation:

A 35-year-old woman with history of IV drug use presented to the retina clinic with one month of bilateral photopsias and two to three weeks of severe worsening blurry vision in both eyes. She reported that with both eyes open she could see a crescent shaped area of relatively clearer vision. Extensive review of systems revealed recent aches in her back and migranous headaches but no fever, rash, sores, headaches, acute illness, or recent travel. She lived near the woods but was not aware of any tick bites. Of note, her sexual partner of the past six months was treated for primary syphilis a few months prior. She had not been tested or treated, as she had no symptoms, and had not seen a physician in years. She reported she was currently in recovery for her drug abuse. She had not been tested for any infections recently, including HIV, and had not received a vaccination for COVID-19.

Figure 1. Funduscopic images showing bilateral hypopigmented oval-shaped placoid lesions.

Examination and Testing:

Exam revealed best corrected visual acuity of 20/60 OD and finger counting at 3 feet OS, intraocular pressures of 10 and 11 in the right and left eyes, respectively, and pupils reactive to light and accommodation without anisocoria or a relative afferent pupillary defect. The anterior segment exam was unremarkable, but funduscopic exam was notable for bilateral superior macular hypopigmentation in an oval shape involving the outer retina, RPE, and choroid. There were no anterior chamber or vitreous cells, and the discs appeared sharp. See Figure 1.

Formal visual field testing showed dense defects involving central fixation bilaterally, as seen in Figure 2. Spectral domain optical coherence tomography (SD-OCT) showed severe ellipsoid zone disruption and nodular irregularities in the retinal pigment epithelium (RPE) (Figure 3). Fluorescein angiography showed late staining in a placoid pattern posteriorly corresponding with the areas of pigmentary change and ellipsoid zone disruption (Figure 4), and fundus autofluorescence showed significant hyperautofluorescence corresponding to the same areas bilaterally (Figure 5).

Diagnosis:

The differential diagnosis includes acute syphilitic posterior placoid chorioretinitis (ASPPC), tuberculous chorioretinitis, lymphoma, HIV retinopathy, Lyme disease, viral retinitis, endogenous infectious chorioretinitis, AZOOR, or unlikely a white dot syndrome. However, the findings in this case were quite characteristic of ASPPC (see Discussion). Laboratory testing was performed, with RPR reactive and with a high titer (1:256), and confirmatory FTA-ABS also reactive, indicating an active syphilis infection. HIV-1 PCR testing was negative. Other testing was all within normal limits.

Management:

The patient was admitted to a hospital, where a lumbar puncture was performed, with cerebrospinal fluid studies positive for VDRL, which confirmed a concomitant diagnosis of neurosyphilis. The patient was started on intravenous penicillin. However, the patient left against medical advice after about 4-5 days prior to any definitive improvement in symptoms.

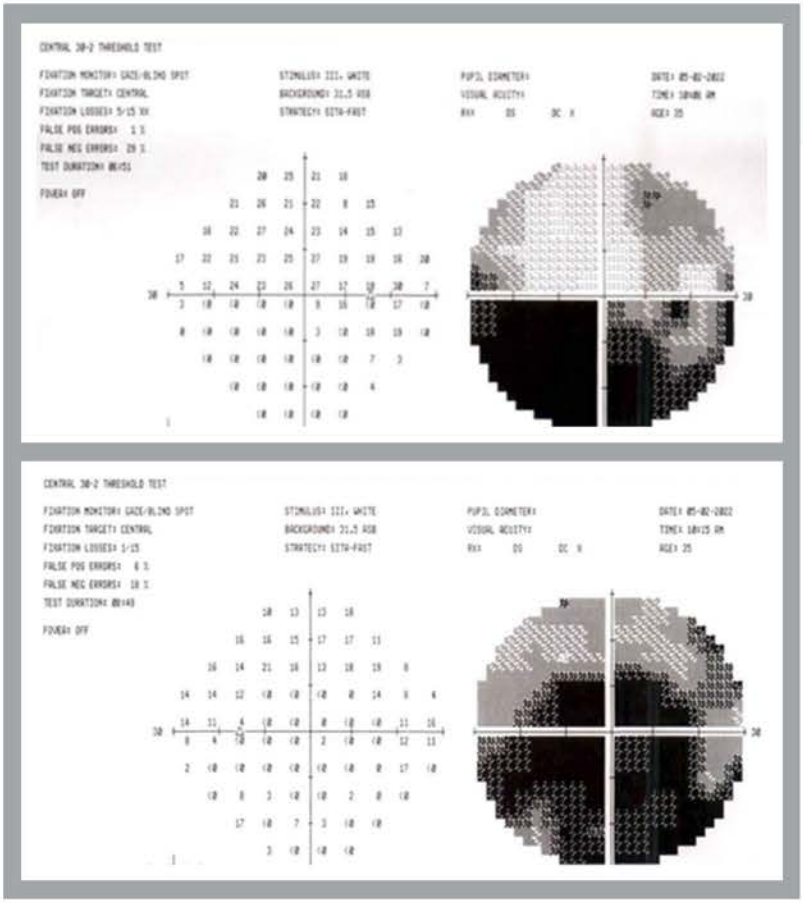

Figure 2. Visual field testing demonstrating dense defects involving central fixation bilaterally.

Discussion:

Ocular manifestations of syphilis are varied and can involve nearly any structure of the eye.[1] Posterior segment involvement includes vitritis, retinal vasculitis, optic neuritis, superficial retinal infiltrates, multifocal retinitis, or chorioretinitis as in ASPPC. Known as the great masquerader, ocular syphilis may mimic nearly any inflammatory or infectious ocular disease, and it is often comorbid with other sexually transmitted infections, particularly HIV. Ocular involvement may occur with any stage of systemic syphilis, and serologic testing with treponemal and non-treponemal tests is important to establish a diagnosis.[2] Patients with ocular syphilis should be worked up for involvement of the central nervous ssystem by a combination of neuroimaging and CSF studies. Neurosyphilis may be asymptomatic but can also manifest as meningitis, dementia paralytica, and tabes dorsalis, and may occur concomitantly with ocular and otologic syphilis.[3] They require intravenous penicillin and management with an infectious disease specialist.[2]

ASPPC in particular can present dramatically, with rapid onset of severe central vision loss and scotoma bilaterally. Diffuse ellipsoid zone disruption with nodular irregularities in the outer retina and RPE are commonly seen, as well as characteristic hypopigmented deep placoid lesions centered on the macula, which show hyperautofluorescence, as seen in this case.[4-6] As classically described, if treated promptly with parenteral penicillin, these eyes may show significant improvement in visual functioning, although long-term atrophic changes may lead to decreased visual acuity and scotomata.[4,7]

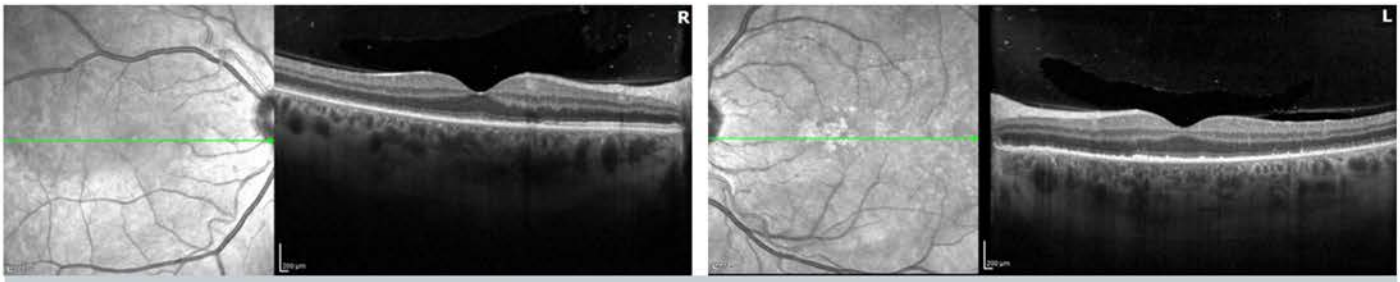

Figure 3. SD-OCT with ellipsoid zone disruption and nodular RPE irregularities bilaterally.

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4. Eandi CM, Neri P, Adelman RA, Yannuzzi LA, Cunningham ET, Jr. (2012) Acute syphilitic posterior placoid chorioretinitis: report of a case series and comprehensive review of the literature. Retina 32: 1915-1941. DOI 10.1097/IAE.0b013e31825f3851

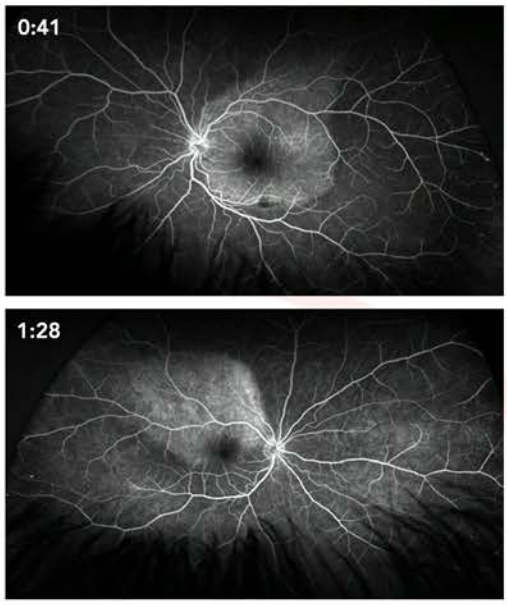

Figure 4. Fluorescein angiography images showing late staining in placoid pattern bilaterally.

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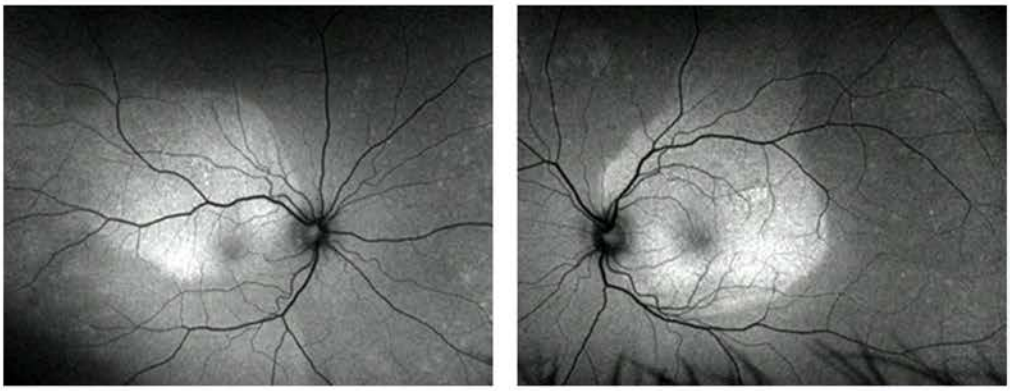

Figure 5. Fundus autofluorescence showing marked hyperautofluorescence in the affected lesions bilaterally.

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