Multimodal Imaging of Atypical Acute Syphilitic Posterior Placoid Chorioretinitis Mimicking a White Dot Syndrome

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ABSTRACT: As rates of infectious syphilis continue to rise in the U.S., it is important to be familiar with known manifestations of ocular syphilis as well as report presentations not previously described in the literature. Here, the authors report a case of a 49-year-old myopic woman presenting with bilateral white dots characteristic of a white dot syndrome; these white dots were not evident on slit-lamp examination and became obvious on fundus autofluorescence. She tested positive and was successfully treated for syphilis. This case demonstrates that ocular syphilis can present with white dots and should be on the differential diagnosis of white dot syndromes.

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INTRODUCTION

Syphilis, a well-known masquerader, is notorious for its myriad of ocular manifestations. We report a case of acute syphilitic posterior placoid chorioretinitis presenting with features of a white dot syndrome on multimodal imaging.

CASE REPORT

A 49-year-old myopic woman presented with bilateral decreased vision and photopsias for 2 weeks in the setting of a recent upper-respiratory infection. Her medical history was significant for diabetes, hypothyroidism, and migraines. She was a former smoker, consumed alcohol socially, and denied any recreational drug use, animal exposure, or risky behavior for sexually transmitted infections.

Best-corrected visual acuity (BCVA) was 20/100 in the right eye (OD) and 20/50 in the left eye (OS). Slit-lamp examination showed quiet anterior chambers. Funduscopic examination was unremarkable in both eyes (Figures 1A and 1B). Ultra-widefield fundus autofluorescence (FAF) imaging showed irregular hyperautofluorescent plaques within the central macula and surrounding the optic discs as well as multiple hyperautofluorescent spots extending to the mid-peripheral fundus (Figures 1C and 1D). Spectral-domain optical coherence tomography (SD-OCT) through these involved areas showed disruption of the ellipsoid zone (EZ), with retinal pigment epithelium (RPE) clumping in the macula bilaterally (Figures 1I and 1J). Though not observed on slit-lamp examination, vitreous cells were readily detectable in both eyes on swept-source OCT (SS-OCT) scans (Figures 1K and 1L). En face SS-OCT scans reconstructed at the level of the outer retina showed discrete areas of hyperreflectivity correlating with areas of hyperautofluorescence in the macula on FAF (Figure 1C, insets). Fluorescein angiography (FA) showed late staining of the optic discs bilaterally and subtle staining of the outer retina and retinal pigment epithelium (Figures 1E and 1H). Indocyanine angiography (ICGA) was remarkable for late macular hypocyanescence in both eyes (Figures 1F and 1G).

An infectious disease evaluation revealed a high rapid plasma reagin (RPR) titer of 1:64 and positive fluorescent treponemal antibody absorption. Quantiferon-TB, Lyme, and HIV testing was negative. The patient was administered standard-of-care treatment, after which her RPR titers came down to 1:2. Although she tested negative for *Bartonella* immunoglobulin M (IgM), she was highly positive (>1:1024) for IgG and received a 4-week course of azithromycin and rifampin. At 5-week follow-up,

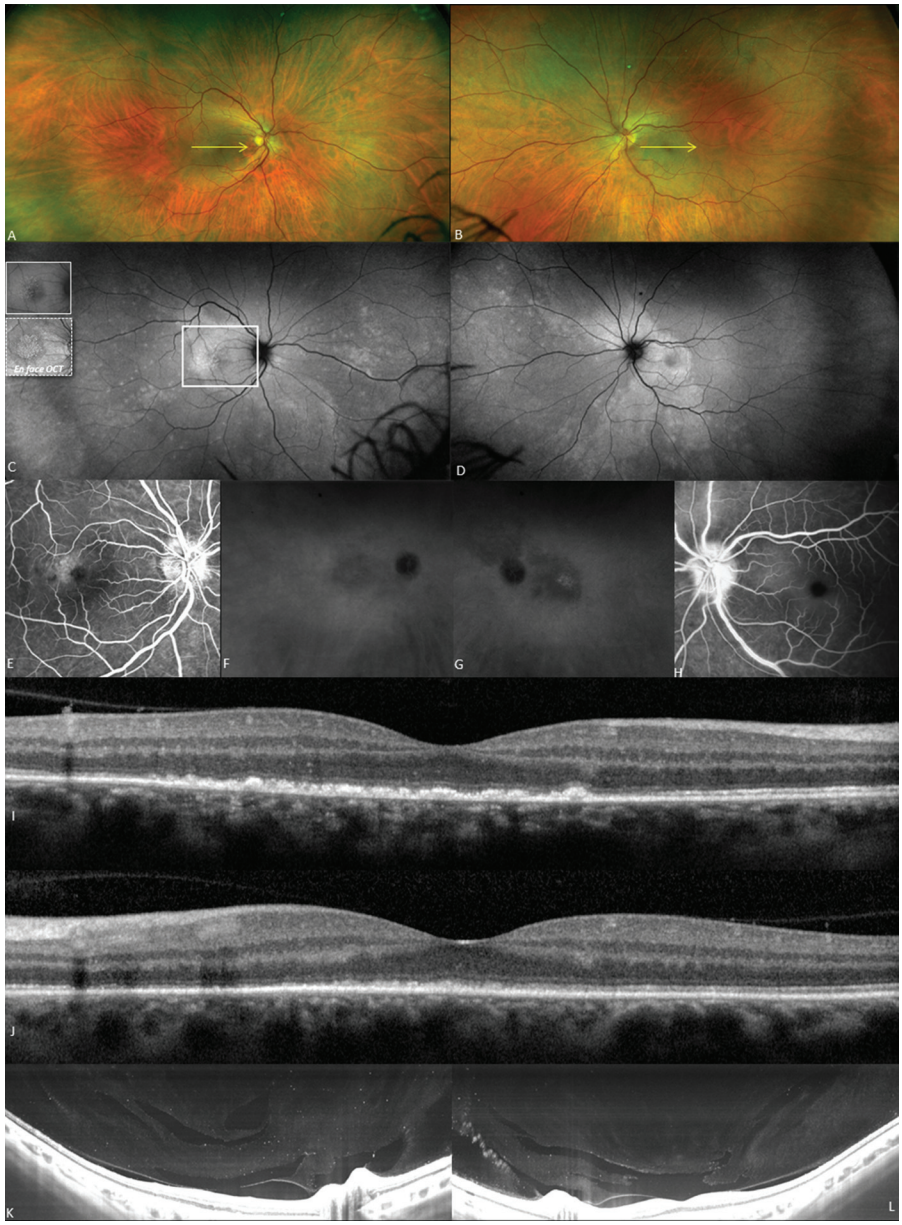

Figure 1. Initial presentation. (A, B) Ultra-widefield color fundus photography. (C, D) Ultra-widefield fundus autofluorescence reveals placoid lesions bilaterally surrounded by hyperautofluorescence spots. In the right macula hyperautofluorescent nodules correlate with hyperreflective spots on en face optical coherence tomography (OCT) of the outer retina (C, inset). (E, H) Fluorescein angiography reveals late staining of the macular region of the right eye and of the optic discs bilaterally. (F, G) Late-phase indocyanine green angiography reveals macular hypocyanescence. (I) OCT of the right eye reveals ellipsoid zone disruption with retinal pigment epithelial clumping. (J) OCT of the left eye reveals ellipsoid zone disruption. The location of the spectral-domain OCT B-scans correspond to the yellow arrows on the corresponding color fundus photographs. (K, L) Swept-source OCT with contrast adjustment reveals the presence of vitreous cells.

she reported near complete resolution of her visual symptoms, with BCVA measuring 20/25 OD and 20/20 OS. FAF imaging (Figures 2A and 2B) revealed resolving hyperautofluorescent spots and normalization of the EZ on OCT in both eyes with few remaining RPE clumps mainly observed in the right eye.

DISCUSSION

Syphilitic chorioretinitis is known to have a myriad of presentations that can be broadly classified into inner and outer retinitis. Syphilitic inner retinitis has been reported to show white dots on color fundus photographs in immunocompetent homosexual men.[1] Outer syphilitic retinitis has previously been reported to occur as either an acute syphilitic posterior placoid chorioretinitis (ASPPC) or syphilitic outer retinitis (SOR) showing zonal disturbance of the ellipsoid and the RPE, but not with white dots.[2]

Matsumoto and Spaide showed that in ASPPC, the placoid lesion consists of two hyperautofluorescent components: a diffuse area of hyperautofluorescence at the level of the RPE corresponding to the placoid lesion and intensely hyperautofluorescent spots that lie on the RPE within the placoid lesion.[3] No OCT or widefield imaging was provided in that report. Though nodular elevations of the

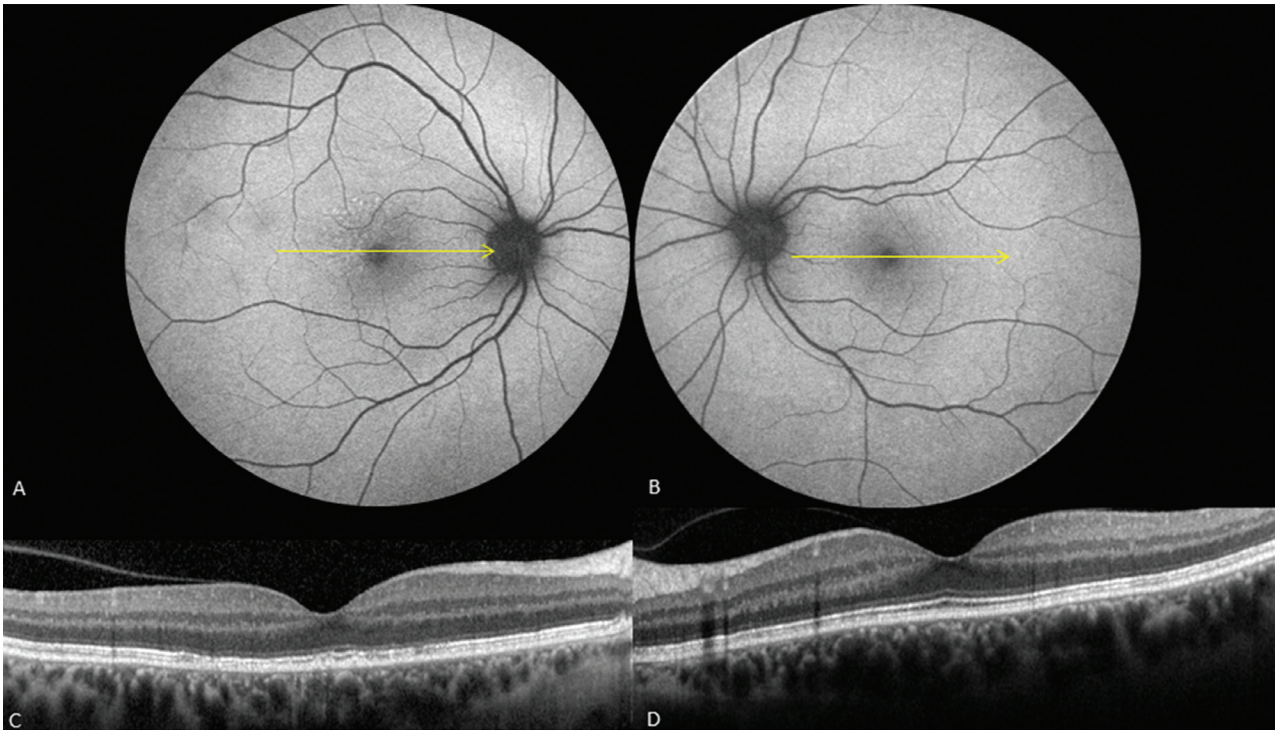

Figure 2. Post-treatment 5-week follow-up. (A, B) Fundus autofluorescence reveals near complete resolution of the hyperautofluorescent spots. (C, D) Optical coherence tomography (OCT) restoration of the ellipsoid zone bilaterally, and improvement in retinal pigment epithelium clumps in the right eye. The location of spectral-domain OCT B-scans is shown by yellow arrows on the corresponding fundus autofluorescent scans.

RPE appearing as speckled hyperautofluorescence has been reported on SD-OCT within the ASPPC lesion, our case shows that syphilitic chorioretinitis can also manifest on FAF as isolated white dots in the midperiphery separate from the ASPPC lesion.[4] Given the lack of cat exposure, lymphadenopathy, and stellate maculopathy, as well as negative IgM and high IgG titers, we do not believe that this is a presentation of acute cat-scratch neuroretinitis.

The Centers for Disease Control and Prevention recently reported a substantial increase in infectious syphilis in the United States from 2.1 cases per 100,000 in 2001 to 8.7 cases per 100,000 in 2016, the highest rate since 1993.[5] Syphilis screening — testing in asymptomatic persons — in those at increased risk (eg, men who have sex with men and engage in high-risk sexual behavior, those living with HIV infection, sex workers, persons who exchange sex for drugs, and those in adult correctional facilities) is recommended.[6]

In symptomatic patients, however, like those with characteristic syphilitic genital ulcers or maculopapular skin rashes and in those with atypical neurological and ophthalmological findings, syphilis diagnostic testing is strongly recommended. Typical serological testing includes a non-treponemal assay (RPR or Venereal Disease Research Laboratory test) followed by a treponemal assay (Treponemal Pallidum Particle Agglutination or Fluorescent Treponemal Antibody Absorption test). Increasingly, laboratories are offering the "reverse-algorithm," whereby the treponemal assay is performed first, followed by the non-treponemal assay.

Even as penicillin approaches 75 years in clinical use, penicillin G remains the treatment of choice for syphilis infection, either as an injectable long-acting formulation of benzathine penicillin G 2.4 million units single dose for early syphilis of less than 1 year's duration or as aqueous crystalline penicillin G 16 million to 24 million units administered intravenously for 10 to 14 days for ocular or neurosyphilis followed by a single additional dose of the long-acting formulation. Post-treatment follow-up includes repeat non-treponemal titers every 3 months until a four-fold titer decline is documented.

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