



A Young Female Patient Develops a Dark Spot in Her Vision

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Abstract

Over the last two decades the incidence of syphilis has increased. Ocular syphilis can occur during any stage of syphilitic infection and Acute Syphilitic Posterior Placoid Chorioretinopathy (ASPPC) is a rare but pathognomonic manifestation of ocular syphilis. Key ophthalmologic findings of this disease can be demonstrated on dilated retinal exam and optical coherence tomography with supplemental findings seen on fundus auto fluorescence imaging and intravenous fluorescein angiography. Confirmation of syphilitic infection is made by serum and cerebrospinal fluid serologies. Treatment of ocular syphilis, including ASPPC is treated as a variant of neurosyphilis, requiring intravenous or intramuscular penicillin. The role of adjunctive steroids following antibiotic treatment is not clearly defined. With proper treatment, many but not all patients with ASPPC regain excellent visual acuity.

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Case report

A Caucasian 43 year-old female presented with a five day history of a non-progressive dark spot in her vision in the left eye. She reports no past ocular history or trauma. When asked about systemic illnesses she endorses for the last 4 months she has had rashes over her body which have been slowly progressive. She also endorses a history of recent onset nasal sores. She was evaluated by her primary physician regarding her rash and started treatment with prednisone and terbinafine with-

out improvement. Treatment was modified to fluconazole and prednisone without resolution. She has a history of anxiety and current medications included citalopram, hydroxyzine, and naproxen. She works as a school custodian with a pet dog at home and denies recent travel. Only life stressor is that she is currently in the middle of a divorce. She is sexually active with only one partner and reports no history of sexually transmitted disease.



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The patient's best-corrected visual acuity was 20/20 in the right eye and 20/50 in the left, pupils and intraocular pressure were normal. The patient had full extraocular motility. The anterior segments were unremarkable and vitreous clear in both eyes. The optic nerve, retinal vasculature, macula and periphery were normal in the right eye. The left optic nerve had grade 1 optic nerve head swelling. A yellowish lesion with Retinal Pigment Epithelial (RPE) disruption in the macula was noted in the left eye (Figure 2A). Optical Coherence Tomography (OCT) confirms this change (Figure 2B). Humphrey visual field testing was reliable showing a full field in the right eye and non-specific central defects in the left eye. OCT of the optic nerve head showed average Retinal Nerve Fiber Layer (RNFL) thickness of 108 μm OD and 124 μm OS. This represents a normal thickness in the right optic nerve and a slightly elevated thickness in the left.

Given the above ocular findings there was concern for ocular syphilitic infection, specifically Acute Syphilitic Posterior Placoid Chorioretinopathy (ASPPC). In addition, the patient's rash with palmar involvement and systemic symptoms were further concerning for secondary syphilitic infection. The patient was hospitalized for treatment of suspected neurosyphilis. After admission she had serologic testing revealing a positive serum RPR titer at 1:256, negative HIV testing, lumbar puncture showing

an opening pressure of 19.2 mmHg and reactive VDRL CSF titer at 1:1. PCR testing for *Chlamydia trachomatis* and *Neisseria gonorrhoea* was negative. The inpatient infectious disease team evaluated the patient and started treatment for neurosyphilis with intravenous penicillin G, 12 million units every 12 hours with plans to complete a 14-day course. On the second day of hospitalization the patient received a Peripherally Inserted Central Catheter (PICC) line and was discharged to complete the remainder of her antibiotic treatment at an outpatient infusion clinic.

The patient was seen in outpatient ophthalmology clinic 8 days after beginning penicillin treatment and was noted to have stable vision in her affected eye compared to presentation, 20/50 in the left eye. On dilated exam the retinal placoid lesion in the left eye was nearly resolved and the patient reported that her body rash was much improved as well. She followed up again in outpatient ophthalmology clinic 3 weeks after completion of penicillin treatment on at this time her vision in the affected, left eye had improved to 20/20. In the left eye, on dilated exam the retinal placoid lesion had completely resolved, and the optic nerve edema had resolved with only optic nerve hyperemia remaining. OCT through the macula of the left eye showed resolution of the RPE hypertrophy and nodularity seen on presentation.



Figure 1: Color photograph of the patient's rash on back and palms of hands.

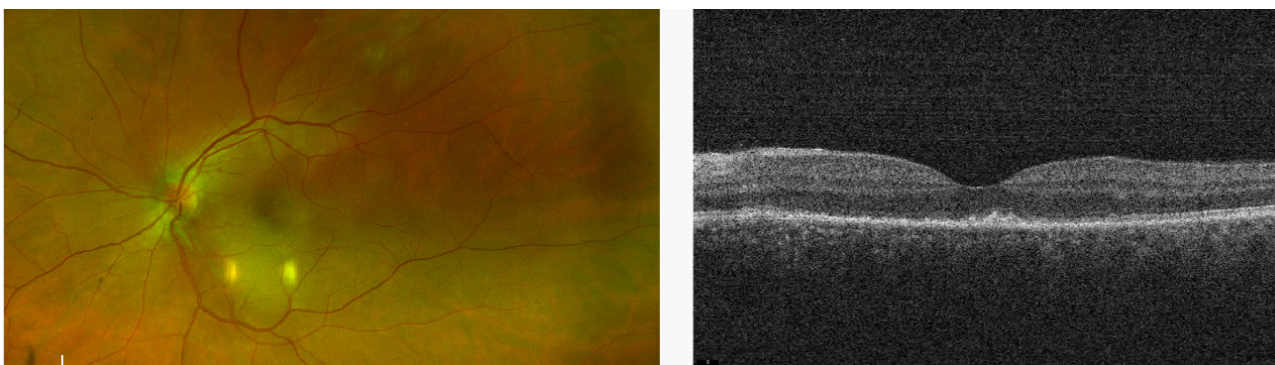


Figure 2: (A) Fundus photograph of the right eye demonstrates yellow, placoid lesion with pigmentary disruption in superior macula extending to the superior arcade. 270° degrees of optic disc edema with slight hyperemia is also noted. (B) Spectral-Domain OCT through the left macula demonstrates a normal foveal contour and is remarkable for RPE disruption and thickening in a granular configuration as well as focal disruption of the ellipsoid zone. No subretinal or intraretinal fluid is present.

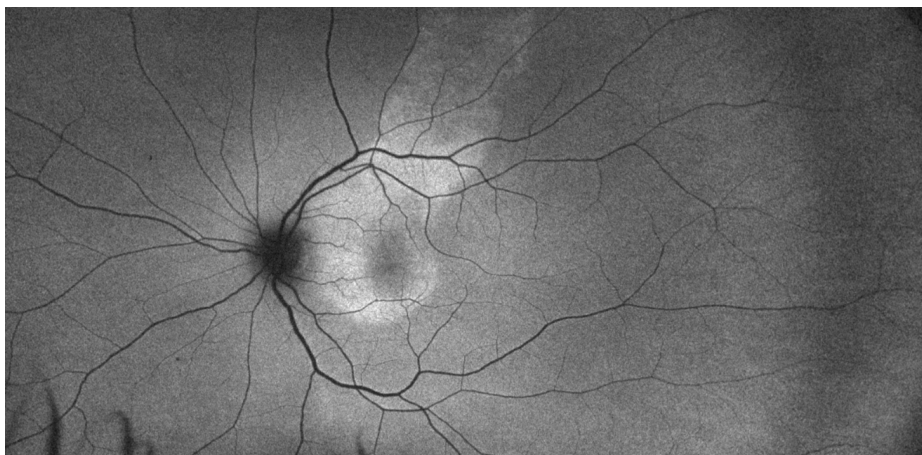


Figure 3: Fundus auto fluorescence shows hyperautofluorescence in the macula corresponding to the yellowish, placoid lesion seen on color fundus photograph in Figure 1.

Discussion

Syphilitic uveitis is a rare complication of acquired syphilitic infection and in this case, our patient had classical systemic findings including syphilitic rash with palmar involvement consistent with secondary disease as well as fundus and OCT findings demonstrating Acute Syphilitic Posterior Placoid Chorioretinopathy (ASPPC). Appropriate next steps include confirming the diagnosis with serologic testing followed by lumbar puncture to assess for neurosyphilis. Treatment will include a 10-14 day course of IV penicillin. The chorioretinal lesions seen in ASPPC tend to be yellowish, oval, placoid lesions often limited to the posterior pole without frank retinal necrosis on exam or OCT. White dot syndromes include similar appearing posterior uveitides to infectious masqueraders [1]; the patient's rash and systemic symptoms lend support to an infectious etiology.

The incidence of syphilis in the United States increased from the early 2000s to the present, though remains lower than levels in the 1990s [2]. Ocular syphilis can present during any stage of the disease and in any layer of the eye, thus often referred to as the great masquerader. Prior survey data from uveitis specialists suggests syphilitic uveitis most often presents during the secondary stage of the disease (40.2%). In addition, the most common syphilitic uveitis seen include posterior uveitis (60.8%) followed by pan uveitis (22.5%) [3].

ASPPC is a previously described rare but pathognomonic manifestation of ocular syphilis first described in 1990.⁷ Patients develop one or more oval or circular yellowish placoid lesions at the level of the RPE within or near the macula [4,2]. It is suspected treponema reach the choroid and outer retina via the choroidal blood vessels with abundant circulation near the macula, accounting for posterior location of plaques in this disease. ASPPC can be associated with mild Sub Retinal Fluid (SRF) in the first 1-2 days after symptom onset, however patients imaged after 7 days of symptoms, SRF was not present in any cases [2]. Presenting acuity ranges from 20/20 to count fingers with a mean of 20/80 [5]. OCT findings include a disrupted ellipsoid zone and thickened, granular RPE. Fundus auto fluorescence typically shows hyper auto fluorescence in the area of ASPPC [1] (Figure 3). Small, punctate hyper auto fluorescent dots can also be seen associated with the plaque, thought to represent photoreceptor debris accumulating over dysfunctional RPE. Plaque lesions can also be imaged with intravenous fluorescein angiography with typical findings showing early phase hypo fluorescence over the lesions followed by late phase staining of the lesions with or without leakage [6].

The majority of patients receiving antibiotic treatment have resolution of imaging and OCT findings of ASPPC and improvement in vision [4,2,5]. The utility of steroids in the course of syphilitic uveitis treatment is not well defined. Only one meta-analysis has been performed investigating adjunctive steroid use in syphilitic uveitis and all but one of the included studies were retrospective in nature [7]. Results showed that adjunctive steroids or immune suppressives had no additional benefit over antibiotic monotherapy, however given most of the data was retrospective in nature it is unclear if this conclusion is applicable to all patients with syphilitic uveitis. In a survey of the International Ocular Syphilis Study Group, 31.4% of providers routinely used adjunctive steroids in the treatment of syphilitic uveitis and 66.7% used them in selected cases [3]. Nearly all providers agree steroids should be started only after appropriate antibiotic coverage and the majority of providers choose topical steroids which can be titrated quickly based on a patient's disease course. Our patient had minimal intraocular inflammation at presentation and did not receive steroid treatment during her treatment.

Interestingly our patient was also noted to have optic disc edema only on the left. Opening pressure was normal on lumbar puncture. The quick onset of symptoms coupled with absence of pain suggest our patient likely had optic perineuritis due to neurosyphilis as opposed to anterior optic neuropathy, though both are possible [7]. The patient's disc edema resolved with her above course of antibiotic treatment.

Conclusion

Ocular syphilis can occur in any stage of syphilitic disease and any segment of the eye. ASPPC is a rare but pathognomonic manifestation of ocular syphilis with distinct imaging and OCT findings whereby eye providers can be the first to suspect syphilitic disease and help guide patients to proper testing and treatment. Management of ocular syphilis requires serum and CSF evaluation followed by penicillin treatment, close monitoring and potentially adjunctive corticosteroids.

References

1. Raven ML, Ringeisen AL, Yonekawa Y, Stem MS, Faia LJ, et al. Multi-modal imaging and anatomic classification of the white dot syndromes. *Int J Retina Vitreous*. 2017; 3: 12.
2. Pichi F, Neri P. Multimodal imaging patterns of posterior syphilitic uveitis: A review of the literature, laboratory evaluation and

-
- treatment. *Int Ophthalmol.* 2020; 40: 1319-1329.
3. Oliver GF, Stathis RM, Furtado JM, et al. Current ophthalmology practice patterns for syphilitic uveitis. *Br J Ophthalmol.* 2019; 103: 1645-1649.
 4. Pichi F, Ciardella AP, Cunningham ET Jr, et al. Spectral domain optical coherence tomography findings in patients with acute syphilitic posterior placoid chorioretinopathy. *Retina.* 2014; 34: 373-384.
 5. Eandi CM, Neri P, Adelman RA, Yannuzzi LA, Cunningham ET Jr; International Syphilis Study Group. Acute syphilitic posterior placoid chorioretinitis: report of a case series and comprehensive review of the literature. *Retina.* 2012; 32: 1915-1941.
 6. Franco M, Nogueira V. Severe acute syphilitic posterior placoid chorioretinitis with complete spontaneous resolution: The natural course. *GMS Ophthalmol Cases.* 2016; 6: Doc02.
 7. Smith GT, Goldmeier D, Migdal C. Neurosyphilis with optic neuritis: an update. *Postgrad Med J.* 2006; 82: 36-39.
 8. Gass JD, Braunstein RA, Chenoweth RG. Acute syphilitic posterior placoid chorioretinitis. *Ophthalmology.* 1990; 97: 1288-1297.
 9. Zhang T, Zhu Y, Xu G. Clinical Features and Treatments of Syphilitic Uveitis: A Systematic Review and Meta-Analysis. *J Ophthalmol.* 2017; 2017: 6594849.