Painless syphilitic uveitis in the Emergency Department

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INTRODUCTION
Syphilis is a sexually transmitted, systemic, chronic infection that can affect all structures of the eye from cornea to optic nerve.1,2 The British Ocular Syphilis Study (BOSS) reported an annual incidence of intraocular syphilis of 0.3 per million UK adult population based on 2014 data. Bilateral ocular involvement was present in 56%, and the mean presenting Snellen visual acuity was 20/63. Pan-uveitis was diagnosed in 41.3% of cases, followed by isolated anterior uveitis (9.5%) as the next most common diagnosis.3 Epidemiological data suggests that ocular findings of syphilis occur in 2.5-5% of patients with tertiary syphilis and may be a presenting manifestation of syphilis.4 While 90% of syphilis cases occur in developing countries, an increasing trend of syphilis cases is present in young men in industrialized nations.5 While commonly regarded as a painful entity, an exact incidence of painless syphilitic uveitis remains unclear.

CASE REPORT
A 41-year-old man with a history of chronic obstructive pulmonary disease, stroke, hypertension, and congestive heart failure presented to the ED for evaluation of progressive painless blurry vision and redness of the right eye for the previous two weeks. He denied tearing, discharge, photophobia, or headache. The patient did not receive relief from napazoline-pheniramine ophthalmic drops for presumed allergic conjunctivitis. Medications include: amlodipine, atorvastatin, budesonide-formoterol, carvedilol, hydrochlorothiazide, lisinopril, and tiotropium. His blood pressure was 149/97, heart rate 75, temperature was 37.2°C, respiratory rate 15, and pulse oximetry of 97% on room air. Gross examination revealed minimal paralimbic injection of the right eye, with no injection of the left eye. Eyes were not tender to palpation, and no photophobia was appreciated. His corrected visual acuity was 20/200 in the right eye and 20/30 in the left eye. The right pupil was 3 mm at baseline and minimally reactive to 2.5 mm in response to light. The left pupil was 4 mm at baseline and 3 mm in response to light. Extraocular muscle testing was normal. Corneal staining and slit lamp examination were not performed in the Emergency Department. The remainder of the physical examination was unremarkable. There was no adenopathy or rash. The patient had a scheduled appointment in two months, but an urgent ophthalmologist’s slit lamp assessment that day revealed 3+ cell in the anterior chamber and extensive posterior synechiae on the right. No abnormalities were noted on slit lamp assessment of the left eye. Tonometry revealed an intraocular pressure of 11 mm Hg in the right eye and 16 mm Hg in the left eye. Fundoscopic examination revealed normal discs bilaterally. Synechiae observed in the right eye created a hazy view of the macula and vessels. Synechiae were broken with 10% neosynephrine, and the patient was started on prednisolone acetate and tropicamide ophthalmic drops. Two days later, lab evaluation revealed unequivocal rapid plasma reagent (RPR), HLA-B27 positivity, and fluorescent treponemal antibody absorption (FTA-ABS) positivity. Further history revealed a latent history of syphilis requiring doxycycline because of a penicillin allergy. Two days after presentation, the patient was hospitalized for penicillin desensitization for syphilitic uveitis. He tested negative for HIV. Inpatient lumbar puncture venereal disease research laboratory (VDRL) testing of the cerebrospinal fluid (CSF) was non-reactive. Two months after presentation and appropriate treatment with IV penicillin for 14 days with 24 Million U daily, the patient had normal visual acuity and no cells or flare on slit lamp examination. While the patient had negative VDRL testing of the cerebrospinal fluid (CSF), the inpatient team and Infectious Diseases consultants treated him empirically for early neurosyphilis due to his ophthalmic complaints. Uncertainty still remains as to the stage of this case.

DISCUSSION
Inclusion of syphilitic uveitis into the differential diagnosis for the presentation of red eye has significant education impact for the emergency medicine practitioner. The diagnosis of red eye can be divided into two components: painless and painful etiologies. Painful causes include corneal ulcer, abrasion, herpes simplex keratitis, conjunctivitis, acute glaucoma, scleritis, or uveitis. Painless causes include blepharitis, ectropion, or subconjunctival hemorrhage.6 Syphilitic uveitis and scleritis usually presents as a painful, red eye with decreasing visual acuity, and it is unclear why this patient had a painless presentation.1,7
In the absence of systemic signs or symptoms, nonspecific ocular findings make the diagnosis of syphilis difficult, and support its image as the “great masquerader.” The disease can also easily be mistaken for idiopathic uveitis, and requires clinical assessment for associated signs of adenopathy, rash, or genital lesions. Consideration of syphilis as a diagnosis requires a serologic treponemal test (FTA-ABS), as the nontreponemal tests (RPR and VDRL) may be nonreactive in 30-75% of syphilitic uveitis cases. Current literature is divided as to which patients should have a lumbar puncture. Recent studies have shown that those with syphilis and serum RPR $\geq 1:32$ are at higher risk for early neurosyphilis. Other authorities suggest that since neurosyphilis cannot be diagnosed by serologic tests, cerebral spinal fluid examination is required in all presenting patients due to the risk of recurrence if improperly treated. While clinical signs of neurosyphilis (cranial nerve dysfunction, ophthalmic abnormalities) may suggest the disease, diagnosis can only be made with serologic and cerebrospinal testing. The CDC suggests that positive VDRL on CSF be classified as clinically confirmed neurosyphilis and an abnormal elevation in white blood cells or protein to be clinically probable neurosyphilis. With negative CSF findings in this patient’s case, the consultants and primary team deemed the ocular findings concerning for neurosyphilis and treated accordingly. Additionally, all patients with ocular signs and symptoms of syphilis should be tested for HIV, as co-infection is present in 33% of patients and HIV-positivity suggests accelerated disease progression and increased risk for relapse.

References

Disclaimer
The views expressed herein are those of the author and do not necessarily reflect the views of the Emergency Medicine Residency program at Brown University.

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