

# Ocular syphilis: a rare manifestation in a low risk population

## Abstract

Syphilis has been called “The Great Masquerader” due to its ability to affect any organ system and its propensity to produce a diverse array of signs and symptoms. Ocular syphilis is one of many possible expressions of this disease. The overwhelming majority of patients with syphilis present with signs or symptoms preceding, or in concurrence with, ocular manifestations. Afflicted individuals also usually fall into one or more high-risk categories. However, as this case highlights, syphilis can reveal itself solely in the eye and in lower risk populations.

Volume 8 Issue 3 - 2018

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Received: April 10, 2018 | Published: May 21, 2018

## Introduction

Ocular syphilis is a rare manifestation of a centuries-old disease that has had a resurgence in the US in the past decade, with a steadily increasing incidence.<sup>1</sup> However, the vast majority of cases occur in patients that are HIV positive, men who have sex with men (MSM), or both.<sup>1</sup> Patients often present with physical indicators in an organ system other than neurological. We present a case of a person that self-identified as a heterosexual male who does not have sex with men, is HIV negative, and whose only presenting symptom was vision change. The infrequent incidence of ocular syphilis can make this a challenging diagnosis that can be overlooked, especially in lower risk populations or in patients with no other manifestations. Furthermore, ocular syphilis' ability to affect any aspect of the eye coupled with its lack of specific findings creates extra obstacles in the diagnosis of these rare patients.

## Case report

A 41 year old Caucasian male with no significant past medical history presented to the ED with two days of painless vision change in the left eye. The vision change was described as a “hazy shadow”, located in the superior medial/nasal aspect of his visual field. He also had difficulty appreciating color, specifically the difference between red-green and orange-purple. The patient has never experienced these symptoms or other vision changes previously. The patient had no other complaints, was afebrile and hemodynamically stable. On physical exam, the only significant finding was on dilated fundoscopic exam which displayed cotton wool spots and macular edema.

Because branch retinal artery occlusion could not be ruled out, the patient was admitted for the following lab work: complete blood count, Hemoglobin A1C, ANA, lipid panel, RPR/FTA-ABS, HIV, PT/PTT. Imaging was also obtained which included CT (Figure 1) and MRI (Figure 2) as well as echocardiogram of the heart. No imaging abnormalities were found, and there were no abnormal labs besides the positive RPR, including a non-reactive HIV test. When the patient was notified of his positive RPR titer (1:64) with confirmatory FTA-ABS, he returned to the hospital for lumbar puncture which did not

reveal any treponemal organisms in his CSF. The patient was initiated on a 14-day course of IV Penicillin, which he completed without complication. The patient's vision changes have since resolved with no permanent damage noted on fundoscopic exam.

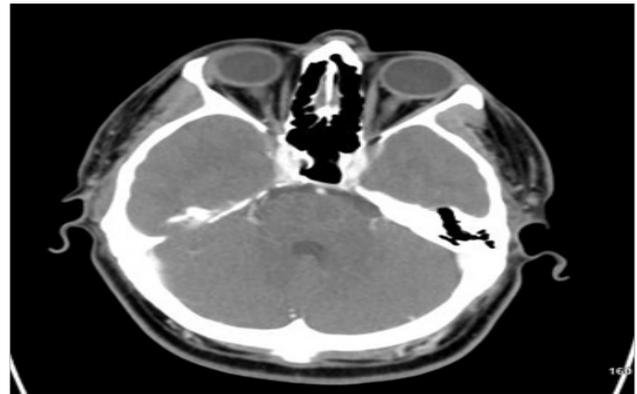


Figure 1 CT head with contrast.



Figure 2 MRI brain without contrast.

## Discussion

Although the incidence of syphilis is highest in MSM, the rates of congenital syphilis in the US have also continued to rise in the past decade, which highlights the need to screen high risk populations and pregnant women to prevent vertical transmission. Within the MSM community, black and Latino males between the ages of 20 and 34 seem to be most at risk for ocular syphilis. However, as this case emphasizes, providers need to be vigilant about assessing populations outside of high risk categories. Per the Center for Disease Control, between 2014 and 2015 388 patients were screened for ocular symptoms that were suspicious for treponemal infection. Only 0.53% in 2014 and 0.65% in 2015 of these patients were found to have ocular syphilis.<sup>1</sup> The epidemiological demographics were consistent with syphilis rates in the US, in the sense that most cases were male, were MSM, and HIV positive. Patient's all had high RPR titer, with an average of 1:128. No specific shared strain was identified between the cases, as has been confirmed with other studies.<sup>2</sup> It is unclear if this particular ocular finding is occurring in a subset of patients as a result of undetermined risk factors.

Of note, there has been a trend in most major medical centers to complement a screening exam for syphilis, such as RPR, with a confirmatory test, like FTA-ABS. This not only eliminates issues regarding false positives with RPR, but will help prevent delays in treatment by conducting the confirmatory test simultaneously with the initial screening test. For those patients in whom non-treponemal testing has been non-reactive due to latency, confirmatory tests have been able to detect disease in patients with secondary and tertiary syphilis. Uveitis, in all of its expressions, seems to be the most prevalent manifestations of ocular syphilis.<sup>3-6</sup> The British Ocular Syphilis Study (BOSS) found that while bilateral ocular syphilis was more common (56%), unioocular syphilis was more likely to be located in the left eye, which was also true of this patient, though no reason for this phenomenon was identified.<sup>5</sup> Inflammation, although not unique to syphilis, has also been noted in most cases of ocular syphilis.<sup>3</sup> Other signs and symptoms include erythema, constriction of the pupil, and floaters seen by the patient. In addition to evidence of uveitis, in one study a patient was found to have imminent central retinal artery occlusion as a presenting sign, highlighting the fact that any ocular manifestation can occur.<sup>7</sup> Luckily, for those presenting with vision loss, most patients regain their vision with standard treatment for neurosyphilis, IV or IM penicillin for 2 weeks.<sup>5</sup> However, instances of delayed treatment have been associated with further visual loss.<sup>3</sup>

Cases have also been noted where re-treatment has been necessary due to new and unresolved ocular symptoms. Steroids have been found to be important in the treatment of particularly severe effects of uveitis but have also been linked to worsening visual conditions, which again highlights the importance of early diagnosis.<sup>7</sup> Patients with delayed diagnosis, especially those with chorioretinitis in the macula, tend to have more complications with vision after therapy.<sup>6</sup> However, methotrexate is starting to be explored to play a potential role in those individuals with residual macular edema after penicillin therapy.<sup>8</sup> Studies have also indicated that although other neurological symptoms may not be present, HIV negative males with ocular syphilis will tend to have abnormal CSF studies, indicating

the importance of lumbar puncture and imaging to rule out other neurological manifestations.<sup>9,10</sup>

## Conclusion

This case of ocular syphilis in a low risk patient with no other symptoms demonstrates three essential points. First, the importance of developing a broad differential diagnosis that includes syphilis when presented with signs or symptoms related to the eye or when symptoms do not point to a singular diagnosis. This is especially important as there is no "typical" systemic or ocular syphilis presentation. Second, if syphilis is suspected or cannot be ruled out, early screening and confirmatory lab work is recommended, as these are low cost tests that could identify an otherwise unlikely culprit. Finally, as exemplified with this patient, ocular symptoms may be the only presenting sign for this easily treatable disease. Although most visual symptoms resolve with treatment, early diagnosis remains crucial for a patient's prognosis.

## Acknowledgements

None.

## Conflict of interest

The author declares that there is no conflict of interest.

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