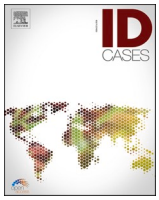
IDCases 37 (2024) e02037

Contents lists available at ScienceDirect

IDCases

journal homepage: www.elsevier.com/locate/idcases



Case report

Acute vision changes as the presenting symptom of ocular syphilis – A case series of two

Sean Leary *, Alyse Volino, Cary Lubkin

Department of Emergency Medicine, Cooper University Hospital, Cooper Medical School of Rowan University, Camden, NJ, United States

ARTICLE INFO

Keywords:
Ocular syphilis
Uveitis
Vision changes

ABSTRACT

Introduction: Syphilis, a bacterial infection caused by Treponema pallidum, can affect a wide variety of organ systems of its host. We aim to present the cases of two patients who presented to an urban, tertiary care, academic Emergency Department with ocular symptoms who underwent imaging, laboratory, and specialist evaluation and were ultimately diagnosed with ocular syphilis.

Case Report: The first patient is a 46-year-old female who presented to the Emergency Department with painless complete vision loss in a unilateral eye. Her exam was significant for bilateral papilledema, and further workup revealed a retinal detachment in the affected eye. Rapid plasma reagin (RPR) titers resulted positive with 1:128 and a reactive fluorescent treponemal antibody (FTA). Further workup including lumbar puncture and magnetic resonance imaging were unable to be obtained due to patient refusal and multiple discharges against medical advice. The second patient is a 38-year-old female with a history of intravenous drug use who presented for bilateral circumferential peripheral vision loss with central sparing. The examination showed bilateral papilledema. Lumbar puncture was performed with normal intracranial pressure. RPR titers resulted positive with 1:128. MRI and ophthalmology evaluation did not reveal any other etiology or explanation for papilledema.

Conclusion: This case series highlights the importance of considering syphilis as a possible cause of ocular symptoms in high risk patient populations and the need for prompt and appropriate treatment given the increasing prevalence of syphilis worldwide.

Introduction

Cases of syphilis or its hypothesized evolutionary precursors have been identified as far back as 15,000 BC [1] A sexually transmitted infection caused by the spirochete *Treponema pallidum*, syphilis cases have been increasing significantly every year since 2000 with almost 134,000 US cases diagnosed in 2020 [2]. This increase has led to various public health campaigns in recognizing, treating, and preventing the condition.

Divided into three stages - primary, secondary, and tertiary - syphilis can affect a wide variety of body systems, thus making diagnosis fairly elusive on first presentation. Primary syphilis involves painless sores on the penis, vagina, anus or other superficial areas. Secondary syphilis leads to more disseminated symptoms including rash most notably on the palms and soles, lymphadenopathy, fevers and malaise. If untreated, symptoms can progress to tertiary syphilis and involve the nervous system leading to a wide variety of neurological and psychiatric symptoms. Syphilis can present with primary ocular symptoms with or without other central nervous system involvement, during any stage of syphilis [3].

Ocular syphilis can pose difficulty in diagnosis given its varied symptoms as well as its potential to mimic other ocular pathology. Most cases of ocular syphilis manifest as uveitis, though all parts of the eye are at risk of damage during infection. Patients can complain of eye redness, pain, vision loss, floaters, and photophobia depending on the structure of the eye that is affected [3]. Consideration of this pathology as a cause of acute vision changes can lead to prompt recognition and diagnosis resulting in potentially vision saving treatment.

Case one

The first patient is a 46-year-old female with a history of bipolar disorder with multiple prior psychiatric unit admissions, homelessness, injection polysubstance use, hepatitis C but notably HIV negative and

* Correspondence to: Cooper University Hospital, Department of Emergency Medicine, Education and Research Building, Second Floor, 401 Haddon Avenue, Camden, NJ 08103, United States.

E-mail address: Leary-sean@cooperhealth.edu (S. Leary).

https://doi.org/10.1016/j.idcr.2024.e02037
Received 28 September 2023; Received in revised form 21 July 2024; Accepted 29 July 2024
Available online 2 August 2024

fully immunocompetent who presented for evaluation to the Emergency Department for sudden painless vision loss in the right eye. The patient was otherwise asymptomatic without other prior ocular symptoms. Notably, the patient's husband was treated for syphilis two to three years prior. She did have a history of high-risk sexual behavior as a previous sex worker, but it is unclear if she continued to engage in this activity. Exam showed 20/50 vision in the left eye with no vision in the right eye. The patient had no motion or light perception and did not blink to threat in the right eye. Ocular pressures were equal and within normal range bilaterally. There was no redness or signs of swelling or trauma. The right pupil was minimally reactive.

Computed tomography (CT) of the head as well as computed tomography angiography (CTA) of the head and neck was performed without acute abnormality. Point of care ocular ultrasound showed concern for retinal detachment which was confirmed by ophthalmology on dilated exam performed at the bedside. Additionally, bilateral optic disc swelling suggestive of papilledema was noted. Significant labs revealed positive RPR with titers 1:128 with reactive fluorescent treponemal antibody. Ophthalmology as well as neurology consultants both recommended obtaining magnetic resonance imaging (MRI) and magnetic resonance venogram (MRV) of the brain as well as lumbar puncture given that ocular syphilis could cause optic neuritis, uveitis, exudative retinal detachment, as well as potentially contribute to her underlying psychiatric symptoms. Due to repeated patient refusal, none of the above studies were able to be obtained.

The patient had a lengthy hospital stay secondary to refusal of obtaining the MRI/MRV and lumbar puncture, as well as refusal of administration of parenteral penicillin. She received intermittent dosing of penicillin with multiple elopements, but was ultimately switched to suboptimal oral doxycycline. There was also extensive interdisciplinary discussion regarding the patient's medical capacity due to encephalopathy from presumed neurosyphilis. It was deemed that she did, in fact, lack capacity, but she eloped from the hospital prior to full treatment.

Case two

The second patient is a 38-year-old female with a history of bipolar disorder, depression, injection polysubstance use, prior endocarditis, Hashimoto's thyroiditis, homelessness, hepatitis C but notably HIV negative and fully immunocompetent who presented with one week of decreased peripheral vision. She had a history of high-risk sexual behavior as well as sexual assault but refused to further elaborate. Further questioning revealed intact central vision, but circumferential loss of peripheral vision when each eye was isolated. She denied any neurologic or ocular symptoms prior to this event.

Neurological exam was overall unremarkable. Vision was tested at 20/40 in each eye and bilaterally. Dilation was performed at the bedside by ophthalmology which revealed significant bilateral optic disc edema which raised concern for possible elevated intracranial pressure leading to this pathology. MRI/MRV was overall unremarkable with the exception of mildly narrowed anterior lateral ventricles, though lumbar puncture was performed with normal opening pressure. Cerebrospinal fluid infectious workup including cell studies, glucose, and CSF VDRL did not reveal an etiology for possible papilledema. The patient eloped from the medical floor soon after the lumbar puncture was performed. After elopement, serum RPR resulted positive with a titer of 1:128. Chart review showed a negative RPR one year prior.

A few weeks later, the patient re-presented to the Emergency Department for a separate, unrelated complaint. The patient was ultimately admitted for further management of untreated positive RPR as well as concern for bacteremia. The patient was subsequently diagnosed with gram positive cocci bacteremia as well as fungemia. Ophthalmology re-evaluated the patient which redemonstrated papilledema with visual field loss. Ophthalmology and infectious disease evaluations stated that due to normal imaging and opening pressure as well as ocular exam without signs of anterior uveitis or candidal fungal infection, the most likely explanation for patient's papilledema was ocular syphilis, and that she should be treated for ocular syphilis in the context of a newly positive RPR. The patient ultimately left against medical advice prior to initiation of treatment.

Discussion

One study of 35,000 cases of syphilis in the United States show that roughly 0.53 % to 0.65 % had suspected ocular syphilis, while another study suggests this number could be as high as 2.6 %, though this number is likely underreported due to lack of full ophthalmic evaluation of every patient [4,5]. Overall, there is scarce documentation of cases of ocular syphilis through the lens of emergency medicine. Previous cases of ocular syphilis have been described in HIV co-infected individuals as the two infections often have a close infective association in patients [6]. Neither patient presented in this case series were positive for HIV or immunocompromised, further highlighting the importance of considering this diagnosis with significant morbidity in a broader range of non-immunocompromised individuals who present to the Emergency Department. Special consideration should be given to those who engage in high-risk sexual activity as both patients in this case series had. This can include sex work, frequent new partners, lack of barrier contraception, or those at risk of assault including patient's suffering from homelessness and substance use. Given the overall rising trends in the incidence of syphilis in the United States, the recognition and diagnosis of ocular symptoms of syphilis are paramount to the comprehensive evaluation of those in at-risk populations.

Syphilis has the potential to affect all ocular structures, though uveitis, mostly posterior uveitis or panuveitis, is the most common manifestation [7]. Another common ocular manifestation of syphilis is optic nerve involvement, including papilledema, papillitis, or optic neuritis. Other presentations can include chorioretinitis, necrotizing retinitis, neuroretinitis, retinal vasculitis, retinal artery and vein occlusion, and exudative retinal detachment. More uncommonly, syphilis can present with scleritis or episcleritis [3]. Papilledema, which was seen in both of the cases we present, refers to swelling of the optic discs that can be caused by elevated intracranial pressure. The exact mechanism of syphilis causing papilledema is unknown, though it has been hypothesized that meningeal irritation secondary to infection with syphilis can cause secondary intracranial hypertension. Alternatively, it has been hypothesized that the mechanism of syphilis causing optic disc edema is independent of elevated intracranial hypertension, in which case the optic disc edema would be described as papillitis rather than papilledema [8]. This hypothesis could explain the normal opening pressure on lumbar puncture in the patient we present who had bilateral optic disc edema.

Diagnosis of ocular syphilis can be difficult due to the vague and nonspecific symptoms with which it can present and variable response to our standard testing. Systemic evaluation with nontreponemal tests including rapid plasma reagin (RPR) and Venereal disease research laboratory (VRDL) can be helpful in those with ocular complaints. Lumbar puncture should be performed in patients with a history of or current syphilis who present with new ocular complaints. Attention should still again be drawn to the caveat that patients can present with ocular syphilis with or without CSF abnormalities [9]. Color fundus photography and fundus autofluorescence are used to document inflammatory changes at the level of the retina, choroid, and optic nerve. CT and MRI of the brain as well as lumbar puncture can be used to exclude alternative etiologies of ocular symptoms.

Delayed treatment has the potential to result in long term visual impairment due to a myriad of complications including cataracts, posterior synechiae, ocular hypertension, optic nerve atrophy, glaucoma, retinal detachment, and retinal vascular occlusions, among others [3]. Current guidelines recommend ocular syphilis be treated as neurosyphilis with penicillin G 3–4 million units intravenous every four hours for 10–14 days, regardless of the presence or lack of CSF abnormalities.

Alternatives would include ceftriaxone 2 g intravenously daily for 10–14 days or more suboptimal oral doxycycline 200 mg twice daily for 21–28 days, though the efficacy for this remains unclear. Those with penicillin allergies should be desensitized and then treated with penicillin [10]. The prompt recognition, evaluation, diagnosis, and treatment is critical to prevent these potentially permanent complications.

Conclusion

The cases described above highlight the importance of giving consideration to syphilis as a potential cause of patients with acute vision changes in the Emergency Department population. Patients can present with a wide variety of ocular complaints or symptoms and with a thorough history and proper ocular exam, the diagnosis can be made to facilitate proper treatment to limit the risk of vision-threatening complications.

Ethical approval

N/A.

Funding

This research did not receive any specific grant from funding agencies in the public, commercial, or not-for-profit sectors.

CRediT authorship contribution statement

Sean Leary: Conceptualization, Data curation, Writing – original draft. **Alyse Volino:** Supervision, Writing – review & editing, Conceptualization. **Cary Lubkin:** Conceptualization, Supervision, Writing – review & editing.

Consent

Unable to be obtained due to patients being untraceable. Identifying information removed from manuscript and patients anonymised.

Declaration of Competing Interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

References

[1] Tampa M, Sarbu I, Matei C, et al. Brief history of syphilis. J Med Life 2014;7(1):4–10.
[2] Division of STD Prevention, National Center for HIV, Viral Hepatitis, STD, and TB Prevention, Centers for Disease Control and Prevention.
[3] Furtado JM, Simões M, Vasconcelos-Santos D, et al. Ocular syphilis. Surv Ophthalmol 2022;67(2):440–62.
[4] Oliver SE, Aubin M, Atwell L, et al. Ocular syphilis – eight jurisdictions, United States, 2014-2015. MMWR Morb Mortal Wkly Rep 2016;65(43):1185–8.
[5] Gupta V, Shoughy SS, Mahajan S, et al. Clinics of ocular tuberculosis. Ocul Immunol Inflamm 2015;23(1):14–24.
[6] Lee MI, Lee AW, Sumsion SM, Gorchynski JA. Don't forget what you can't see: a case of ocular syphilis. West J Emerg Med 2016 Jul;17(4):473–6.
[7] Yang P, Zhang N, Li F, et al. Ocular manifestations of syphilitic uveitis in Chinese patients. Retina 2012;32(9):1906–14.
[8] Chen John Jing-Wei, Thurtell Matthew. Characterizing optic disc edema in the setting of neurosyphilis. Invest Ophthalmol Vis Sci 2015;56(7):3881.
[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.
[10] Hamill MM, Ghanem KG, Tuddenham S. State-of-the-art review: neurosyphilis. Clin Infect Dis 2024;78(5):e57–68.