A case of ocular syphilis in a patient with unilateral progressive loss of vision

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SUMMARY

Ocular syphilis is a rare manifestation of systemic infection by Treponema pallidum, capable of affecting multiple ocular structures and causing irreversible vision loss if not promptly treated. We report a case of progressive bilateral visual impairment caused by ocular syphilis. A 49-year-old woman presented with progressively worsening blurry vision in both eyes over one month. There was no history of oral or genital ulcers, joint pain, tuberculosis contact, or high-risk sexual behaviour. Visual acuity was counting fingers in the right eye and 6/36 in the left. Relative afferent pupillary defect (RAPD) was noted in the right eye. Fundus examination revealed a swollen optic disc in both eyes. The right eye showed superonasal retinitis, retinal hemorrhages, vitritis, and signs of vasculitis. The left eye had no retinitis, vasculitis or hemorrhage. Syphilis serology was positive with an elevated erythrocyte sedimentation rate (ESR). Brain imaging revealed incidental finding of a cystic sellar lesion. Fundus fluorescein angiography showed areas of non-perfusion in the right superonasal retina. The patient received intravenous penicillin G 4 million units every 4 hours for 14 days and underwent sectoral laser photocoagulation in the right eye. Visual acuity improved to 2/60 in the right eye and 6/12 in the left. This case highlights the importance of early recognition and prompt treatment of ocular syphilis to prevent long-term vision loss, as severe or uncommon presentations may be associated with poorer outcomes.

INTRODUCTION

Syphilis caused by treponema pallidum (TP) remains a major global health problem. According to CDC, in national overview of STI in 2023, 209,253 cases of syphilis all stages including congenital syphilis were reported which is the greatest number of cases reported since 1950 and an increase of 1.0% since 2022.[1] Ocular syphilis is an uncommon but important complication of syphilis. Untreated ocular syphilis may lead to permanent vision loss.[2]

Ocular involvement can occur at any stage of syphilis in both HIV-positive and HIV-negative individuals. Eye involvement may be asymptomatic or present as an anterior, intermediate or posterior uveitis, a retinal vasculitis, retinitis, optic neuritis or scleritis.[3] Most cases of ocular syphilis present as uveitis, and the visual acuity depends on structures involved.[4]

As ocular syphilis can affect most structures in the eye and is known as The Great Masquerade, this makes diagnosis challenging. Thus, a high index of clinical suspicion is required to identify *Treponema pallidum* as the causative agent.

CASE PRESENTATION

A 49-year-old woman with no prior known medical illnesses presented with progressively worsening unilateral blurry vision for one month. She reported no associated symptoms such as eye redness or ocular pain. There was no relevant family history, no known contact with tuberculosis patients, and no joint pain, oral or genital ulcers. She denied any high-risk behaviours. She had been married for 23 years and has three children. Her husband also denied any high-risk behaviours or history of promiscuity.

On general examination, she appeared healthy, conscious, and well-oriented to time, place, and person. There were no skin lesions noted. Neurological examination, including other cranial nerves, was unremarkable. Respiratory and cardiovascular examinations were normal. No lymphadenopathy or hepatosplenomegaly was elicited.

Ocular examination revealed a visual acuity of counting fingers in the right eye and 6/36 in the left eye. Positive RAPD was noted in the right eye, along with reduced light brightness and red desaturation. Ishihara colour vision testing revealed 0/21 in the right eye and 10/21 in the left eye. The anterior segments of both eyes were normal. Anterior vitreous cells were present bilaterally, suggesting intraocular inflammation.

Fundus examination of the right eye revealed a swollen optic disc with obscuration of the major vessels. There was a patch of retinitis with surrounding retinal haemorrhage located superonasally, approximately 3-disc diameters in size, along with vitritis. The retinal vessels were slightly tortuous, with evidence of vasculitis and sclerosed vessels in the superonasal quadrant. There was no macular star (Figure 1a). Fundus examination of the left eye showed a swollen optic disc with vessel obscuration, although no vitritis, retinitis, or vasculitis was seen. The vessels were slightly tortuous, and the macular reflex appeared dull (Figure 1b). Intraocular pressure was within normal range in both eyes.

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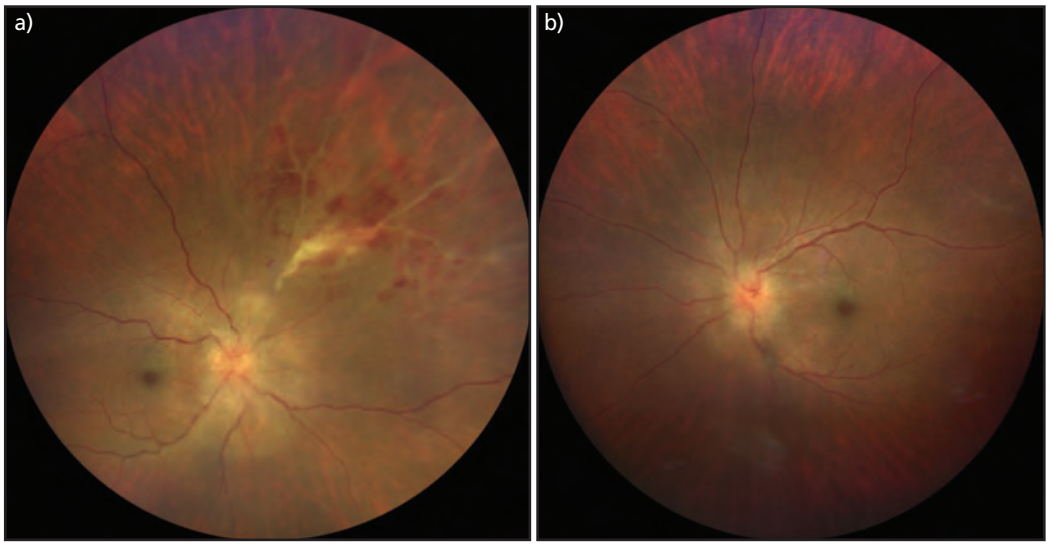

Fig. 1: a) Right eye fundus showed swollen optic disc and presence of retinitis at superonasal area surrounded by retinal haemorrhage with vasculitis and sclerosed vessels
b) Left eye fundus showed swollen optic disc with no retinitis or vasculitis

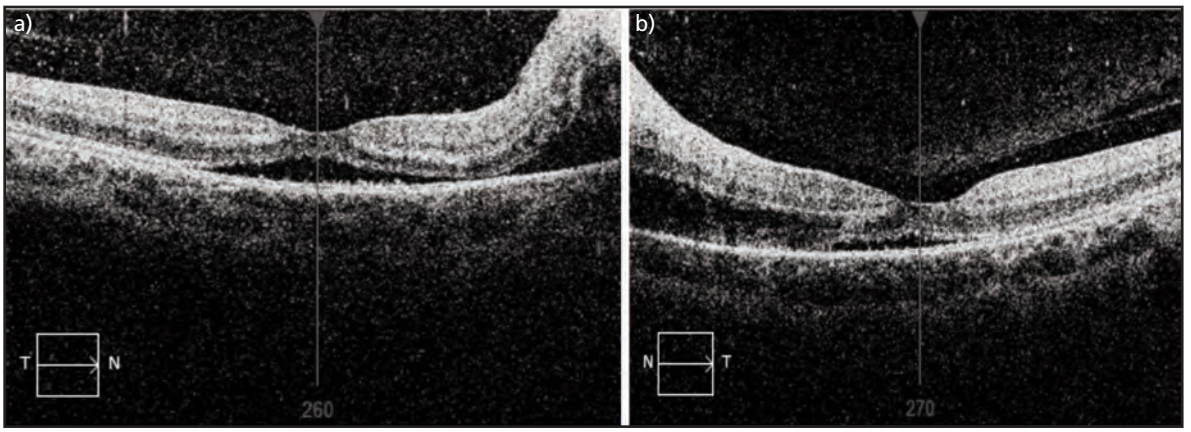

Fig. 2: Optical coherence tomography of the macula showed presence of peripapillary subretinal fluid with irregular protrusion of RPE in both eyes

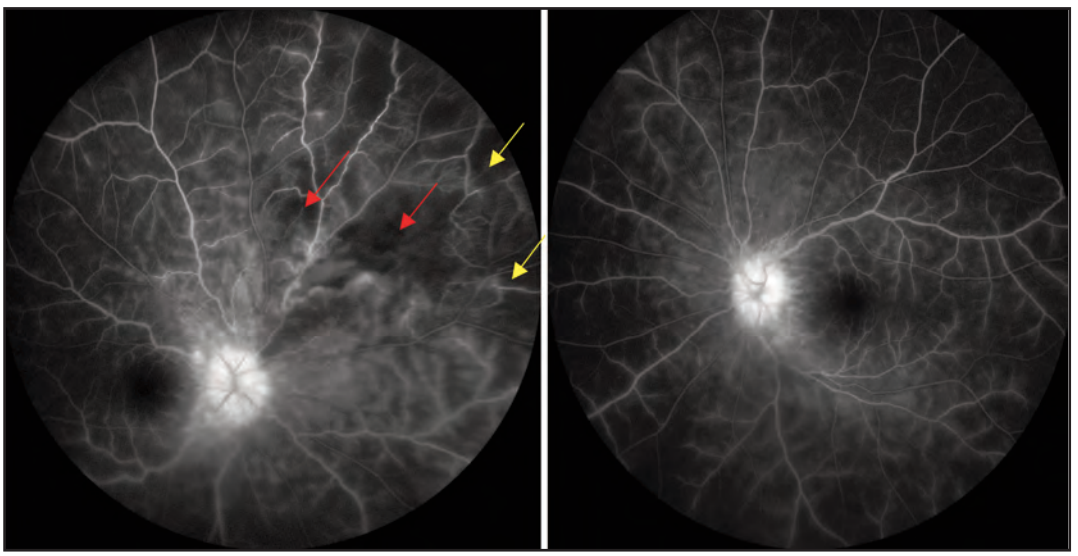

Fig. 3: FFA demonstrated masking from the haemorrhage retinitis (red arrow) with small areas of non-perfusion (yellow arrow) distal to it over superonasal quadrant of the right eye eye without leakage. FFA of the left eye was normal

Laboratory investigations revealed reactive syphilis serology, with an ESL titre of 219 and a rapid plasma reagin titre of 1:32, indicating active syphilitic infection. The ESR was elevated at 81 mm/h. Screening tests for HIV and hepatitis were non-reactive. A contrast-enhanced CT scan of the brain showed incidental finding of cystic sellar lesion thus she was referred to neurosurgical team. Optical coherence tomography (OCT) macula of the right eye showed presence of peripapillary subretinal fluid extending subfoveally with irregular protrusions of the retinal pigment epithelium (RPE) at macula and hyper reflective dots in the vitrous. Whereas for the left eye showed peripapillary fluid extending towards macula with subfoveal fluid and irregular protrusions of the RPE. (Figure 2). FFA showed masking from the haemorrhage retinitis with small areas of non-perfusion distal to it over superonasal quadrant of the right eye, however there was no dye leakage that indicate retinal neovascularization (Figure 3).

The patient was treated with IV Crystalline Penicillin G at a dose of 4 megaunits every 4 hours for a total duration of two weeks. In addition, sectoral laser photocoagulation was performed over the superonasal quadrant of the right eye. Upon completion of treatment, her vision showed only slight improvement, with visual acuity improving to 2/60 in the right eye and 6/12 in the left eye. The patient was discharged after completing her treatment course.

DISCUSSION

Syphilis is a systemic, bacterial infection caused by the spirochete Treponema pallidum. Due to its many protean clinical manifestations, it has been named the "great imitator and mimicker." Syphilis remains a contemporary plague that continues to afflict millions of people worldwide.[5] Most cases are sexually transmitted, but it also can be acquired congenitally. It can be recognized both in immunocompetent and immunocompromised individuals.

Ocular syphilis presents with varied phenotypes, including anterior uveitis, interstitial keratitis, scleritis, intermediate uveitis, posterior uveitis such as retinitis, chorioretinitis, and retinal vasculitis, as well as the distinctive acute syphilitic posterior placoid chorioretinitis. Optic nerve involvement may manifest as optic neuritis, papillitis, or optic peri neuritis, while panuveitis is also common. A study from Mohd Fadzil et al and Yang et al found that panuveitis is the most common presentation.[4,6] Other rarer presentations of ocular syphilis, such as stromal keratitis, iridocyclitis, necrotizing retinitis, optic atrophy, and retinal vasculitis, are only found in patients that have long-standing tertiary syphilis.[7]

As for our patient, she presented with right posterior uveitis with optic nerve involvement, while the left eye demonstrated isolated optic nerve involvement. The diagnosis was confirmed by reactive syphilis serology. Other than that, we also need to rule out other causes such as sarcoidosis, tuberculosis and other viral retinitis. A CT scan of the brain was performed and revealed a cystic lesion in the sellar region. To further evaluate the nature of this lesion and to rule out the possibility of a syphilitic cerebral gumma, an MRI of the brain was conducted. The MRI findings of empty sella made a syphilitic gumma appear unlikely.

It is important to note that ocular syphilis is a recognized variant of neurosyphilis and may present with similar features. The diagnosis of neurosyphilis remains a challenge due to the lack of any existing definitive standardized testing. It is, therefore, based on a combination of history, clinical findings, serological testing, and cerebrospinal fluid (CSF) analysis results. Although a definitive diagnosis of neurosyphilis requires CSF examination, it has been argued that this may not be essential if the results will not alter the management plan, as the CDC recommends the same treatment regimen for both ocular syphilis and neurosyphilis. In our case, the patient declined lumbar puncture.

Our patient was treated with IV Crystalline Penicillin G of 4 million units every 4 hours for 2 weeks duration as recommended by CDC. For treatment of ocular syphilis and neurosyphilis, the CDC recommends 18– 24 million units of IV penicillin for 10–14 days, with limited data to support IV ceftriaxone 1–2 grams for 10–14 days as an alternative therapy for a penicillin allergy.[8] The patient was also treated with sectoral laser pan-retinal photocoagulation due to the presence of capillary non-perfusion area without evidence of leakage on FFA. Some articles suggested that combination of laser pan retinal photocoagulation (PRP) and intravitreal anti-vascular endothelial growth factor (anti-VEGF) therapy is an effective treatment for occlusive retinal vasculitis with neovascularization.[9,10]

Ocular syphilis is a treatable condition, and visual prognosis is generally favourable with appropriate antibiotic therapy, particularly when initiated before significant loss of visual acuity occurs.[11] In our patient, however, visual recovery was limited, predominantly in the right eye. This poor outcome may be explained by several factors highlighted by Zhang et al, who reported that delayed initiation of treatment (>12 weeks after onset of uveitis), prolonged ocular symptoms (>28 days), presence of macular oedema or chronic optic neuropathy, HIV co-infection, and poor baseline visual acuity are associated with worse visual prognosis.[12] At follow-up, Ishihara colour vision testing demonstrated minimal improvement, with OCT macula showing resolution of peripapillary sub foveal fluid. Nevertheless, temporal pallor of the right optic disc was noted, indicating optic atrophy.

CONCLUSION

Syphilis may go undetected without a high index of clinical suspicion due to its nonspecific presentations. All patients with vision loss and ocular inflammation should have syphilis testing as a part of their infectious disease workup. Early diagnosis and prompt treatment after onset of symptoms may contribute to a more favourable prognosis for ocular syphilis.

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