

A case of ocular neurosyphilis in a patient with HIV

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ABSTRACT

Neurosyphilis, although very uncommon, still does occur, especially in patients with HIV infection. The clinical presentation is variable and can include abnormal gait, numbness, poor concentration, neuropsychiatric symptoms, and meningitis; very rarely it can lead to ocular syphilis. We report a patient with HIV who presented with ocular syphilis leading to blindness. He was started on IV antibiotics, and his vision recovered almost to normal. Although the treatment is very straightforward, the timing of diagnosis and starting antibiotics early in the course are crucial to prevent serious consequences, such as permanent loss of vision.

Key words: Neurosyphilis, HIV, blindness

INTRODUCTION

Neurosyphilis in immunocompetent individuals is very rare now compared to the pre-antibiotic era. The incidence is still several fold higher in immunodeficient individuals, especially in patients with human immunodeficiency virus (HIV) infection. The cases we found in a literature review involved mostly the brain; eye involvement occurred infrequently. Ocular syphilis is a type of neurosyphilis that can threaten vision leading to blindness if not treated promptly. Its diagnosis can be challenging, but it should be considered if the neuro-ophthalmological findings cannot be explained by any other commonly encountered disease, and diagnosis by serology should be considered adequate since it is a potentially treatable condition.¹

CASE

We report a 27-year-old man with HIV, not on any therapy, who came to the hospital with complete loss

of vision in the right eye for 4 months and partial loss of vision in the left eye for 5 weeks. He also had a 6-month history of maculopapular rash over his trunk that extended to involve the entire body, including the palms, prior to presentation. He denied any redness, watering, or itchiness of the eyes and focal neurological deficits. He also denied a history of diabetes, sexually transmitted infections (STIs), and recent trauma to the eyes.

On physical examination, he was cachectic. His visual acuity was less than 20/200 in his right eye and 20/100 in his left eye. There was desquamative rash on his palms but not on his trunk or genitals, and he had no lymphadenopathy (Figure 1).

Normal laboratory tests included complete blood counts, hepatic, renal, and thyroid function tests,



Figure 1. Desquamative rash on the palms.

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fasting blood glucose, electrolytes, erythrocyte sedimentation rate, and C-reactive protein. Cerebrospinal fluid analysis showed a lymphocytic pleocytosis (WBC: 110/ μ L and lymphocyte count: 98/ μ L), a low glucose level (29 mg/dL), and a high protein level (82 mg/dL). A diagnosis of neurosyphilis was confirmed with a reactive serum rapid plasma Reagin (RPR) test and a positive CSF venereal disease research laboratory (VDRL) test. His CD4 count was 418/mL, and his viral load was 289,000/mL. He tested negative for other STIs, including genital herpes and gonorrhea, tuberculosis and hepatitis. Intravenous penicillin G was administered for 10 days. HIV treatment was held to avoid the immune reconstitution inflammatory syndrome.

He was discharged with better vision to follow-up with ophthalmology and primary care to initiate anti-retroviral therapy. He was advised to observe safe sex practices. When he followed up with outpatient ophthalmology, slit lamp examination showed 270 degrees synechia in the left iris and trace cataracts and nuclear sclerosis in the lenses of both eyes. On fundus examination, no detailed view could be obtained in the right eye, but the left eye had trace vitritis, scatter granules RPE changes in the periphery and chorioretinal scarring. B-scan, fundoscopy, and optical coherence tomography of the macula showed panuveitis, chorioretinitis, nuclear sclerosis cataract, and disc hyperemia without signs of raised intraocular pressure. In three months, he completely regained vision in his left eye and had partial recovery of vision in his right eye.

DISCUSSION

Painless vision loss without accompanying symptoms or trauma in a young individual without any obvious cause, such as diabetes, presents a wide differential diagnosis that can be categorized into compressive, mitochondrial, vascular, infectious versus noninfectious optic neuritis, and infiltrative etiologies. Pituitary lesions, meningioma, slowly growing aneurysm, leukemia, and lymphoma were considered but were less likely due to absence of raised intraocular pressure. Noninfectious causes, including sarcoidosis, and infectious causes like tuberculosis were considered, but were not likely given the absence of

constitutional symptoms. Bilateral anterior ischemic optic neuropathy was also considered, but our patient lacked the typical acute onset of vision loss commonly associated with this disease.

Our patient's HIV history pointed toward an infectious etiology. A reactive serum RPR and CSF VDRL confirmed neurosyphilis; disc hyperemia on ophthalmoscopy and the positive response to IV antibiotic eventually validated the diagnosis. Ocular syphilis commonly causes posterior uveitis and panuveitis but may occasionally cause anterior uveitis, optic neuropathy, retinal vasculitis, and interstitial keratitis. It can occur in any stage of syphilis, including primary and secondary syphilis and can cause decreased visual acuity and eventually permanent blindness.²

The diagnosis requires clinical suspicion since a negative CSF VDRL does not rule out neurosyphilis, and only 35% cases of ocular syphilis have neurosyphilis. The recommended adult regimen is IV aqueous crystalline penicillin G 18–24 million units per day (either as continuous infusion or 3–4 million units every 4 hours) for 10–14 days. An alternative regimen for adults is procaine penicillin 2.4 million units IM per day and oral probenecid 500mg four times per day, also for 10–14 days.³ Co-administration of corticosteroids or immunosuppressant drugs with antibiotics is controversial. Oral corticosteroids with topical steroids, NSAIDs, mydriatics like atropine, and lubricating agents are often required to control ocular inflammation. The use of intravitreal dexamethasone implant for refractory macular edema in syphilitic uveitis has also been reported.⁴ However, corticosteroids should not be started before starting antibiotics as this could worsen the disease to a sight and/or life-threatening degree.⁵ Patients treated for syphilis should have the VDRL test repeated every three months for a period of one year post treatment since titers should become nonreactive within a year after therapy. Patients should be retreated, if an initially high-titer VDRL test does not decrease fourfold within a year, or if a previously non-reactive VDRL test becomes reactive again.⁶

Factors associated with poor visual prognosis include the time between onset of uveitis and treatment (>12 weeks), longer duration of ocular symptoms (>28 days) as seen with our patient, the presence of

macular edema or long-standing optic neuropathy, coinfection with HIV, and poor initial visual acuity. Factors associated with higher success rates include the presence of vasculitis (detected by fundus fluorescence angiography) or anterior uveitis. Common long-term complications of syphilitic uveitis include the development of glaucoma, cataracts, epiretinal membranes, and macular edema. Choroidal neovascularization and widespread chorioretinal scarring can occur in some patients. The Jarisch-Herxheimer reaction following initiation of antibiotic therapy may result in fever, malaise, and headache, as well as a worsening of ocular manifestations and may be prevented by administration of systemic corticosteroids concurrent with antibiotic treatment.⁷

CONCLUSION

Ocular syphilis, although a rare entity, if overlooked in the early stages, can lead to irreversible damage to the eyes. Early diagnosis and treatment can prevent permanent blindness.

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