

## CASE REPORT

# Ocular Syphilis – Uncommon but not Rare

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### ABSTRACT:

Syphilis is a multisystem bacterial infection caused by the spirochete *Treponema Pallidum*, which has regained attention as a new epidemic due to worldwide increase in the cases of this disease. Ocular syphilis is an unusual manifestation of disease, mostly occurring in the secondary or tertiary stage of disease. Ocular involvement may be the presenting manifestation of syphilis and it is often associated with delayed diagnosis and treatment, which may result in irreversible structural and functional ocular damage. Although, ocular syphilis can affect any part of eye, uveitis is the most common ocular manifestation of the disease. A high index of suspicion is mandatory as ocular syphilis can mimic variety of other ocular inflammations. We report two cases of syphilitic panuveitis where high index of suspicion resulted in the correct diagnosis and management.

**Keywords:** Ocular Syphilis, Uveitis, Panuveitis, Neurosyphilis.

### INTRODUCTION:

Due to its highly variable ocular manifestations, syphilis is known as “the great masquerader” resulting from infection by the spirochete *Treponema Pallidum*. During the last two decades, there is resurgence of the disease with a WHO estimate of about 11 million new cases of syphilis worldwide every year, over 90% of which occur in developing countries.<sup>1</sup> The infection has also re-emerged in developed countries in homosexual population with high number of cases co-infected with HIV.<sup>2,3</sup> Ocular syphilis is an uncommon presentation of syphilis that can occur in any stage of the disease and involve any ocular tissue, but typically occurs in secondary and tertiary stage.<sup>1,4</sup> Common ocular manifestations of syphilis include interstitial keratitis, anterior, intermediate and posterior uveitis, vitritis, chorioretinitis, retinitis, retinal vasculitis, secondary glaucoma, cataract, and cranial nerve and optic neuropathy.<sup>5,6,7</sup>

Uveitis is the most common ocular manifestation of syphilis and poses a diagnostic and therapeutic challenge as it is resistant to the conventional treatment of uveitis. Diagnosis can be confirmed on serological analysis that includes non treponemal tests [Venereal Disease Research Laboratory (VDRL) and Rapid Plasma Reagin (RPR) tests] and treponemal tests [Fluorescent treponemal antibody absorption (FTA-ABS) and *Treponema Pallidum* Haemagglutination assay (TPHA) test]. We report two cases of syphilitic uveitis where high index of suspicion resulted in the correct diagnosis and timely management with favorable outcome.

### CASE REPORT:

**Case 1:** A 42-year-old male presented with bilateral progressive painless visual deterioration and floaters for the last two years. Available medical record revealed that he has been treated with topical steroids for recurrent uveitis by various ophthalmologists. Systemic inquiry revealed history of frequent extra marital sexual contacts and penile ulceration 3 years ago. General physical and systemic examination did not reveal any abnormality. On ocular examination, his best corrected visual acuity (BCVA) was 6/18 in both eyes. Anterior segment examination showed cells (++) in both eyes with intraocular pressures (IOP) of 19 and 16 mm Hg. Posterior segment examination revealed bilateral vitreous haze and snow ball opacities in inferior vitreous along with dull foveal reflex. Serological studies revealed positive TPHA test (1/320 titer) and Hepatitis Anti HCV antibody, while VDRL test and HIV test were negative. CSF examination revealed elevated protein levels of 100 mg/dl while CSF VDRL and TPHA were negative. Once the diagnosis of ocular syphilis was established, he was given intravenous injection Penicillin 1 Million units 6 hourly for two weeks followed by 3 weekly doses of intramuscular injection Benzathine Penicillin (2.4 M units) under supervision of consultant dermatologist. Concomitant treatment with topical 1% Prednisolone acetate eye drops, Nepafenac eye drops and 1% Cyclopentolate eye drops was continued for ocular inflammation. At 6 months follow up visit patient was asymptomatic with BCVA of 6/6 in right eye and 6/7.5 in left eye with negative VDRL and TPHA reports.

**Case 2:** A 40-year-old male reported in eye OPD with a history of pain, redness and decrease vision in both eyes for the last 3 months. Detailed systemic history revealed appearance of skin rashes about 3 months ago but he denied any extra marital sexual exposure. General physical examination showed fading papulo squamous rashes on palms and soles (Figure 1a). At presentation, his BCVA was counting fingers at 1 meter in right eye and 6/18 in left eye. Anterior segment examination showed cells (+++), posterior synechiae and pigment deposit on lens in both eyes with IOP of 10 mm Hg in each eye. Posterior segment examination of right eye showed vitreous haze and opacities along with macular edema, whereas left eye revealed vascular sheathing

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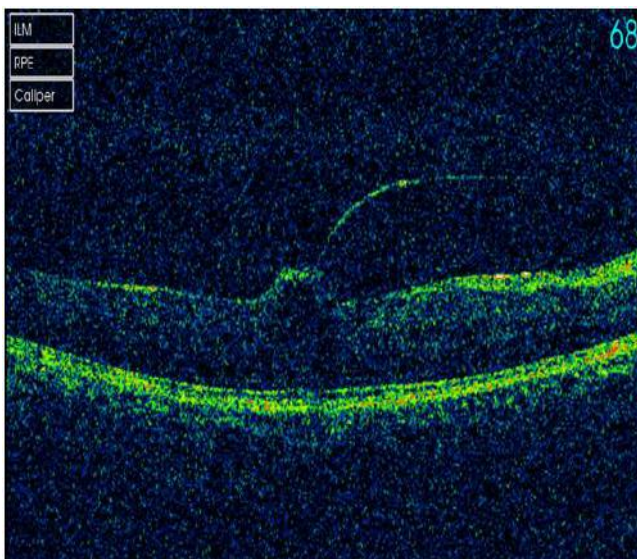
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Received: 15-10-2015  
Revised: 27-10-2015  
Accepted: 01-11-2015

along with vitreous cells and opacities. Serological studies revealed positive TPHA test (1/320 titer) and VDRL test (1/8 titer). However, CSF examination did not reveal any abnormality. Other investigations including HIV test were negative. Initially he was managed with topical and posterior sub-tenon steroid injections by eye specialist. Subsequently, on confirmation of ocular syphilis on serology, consultant dermatologist started him on intravenous injection Penicillin for 14 days followed by 3 weekly doses of injection Benzathene Penicillin 2.4 Million units intramuscularly. At 3 month follow up, he had BCVA of 1/60 in right eye and 6/9 in left eye. Posterior segment examination of right eye showed a few vitreous opacities along with vitreomacular traction and thin cystic retina, whereas left eye showed scattered vitreous opacities and few patches of chorioretinal atrophy and cellophane maculopathy (Figure 1b).

Figure: 1a  
Syphilitic palmer Rash



Figure: 1b  
Vitreomacular Traction on OCT



## DISCUSSION:

Although uncommon, ocular syphilis poses diagnostic challenge for the ophthalmologists because of its protean features and lack of any pathognomonic manifestations. Syphilitic uveitis is an uncommon cause of uveitis accounting for 1.6 -4.5% of cases and it may be granulomatous or non-granulomatous, unilateral or bilateral and can involve anterior, intermediate or posterior segment of eye.<sup>4</sup> Birnbaum in a retrospective analysis of etiology of chronic anterior uveitis over a period of 35 years found ocular syphilis as a cause of anterior uveitis in 2.7% of cases.<sup>8</sup> Both of our cases had bilateral panuveitis. Variable presentation of ocular manifestations associated with ocular syphilis has been reported in worldwide literature. Anshu<sup>9</sup> in a series of 22 consecutive patients in Singapore, reported non granulomatous anterior uveitis (81.8%) as the most frequent finding in ocular syphilis, followed by vitritis (65.4%), papillitis (27.5%), scleritis/episcleritis (22.7%), interstitial keratitis (22.7%), granulomatous uveitis (13.7%), vasculitis (13.7%) and chorioretinitis (13.7%). In British ocular syphilis study (BOSS),<sup>2</sup> panuveitis was the commonest diagnosis affecting 41.3% of eyes followed by optic neuritis (22%), posterior uveitis (12.7%) and pure anterior uveitis (9.5%). Amaratunge<sup>10</sup> in his review article reported posterior uveitis as the commonest presentation of ocular syphilis followed by pan uveitis. Hong<sup>6</sup> found pan uveitis in 78.6%, anterior uveitis in 14.3% and posterior uveitis in 7.1% of eyes of patients with ocular syphilis. Awais<sup>11</sup> reported a case of syphilitic pan uveitis in a young Pakistani male who was managed with intravenous Penicillin G and systemic and periocular steroids. One of our patients had papulo-squamous rashes on palms and soles. Mathew<sup>2</sup> in their prospective study found systemic signs of early syphilis in 29.3% of patients with ocular syphilis which include generalized rash, mucosal lesions and lymphadenopathy, whereas, Francesco<sup>1</sup> et al reported history of papulo-macular rash on the trunk and palms in 26.3% of patients of ocular syphilis. It is recommended that all patients with ocular syphilis should undergo CSF examination and managed for neurosyphilis by giving intravenous Penicillin G (18-24 million units daily) for 10-14 days as per United States center for disease control (CDC) sexually transmitted disease treatment guidelines. Favorable visual and anatomical outcome in cases of ocular syphilis depends on early diagnosis and intervention. Three out of four (75%) eyes in our study achieved final BCVA of 6/12 or better. Mathew<sup>2</sup> reported BCVA of 6/12 or better in 92.1% of eyes after treatment, whereas, Francesco<sup>1</sup> et al found improved BCVA shortly after therapy in 93.3% of cases.

## CONCLUSION:

Diagnosis of ocular syphilis may remain elusive due to extreme variation in presentation and mimicking other ocular inflammations. Awareness of the re-emergence of syphilis and keeping a high index of clinical suspicion in cases of uveitis can allow ophthalmologists to diagnose and treat the disease early, with favorable outcome and

reduced ocular morbidity.

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