Case Report

Bilateral Toxoplasma Retinochoroiditis with Franceschetti's Syndrome in an Immunocompetent Patient - An Unusual Presentation

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Abstract

Toxoplasma gondii is a major cause of chronic ocular parasitic infection which leads to severe visual impairment. Toxoplasma retinochoroiditis is considered to be one of the most common causes of posterior uveitis and is usually unilateral in immunocompetent patients. Toxoplasmosis in immunocompromised patients may be bilateral with a fulminant course. Ocular toxoplasmosis can have protean manifestations, some of which may be atypical, posing diagnostic challenge. We report a case of ocular toxoplasmosis in a 24-year-old female with healthy immune system who presented with bilateral recurrent panuveitis. The fundus examination revealed bilateral active retinochoroiditis with traction band involving the disc and macula (Franceschetti's syndrome). The battery of investigations for granulomatous uveitis were normal. Serology showed evidence of toxoplasmosis and PCR of intraocular fluid confirmed the diagnosis. The patient was treated with anti toxoplasma medications and steroids. Atypical presentations of toxoplasmosis could be a diagnostic dilemma. Toxoplasmosis may present with simultaneous bilateral involvement in the absence of immunosuppressed status. Early diagnosis and prompt treatment could prevent vision threatening complications like traction bands, which portend a poor visual outcome. Toxoplasmosis should be considered in the differential diagnosis of bilateral pan uveitis in an immunocompetent patient and retinal traction bands could be an unusual manifestation of toxoplasma retinochoroiditis.

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Introduction

Toxoplasma gondii is an obligatory intracellular parasite, the asexual form of which produces oocysts in the gut of cats. Toxoplasmosis affects about one third of the human population.[1] Toxoplasma retinochoroiditis is considered to be one of the most common causes of posterior uveitis and is usually unilateral.[2,3] Bilateral and multifocal involvement with fulminant vitritis and retinitis is seen in immunosuppressed patients.[3] Retinal traction bands extending from a healed choroiditis scar to the optic disc, known as Franceschetti's syndrome, is an unusual presenting feature of ocular toxoplasmosis.[4,5]

Case Report

A 24-year old female presented with gradual diminution of visual acuity in both eyes for four years. The patient also had complaints of pain, redness and photophobia. There was no history of penetrating injury to the eye and the patient denied contact with pets. There was no history of collagen vascular disease or systemic illness. There were recurrent episodes in the past and she was on irregular treatment with systemic steroids. She had no features suggestive of underlying immunosuppressive illness.

The best corrected visual acuity was 3/60 in OD and PL+ in OS. Slit lamp examination revealed circumcorneal congestion, mutton fat keratic precipitates, significant anterior chamber reaction, iris nodules, posterior synechiae and festooned pupil in both eyes. Intraocular pressure was normal. (Figure 1-4)

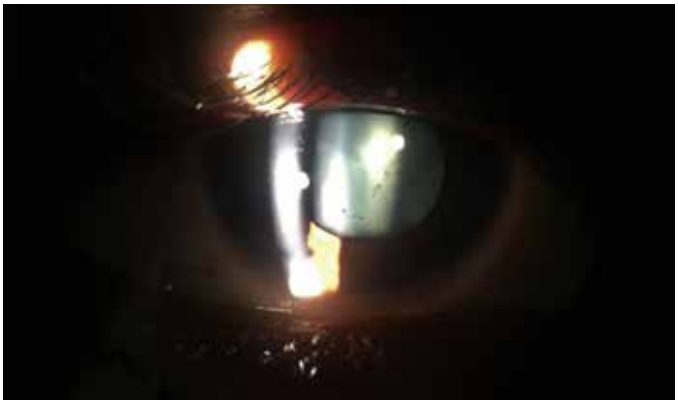

Figure 1: *Right eye: Anterior segment showing mutton fat KP's.*

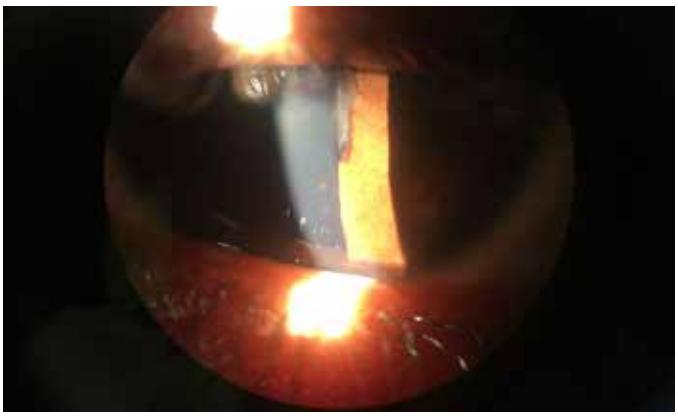

Figure 2: *Left eye: Anterior segment showing mutton fat KP's*

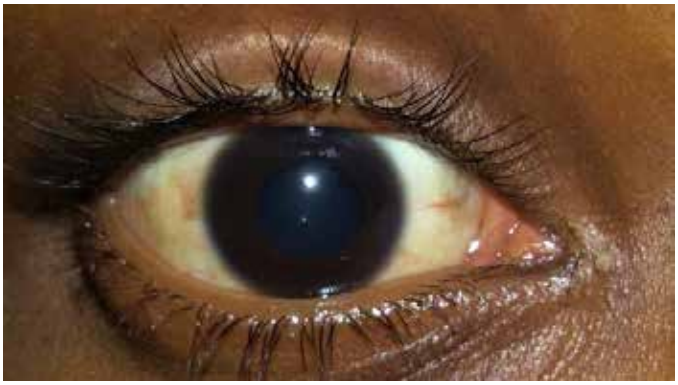

Figure 3: *Right eye: Posterior synechiae*

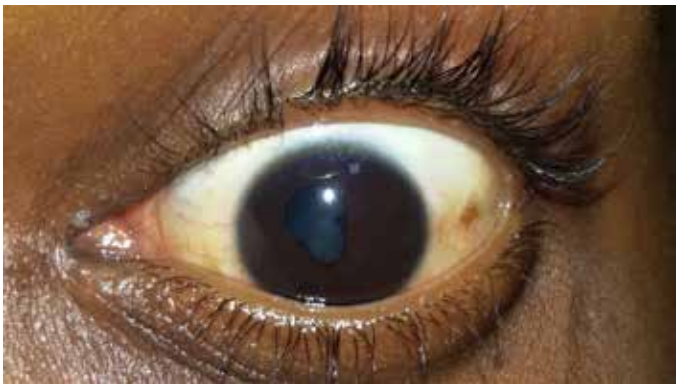

Figure 4: *Left eye: Posterior synechiae*

The posterior segment examination of the right eye revealed severe vitritis, disc edema and a traction band extending from the disc to macula. There was a satellite lesion at the macula, adjacent to the traction band. (Figure 5) The optic disc was barely visualized in the left eye due to severe vitritis, which obscured other details of the fundus.

A battery of investigations were done to exclude granulomatous uveitis including complete blood cell count, chest x-ray, C-reactive protein, erythrocyte sedimentation rate, serum calcium, all of which were unremarkable. Mantoux test was negative and serology for HIV, RA factor, ANA and VDRL were non-reactive. ELISA for toxoplasmosis unveiled IgM antibody to be non-reactive (0.06 Col), equivocal laboratory values being 0.50 - 0.60. IgG antibody was strongly positive (10.7 IU/ml), equivocal laboratory values being 1.6 – 3.0. These values strongly suggest prior exposure to Toxoplasma gondii. PCR of aqueous sample was positive and confirmed the diagnosis of ocular toxoplasmosis.

Pharmacotherapy was started with co-trimoxazole and pyrimethamine, supplemented with oral folinic acid. Oral steroid was started after two days of initiating antibiotics and was gradually tapered over six weeks. The patient was reviewed two weeks later and exhibited a good response to treatment. The visual acuity improved to 5/60 and 4/60 in the right and left eyes respectively. There was reduction in keratic precipitates and anterior chamber reaction in both eyes. The severity of vitritis had remarkably declined and the initial fundus findings were confirmed in the right eye (Figure 6). The vitreous haze in the left eye precluded adequate visualization of the fundus except for the presence of disc edema and central choroiditis, with an adjacent healed retinochoroiditis scar. Traction bands were not visualized in the left eye. There was no view of the periphery (Figure 7).

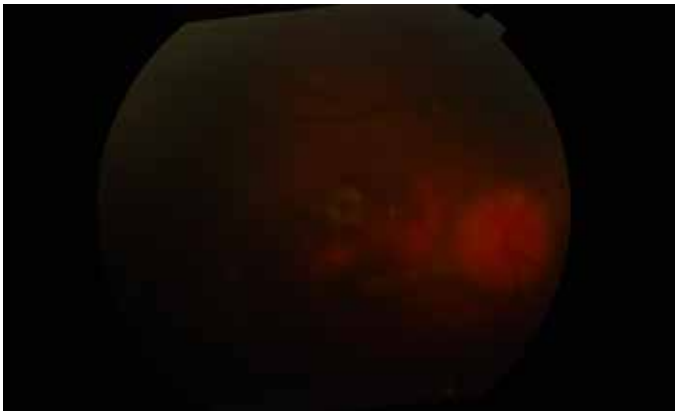

Figure 5: *Right eye: Fundus showing hazy view of satellite scar and traction band (pre-treatment)*

Figure 6: *Right eye: Fundus showing satellite scar and traction band from optic disc towards macula (post-treatment)*

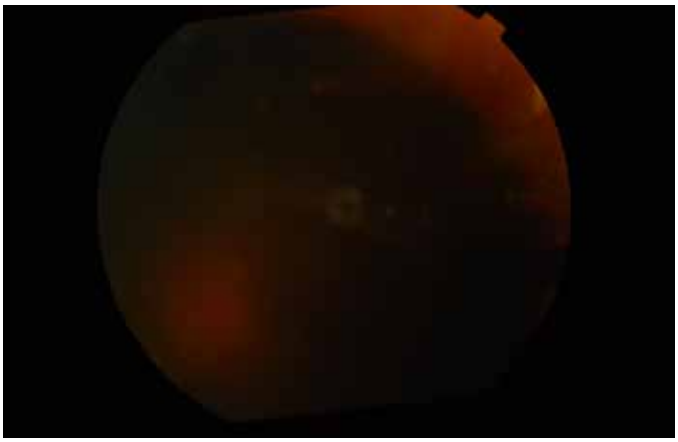

Figure 7: *Left eye: Fundus shows hazy view of optic disc due to intense vitritis (post-treatment)*

Discussion

Toxoplasmosis is an obligate intracellular parasite that has a predilection for immunocompromised individuals. Reports show that the prevalence of seropositivity in India may be

as high as 20-40%.[6] However all seropositive patients do not develop symptomatic disease.[7] 80% of immunocompetent hosts do not show any clinical signs.

Ocular toxoplasmosis a major cause of infectious posterior uveitis worldwide.[6] It accounts for 2.8 to 4.2% of uveitis.[8] Toxoplasmosis is acquired by ingestion of food contaminated with oocysts shed by cats or undercooked meat. The parasite is speculated to reach the retinochoroidal layers through the ciliary vessels or central retinal vessels.[6]

Ocular toxoplasmosis most commonly manifests in second and third decade of life, usually manifesting between 25 and 31 years of age.[2,9] Toxoplasma retinochoroiditis occurs as an unilateral presentation in the majority, either as a primary or reactivated disease. In immunocompromised hosts, the infection tends to be bilateral and multifocal with a fulminant retinitis. 3 20 to 80% are observed to have recurrent episodes and recurrences are more common in immunocompromised individuals. 10 To the best of our knowledge, the occurrence of bilateral acquired ocular toxoplasmosis has been reported only once in an immunocompetent patient. Bilateral ocular toxoplasmosis is atypical and has been described in congenital cases and immunosuppressed individuals.[11]

The most common presenting feature of ocular toxoplasmosis is blurred vision, floaters and metamorphopsia.[12] The other symptoms like pain and photophobia, similar to this case, may be variably present depending on the severity of inflammation.[2]

Toxoplasma typically affects the posterior pole producing a foci of necrotizing retinochoroiditis adjacent to an old pigmented retinochoroidal scar and vitreous inflammation, leading to tissue destruction.[13] The presence of satellite lesion signifies reactivation of toxoplasmosis, which is the commonly described mode of infection in the overwhelming majority of acquired toxoplasmosis, in immunocompetent hosts.[5]

Toxoplasmosis can have protean manifestations. Atypical presenting features include including Coats-like retinopathy, pigmentary retinopathy, retinal vascular occlusion, serous retinal detachment, scleritis, and panuveitis in the absence of overt retinitis.[3] Vitreoretinal traction bands have been described as an uncommon manifestation.[5,14] The retinochoroiditis in this patient was exemplified by the presence of such a traction band, which we consider as a variant of Franceschetti's syndrome.

When the active infection subsides, a well-defined punched out atrophic scar with hyper pigmentated border appears. 10 Retinochoroiditis is associated with retinal arteritis, phlebitis, cystoid macular edema and subretinal neovascular membranes which lead to irreversible damage to the photoreceptors. The outcome is directly related to the extent of residual macular scar.[15] The poor visual outcome in this case is reflected by the presence of scarring and traction band at the posterior pole.

Toxoplasmosis can present with posterior hyaloid thickening and vitreomacular traction bands, where the release of these bands with complete removal of posterior hyaloid membrane is known to improve visual prognosis.[15,16] Such an intervention may be necessary for complete visual recovery in this case.

The diagnosis is confirmed by history, clinical findings and serology. Serological tests include the detection in the serum of immunoglobulins IgM and IgG.[10] Serology has limited value in diagnosis, considering the high seropositivity of the infection in the population. PCR of intraocular fluid is required to establish the diagnosis in atypical cases.[2,3] Though gold standard technique for toxoplasmosis is isolation of T.gondii by bioassay, PCR of aqueous and vitreous may be tested with high sensitivity and specificity for the presence of DNA sequences.[5]

The treatment of ocular toxoplasmosis consists of pyrimethamine, sulfadiazine, folinic acid and systemic corticosteroids. Ocular toxoplasmosis treated with timely antimicrobials and steroids shows good visual and structural outcome.[15] Early diagnosis and prompt treatment can prevent vision threatening complications.

Conclusion

The case is highlighted for its bilateral presentation in an immunocompetent patient. Toxoplasmosis should be considered in the differential diagnosis of bilateral panuveitis and bilaterality can occur in the absence of immunosuppressed status. Unusual presentations like retinal traction bands poses a diagnostic challenge. PCR plays a substantial role in confirming the diagnosis of atypical cases.

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