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A Rare Presentation of Outer Retinal Toxoplasmosis in an Immunocompromised Individual

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Abstract: The immunocompromised state of an individual can give rise to many devastating opportunistic diseases. Outer retina toxoplasmosis is such an example, which is seen mainly in immunocompromised patients. Rapid and precise diagnosis is of utmost importance in treating and preventing complications of the disease. Here we report a rare presentation of outer retinal toxoplasmosis in a patient, immunocompromised due to chemotherapy.

Keywords: Toxoplasmosis, outer retina, immunocompromised, retinitis

INTRODUCTION

Toxoplasmosis is a parasitic disease caused by Toxoplasma *gondii* which can cause severe eye problems. It is one of the commonest organisms which inflict the immunocompromised [1]. The diagnosis of ocular toxoplasmosis is mainly based on clinical findings, as it is difficult to isolate the organism from eye samples. Thus, it is crucial to know the various clinical presentations and subtypes of ocular toxoplasmosis in order to accurately diagnose and to commence treatment promptly. The typical lesion is characterized by a discrete area of retinitis, near the border of a pre-existing retinochoroidal scar [1]. Classical ocular toxoplasmosis usually affects the inner retinal layer with marked vitritis [2]. However deep retinal involvement may also occur, termed outer retinal toxoplasmosis, and is seen more commonly in patients with low immunity. As to our knowledge, there have been no reports of patients, immunocompromised due to chemotherapy, who presented with outer retinal toxoplasmosis and marked vitritis and anterior chamber reaction.

CASE REPORT

An 80-year-old Indian lady, with an underlying past ocular history of left ocular lymphoma presented with a three months history of right eye (RE) painless and progressive blurring of vision. She was noted to be immunocompromised with a white cell count of $0.8x10^9$/L due to the maintenance chemotherapy for her left eye (LE). Vision at presentation was 3/60, pinhole (PH) 6/24, N 36 and 6/36, PH 6/24, N24 in the RE and LE respectively. On examination of the RE, she was pseudophakic with anterior chamber cells of 2+, and vitritis 2+ with sheaths of vitreous condensation whereas, the LE, already proptosed due to the primary ocular lymphoma, was quiet. On examination of the fundus, chorioretinal scars were noted nasal to the optic disc in the RE and temporally in the LE. A vitreous tap was done which was negative for malignant cells. The inflammation in the RE was persistent even on topical steroid treatment, and thus, the patient was referred to the medical retina (MR) team for further management.

On examination with the MR team, her right eye vision was 6/60, PH same, N 24 and left eye vision was 6/36, PH same, N18. Giant cells were noted on the intraocular lens in the right eye with anterior chamber cells of 2+, vitritis 2+ and sheaths of vitreous condensation in the RE, typical of ocular lymphoma (Figure 1). There were no signs of retinitis or vasculitis noted. The LE which was originally quiet also revealed occasional anterior chamber cells with vitritis. She was started on oral and topical steroids and was co-managed with the haematology team. Unfortunately, 1 month later, on top of the persistent anterior chamber inflammation and worsening vitritis, funduscopy showed diffuse yellow- white area of retinitis nasal to the optic disc adjacent to the old chorioretinal scar bilaterally (Figure 2).

Fundus fluorescein angiography (FFA) showed hyperfluorescence over the lesions with diffuse vasculitis bilaterally and hot discs in both eyes (Figure 3). Optical Coherence Tomography over the lesions showed intraretinal lesions of the outer retina layer

(Figure 4a and b). Infectious screening was done including serum Tuberculosis Quantiferon, Cytomegalovirus (CMV), Epstein Barr virus (EBV), Varizella Zoster virus (VZV), Syphilis (VDRL) and vitreous tap for HSV 1 and 2, CMV, EBV and VZV which all produced negative results. Serum Toxoplasma investigations, however, yielded positive results for Toxoplasma IgG but Toxoplasma IgM was equivocal.

A clinical diagnosis of toxoplasmosis-related chorioretinitis was made. She was subsequently treated with oral Azithromycin 500mg OD which showed immediate positive response as evidenced by reduced vitritis and retinitis.

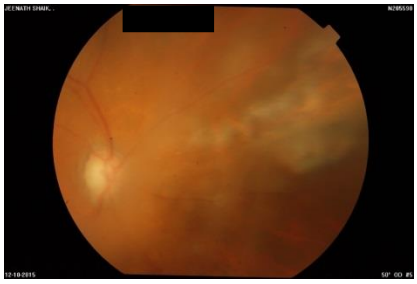

Fig-1:Hazy fundus view due to the sheaths of AC cells and vitritis

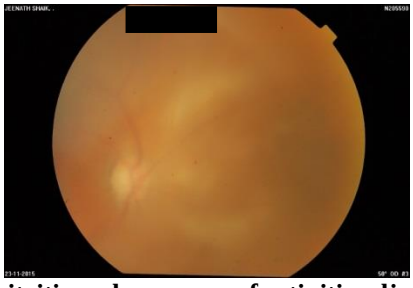

Fig-2: Worsening vitritis and new area of retinitis adjacent to the old scar

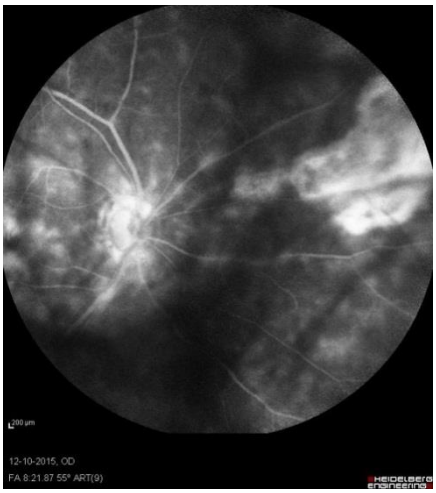

Fig-3: FFA of the RE showing hot disc and hyperfluorescence of the retina. Overlying dense vitreous condensation is also seen

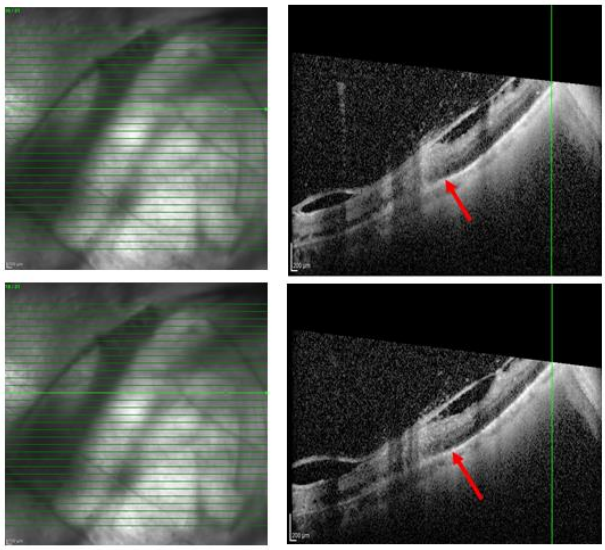

Fig-4a and b: OCT showing outer retina layer involvement

DISCUSSION

Toxoplasma gondii is an organism which has a predilection for the central nervous system, thus making it the commonest infectious cause of retinal inflammation especialy in immunocompromised hosts [1]. Outer retinal toxoplasomosis is an atypical manifestation of toxoplasmosis, which was first described by Gass in 1968 and further described by Friedmann and Knox in 1969 [3]. It is a subset of ocular toxoplasmosis that initially and primarily affects the outer retinal layers, and was described as having only mild or no vitreous reaction [3]. It was proposed by Doft and Gass, that the changes in outer retinal toxoplasmosis were encysted Toxoplasma gondii or focal outer retinal gliotic scars [5]. Further postulation is that the involvement of the outer retina may, in fact, be just an early manifestation of the disease, but as it usually occurs near the macula, visual symptoms occur earlier, thus, leading to earlier detection, before involvement of the inner retina [2].

In our patient, the vitritis was noted to be marked, and not mild. One possible explanation for the severe reaction is that, outer retina toxoplasmosis may have a more pronounced reaction in immunocompromised individuals, thus leading to the severe vitritis and anterior chamber (AC) reaction. In a case series of patients with ocular toxoplasmosis who were immunocompromised due to AIDS, it was observed that the reaction was more severe and usually not even associated with scars [1]. One case of outer retinal layer toxoplasmosis in an HIV patient had similar findings of AC inflammation of 1+ with condensations in the vitreous of both eyes and diffuse, yellow-white retinitis [1]. Based on this case series by Holland *et al*. it was found that AIDS related ocular toxoplasmosis may have several clinical manifestations [1]. Prominent inflammatory reactions in the anterior chamber and vitreous are common due to the immunocompromised state [1].

Studies done in nonhuman primates, suggest that immunodeficiency alone does not cause reactivation of old lesions, as evidenced by some patients with AIDS who have toxoplasma scars but no signs of reactivation [1]. Thus, it is postulated that if organisms are reactivated by other undetermined factors, the immunodeficient status of the patient, may allow the organism to proliferate unchecked and cause severe disease [1]. Besides this, it is known, that aging itself leads to changes in both innate and adaptive immunity, thus increasing the prevalence and severity of many diseases in the elderly [6]. The absence of retinochoroidal scarring, on the other hand, should not rule out ocular toxoplasmosis as it has been shown that, in immunocompromised patients with AIDS, most ocular lesions are due to newly acquired diseases or organisms, and have thus, no pre-existing scars [1]. On top of that, cyst has also been seen in immunocompetent individuals retinal tissue, which appear normal [1].

Although serology is important in the diagnosis of toxoplasmosis, it has been shown that it may be unreliable in immunocompromised AIDS patients. It has been noted that IgM titers are usually negative or low and IgG titers are rarely more than 1:1024 [1]. The severe immunodeficiency due to the disease, may be the reason there is a lack of IgM response [1], explaining the equivocal IgM results seen in this case. A beneficial test, besides serology, is the Optical Coherence Tomography which is a key investigation in the diagnosis of outer retinal toxoplasmosis. Lesions can be scanned to see if they involve the outer retinal layers, hence aiding in the diagnosis, as seen in our case.

The treatment of disseminated toxoplasmosis in immunocompromised patients, is shown to have an 80% response rate and it is therefore, important to treat

them as soon as possible [1]. Patients with AIDS-related ocular lesions responded well to a standard treatment of pyrimethamine with an antimicrobial such as sulfadiazine. However, the most effective treatment in these immunocompromised patients was shown to be a combination of pyrimethamine with clindamycin [1]. Newer antimicrobials such as atovaquone and azithromycin, which was used in this case, have been shown to reduce the number of tissue cysts in animals [6]. These, though, have not been proven to prevent recurrences with short term therapy [6]. Immunocompromised individuals show high recurrences of about 30% when therapy is discontinued [1]. Thus, indefinite continuation of the treatment maybe necessary to control the disease. Corticosteroid therapy has also been used in the treatment of toxoplasmosis in reducing inflammation. That said, the use of corticosteroids without the concurrent use of antimicrobials can lead to severe tissue destruction [6].

The diagnosis of ocular lymphoma in the RE was entertained at early presentation, however, the cells in the vitreous and AC were fine and not malignant looking. There was no noticeable mass either. The FFA findings of retinitis with bilateral hot disc were also not in favour of this diagnosis. More importantly, the results of the vitreous biopsy was negative for malignant cells. Cytomegalovirus retinitis was also considered but ruled out as there was no retinal haemorrhages or granular appearance of the lesion. Vitreous tap and serum were also negative for CMV.

CONCLUSION

To conclude, toxoplasmosis is a disease which can cause retinochoroiditis in both immunocompetent and immunocompromised patients, by either congenital infection, reactivation or acute infection. The clinical picture may differ in terms of the reaction produced, especially in the immunocompromised. Clinical correlation is, thus, paramount in the diagnosis and treatment of this disease and it is important to note the various different presentations.

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