Case Report

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Surgical management of complications in a case of progressive outer retinal necrosis

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Abstract:

We report a case of bilateral progressive outer retinal necrosis (PORN) in a patient with acquired immune deficiency syndrome with CD4 count 50 cells/μL. He was treated with standard intravenous and intravitreal antivirals but ultimately developed complications such as retinal detachment and epiretinal membrane. His vision was preserved with early pars plana vitrectomy. This case demonstrates that prompt clinical diagnosis of PORN with its successful medical and surgical management can help prevent progression of this frightening disease.

Keywords:

Cracked mud, ganciclovir, progressive outer retinal necrosis, viral retinitis

Introduction

Progressive outer retinal necrosis (PORN) is a herpetic viral retinopathy with multifocal lesions in deep retinal layers, lesions in the peripheral retina with or without macular involvement, rapid progression, absence of vascular inflammation, and minimal intraocular inflammation.[1,2] This condition typically affects immunocompromised patients.[1] PORN may be caused by herpes simplex virus-1 (HSV) and *Cytomegalovirus* (CMV) although varicella zoster virus is most commonly seen.[1,3]

Case Report

A 43-year-old male who was a known case of acquired immune deficiency syndrome for the last 1 year came with chief complaints of diminution of vision (DOV) in his right eye (OD) for 45 days. He was on highly active antiretroviral therapy (HAART). His best-corrected visual acuity (BCVA) in OD was counting fingers 3 m and 6/12 in the left eye (OS). His anterior segment was unremarkable except for cortical cataract in both eyes (OU). Fundus examination in OD showed clear media and pale disc and multiple patches of whitish creamy retinitis lesions with illdefined margins involving the macula and peripheral outer retina [Figure 1a] and that in OS showed single similar lesion superotemporal to the macula [Figure 2a]. Optical coherence tomography OD showed lost foveal contour with loss of differentiation of retinal layers and fairly normal foveal contour in OS [Figure 1c]. Fluorescein angiography showed multiple areas of hyperfluorescence with leakage and staining corresponding to the lesions in OU [Figure 1d and e]. Based on the clinical picture and history, a diagnosis of PORN was made. He was referred to an infectious disease specialist and was started on intravenous (IV) acyclovir (12.5 mg/kg) three times a day and twice weekly intravitreal (I/V) ganciclovir (2 mg/0.1 ml) was advised in OU for 2 weeks. Serum immunoglobulin G was positive for HSV-1 and CMV. After 2 weeks, he was started on oral valacyclovir and IV acyclovir was stopped. Fundus showed perivascular

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clearing of retinal opacification in OD with typical cracked-mud appearance [Figure 1b], and the patient felt symptomatically better. After 1 month, he presented with decline in vision and peripheral rhegmatogenous retinal detachment (RRD) in OD [Figure 3a and c]. Pars plana vitrectomy (PPV), silicone oil injection along with I/V ganciclovir, was performed. After 1 month of surgery, BCVA improved to 6/60 with healed retinal lesions [Figure 3b and d]. At this time, he also complained of DOV in the OS. On examination, BCVA was 6/36 along with cataract and epiretinal membrane (ERM) [Figure 2a and c]. Cataract surgery and PPV with ERM removal was performed, and BCVA improved to 6/12 [Figure 2b and d]. He was then continued on HAART only and followed up for 6 months. He had stable BCVA in the OU without any recurrence till the last follow-up.

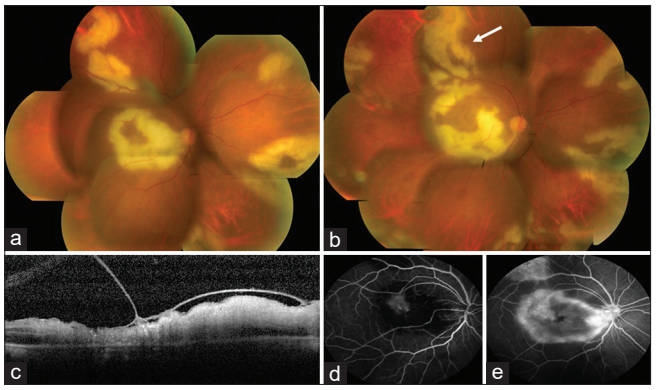

Figure 1: Fundus picture (a and b) showing multiple patches of whitish creamy retinitis lesions involving the macula and periphery at presentation and after 2 weeks of treatment, respectively (arrow shows cracked-mud appearance). Optical coherence tomography (c) showing lost foveal contour with loss of retinal layer differentiation. Fluorescein angiography (d and e) showing leakage corresponding to the site of retinitis lesions

Discussion

PORN is a devastating ocular complication, typically occurring in severely immunosuppressed patients. Timely recognition and its prompt treatment is crucial, as BCVA deteriorates rapidly. Most patients who present have an initial loss of vision, and in a descriptive series published by Engstrom *et al*. in 1994, two-thirds of eyes progressed to no light perception within 4 weeks of onset.[4]

The preferred regimen for I/V treatment is to inject intravitreous ganciclovir sodium (2 mg/0.05 ml) three times weekly for 2 weeks, followed by injections once or twice a week until the retinitis is stabilized. High doses of IV antiviral therapy are required to protect the other eye and the central nervous system at induction doses for 3 weeks, followed by maintenance antiviral therapy orally until complete healing.[5] Our patient was treated twice a week with I/V ganciclovir injections and IV high doses of acyclovir for 2 weeks and then continued on oral valacyclovir for 1 month and maintained on HAART later on.

CD4 count of <100 cells/μL in HIV-positive patients is known to be associated with microvasculopathy, which tends to be retinal or conjunctival. In HIV-positive patients, HAART has also been suggested to modify prognosis by reducing viral load and increasing CD4 count. A study reported by Woo *et al*.[6] suggested that CD4 counts improved and retinal lesions regressed after the commencement of HAART. It has also been postulated that if patients were receiving HAART at the onset prior to the necrosis, the retina was less likely to detach, as it regresses progression. Our patient had CD4 count of 50 cells/μL at presentation and was maintained on HAART from the beginning of the treatment. The visual outcome in HIV-positive patients is poor: 49% of patients had a final BCVA of "no light perception" within

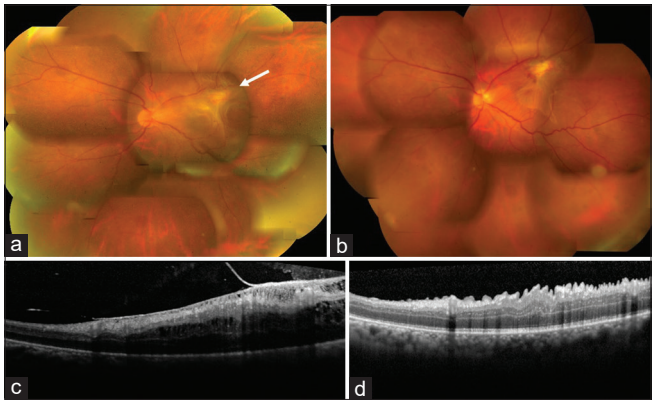

Figure 2: Fundus picture (a and b) showing epiretinal membrane (arrow) and status post cataract surgery with epiretinal membrane removal, respectively. Optical coherence tomography (c and d) pre- and postepiretinal membrane removal, respectively

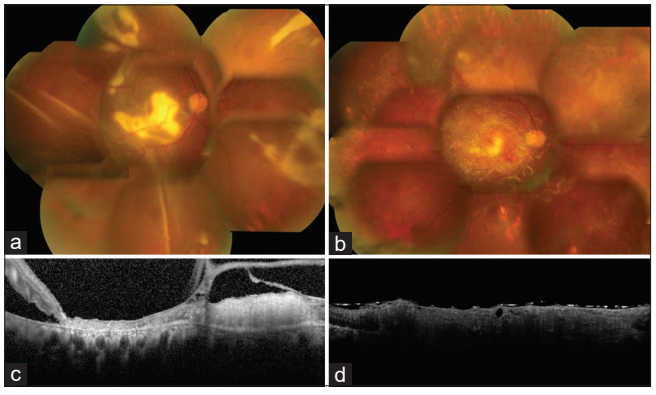

Figure 3: Fundus picture (a and b) showing retinal detachment and status post pars plana vitrectomy with silicone oil injection, respectively. Optical coherence tomography (c and d) pre- and postpars plana vitrectomy, respectively

4 weeks after the initial diagnosis. Indeed, about 70% of eyes develop RRD within a median time of 30 days to 3 months after presentation.[4,7] In the patient's right eye, the extent of the necrosis halted with treatment.

Without laser treatment, our patient developed RRD after 1 month, although prophylactic demarcating laser treatment is controversial in this setting.[5] One of the aims of the treatment in our patient was to prevent further progression and complication in OS and to preserve the vision in OD after RRD.

Conclusion

PORN is a devastating ocular disease, typically occurring in severely immunosuppressed patients (particularly HIV patients) and characterized by multifocal peripheral retinal lesions which rapidly coalesce without anterior inflammation and vasculitis. IV and I/V antivirals resulted in the regression of retinitis lesion, however development of RRD and ERM is common in these patients despite aggressive treatment. This case highlights the importance of early clinical diagnosis of PORN, and timely surgical management for the complications developing in the natural course of the disease coupled with antivirals can help improve visual acuity and prevent recurrences.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given his consent for his images and other clinical information to be reported in the journal. The patient understands that his name and initial will not be published, and due efforts will be made to conceal his identity, but anonymity cannot be guaranteed.

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Conflicts of interest

The authors declare that there are no conflicts of interests of this paper.

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