Case report

Atypical posterior pole retinitis

Parthopratim Dutta Majumdera, Saurabh Mistrya, Saurabh Luthrab, Shrutanjoy M Dasb, Varsha Sekarc, Ekta Rishia, Jyotirmay Biswasa,∗

a Sankara Nethralaya, Chennai, India
b Drishi Eye Institute, Dehradun, India
c Military Hospital, Dehradun, India

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ABSTRACT

Purpose: Necrotizing retinopathy is an uncommon, devastating, potentially blinding condition, which can be seen in both immunocompetent and immunocompromised patients. The purpose of this case is to report a new subset of necrotizing viral retinopathy.

Observation: A 34-year old lady presented to our outpatient department with sudden diminution of vision in both eyes following a brief history of viral fever with vesicular rashes. Fundus examination showed areas of necrotic retinopathy in posterior pole involving macula with scattered superficial retinal hemorrhages, cotton-wool spots in both the eyes. She was treated with intravenous acyclovir and oral valacyclovir.

Conclusion and importance: Our case may represent a new subset of necrotizing viral retinopathy, which may intermediate the clinical pictures of acute retinal necrosis and progressive outer retinal necrosis.

1. Introduction

Necrotizing retinopathies are a group of rare disorders in which there is posterior segment involvement associated with viral aetiology. Viral retinitis usually follows acute viral systemic illness and may present as multifocal retinitis. The spectrum of necrotizing viral retinitis commonly includes acute retinal necrosis (ARN), progressive outer retinal necrosis (PORN) and cytomegalovirus (CMV) retinitis. PORN and CMV retinitis are usually seen almost exclusively in severely immunocompromised patients. Posterior pole involvement in ARN is uncommon, but involvement of macula does not exclude the diagnosis of ARN. Various authors have described multifocal, progressive, necrotizing retinitis with vitritis involving posterior pole in a small proportion of patients. Fundus findings of these patients simulated central involvement of ARN or PORN with vitritis. We report a case of presumed viral retinitis in an immunocompetent female who presented with clinical picture like ARN involving posterior pole.

2. Case report

A 34-year-old female presented to our outpatient department with sudden diminution of vision in both eyes for the past one week. Her ocular symptoms were preceded by a history of fever one week back, following which she developed vesicular rashes over forehead (Fig. 1). Associated with the onset of rashes, she also complained of headache, vertigo, neck stiffness and joint pains. She also gave us history of contracting chickenpox in childhood with a relapse four-year back. She was seen by a dermatologist locally and was started on oral antiviral treatment (Oral Acyclovir, 800mg five times a day). Her visual acuity in both eyes was counting fingers close to face. Intraocular pressure, measured with applanation tonometer was 10 mm in both the eyes. Slit-lamp examination revealed quiet anterior chamber and cells in anterior vitreous of both the eyes. Fundus examination showed areas of necrotizing retinitis in posterior pole involving macula with scattered superficial retinal hemorrhages, cotton-wool spots in both the eyes. She was treated with intravenous acyclovir and oral valacyclovir.

Conclusions: Our case may represent a new subset of necrotizing viral retinopathy, which may intermediate the clinical pictures of acute retinal necrosis and progressive outer retinal necrosis.
fluid analysis, normal CT Scan and MRI Brain, normal serum protein electrophoresis, negative antiphospholipid antibody, negative antinuclear antibody, negative anti double stranded DNA (dsDNA) antibody, negative cytoplasmic and perinuclear Anti-Neutrophil Cytoplasmic Antibodies (ANCA), negative Treponema pallidum haemagglutination (TPHA) test and a normal peripheral blood smear. She was tested negative for human immunodeficiency virus (HIV) antibody. Her tuberculin skin test was negative and high resolution computerized tomography of the chest (HRCT-Chest) did not reveal any abnormality. Aqueous aspirate from her right eye was sent for polymerase chain reaction (PCR) for the detection of herpes simplex, varicella zoster, cytomegalovirus and chikungunya genomes, which came out to be negative. She was started on intravenous acyclovir 500mg 8 hourly. She was also started on oral prednisolone 50mg daily after obtaining necessary clearance from physician. Her visual acuity did not improve even after one week of initiation of treatment. She underwent vitreous biopsy and vitreous aspirate from her right eye tested negative for PCR for herpes simplex, varicella zoster, cytomegalovirus and chikungunya genomes. She continued to receive intravenous acyclovir. Her visual acuity deteriorated further in right eye due to increase in macular oedema, which was confirmed by SS-OCT (Fig. 3). The patient was given intravitreal injection of bevacizumab (1.25mg) in right eye. She was started on oral valacyclovir 1 g 8 hourly, the oral corticosteroid was continued in tapering doses and she was discharged from the hospital. She was examined again after one week. Her best-corrected visual acuity in right eye was counting fingers at 50 cm distance and 6/60 in
left eye. Slit-lamp examination of both eyes showed quiet anterior chamber and anterior vitreous. Fundus examination of right eye showed patches of retinitis at various stages of resolution with resolving macular edema. She was advised to continue the same treatment. She was again examined after three weeks. This time she was feeling better symptomatically and her visual acuity in right eye was counting fingers at 1 m distance and 6/24 in left eye. Fundus examination showed further resolution of lesions (Fig. 4). She was continued on oral valacyclovir 1 g 8 hourly and oral steroids were gradually tapered over next three months. Subsequently, she was followed up closely, and at last follow-up at 10 months, her best-corrected visual acuity was right eye counting fingers at 1 m distance and 6/18 in left eye. Slit-lamp examination of both eyes showed quiet anterior chamber and anterior vitreous. Fundus examination revealed completely resolved retinal lesions with macular thinning, more in the right eye as compared to left (Fig. 5). Left eye showed mild optic disc pallor. Visual evoked potential (VEP) was normal in both eyes.

3. Discussion

ARN is characterized by well-demarcated, multifocal patches of yellowish-white infiltrates in the periphery of retina at the level of deep retina and retinal pigment epithelium. Posterior pole involvement in ARN is relatively uncommon, though reported in literature. One of the characteristic feature of ARN is the presence of significant vitritis and vascular involvement. On the other hand, PORN is characterized by multifocal, discrete, white, outer retinal lesions at the posterior pole that progress rapidly to confluence and absence of inflammation in vitreous. PORN is almost exclusively seen in immunocompromised patient, though Carrillo-Pacheco et al. reported a case of PORN in an immunocompetent patient. In a case series of ARN, Margolis et al. described a pattern of retinitis that affected mainly the posterior pole in 12 eyes. These patients showed acute, posterior, multifocal necrotizing retinitis caused by herpetic infection and shared features of both ARN and PORN. Navarro et al. reported a case of presumed bilateral acute retinal necrosis with delayed onset caused by herpes simplex virus type 2, in a 51-year-old patient with history of old retinal detachment in his fellow eye. They suggested a diagnosis of acute retinal necrosis involving the posterior pole and sparing periphery. The causative agent was demonstrated using polymerase chain reaction analysis of the aqueous humor. Intensive medical treatment with intravenous acyclovir, intravitreal foscarnet, and corticosteroids was administered, slowing down but not stopping progression of the disease.

Our patient had minimal vitritis and retinal vessels were minimally involved. The patient was tested negative for HIV antigens and had no history of immunosuppression. Our patient also presented with necrotizing retinitis involving posterior pole which was preceded by a systemic viral illness with vesicular rashes over forehead. We have thoroughly investigated and ruled out any possible association with systemic rheumatic disorders. However, PCR analysis of aqueous and vitreous samples in our patient failed show amplification of any viral genome. Inability to yield any specific result by PCR in presence of clinically confirmed necrotizing viral retinopathy is not uncommon and has been reported in literature. Prior empirical treatment with oral acyclovir may be accounted for the negative PCR both from aqueous and vitreous aspirate in our case. Antiviral agents are known to stop the viral replication below the threshold of PCR sensitivity. CME in viral retinitis is usually known to occur relatively late as a part of breakdown of blood-retinal barrier. In our case, presence of cystoid macular edema with subretinal fluid can be attributed to the severity and location of the inflammation.

We presume that our patient represents a part of the spectrum of viral retinopathies, which may intermediate between ARN and PORN. Further studies are required to understand the clinical features and outcomes of such a clinical entity.

Patient consent

Consent to publish this case report has been obtained from the patient in writing.

Disclosure

Ethics approval and consent to participate: Written informed consent was obtained from the patients for the publication of this report and any accompanying images. Ethics approval was granted to us by our own institute’s IRB.

Conflicts of interest

The authors declare that they have no competing interests.

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Appendix A. Supplementary data

Supplementary data to this article can be found online at https://doi.org/10.1016/j.ajo.2019.100494.

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