Bilateral acute retinal necrosis associated with bilateral uveal effusion in an immunocompetent patient: A challenging association

S Bala Murugan, Girish Bharat Velis, Manavi D Sindal

Bilateral uveal effusion syndrome associated with bilateral acute retinal necrosis is a diagnostic and therapeutic challenge. A 52 year old man presented with bilateral angle closure with choroidal detachment. With restricted fundus view, parenteral steroid was started. During close follow up bilateral discrete areas of peripheral retinitis were noted. Parenteral steroids were promptly stopped and parenteral antivirals with oral steroids were continued. It showed healing response with nil recurrences till last follow up. Aggressive treatment of bilateral uveal effusion with parenteral steroids can cause progression of bilateral acute retinal necrosis leading to phthisis bulbi. However early diagnosis, prompt intervention and close follow up are the key elements to therapeutic success even during diagnostic surprises and avoid costly mistakes.

Key words: Acute retinal necrosis, angle closure, choroidal detachment, uveal effusion

Coexistence of uveal effusion with acute retinal necrosis (ARN) is a challenge both for diagnosis and therapy. In a hazy media with restricted fundus view, peripheral retinitis can be overlooked. Treatment of uveal effusion with parenteral steroids can cause progression of retinitis. We report a unique presentation of bilateral ARN (BARN) with bilateral uveal effusion in a systemically immunocompetent patient.

Case Report

A 52-year-old man presented to us with complaints of painful defective vision of sudden onset associated with redness in both eyes for 3 days. He revealed treatment elsewhere with oral acetazolomide, topical timolol maleate, and intravenous methylprednisolone (IVMP) with no improvement. His best-corrected visual acuity (BCVA) was 1/60 right eye (OD) and 6/36 left eye (OS). Anterior segment examination in both eyes showed lid edema, circumcorneal congestion, corneal epithelial edema, Descemet's folds, multiple, fresh, pigmented keratic precipitates on the corneal endothelium, shallow anterior chamber (AC) (Van Herick Grade 1), fixed mid-dilated pupil, and glaucomflecken [Fig. 1]. The intraocular pressure was 13 mmHg OD and 18 mmHg OS. The angle structures were not discernible on gonioscopy. The fundus view was restricted in both the eyes with faint disc edema in Grade 2 vitreous haze. Ultrasound B scan revealed 360° bilateral choroidal effusion [Fig. 2]. A provisional diagnosis of bilateral uveal effusion syndrome with secondary angle closure was made. Thorough uveitis work up such as mantoux test, chest roentgenogram, *Treponema Pallidum* hemagglutination test, veneral disease research laboratory test, Hepatitis B surface antigen, HIV titers, serum antibody titers for herpes simplex, zoster, and cytomegalovirus were negative. However, he had neutrophilia, lymphocytopenia, and raised erythrocyte sedimentation rate. Peripheral smear showed iron deficiency anemia with no abnormal leukocytes. Systemic evaluation for other causes of immunosuppression beyond HIV included thyroid, renal profile, liver function test, hemoglobin electrophoresis, and bone marrow aspiration biopsy and cytology which were not contributory. We stopped acetazolamide and continued IVMP, topical prednisolone, and timolol maleate.

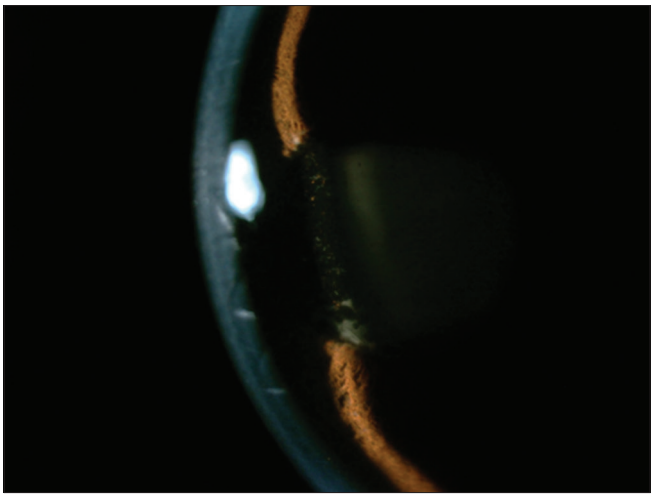

Figure 1: Slit lamp biomicroscopic image showing Descemet's membrane folds, pigments on endothelium, shallow anterior chamber peripherally, small pupil, and glaucomflecken

On day 3, BCVA remained 1/60 OD and 6/36 OS. With status quo anterior segment, the view of posterior segment cleared up in OS with Grade 1 vitreous haze. It had normal disc with multiple peripheral yellowish white retinitis patches [Fig. 3]. Ultrasound B scan showed 360° choroidal detachment. Optical

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coherence tomography revealed fusion of inner retinal layers OD [Fig. 4a] and thinning in OS [Fig. 4b].

Promptly, IVMP was stopped, and parenteral acyclovir in the dosage of 500 mg three times a day was initiated. Oral steroid 60 mg/day was started after 48 h of antiviral therapy. The parenteral antivirals were reduced to twice a day due to intolerable side effects after discussion with our internist as unidentified immunosuppression was initially anticipated. However, detailed work up for it along with CD4 and 8 counts confirmed his immunocompetent status.

After 7 days of parenteral acyclovir, BCVA improved to 6/60 OD and 6/6 OS with complete resolution of retinal necrosis and choroidal detachment in the OS [Fig. 5a and b]. It also showed regressing retinitis lesions and choroidal detachment in the OD [Fig. 6a and b]. We shifted to oral valacyclovir 1 g/day along with tapering doses of oral steroids for 12 weeks. His BCVA improved to 6/24 OD and 6/6 OS, with corresponding cataract of Grade 2 and 1 posterior subcapsular cataracts (PSCC), respectively. The retinitis and choroidal detachment had fully resolved. He was normal till 3 years of follow-up with us. However, he consulted another ophthalmologist and underwent OD cataract extraction with intraocular lens implantation under periocular steroid cover devoid of oral antivirals. Postoperatively,

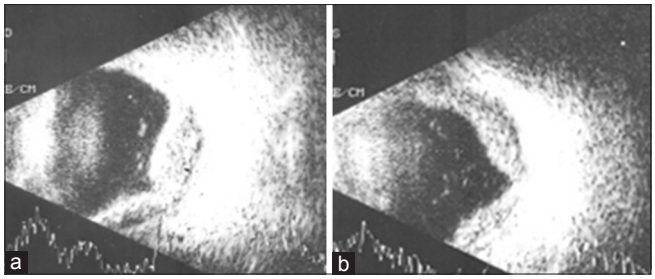

Figure 2: (a) B scan ultrasonography of the right eye showing choroidal thickening with detachment. (b) B scan ultrasonography of the left eye showing choroidal thickening with detachment

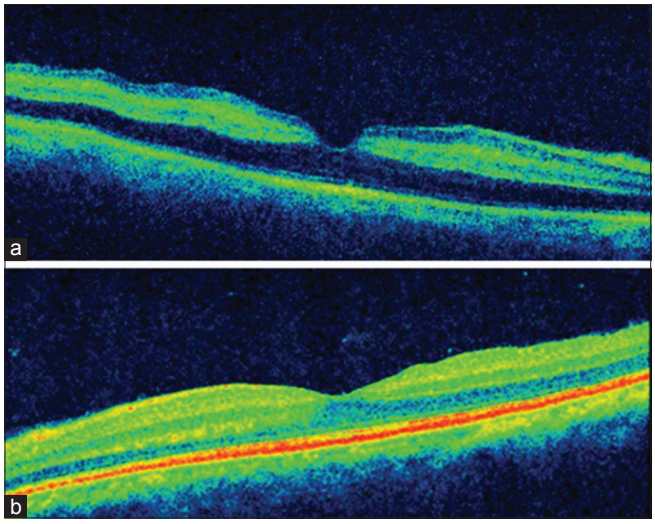

Figure 4: (a) Optical coherence tomography image showing fusion of inner retinal layers right eye. (b) Optical coherence tomography image showing thinned out inner retinal layers left eye

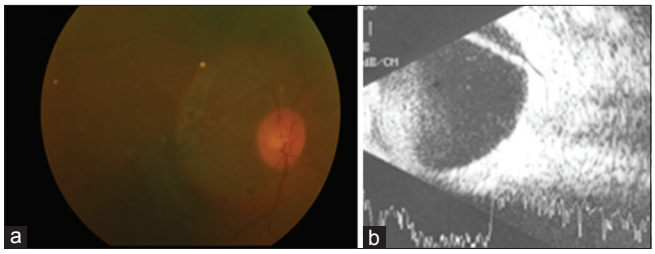

Figure 6: (a) Fundus image of the right eye showing regressing retinitis lesions. (b) B scan ultrasonography of the right eye showing regressing choroidal detachment

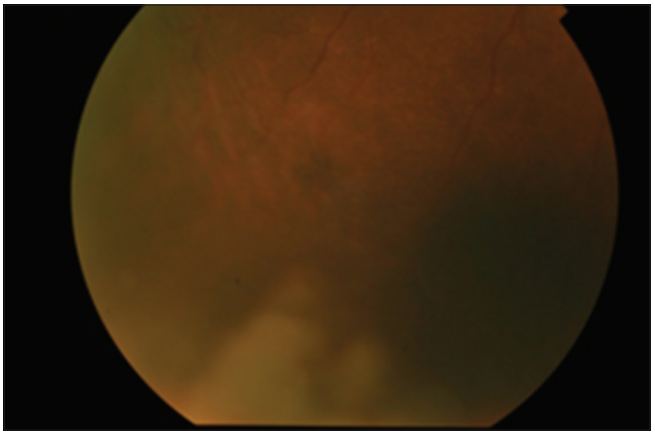

Figure 3: Fundus image of the left eye showing multiple peripheral yellowish white retinitis patches

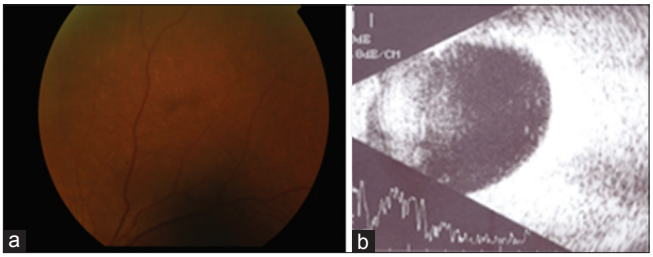

Figure 5: (a) Fundus image of the left eye showing complete resolution of retinal necrosis. (b) B scan ultrasonography of the left eye showing resolved choroidal detachment

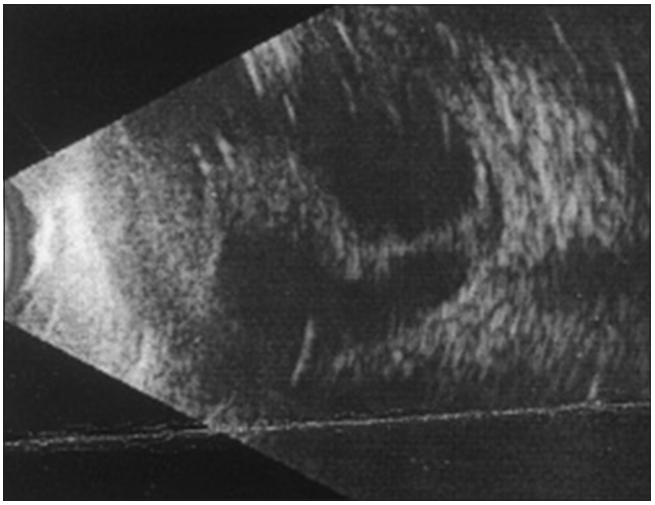

Figure 7: B scan ultrasonography of the right eye showing phthisis bulbi

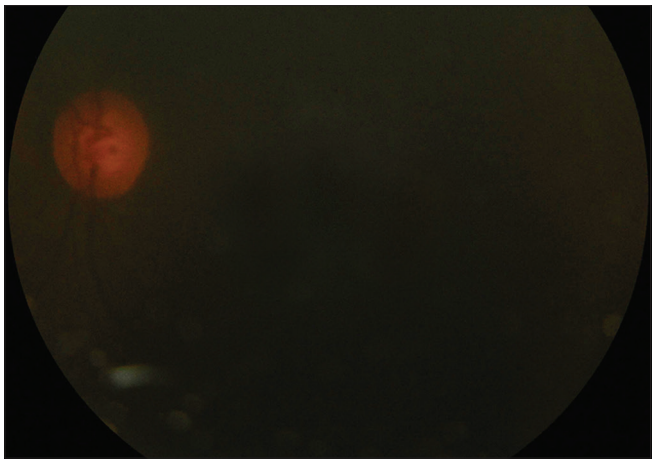

Figure 8: Fundus image of the left eye at the last follow-up showing normal retina in a hazy view

intravitreal steroids were given at 3 periodic intervals. When reviewed again in our institute, he had no perception of light OD and BCVA of 6/12 OS. IOP was 4 mmHg OD and 12 mmHg OS. OD was pthisical with closed funnel retinal detachment (RD) on ultrasound B scan [Fig. 7]. OS had normal fundus with a hazy media due to nuclear sclerosis and PSCC of Grade 2 [Fig. 8].

Discussion

ARN syndrome, characterized by confluent peripheral necrotizing retinitis, retinal arteritis, and vitritis, has been described in literature as unilateral condition common in immunocompetent individuals.[1] However, BARN has been reported in patients with reduced host defence.[2] In contrast, BARN in an immunocompetent patient due to dexamethasone usage has been documented by Bristow *et al*.[3]

We report a case of BARN in a systemically immunocompetent individual with bilateral uveal effusion. In our case, acetazolamide usage led to bilateral simultaneous AAC and uveal effusion. The subsequent administration of methylprednisolone caused a wave of immunosuppression and reactivation of latent virus.[4] The temporal relationship of the events with the drug usage versus the resolution of signs and symptoms with their cessation explains this plausible hypothesis. Our case differs from a similar published case report by Kaushik *et al*.[5] in having longer follow-up for 3 years.

The role of systemic corticosteroids in the management of drug-induced angle closure is controversial. It can also cause choroidal effusion[6] and was ineffective in one of the published cases.[7] However, Mancino *et al*.[8] and Rhee *et al*.[9] postulated that the use of IVMP in their cases resulted in quicker resolution. The restricted view of retinal peripheries in bilateral AAC prompted us to continue the parenteral steroid initially. In BARN, parenteral steroids without antivirals are contraindicated and can end up in phthisis bulbi. Hence, we discontinued parenteral steroids once retinitis was clinically evident. Oral steroids were promptly initiated after 48 h of antiviral therapy.

Conclusion

Clinician needs to be aware of this unique challenging combination of BARN with bilateral uveal effusion even in immunocompetent individuals. The treatment of uveal effusion with systemic steroids alone or with periocular steroids initially may aggravate BARN if careful evaluation of its early signs is overlooked with restricted retinal view in angle closure. Even in BARN, steroids can be given only under antiviral cover. In treating sequelae like cataract, ARN can reactivate with intravitreal steroids progressing to RD and phthisis, if perioperative antivirals are omitted. Early diagnosis, prompt intervention, and meticulous follow-up are the key elements to therapeutic success even during diagnostic surprises and avoid costly mistakes.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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