Bilateral Acute Retinal Necrosis Secondary to VZV in Immunocompetent Patient: A Case Report

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Abstract
We report the case of a French woman with bilateral acute retinal necrosis (BARN) whose disease was treated with intravenous acyclovir, topical prednisolone, topical atropine, intravitreal FOSCARNET, oral valacyclovir and corticosteroid bolus. The corticosteroid was added to her treatment regimen eight days after his initial presentation. The 63-year-old patient presented with a two-week history of bilateral blurred vision). Her best corrected visual acuity (BCVA) was 6/10 in both eyes (OU). Her examination revealed grade 3+ cells, pharmacologically dilated pupils and grade 2+ vitreous cells in the OU. The back of the patient's eye showed vitreous thickening, ghost vessels and retinitis spots in OU. In view of the clinical and additional test results, an assessment of BARN was performed. The patient was initially treated with acyclovir and systemic steroids were administered eight days later. Eventually, there was a significant clinical improvement. At the follow-up visit two months later, our patient developed a retinal detachment in both eyes, which was treated with silicone.
KEYWORDS: Acute retinal necrosis, Bilateral, Immunocompetent, Retinal Detachment,
Silicon.

Introduction

Acute retinal necrosis (ARN) and progressive outer retinal necrosis (PORN) are both classified as necrotizing herpetic retinopathies, which, despite their rarity, represent ophthalmologic emergencies requiring prompt recognition and treatment [1,2]. Herpes simplex virus type 2 (HSV-2) is the most frequently identified causative agent, followed by cytomegalovirus

(CMV) and Epstein-Barr virus (EBV) [3]. Infections associated with varicella zoster virus (VZV) occur sporadically and typically present with clinical features characteristic of ARN [1].
These conditions primarily affect immunocompetent individuals and often present initially as unilateral anterior uveitis accompanied by ocular pain [3]. Necrotizing herpetic retinopathies can occur across all age groups and in both sexes, with an estimated annual incidence ranging from 0.5 to 0.63 new cases per million individuals in the general population [4].

Case Report

A 63-year-old woman with type 2 diabetes was referred to hospital by anophthalmologist for panuveitis that had been present for 15 days and had worsened despite local treatment with prednisolone ophthalmic solution and a subconjunctival injection of Kenacort.
A comprehensive ophthalmologic examination revealed a best corrected visual acuity (BCVA) of 6/10 in both eyes. A slit lamp examination revealed some granulomatous keratic precipitates grade 1+ cells, pharmacologically dilated pupils, and grade 2+ vitreous cells (according to

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the SUN classification) in both eyes Examination of the patient's fundus revealed vitreous opacification in the right eye with peripheral retinal necrosis in a "gauntlet" and a retinal necrosis focus in the superior temporal region of the left eye FIGURE 1

Macular OCT showed mild serous retinal detachment in the right eye and normal findings in the left eye. An OCT scan of the area of necrosis was performed, confirming retinal necrosis in both eyes. FIGURE 2

Fluorescein and indocyanine green angiography confirmed the presence of areas of retinal necrosis in both eyes. FIGURE 3

She was given valaciclovir at a dosage of 3 grams per day. In addition, an anterior chamber puncture was performed to obtain a sample for antibodies to herpes simplex virus (HSV) 1 and 2, varicella zoster virus (VZV), and cytomegalovirus (CMV). In particular, the test results showed a positive result for VZV antibodies, while HSV1, HSV2 and CMV antibodies were all found to be negative.

Cell count, blood chemistry and HIV were negative.

The patient was admitted for intravenous therapy. She received 1,800 mg acyclovir (calculated at 10 mg/kg) administered every eight hours. After 8 days, she was switched to oral valaciclovir and instructed to continue using prednisolone ophthalmic solution and atropine 1% ophthalmic solution three times daily OU. At this point, the patient received an intravitreal injection of foscarnet , with a Corticosteroid bolus for 3 days.

The course was characterised by the regression of the signs of inflammation, in particular the regression of the areas of retinal necrosis in both eyes. FIGURE 4

The patient's BCVA was 3/10 in the right eye and 9/10 in the left eye due to the cataract

At the first follow-up visit two months later, the patient underwent cataract surgery in the right eye. Three weeks after surgery, the patient presented to the emergency room with a sudden decrease in visual acuity in the right eye. The fundus examination revealed a retinal detachment in the right eye. The patient underwent retinal detachment surgery with silicone inlay. Four weeks later, a superior retinal detachment was discovered in the left eye during a follow-up examination. The patient also underwent retinal detachment surgery with silicone insert in the left eye.

Discussion

The acute retinal necrosis (ARN) syndrome was first described by Urayama et al. in 1971 [5]. In 1982, Culbertson et al. [6] identified herpesvirus particles in an enucleated eye affected by ARN using electron microscopy, further supporting a viral etiology.

Subsequent studies employing various diagnostic techniques have consistently confirmed the viral origin of ARN. Among the causative agents, varicella-zoster virus (VZV) is most frequently implicated, followed by herpes simplex virus (HSV) [7]. Typically, VZV and HSV-1 are associated with ARN in individuals over the age of 25, while HSV-2 tends to be the primary cause in younger patients [7,8].

The diagnosis of ARN is based on clinical criteria established by the American Uveitis Society in 1994. These include the presence of characteristic peripheral retinal necrosis, rapid progression, circumferential retinal involvement, occlusive vasculopathy, as well as vitritis and anterior chamber inflammation [8].

In cases with dense vitritis or delayed hospital referral, ARN may be misdiagnosed as idiopathic uveitis, potentially leading to inappropriate treatment with systemic corticosteroids. Additionally, patients presenting with retinal detachment (RD) may undergo vitrectomy without recognition of the underlying viral etiology. Atypical or less pronounced clinical presentations pose a diagnostic challenge, and delayed identification of the disease can result in

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poor visual outcomes and increased risk of involvement of the fellow eye.

Acute retinal necrosis (ARN) typically occurs in immunocompetent individuals following infections with varicella-zoster virus (VZV) or herpes simplex virus (HSV). However, cases of ARN secondary to cytomegalovirus (CMV) have also been documented in immunocompromised patients. Initially, ARN usually affects one eye, but up to 65% of patients may develop bilateral involvement [6]. When the disease progresses to affect the contralateral eye, it is termed bilateral ARN (BARN) [9,10,11,12].

Varicella-zoster virus (VZV) has been identified as the primary causative agent in both ARN and BARN [10,12]. The mechanisms underlying viral spread in BARN remain incompletely understood. Several potential routes, including retrograde transmission through the optic nerve and optic chiasma or via parasympathetic pathways, have been proposed as ways the virus may spread to the contralateral eye [13]. The reasons why some patients develop BARN remain unclear.

Polymerase chain reaction (PCR)-based analysis of intraocular fluid can facilitate the diagnosis of ARN, enabling the prompt initiation of antiviral therapy. Early treatment improves patient outcomes and can help prevent the involvement of the contralateral eye.

It is worth noting that this case represents a rare instance of BARN, with both eyes being affected simultaneously. Most cases reported in the literature describe a typical pattern in which one eye is affected first, followed by the contralateral eye months to years later [14,15].

The first-line treatment for varicella-zoster virus (VZV) retinitis involves the immediate initiation of systemic antiviral therapy—either oral or intravenous—alongside adjuvant intravitreal antiviral injections. Additional therapeutic interventions may include the use of topical and systemic corticosteroids, anticoagulants, prophylactic retinal laser photocoagulation, and pars planavitrectomy [16]. Despite comprehensive management, the visual prognosis remains guarded, particularly in patients with significant immunosuppression [17].

Prompt administration of intravenous antiviral agents is critical in halting the progressive destruction of retinal tissue. Retinal detachment is a well-recognized complication, observed in the majority of patients with necrotizing retinitis. Monotherapy with intravenous acyclovir alone has demonstrated limited efficacy. In contrast, combination antiviral regimens—such as intravenous foscarnet with either ganciclovir or acyclovir—have shown improved outcomes in preserving visual function [18].

In the presented case, the patient was managed with intravenous acyclovir followed by oral valacyclovir and intravitrealfoscarnet. Despite this aggressive approach, she developed retinal detachment two months later. Nevertheless, the early initiation of intravenous antivirals was effective in preserving vision in the left eye and preventing disease progression in the right eye.

Adjunctive intravitreal antiviral therapy is believed to reduce the risk of vision loss in acute retinal necrosis (ARN). However, as reported by Baltinas et al., approximately two-thirds of eyes affected by ARN progressed to retinal detachment, regardless of whether treatment included oral, intravenous, or intravitreal antiviral agents [19].

The use of systemic corticosteroids in the management of infection-related conditions such as bilateral acute retinal necrosis (BARN) is often considered controversial. This is due to the potential for corticosteroids to enhance viral replication and accelerate the progression

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of retinitis [15]. Moreover, systemic corticosteroid use has been implicated in the onset of BARN in some cases [11,12]. Despite this, appropriate timing of corticosteroid administration appears to be a critical determinant of therapeutic success. In the case described, antiviral therapy with acyclovir was initiated first, followed by systemic corticosteroids eight days later, leading to notable clinical improvement.

Acyclovir remains the most commonly reported antiviral agent for treating ARN and BARN, due to its ability to inhibit viral replication [14,15]. The therapeutic benefit of corticosteroids lies in their immunomodulatory effects: they suppress excessive immune responses, stabilize vascular structures, reduce vasodilation, and inhibit the activity of inflammatory mediators such as cytokines and prostaglandins [15,20]. As vitritis and vasculitis are hallmark features of ARN, corticosteroid therapy plays a direct role in controlling these manifestations [7]. Additionally, corticosteroids have been proposed to reduce the risk of secondary retinal detachment in ARN cases [15].

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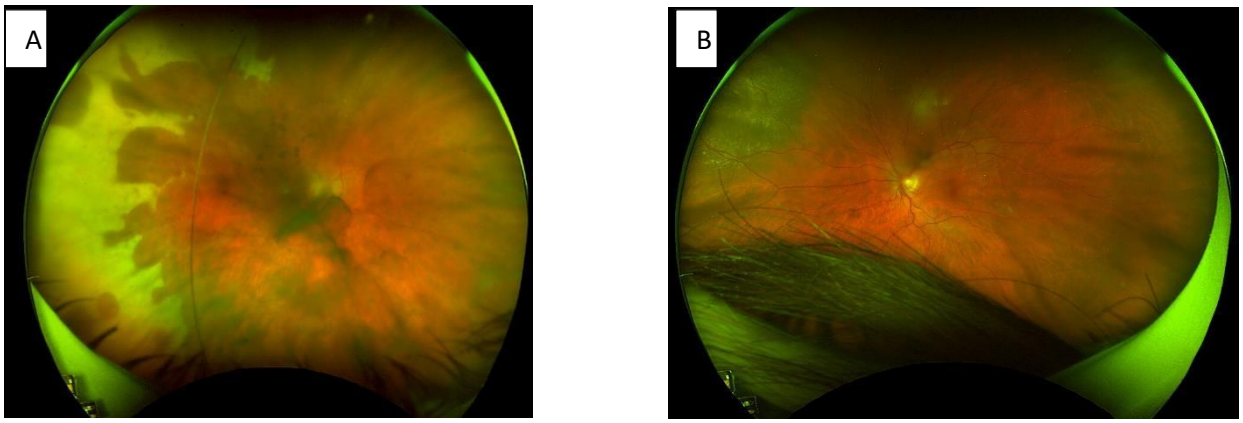

FIGURE1 color fundus photographs of the right (A) and left (B) eye reveal peripheral temporal retinal necrosis in a "gauntlet" pattern with haze in the right eye, focal retinitis superior temporal region of left eye.

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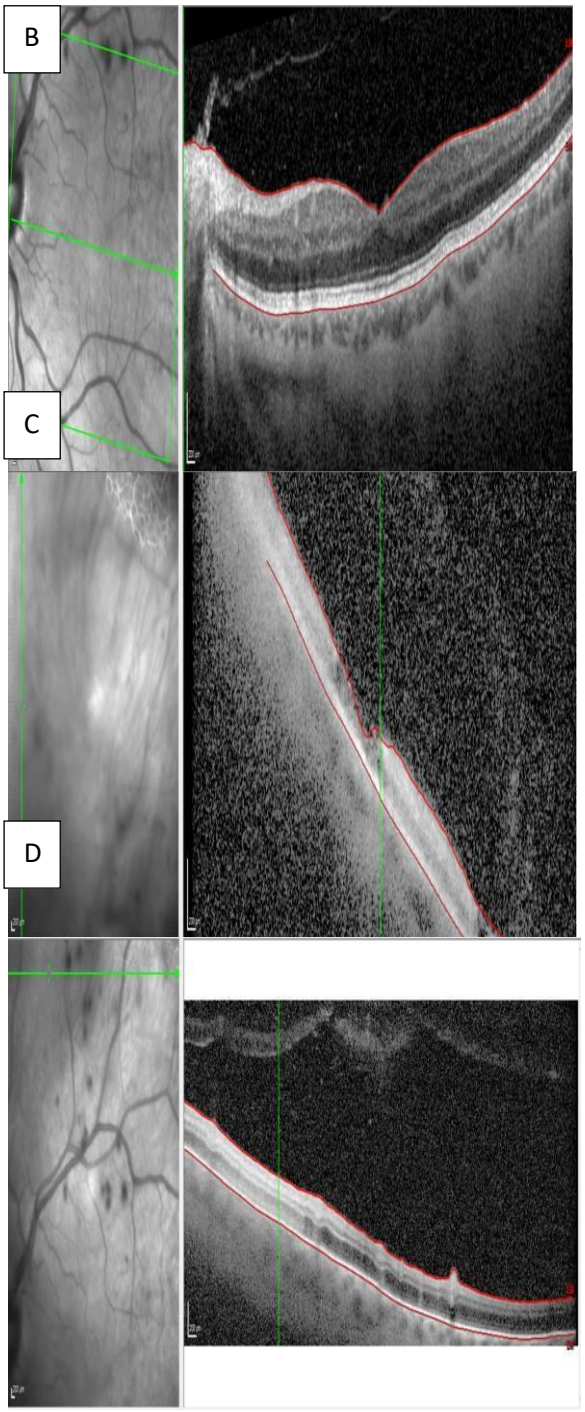

FIGURE 2 OCT show a mild serous retinal detachment in the right eye (A) A and normal OCT findings in the left eye (B). An OCT scan of the area of necrosis was performed, confirming retinal necrosis in both eyes (C.D)

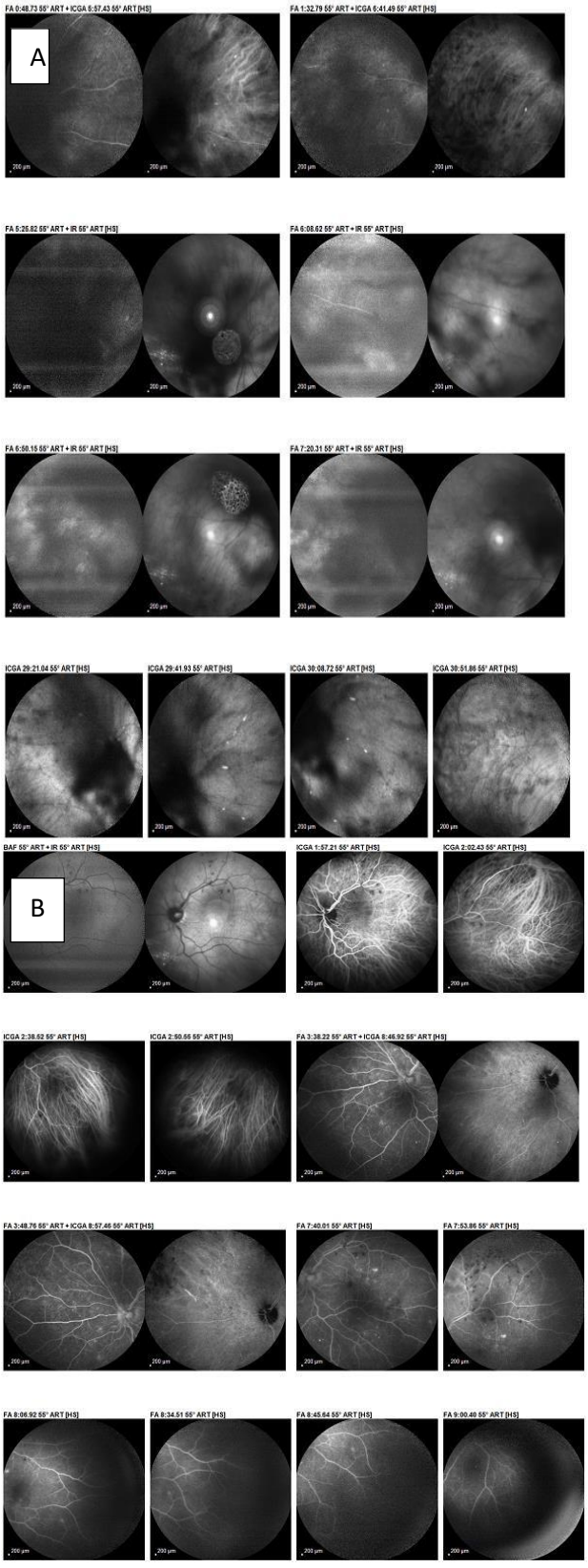

Figure 3 (A.B) Fluorescein and indocyanine green angiography confirm the presence of retinal necrosis in both eyes

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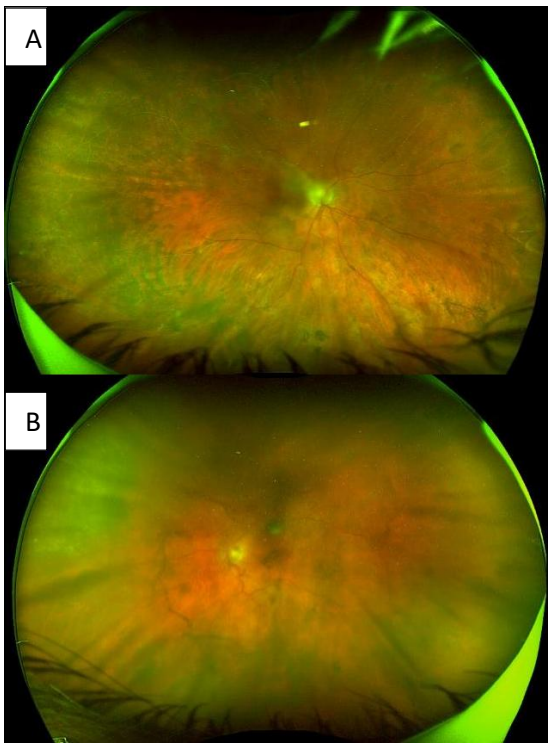

FIGURE 4: fundus photographs of the right (A) and left (B) eye reveal the regression of inflammatory signs, particularly the disappearance of vasculitis and the regression of retinal necrosis in both eyes

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