Journal of the Formosan Medical Association (2012) xx, 1–4

Available online at www.sciencedirect.com

SciVerse ScienceDirect

journal homepage: www.jfma-online.com

CASE REPORT

# Progressive outer retinal necrosis associated with occlusive vasculitis in acquired immunodeficiency syndrome

Chien-Chi Tseng [a], San-Ni Chen [a,b], Jiunn-Feng Hwang [a,b], Chun-Ju Lin [a,b,c,*], Huan-Sheng Chen [d]

[a] *Department of Ophthalmology, Changhua Christian Hospital, Taiwan*
[b] *School of Medicine, Chung Shan Medical University, Taichung, Taiwan*
[c] *Department of Optometry, Chung Hwa University of Medical Technology, Tainan, Taiwan*
[d] *An Hsin QingShui Clinic of Fresenius Medical Care Asia Pacific Co Ltd, Taichung, Taiwan*

Received 8 May 2011; received in revised form 16 April 2012; accepted 16 April 2012

**KEYWORDS**
acquired immunodeficiency syndrome;
occlusive vasculitis;
progressive outer retinal necrosis

A 45-year-old man, a case of acquired immunodeficiency syndrome, received a highly active antiretroviral therapy at the outpatient service for 4 years without regular follow-up. He experienced progressively blurred vision for 6 months and a cutaneous zoster on his back 3 months ago. He was diagnosed with progressive outer retinal necrosis by polymerase chain reaction-restriction fragment length polymorphism using an aqueous humor sample, which revealed an existence of varicella zoster virus. He was given a combination of systemic, intravitreal antiviral and a highly active antiretroviral therapy. Occlusive vasculitis, an unusual finding for progressive outer retinal necrosis, developed in both eyes 1 week after the secondary intravitreal injection. Unfortunately, his vision deteriorated to no light perception in both eyes within 2 weeks. Progressive outer retinal necrosis is characterized clinically as showing minimal or no inflammation in the aqueous and vitreous humors, absence of retinal vasculitis, and patches of yellowish spots located deep in the retina. Physicians should pay attention to this rare case of progressive outer retinal necrosis associated occlusive vasculitis with very poor prognosis in spite of aggressive treatment.


* Corresponding author. Department of Ophthalmology, 135 Nanxiao Street, Changhua County 500, Taiwan.
*E-mail address:* doctoraga@gmail.com (C.-J. Lin).

0929-6646/$ - see front matter Copyright © 2012, Elsevier Taiwan LLC & Formosan Medical Association. All rights reserved.
doi:10.1016/j.jfma.2012.04.005

## Introduction

Progressive outer retinal necrosis is a viral retinitis most commonly caused by varicella zoster virus (VZV) and occasionally by varicella simplex virus (HSV). Both viruses occur in immunocompromised patients, particularly those with acquired immunodeficiency syndrome (AIDS). The CD4 + T-cell counts of these patients are low (20—50 cells/$mm^3$) before the use of highly active antiretroviral therapy (HAART).[1–3] Patients usually have painless and rapid visual loss and may have a cutaneous zoster history. In comparison with acute retinal necrosis (ARN), progressive outer retinal necrosis is characterized by minimal inflammation in the aqueous and vitreous humors and rarely results in occlusive vasculitis. Total retinal detachment (RD) is usually a major consequence resulting from diffuse retinal necrosis. This study reports the fulminant course of progressive outer retinal necrosis in a patient with AIDS, occlusive vasculitis, central retinal vein occlusion, vitreous hemorrhage (VH), and RD despite aggressive and combined treatment that involved both intravitreal antiviral therapy and HAART.

## Case report

A 45-year-old man, a case of acquired immunodeficiency syndrome, presented with progressively blurred vision in both eyes. Previously, he had only mildly blurred vision in the left eye about 6 months ago and was also inflicted with cutaneous zoster 3 months ago. For 4 years he had also received HAART at our outpatient service center for infectious diseases without regular follow-up. On the day of admission, his vision was limited to hand movement in the right eye and light perception in the left eye. Funduscopy showed multifocal, patchy choroidal, and deep retinal opacification with dots and blots of retinal hemorrhage in both eyes (Fig. 1). Anterior chamber and vitreous cavity showed no significant signs of inflammation. Both serum *Toxoplasma gondii*—specific immunoglobulin (Ig) G and IgM and cytomegalovirus (CMV) IgM were negative. The aqueous humor examined by polymerase chain reaction restriction-fragment length polymorphism (PCR-RFLP) detected only VZV. The HSV and CMV DNA were not identified in aqueous humor. Under the impression of VZV-related chorioretinitis of both eyes, he was treated with systemic ganciclovir 250 mg twice-daily plus sulfamethoxazole 400 mg and two trimethoprim 80 mg tablets twice daily. Bilateral intravitreal injections of ganciclovir (2 mg/0.05 ml) were also performed.

He was administered efavirenz and intravenous ganciclovir, followed by continuation of HAART (lopinavir, ritonavir, and efavirenz). On the seventh hospital day, the deep white retinal lesions coalesced and expanded to the peripheral retina with vision worsened to hand movement in the right eye and no light perception in the left eye. Optical coherence tomography (OCT) showed extensive outer retinal necrosis (Fig. 2).

Repeated bilateral intravitreal injections of ganciclovir (2 mg/0.05 ml) were given to the patient after one week, but obliterative vasculitis, an unusual finding for progressive outer retinal necrosis, developed and resulted in central retinal vein occlusion in both eyes 1 week after the secondary intravitreal injection (Fig. 3). The patient also received a dose of acetylsalicylic acid and one tablet daily, and OCT showed almost total loss of identifiable retina layers with retinal necrosis in both eyes (Fig. 4).

To prevent bone marrow suppression in the patient, the intravenous injection of ganciclovir was switched to valganciclovir for 1 day and then to famciclovir. The HAART regimen was also changed to lopinavir and ritonavir two tablets twice daily plus abacavir and lamivudine one tablet once daily due to suspicion of drug resistance.

Panretinal photocoagulation was performed but extensive full-thickness necrosis with VH continued to develop (Fig. 5). His vision deteriorated to no light perception in both eyes within 2 weeks.

## Discussion

Necrotizing herpetic retinopathy is a continuous spectrum of diseases caused by herpes viruses, such as CMV, HSV, and VZV. The clinical presentation depends on the immune state of the patients, ranging from ARN to progressive outer retinal necrosis in patients with normal immunity or severely immunocompromised patients.[4]

Progressive outer retinal necrosis may involve any part of the retina at presentation (including macula) with little or no inflammation in anterior chamber or vitreous. Multifocal opacification of the deep retinal layers is usually seen at onset. Scleritis and occlusive vasculopathy are rarely

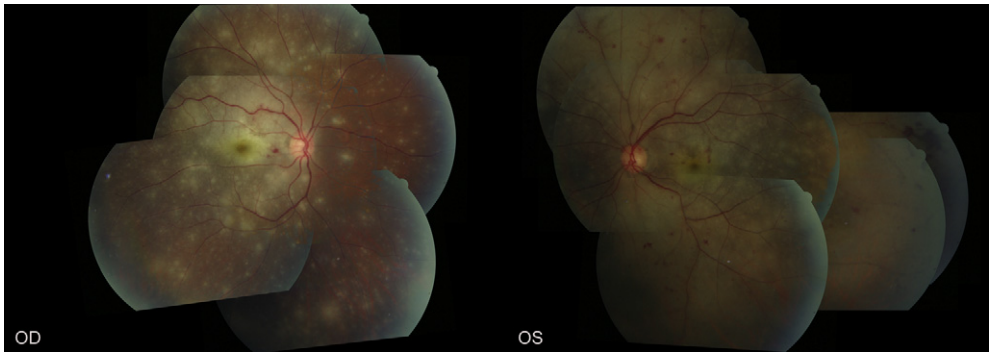


**Figure 1** Funduscopy showed multifocal, patchy choroidal, and deep retinal opacification with dots and blots of retinal hemorrhage in both eyes.

Please cite this article in press as: Tseng C-C, et al., Progressive outer retinal necrosis associated with occlusive vasculitis in acquired immunodeficiency syndrome, Journal of the Formosan Medical Association (2012), doi:10.1016/j.jfma.2012.04.005