

Progressive Outer Retinal Necrosis Leads to Visual Loss despite Early Recognition and Treatment

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Introduction

A 55-year-old Indian male presented with a one week history of rapid deterioration of vision in his right eye. He described it as loss of vision starting from the periphery which eventually encroached on his central vision as well. His visual acuity (VA) on presentation was no light perception in the right eye and 6/6 in the left eye. The patient was later revealed to be positive for human immunodeficiency virus, but had defaulted on treatment. He was found to have a cluster of differentiation (CD4) count of less than 50.

Fundus examination revealed widespread retinitis, perivascular haemorrhage and macular oedema in the right eye. The left eye also had 2 foci of retinitis (Figure 1) seen supero-temporal to the fovea. He underwent bilateral intravitreal injection of Foscarnet (0.1ml), which was repeated about 5 days later. Vitreous samples were positive for Varicella zoster virus (VZV). He was concurrently started on IV Acyclovir and commenced on highly active anti-retroviral therapy (HAART) in consult with the infectious disease team.

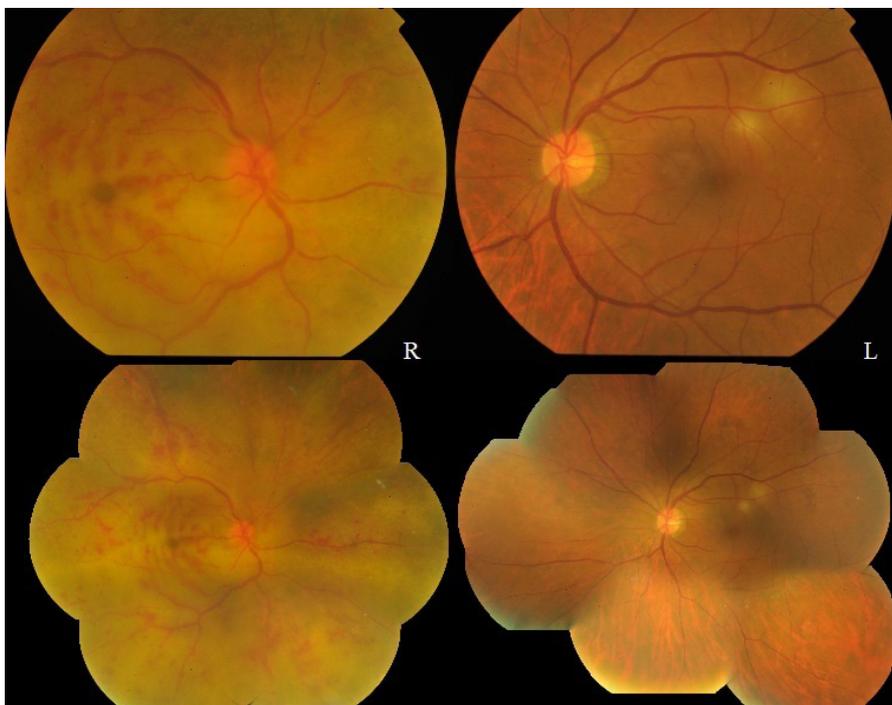


Figure 1: Posterior pole and 9-view photos of the patient taken at presentation

Three weeks later, the patient's right eye VA was still no light perception with extensive scarring at the macula. His left eye VA was stable at 6/12 and there were no new active lesions seen on fundal examination. Unfortunately, the patient subsequently defaulted further treatment and was only seen several months later. By this time, his left eye VA had also deteriorated to light perception due to new areas of scarring at the macula.

Discussion

Progressive outer retinal necrosis (PORN) is characterized by a very rapidly progressive retinitis that begins as multifocal areas of deep whitening throughout the macula or periphery that coalesce within days to confluent sheets. Vitritis, retinal vasculitis, and anterior segment inflammation is usually absent. In some instances, a cracked mud appearance representing perivascular clearing of the retinal necrosis has been described [1].

The natural course of the disease is rapid progression to blindness. Without therapy, the necrosis rapidly progresses circumferentially to involve the entire retina. However, even with anti-herpetic therapy, the prognosis for PORN is guarded with two-thirds of eyes having final visual acuity of no light perception. The antiviral medications used to treat PORN are the same as those used in the treatment of Acute Retinal Necrosis (ARN) [2,3]. Like ARN, the development of optic atrophy and retinal detachments occur in greater than 70% of cases. The outcome is a blind eye with an atrophic retina and a pale optic nerve.

This case once again demonstrates the devastating effects of PORN in an immune compromised patient. It also reaffirms the fact that despite early aggressive antiviral therapy and HAART, patients still have a very guarded visual prognosis.

References

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