CASE REPORT

Progressive outer retinal necrosis: a missed diagnosis and a blind, young woman

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SUMMARY

We present a 33-year-old woman with a history significant for HIV/AIDS (CD4 count of 17) and diabetes mellitus who was diagnosed as having progressive outer retinal necrosis (PORN) after presenting with peripheral vision loss. This case provided a diagnostic challenge and demonstrates the devastating effects of a misdiagnosis as it pertains to PORN.

BACKGROUND

Progressive outer retinal necrosis (PORN) also referred to as varicella-zoster virus retinitis (VZVR) is an aggressive form of necrotising inflammation typically affecting patients with advanced AIDS or those who may be severely immune compromised as a result of chemotherapy. Early detection and treatment is crucial to preservation of vision and undamaged retinal tissue.

CASE PRESENTATION

The patient is a 33-year-old woman with a medical history significant for HIV/AIDS (CD4 count of 17) non-compliant with highly active antiretroviral therapy (HAART) therapy and diabetes mellitus who originally presented to an outside hospital for the loss of peripheral vision of 1 week duration. She noted a headache accompanied with painful eye movement, which preceded the onset of her vision loss by a week. She was discharged directly from the emergency department (ED) and asked to follow-up with her ophthalmologist the next morning. The patient failed to comply and her symptoms progressed. She called her primary care physician regarding her worsening symptoms who advised her to return to the ED. In the ED a lumbar puncture and MRI of her head were performed, which was consistent with a demyelinating process including multiple sclerosis or infectious aetiology. A preliminary diagnosis of multiple sclerosis was made and the patient subsequently received intravenous methylprednisolone and was ultimately discharged. She presented to a tertiary care centre several days following discharge as her vision rapidly began to deteriorate to the point where she only had slight light perception. In addition, she was now febrile, diaphoretic and having severe generalised headaches with accompanying nausea and vomiting.

DIFFERENTIAL DIAGNOSIS

- ▶ Cytomegalovirus (CMV) retinitis
- ▶ Acute retinal necrosis

- ▶ PORN
- ▶ Toxoplasma retinitis

TREATMENT

At the tertiary care centre, for the first time since symptoms began, a funduscopic examination was performed and diffuse haemorrhages were noted. Subsequently, ophthalmology was consulted and a more detailed funduscopic examination revealed extensive intraretinal haemorrhages along the vascular arcades with widespread retinal atrophy (figures 1 and 2). The patient received 2 mg/0.1 ml intravitreal ganciclovir in addition to intravenous ganciclovir induction therapy, which was ultimately transitioned to valganciclovir oral therapy.

OUTCOME AND FOLLOW-UP

Unfortunately the patient was unable to regain any vision and subsequently underwent inpatient rehabilitation prior to discharge.

DISCUSSION

Ocular manifestations of HIV are common, with approximately 70–80% of HIV-infected patients being treated for an HIV-associated eye disorder during the course of their illness. A CD4 count less than 100/ml is associated with retinal or conjunctival microvasculopathy, CMV retinitis, VZVR and toxoplasma retinitis.¹ The syndrome of PORN or VZVR, is a unique type of herpetic retinitis inflammation typically affecting patients with advanced AIDS or those who may be severely immune compromised as a result of chemotherapy. The diagnosis of PORN is a clinical diagnosis with diagnostic criteria including the presence of multifocal lesions without granular borders in deep retinal layers, evidence that the infection started in the peripheral retina with or without foveal involvement, an extremely rapid progression and the presence of minimal intraocular inflammation.²

Prompt diagnosis and treatment of PORN are necessary to limit retinal damage and preserve vision. Ormerod *et al* investigated retrospectively 11 patients (20 eyes) with PORN. They noted rapid progression, 82% having bilateral involvement, relative resistance to intravenous antiviral therapy and 70% rate of retinal detachment. Only 4 of 20 affected eyes retained useful vision at a 2-month follow-up and 70% had no light perception at the conclusion of the study.³ Recently, Sittivarakul *et al* investigated seven patients (11 eyes) with a clinical diagnosis of PORN between

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Figures 1 Detailed fundusoscopic examination illustrating areas of oedema temporal to the fovea with intraretinal haemorrhages originating from venules along all four vascular arcades and oedematous retina, deep retinal atrophy with diffuses haemorrhages more prominent towards the posterior pole.

2003 and 2008. They found a known history of cutaneous zoster in 57% of cases with 57% of patients having bilateral disease, retinal detachment occurring in 54.5%, and visual acuity with no light perception in 45.5% of patients at the final recorded follow-up.⁴

Current treatment options for PORN include antiviral therapy, including intravenous and oral acyclovir, oral famciclovir or intravitreal foscarnet, valacyclovir or famciclovir. In severe cases of PORN, early vitrectomy with laser demarcation of areas of retinal necrosis in conjunction with intravenous antiviral agents ought to be considered.⁵ Gore *et al* performed a retrospective analysis of 39 patients (67 eyes) with intravitreal

ganciclovir started immediately in all salvageable eyes (n=50). They noted that intravitreal ganciclovir may offer a more targeted approach and may result in better visual outcomes when compared with systemic therapy alone.⁶

The incidence of PORN has markedly diminished since the advent of potent HAART therapy, yet a misdiagnosis can be devastating. PORN should be included in the differential when any immune compromised patient, that is, AIDS or as a result of chemotherapy, presents with visual deficits. Fundusoscopic examination is crucial in these patients as early diagnosis and prompt initiation of antiviral therapy is necessary in order to preserve retinal tissue and vision.

Figures 2 Detailed fundusoscopic examination illustrating areas of oedema temporal to the fovea with intraretinal haemorrhages originating from venules along all four vascular arcades and oedematous retina, deep retinal atrophy with diffuses haemorrhages more prominent towards the posterior pole.

Learning points

- ▶ A CD4 count less than 100/ml is associated with retinal or conjunctival microvasculopathy, cytomegalovirus retinitis, varicella-zoster virus retinitis and toxoplasma retinitis.
- ▶ A funduscopy examination is crucial in any immune-compromised patient presenting with visual deficits.
- ▶ Early diagnosis and prompt initiation of antiviral therapy is necessary in order to preserve retinal tissue and vision in progressive outer retinal necrosis.
- ▶ Intravitreal ganciclovir may offer a more targeted approach and may result in better visual outcomes when compared with systemic therapy alone.

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