Case Report: Cytomegalovirus Induced Unilateral Acute Retinal Necrosis with Contralateral Optic neuropathy in an Immunocompetent Adult

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Background: Acute retinal necrosis (ARN) is a spectrum of necrotizing herpetic retinopathies. The most common cause of ARN includes VZV, followed by HSV-1, HSV-2, and rarely CMV. We report an unusual healthy patient presented with CMV induced unilateral ARN and subsequently developed optic neuropathy without retinitis in the fellow eye.

Case: A 27-year-old male presented with a 1-week history of blurred vision OS. He was otherwise healthy. Visual acuity was 20/20 OD and Hm OS. The right eye was unremarkable. Left ocular examination revealed grade IV anterior chamber cells and positive RAPD. Fundus examination demonstrated grade I vitreous haze, occlusive arteritis, optic disc swelling and peripheral retinal infiltration. The patient was diagnosed with ARN OS. Aqueous PCR was performed and the result was positive for CMV. No signs of systemic CMV infection were detected. The anti-HIV test was negative. The T cell function and T cell proliferation assays were unremarkable. He received intravenous ganciclovir with concomitant an intravitreal injection of ganciclovir (2 mg/0.05 mL). After 2 days of therapy, prednisolone (1MKD) and prophylactic barrier laser photocoagulation were administered. The retinitis gradually improved.

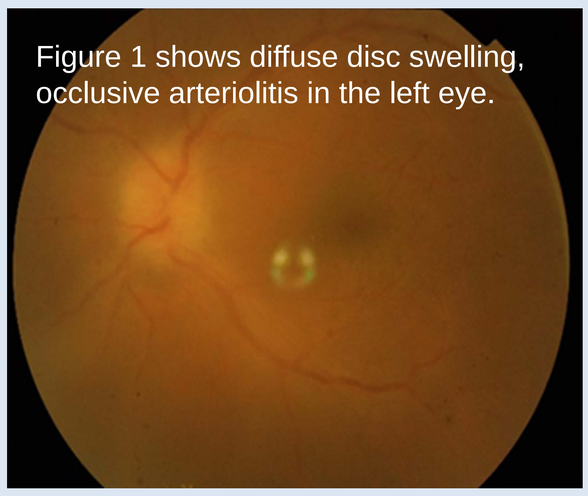

Figure 1 shows diffuse disc swelling, occlusive arteriolitis in the left eye.

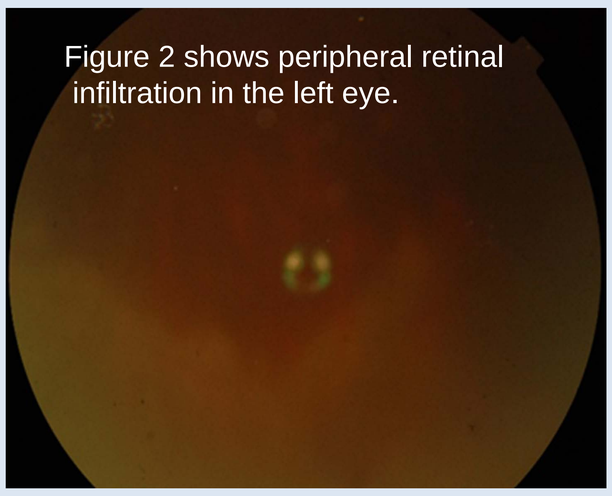

Figure 2 shows peripheral retinal infiltration in the left eye.

On the 12th day, the right eye developed a sudden onset of painless visual loss with a visual acuity of counting fingers. The right eye showed no inflammation, no retinitis and normal optic disc appearance. Both pupils were 8 millimetres which were not reactive to light. The neurological examination was unremarkable. Regarding an abrupt onset of visual loss and abnormal pupil, the occlusive vasculopathy of the right optic nerve was suspected. MRI of the brain and orbits provided swelling with enhancement of the left optic nerve but unremarkable right optic nerve. Computed tomographic angiography of the brain was unremarkable. Blood analysis included ANA,RF,ANCA, anti-beta-2-glycoprotein I antibodies, anticardiolipin antibodies, lupus anticoagulant, homocysteine, proteinC, protein S, antithrombin-III, FBS, VDRL, TPHA and lipid profiles. All of them were normal except lupus anticoagulant was positive. Echocardiogram was unremarkable. Aqueous PCR of the right eye found negative for CMV. The patient then received intravenous methylprednisolone 1 gram daily for 3 days, followed by a gradually tapered course of prednisolone over 2 months. Warfarin was also administered. The retinitis of the left eye improved, however the right vision decreased to no light perception despite anticoagulant therapy.

After 3 weeks of intravenous ganciclovir therapy, valganciclovir 450 mg twice daily was given for 1 year. At a 2-month follow- up, the right optic disc became pale and the left eye developed rhegmatogenous retinal detachment which was successfully treated with pars plana vitrectomy. Lupus anticoagulant became negative at a 5-month follow-up. Warfarin was then discontinued. At a 1- year follow-up, visual acuity was no light perception OD and 5/200 OS.

Comments: After treatment of ARN, the patient subsequently experienced visual loss from optic neuropathy in the fellow eye. No retinitis was detected. Regarding an abrupt onset of visual loss and abnormal pupil, occlusive vasculopathy of the optic nerve was suspected. The possible hypothesis of right optic neuropathy included immune related vasculitis and thromboembolic phenomenon such as secondary from CMV infection, cardiac emboli, antiphospholipid antibody syndrome and other hypercoagulable conditions. Blood examination showed elevation of lupus anticoagulant. It was first thought to be caused by antiphospholipid antibody syndrome. However, lupus anticoagulant became negative 5 months apart so it did not meet the diagnostic criteria of antiphospholipid antibody syndrome. Therefore, it was possible that CMV induced transient elevation of lupus anticoagulant can be a cause of vascular thrombosis of the optic nerve, which led to contralateral optic neuropathy. CMV infection can trigger arterial and venous thrombosis by various mechanisms such as facilitating leukocytes and platelets adhesion to the infected endothelial cells. The envelope of CMV also has intrinsic procoagulant properties, which may promote thrombin generation via factor X activation, surface tissue-factor activity, and procoagulant phospholipid expression. CMV can enhance circulatory levels of Von-Willebrand factor and factor VIII. In addition, CMV can transiently increase antiphospholipid antibodies including lupus anticoagulant. This patient developed an unusual event, possibly caused by vascular thrombosis after ocular CMV infection.

Conclusions: This case emphasizes the need to include CMV in the causative organism of ARN, even in immunocompetent patients. Ophthalmologist should remain vigilant that ocular CMV infection can induce vascular thrombotic phenomenon in patients