Simultaneous cytomegalovirus uveitis and HIV retinopathy

Uveíte por citomegalovírus e retinopatia por HIV simultâneas

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ABSTRACT

A 41 year-old man presented with unilateral vision loss for a week and constitutional symptoms for 3 months. Ophthalmic evaluation revealed cotton wool spots on the right eye and widespread retinal necrosis and hemorrhage on the left eye, suggestive of cytomegalovirus uveitis and HIV infection and retinopathy, and confirmed by serology. The patient was treated with ganciclovir and highly active antiretroviral therapy and preserved contralateral vision. Clinicians should be aware of uveitis and retinopathies to prevent irreversible vision loss and systemic conditions.

RESUMO

Paciente do sexo masculino, 41 anos, com queixa de perda de acuidade visual unilateral por 1 semana e sintomas constitucionais por 3 meses. Ao exame oftalmológico, apresentava exsudatos algodonosos, em olho direito, e áreas de necrose e hemorragias retinianas, em olho esquerdo, com suspeita de uveíte por citomegalovírus e retinopatia por HIV, confirmadas por sorologias. O paciente foi tratado com ganciclovir e terapia antirretroviral e preservou a visão contralateral. Os oftalmologistas devem estar atentos para casos de uveítes e retinopatias, para prevenirem perda visual irreversível e condições sistêmicas.

INTRODUCTION

Human immunodeficiency virus (HIV) is an endemic disease and a significant cause of vision loss, with affections like HIV and cytomegalovirus (CMV) retinopathy. Cytomegalovirus retinitis is the most common cause of retinal infection on AIDS patient and presents in a progressive and often blinding fashion.[1,2] The diagnosis is clinical and associated with severe immunosuppression, requiring investigation.[2]

This case report was approved by the Research Ethics Committee of Irmandade da Santa Casa de Misericórdia de São Paulo, CAAE 59515522.1.0000.5479.

CASE REPORT

A 38-year old man presented with a history of blurry left eye (OS) vision for a week. His uncorrected visual acuity was 10/20 in his right eye (OD) and hands motion in the OS. Examination of his OD revealed cotton wool exudates (Figure 1), and OS had fine keratic precipitates, anterior chamber reaction 2/4+, afferent pupillary defect, mydriasis and widespread necrotic and hemorrhagic retinitis (Figure 2). The patient presented unremarkable medical history except for headaches, diarrhea, and weight loss in the past 3 months.

The patient had a clinical diagnosis of HIV and CMV uveitis and was admitted for medical treatment. His exams revealed HIV virus, a CD4 of 68, decreased liquor glucose, and elevated protein; colonoscopy light colitis and retitis and cranial magnetic resonance imaging (MRI) showed multiple focal alterations of signal intensity on basal nucleus, thalamus, and encephalitis trunk, suggestive of CMV encephalitis.

He started being treated with endovenous (EV) ganciclovir 5mg BID, highly active antiretroviral therapy (HAART) with lamivudine, dolutegravir and tenofovir, and clinical prophylaxis to opportunistic diseases. After ending the 21-day ganciclovir course and being clinically stable, he was discharged and referred to a day hospital for further prophylaxis.

Left eye evolved to no light perception and consensual pupillary defect at the OD. The CMV-affected eye evolved with complete necrosis as expected within a fulminant pattern (Figure 3). The OD showed a normal fundus without lesions (Figure 4) and a 20/20 visual acuity.

DISCUSSION

HIV infection is a widespread condition that shows variable manifestation according to disease stage, such as toxoplasmosis, CMV, and pneumocystis. Viral load and CD4 counts are directly associated with prognosis. Cytomegalovirus is a ubiquitous DNA herpesvirus, potentially devasting in immunosuppressed individuals.[3,4]

Figure 1. Cotton wool exudates on retinography and OCT on the right eye.

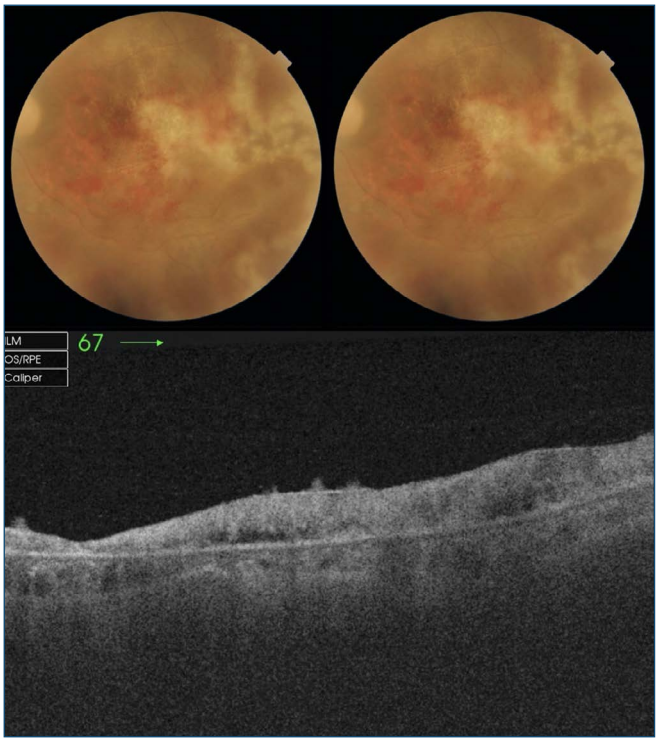

Figure 2. Left eye showing widespread necrosis and hemorrhagic retinitis, with retinal disorganization on OCT.

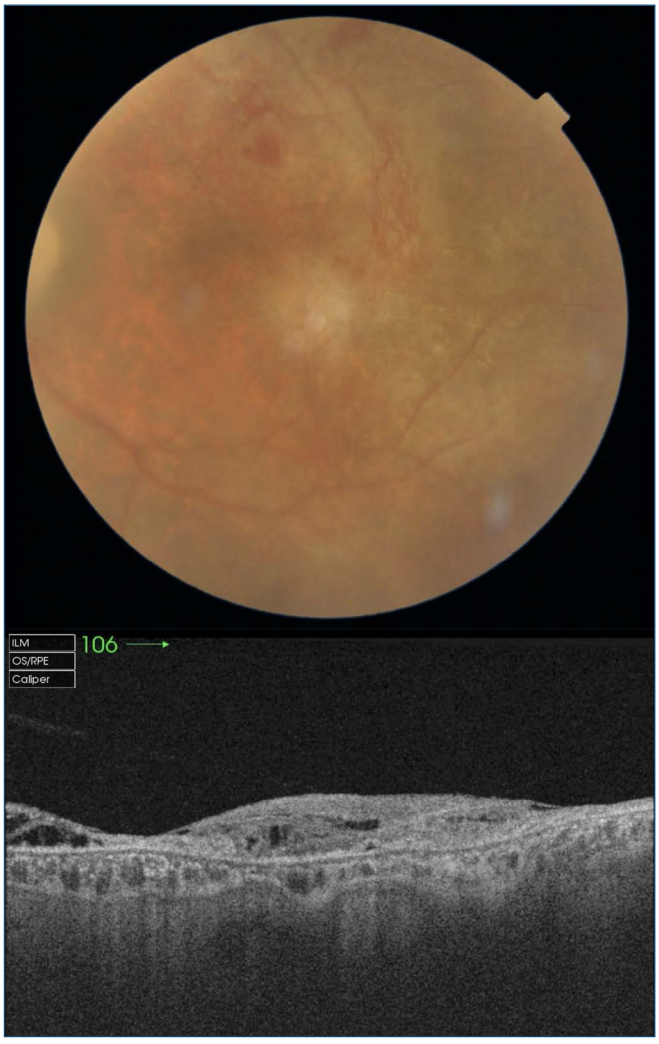

Figure 3. Complete necrosis and retinal disorganization after CMV retinitis.

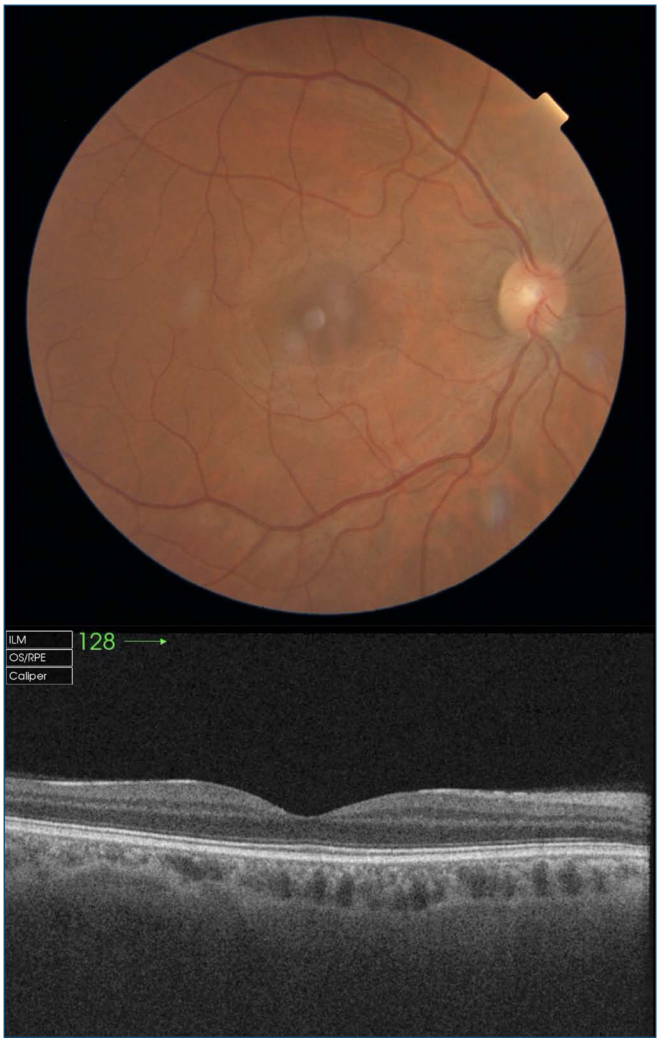

Figure 4. Normal fundus and OCT showing total remission of HIV retinopathy on the right eye.

Therefore, a suspected case should be tested for HIV due to the clinical association and severity.

Cytomegalovirus encephalitis is a central nervous system infection associated with immunodeficiency, characterized by signs such as headaches, seizures, and blurry vision due to its capacity to affect the whole neuroaxis, commonly involving the retina or causing cognitive dysfunction.[5] Fortunately for our patient, signs were limited to headaches and eye involvement, without seizure focus, and had a good evolution without further symptoms.

Cytomegalovirus retinitis is an opportunistic infection, quite common (>55%) in untreated HIV patients with a CD4<50. The affection has full thickness retinal necrosis and may lead to retinal detachment, atrophic scar tissue, vasculitis and choroiditis.[1,3] Diagnosis is clinical and may be further supported by vitreous polymerase chain reaction. Acute retinal necrosis is an important differential diagnosis, but ARN typically has more vitritis, more peripheral involvement, and less hemorrhage, and is treated with acyclovir or valaciclovir.[6]

Photopsias, scotomas, and blurry vision are the main symptoms of CMV, but 54% of the patients are asymptomatic. The initial presentation shows small yellow peripheral lesions that follow the vasculature centripetally, but the classic sign consists in white granular lesions and retinal hemorrhage. Vitritis is minimal; rhegmatogenous retinal detachment can happen in extensive disease and keratic precipitates suggests uveitis.[2,7] In our case, a typical fulminant pattern and uveitis prompted a clinical diagnosis, confirmed by serology. In resource constrained developing country or at inner city settings, a clinical diagnosis may be a key for early treatment and a less damaging final scenario.

The main patterns are: the fulminant, with classic ischemic hemorrhagic lesions ('scrambled eggs and ketchup') with a grim prognosis, leading to retinal tears

and detachment, the granular, peripheral 'brush fire' pattern, and the rare, frosted branch angiitis surrounding vessels. Diagnosis is clinical. The first line therapy is either valganciclovir or ganciclovir, the former being cheaper but requiring hospital admission; foscarnet or cidofovir are secondary options. Patients need a maintenance dose until sustained CD4>100 for a minimum 3 months. Relapse is usually a sign of worsening immune system, inadequate drug levels or resistance.[7-9] Due do its gravity, the preservation of the contralateral eye is the main treatment goal, such as in our report, where the patient maintained a contralateral 20/20 vision and an independent life.

HIV retinopathy is a microangiopathy characterized by small temporary wooly exudates. Its pathophysiology is not well understood and lies on immune complex deposition, virus mediated invasion of vascular endothelium or increased blood viscosity. Most patients evolve with visual acuity of 20/40 or better, but structural damage can happen, and visual field and contrast sensibility loss are relatively common. It usually does not require specific treatment and improves with HAART introduction, as seen with a total structural and functional remission in 4 months even before recovery of the CD4 counts.[10]

The case illustrates a patient with bilateral HIV and CMV uveitis and CMV encephalitis as the first manifestation of AIDS. The patient was unaware of its seropositive status and on imminent systemic infection and sight loss. Even with widespread testing and HAART easily available, undiagnosed HIV is resurging and it is a source of morbidity and new infections and shows the importance of regular health care. Furthermore, we cannot stress enough the importance of retinal mapping in order to detect initial peripheral lesions and prevent macular involvement in HIV positive patients and the subsequent reserved prognosis. Even though the CMV affected eye had disease progression, the contralateral one showed a complete recovery and averted being affected by the same affliction, as it would have without treatment, and the encephalitis had its course subdued before it could inflict widespread damage.

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