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SHORT COMMUNICATION

Case report

Herpes simplex virus type 2 acute retinal necrosis in an immunocompetent patient

E. MISEROCCHI[1], G. MODORATI[1], C. AZZOLINI[1], C.S. FOSTER[2], R. BRANCATO[1]

[1] Department of Ophthalmology and Visual Sciences, University Hospital San Raffaele, Milano - Italy
[2] Immunology and Uveitis Service, Department of Ophthalmology, Massachusetts Eye and Ear Infirmary, Harvard Medical School, Boston - USA

PURPOSE. *To report a case of acute retinal necrosis caused by herpes simplex virus 2 in an otherwise healthy patient.*

CASE REPORT. *A 45-year-old man presented with one month's history of decreased vision in the right eye. He had previously received a course of intravenous gancyclovir because of a clinical suspicion of cytomegalovirus retinitis. The patient's ocular history was remarkable for a similar episode in the left eye thirty years earlier, resulting in important visual impairment. System and laboratory investigations were unremarkable. Ocular examination showed severe anterior granulomatous uveitis, vitreous haze, areas of necrosis and retinal exudates. The anterior chamber tap disclosed the presence of HSV type 2, and oral steroids and acyclovir were instituted. Two weeks after the patient had been discharged, a retinal detachment occurred in the right eye, necessitating surgical repair. The presence of HSV type 2 was confirmed in the vitreous. Visual acuity recovered completely after surgery and the patient was placed on a maintenance dose of oral acyclovir.*

CONCLUSIONS. *HSV type 2 is a rare cause of acute retinal necrosis in healthy patients. Bilateral involvement can occur in the fellow eye, even with a long delay. Acute retinal necrosis is a severe ocular inflammatory syndrome associated with a very poor visual outcome. It is caused by VZV, HSV type 1 and, less commonly, by HSV type 2. The disease can affect healthy patients and cause bilateral involvement in the fellow eye, even with a long delay.. (Eur J Ophthalmol 2003; 13: 99-102)*

KEY WORDS. *Acute retinal necrosis, Cytomegalovirus, Herpes simplex virus, Herpes zoster virus, Retinitis, Retinal detachment, Vitrectomy*

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INTRODUCTION

Acute retinal necrosis syndrome is characterized by peripheral necrotizing retinitis and retinal vasculitis, frequently with anterior chamber inflammation, vitritis and papillitis. It is caused by herpes (VZV) virus, herpes simplex virus type 1 and, less frequently, herpes simplex virus type 2 and cytomegalovirus. It has a rapidly progressive course without prompt antiviral treatment (1).

We report a case of acute retinal necrosis caused by herpes simplex type 2 in an otherwise healthy patient.

Case report

A 45-year-old Caucasian man presented to the Immunology and Uveitis Service at the San Raffaele Hospital, Milan, in January 2001 with a one-month his-

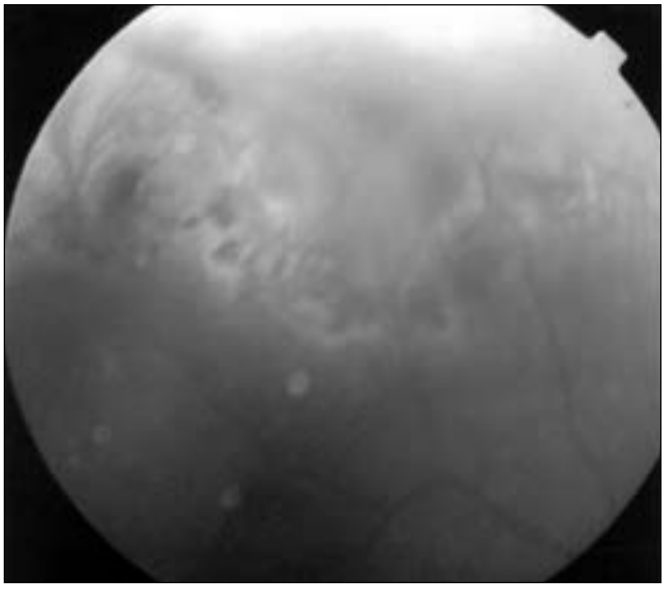

Fig. 1 - *Right eye: whitish areas of retinal necrosis and exudates with scattered pigmentary changes. There is severe vitreous haze.*

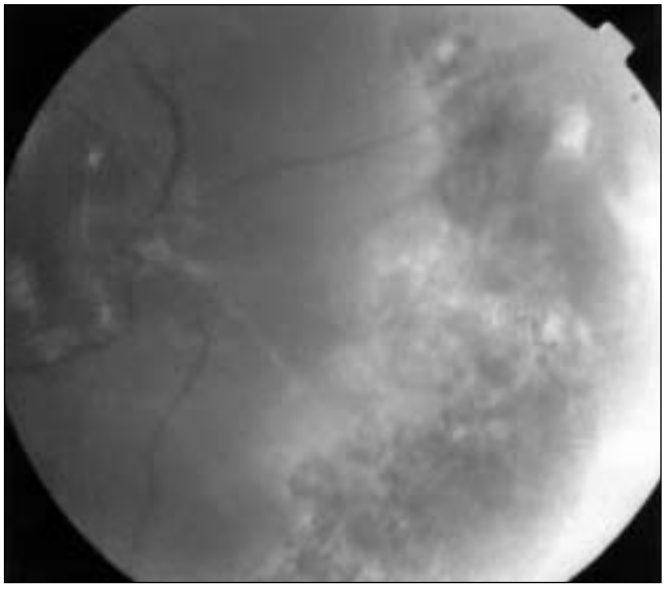

Fig. 2 - *Right eye: five months after surgery the vitreous is clear and the retina is attached. There are pigmentary changes on 360 degrees in the peripheral retina.*

tory of decreased vision in the right eye. He had been referred from a different hospital, where he had been admitted in December 2000 after sudden onset of pain and visual loss in his right eye; on the basis of a clinical suspicion of cytomegalovirus retinitis, the patient had received a three-week course of intravenous ganciclovir.

His ocular history was relevant for a similar episode of acute loss of vision and pain thirty years earlier in his left eye, clinically diagnosed as viral retinitis. The patient had then been treated with steroid pulse and intravenous antiviral agents, but no specific etiological diagnosis for the retinitis had been established. Unfortunately visual acuity in the left eye never recovered.

The patient's medical history and system review were unremarkable. At our first ocular examination, his visual acuity was 20/300 in both eyes. Anterior segment examination of the right eye showed moderate conjunctival hyperemia, mutton-fat keratic precipitates and 3+ cells in the anterior chamber. The left eye had a band keratopathy and a dense nuclear cataract 3+. Intraocular pressure was 14 mmHg in both eyes.

Fundus examination of the right eye disclosed severe vitritis and a swollen optic nerve. In the peripheral retina, which was difficult to visualize because of the vitreous haze, some whitish confluent areas of necrosis and exudates were detectable on 360 degrees, with scattered pigmentary changes (Fig. 1).

The limited view of the posterior pole of the left eye due to the dense cataract revealed a pale atrophic optic disc and a large fibrotic vitreo-retinal strand extending from the optic disc to the nasal retina. Chorioretinal scars were also present throughout the posterior pole. A clinical diagnosis of acute retinal necrosis in the right eye was made.

Considering the visual acuity of the affected eye, we decided to do an anterior chamber tap rather than a vitreous biopsy in an effort to confirm the clinical suspicion; polymerase chain reaction (PCR) analysis of the harvested fluid was positive for HSV type 2. In order to exclude any associated systemic condition or autoimmune disease, we requested all the routine laboratory tests for patients with uveitis. The results were all normal except for an elevated ESR (31), elevated CRP (10), elevated alpha 1-acid glycoprotein (1.19) and the presence of antinuclear antibodies (titer 1:160, granular pattern). Convalescent serum titers for HSV type 1, type 2 and CMV were consistent with

past infection. CD4/CD8 ratio was normal, as was the total CD4 T cell count.

Since the patient's neurologic examination was normal, no cerebrospinal fluid tap or diagnostic imaging were done.

The patient was discharged with systemic oral prednisone (50 mg a day) and oral acyclovir (800 mg 5 times a day), hourly prednisolone acetate 1% drops and atropine drops twice a day. One week after discharge, visual acuity in the right eye was 20/70; there were no cells in the anterior chamber but the vitreous still contained 3+ cells.

The patient returned one week later to the emergency room for sudden visual loss in his right eye: visual acuity was counting fingers and ophthalmoscopic examination disclosed a peripheral retinal detachment, 360 degrees, with sparing of the macula. A total vitrectomy, endolaser photocoagulation and a scleral buckle were performed; the vitreous was sent for PCR analysis which confirmed the presence of HSV type 2 DNA.

In July 2001, five months after surgery, visual acuity in the right eye was 20/40, the retina was attached and the vitreous was clear (Fig. 2). The patient was able to taper the oral steroid, and remained on a maintenance dose of acyclovir (800 mg/day).

DISCUSSION

Acute retinal necrosis is an ocular inflammatory syndrome that includes necrotizing retinitis predominantly of the peripheral retina, occlusive vasculitis and an inflammatory reaction of the vitreous and anterior chamber. Even with prompt diagnosis and treatment, the visual outcome may still be poor (1). The process can be bilateral, involving the fellow eye within weeks, but delay up to 20 years has been reported (2).

Acute retinal necrosis is most commonly caused by VZV or HSV type 1. However, sporadic cases associated with herpes simplex type 2 have been documented and reactivation of congenital HSV type 2 infection has been implicated as a possible cause of acute retinal necrosis (ARN) (3-5). It is still not clear how the virus gains access to the retina in an otherwise healthy patient; it has been suggested that this can occur by vascular dissemination or reactivation of a latent source of infection. Activation of a latent source of herpes virus confined within the retina, trigeminal ganglion, or central nervous system might explain why many ARN patients present no serologic, systemic or other focal evidence of herpetic infection (6).

The patient described is one of those rare cases of HSV type 2 ARN with no nervous system involvement such as encephalitis, history of congenital HSV type 2 infection or prematurity. Direct demonstration of HSV type 2 in the eye by PCR has been reported only in a few cases (3). Although a specific pathogen for the retinitis in the patient's left eye 33 years earlier could not be confirmed by laboratory techniques such as PCR analysis, the ophthalmoscopic findings were compatible with a viral process. Since HSV type-2 was found in the right eye, it is possible that the same disease occurred in the fellow eye, i.e. our patient had bilateral acute retinal necrosis with an unusual delay of the fellow eye.

We believe that HSV type 2 ARN in an immunocompetent patient without CNS involvement must be considered in the differential diagnosis of ARN. Moreover, given the possibility of a long interval before involvement of the fellow eye and the importance of early treatment, patients - and their ophthalmologists - should be alert for ocular symptoms after an episode of ARN, for the rest of their lives.

Reprint requests to:
Prof. Rosario Brancato
Department of Ophthalmology
and Visual Sciences
University Hospital San Raffaele
Via Olgettina, 60
20132 Milano, Italy
brancato.rosario@hsr.it

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