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A Case Report of Acute Viral Retinal Necrosis: ARN Syndrome

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Abstract **Case Report**

ARN syndrome or acute retinal necrosis is a clinical syndrome following a viral disease in an immunocompetent patient and which may jeopardize the visual prognosis if diagnosis and therapy delay, due to the Herpes group viruses. In this report, we present a case of acute viral retinal necrosis in a young immunocompetent patient whose antiviral and anti-inflammatory therapy was favorable.

Keywords: acute retinal necrosis (ARN), diagnosis, immunocompetent patient, varicella zoster virus (VZV).

INTRODUCTION

ARN syndrome or acute necrotizing retinitis was first described in 1971 by Urayama *et al.*, [1]. It is a clinical syndrome following a viral disease in an immunocompetent patient and which may endanger the visual prognosis if diagnosis and therapy delay. This is due to its severity and risk of bilateralization. Herpes group viruses, more often varicella zoster virus (VZV) and herpès simplex virus 1 and 2 (HSV-1 and 2) [2-5], rarely cytomegalovirus (CMV) [7], are involved in ARN syndrome. It combines a retinal necrosis; well limited, initially located in the middle periphery, with a tendency to extend circumferentially of full-thickness; then towards the posterior pole, an occlusive vasculitis predominant on the arterial network and marked inflammation of the anterior and posterior segment [6].

CASE REPORT

We report the case of a 22-year-old patient, with no specific pathological history, who visited for reduced visual acuity progressing 3 days before admission without other associated ocular or extraocular signs. The admission exam objectified at the right eye: visual acuity corrected to 12/20, eye pressure at 12mmHg, a clear cornea, a Tyndall effect in the anterior chamber at 2+, a clear lens and the fundus examination detected a hyalitis, the appearance of a Retinal necrosis associated with extensive temporal subretinal hemorrhage (Figure 1). The examination of the other eye was normal with visual acuity corrected to 20/20. The HSV (1 + 2), VZV, EBV, HIV (1 + 2), CMV serologies were negative. Retinal angiography showed areas of ischemia supero-temporally with extensive temporal subretinal hemorrhage sparing the macula (figure 2) and macular optical coherence tomography was normal (figure 3). Intravenous acyclovir 800 mg was set for 3 weeks with local (1 drop*8/ day) and oral (2 mg / kg / day) corticosteroid therapy associated with good clinical outcome marked by the disappearance of tyndall effect from the anterior chamber and hyalitis, decrease of subretinal hemorrhage extent and ischemia area steadiness. Visual acuity recovered to 10/10 after a 3 week-treatment. Maintenance antiviral treatment (valaciclovir 500mg * 3 / day) as well as topical corticosteroid therapy (one drop twice/ day) was maintained for 6 months with gradual regression. The patient also underwent retinal photo-coagulation in areas of ischemia. The patient is followed up to this day to watch for a recurrence.

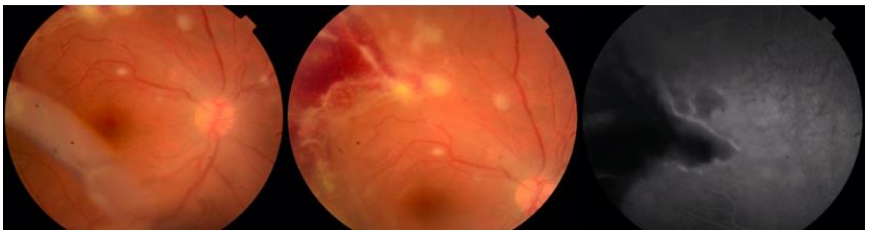

Figure 1: Initial appearance on retinal angiography of retinal necrosis, subretinal hemorrhage, and hyalitis with an intact macula

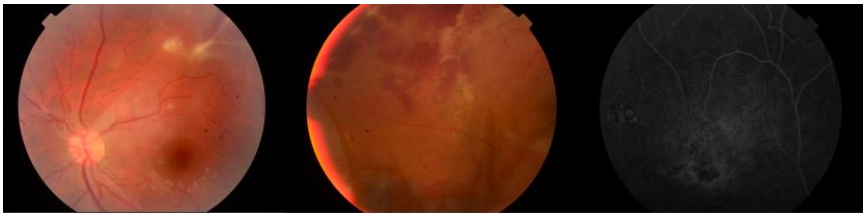

Figure 2: Appearance on retinal angiography of retinal necrosis with regression of subretinal hemorrhage with an intact macula 2 weeks after treatment with injectable aciclovir

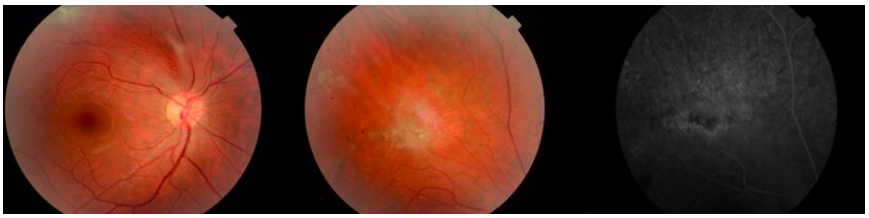

Figure 3: Appearance on retinal angiography of the scarred retinal focus after 6 months of treatment

DISCUSSION

The five diagnosis criteria for acute retinal necrosis syndrome were described by Holland *et al.*, in 1994 and which include: anterior uveitis, vitreous reactions, retinal necrosis in disseminated or confluent areas, retinal arteritis, and variable intensity papillary edema [6]. This pathology remains the prerogative of the immunocompetent patient [12], with a few cases reported in the immunodepressed [6-8].

Acute retinal necrosis occurs in immunocompetent patients, due to the high virulence of the HSV and VZV viruses, which accounts for the sudden and rapidly progressive nature of retinal damage despite an effective immune system. The consequences of this antiviral control are deleterious by the importance of the cytopathogenic effects and the brutality of the inflammatory phenomena which are involved [1-12].

The diagnosis of viral retinitis is above all clinical, and supported by a favorable response under antiviral treatment. However, the clinical picture is not always typical and it is sometimes difficult to make a positive diagnosis. This is the reason why it is essential to take eye sampling in order to search for the causal aetiology, without delaying the therapy [10]. The determination of the viral etiologic agent in intraocular fluids (aqueous humor, vitreous) may be indicated in certain atypical cases and involves PCR and the ratio of immune load (Golmann-Witmer coefficient) [11].

Antiviral therapy should be launched as soon as possible and the aetiological research should not delay the initial treatment. It consists of the systemic administration of an antiviral treatment generally combined with an anti- inflammatory treatment. The treatment aims to control viral replication but also to fight against inflammatory and vascular phenomena associated with it. At present, the existing antivirals are only virostatic and non-virucidal drug; so they cannot eradicate the virus from the body, which requires an attack treatment followed by a maintenance treatment which avoids the symptoms and subsequent relapses [10]. The standard regimen traditionally involves an induction phase in the form of intravenous acyclovir with a dosage of 10 mg / kg every 8 hours for 7 to 14 days, followed by a maintenance phase with acyclovir, valaciclovir or famciclovir per os. The duration of maintenance treatment is usually 3 months, but may be longer in the case of immunosuppression or multiple relapses [12].

Viral retinal necrosis is both a therapeutic and diagnostic emergency [13]. In fact, in the absence of an appropriate diagnosis and treatment, the visual prognosis is extremely poor with 65% of patients having a final visual acuity of less than 1/10 [14]. Besides, within 2 years, the contralateral eye is affected in 65% of patients without acyclovir treatment [15]. During mild ARN, which is defined by retinal necrosis limited to 25% of the surface of retina as is the case in our patient, treatment with intravenous acyclovir seems to be sufficient and the visual prognosis generally

remains faorable. If treatment were launched earlier, more than 90% of patients would recover visual acuity greater than 2/20 [16].

The immediate prognosis of these conditions depends on three factors: the early diagnosis, the aggressiveness of the medical treatment initiated, and also the causative virus [12]. Recurrences and retinal detachment remain the dredfull complications of the disease.

CONCLUSION

Viral retinal necrosis is both a therapeutic and diagnostic emergency. Indeed, in the absence of an appropriate diagnosis and treatment, the visual prognosis is extremely poor with a very high morbidity rate. Progressive follow-up should last for many years, with long-term preventive treatment in order to avoid bilateralization.

CONFLICTS OF INTEREST

The authors declare no conflicts of interest.

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