Bilateral Acute Retinal Necrosis in a 12-year-old Girl

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Acute retinal necrosis (ARN) is a severe ocular syndrome consisting of a moderate-to-severe anterior uveitis, vasculitis, and vaso-occlusive retinal necrosis. It can occur in healthy individuals at any age, but reports of this condition in children are rare.

A 12-year-old girl presented at our department with bilateral visual loss due to an anterior and posterior uveitis with retinal hemorrhages and necrosis, compatible with the diagnosis of ARN. We report upon the treatment, complications, and final visual outcome in this potentially blinding disease.

CASE REPORT

A 12-year-old girl presented with red, painful eyes and blurred vision present for 10 days. The past medical history revealed chickenpox at the age of 3. Over the last year she had been suffering from frequent respiratory infections. She was not recently exposed to chickenpox or shingles. Visual acuity was 20/100 on the right and counting fingers (CF) on the left. Slit-lamp examination showed severe bilateral anterior uveitis and vitritis. Funduscopy showed bilateral peripheral circumferential retinal hemorrhages with areas of retinal necrosis underneath, macular exudation, and optic disk edema (Figure 1). Goldmann visual field testing showed peripheral limits to 40° in both eyes.

As bilateral ARN was strongly suspected, treatment with intravenous Acyclovir 50 mg/kg/day and topical steroids were started immediately. Pediatric examination and laboratory testing, including immunologic tests, were normal. Serum antibodies were negative for all herpesviruses, human T-cell lymphotropic virus, and toxoplasmosis. On the third day polymerase chain reaction (PCR) analysis applied to an aqueous sample was negative. After 10 days, treatment was changed to oral Acyclovir 200 mg five times daily and methylprednisolone 36 mg daily. Topical treatment was tapered. Because of the extensive retinal hemorrhages, laser treatment to prevent retinal detachment was not applied.

Severe bilateral optic disk and peripheral superior retinal neovascularization developed over the next month and diode laser coagulation was applied in both eyes (Figure 2). One week later vision dropped to CF on the right and 20/200 on the left due to bilateral subhyaloidal hemorrhages and preretinal fibrous membranes. Vitrectomy with peeling of membranes and retinal tamponade with gas on the right and silicone oil on the left was performed. PCR of the vitreous sample was negative for viral antigen. After 2 months, the oil was removed and intraocular lens implantation on the left was necessary because of cataract formation. All medical treatment was discontinued. One year later central vision recovered to 20/20 on the right and 20/40 on the left with no change in peripheral visual field (Figure 3).

DISCUSSION

ARN was first described by Uruyama and coworkers in 1971.1 Patients with ARN usually present with an insidious onset of a red, irritated eye. Investigation reveals panuveitis with vasculitis, retinal hemorrhages, and predominating retinal necrosis.2 ARN is currently caused by a member of the herpes family of viruses.2 The detection of antibody titers in serum or ocular fluids and cultures for the causative agent are time-consuming and often false-negative. PCR applied to an aqueous or vitreous sample is a more recent, fast, and sensitive technique.3 Treatment of ARN consists of antiviral, antiinflammatory, and antithrombotic therapy. Acyclovir is given intravenously 1 week and orally for 12 to 14 weeks to obtain regression of necrosis and to avoid the development of new lesions in both eyes. Antiinflammatory treatment consists of systemic steroids 1 to 2 mg/kg/day during the first week with gradual tapering over the next weeks. Antithrombotic therapy with aspirin 500 mg/day is known to have a beneficial effect. Complications in ARN are frequent and severe: retinal neovascularization, holes and detachment, and optic disk atrophy.2 Treatment consists of laser therapy and
retinal surgery. Reports in literature of children with ARN describe a severe unilateral retinal necrosis in all cases, treated with Acyclovir only. Two-thirds of these children developed a retinal detachment, with a poor final visual outcome after laser therapy.\textsuperscript{4} Our case differs from other reports of ARN in children with bilateral involvement and predominance of retinal hemorrhages without detection of viral antigen in serum, aqueous or vitreous. Therefore, acute multifocal hemorrhagic retinal vasculitis, described by Blumenkranz and coworkers, needs to be considered in the differential diagnosis.\textsuperscript{5} Treatment in our patient consisted of Acyclovir and prednisone, but no aspirin because of the presence of extended retinal hemorrhages.
Intensive follow-up and rigorous treatment with diode laser, vitrectomy, and cataract surgery can result in a good central visual acuity in children with ARN.

References

An Eye on the Arts – The Arts on the Eye

Gómez argues that the mental state of visual attention invariably requires a human mother to turn her mind to whatever she is paying attention to. But when she does that, she usually turns some parts of her body toward it as well. In other words, she looks at it. An infant who lacks the ability to metarepresent won’t know that another mind is spinning inside his mother. He may, however, perceive her mental state of visual attention not as something coming from a mind that he doesn’t know is there but as part of her outer behavior of looking. The same might go for the ape. Although a chimpanzee infant might lack the ability to metarepresent his mother’s mind, he might still be able to interact with her and understand that she can interact with him in the same way that the human infant can “feel” some of his mom’s mental states by experiencing them as inseparably linked to her behaviors.

When two visual attentions contact, we call it a “gaze.” Gómez says that infants and animals unable to metarepresent can, in that case, understand that another is behaving like a subjective entity with intentions as well and, indeed, that is what human adults usually do. A mutual awareness exists. Locked in a gaze, we rarely mentally represent the other’s mental representation of us.

—Steven M. Wise (from Rattling the Cage, Toward Legal Rights for Animals, Perseus)