Magnetic Resonance Imaging of Diffuse Cerebral Vasculitis Associated With Acute Retinal Necrosis

Acute retinal necrosis (ARN) occurs more often in immunocompetent individuals and is defined clinically by discrete areas of peripheral retinal necrosis with rapid confluence, vascular sheathing, and prominent inflammation in the vitreous and anterior chamber. Pathologically, full-thickness necrosis of the retina is seen in the setting of an obliterator arteritis. Fluorescein angiography and histopathologic studies have shown that the arteritis is not confined to the retinal vessels but is seen in virtually all tissues of the eye including the iris, ciliary body, choroid, and optic nerve. This vasculitic process extending beyond the ocular vessels has not been well documented. We report a case of ARN and subclinical, diffuse cerebral vasculitis that was discovered after 3.0-T magnetic resonance imaging and magnetic resonance angiography were performed.

Report of a Case. A 46-year-old man had a 2-week history of decreasing vision in the right eye. He was initially diagnosed with anterior uveitis and treated with topical prednisolone and cycloplegic drops. His vision worsened to hand motions. Fundus examination at that time showed vitritis, vascular sheathing with hemorrhages, and patchy retinal opacities that at times coalesced. The diagnosis of ARN was made. The patient was generally healthy but had suffered recently from some nonspecific, diffuse headaches and problems with forgetfulness. However, he reported no focal neurologic symptoms or signs. He had no recent skin pain or lesions.

Intravenous acyclovir was given for 10 days. Optic nerve involvement was suspected, and the patient was transferred to our hospital where high-resolution magnetic resonance imaging and magnetic resonance angiography of the orbit and brain were performed using a 3.0-T scanner. There was no optic nerve sheath distension. No abnormalities of the visual nuclei or radiations were seen. However, signal abnormalities in the right basal ganglia and right thalamus were consistent with subacute strokes of different ages (Figure 1). The angiography revealed multiple focal abnormalities in flow enhancement of the vessels throughout the brain including those supplying the areas of the stroke (Figure 2). These observations were consistent with a diffuse vasculitis. Finally, the ophthalmic arteries demonstrated irregular flow enhancement that was more severe on the right side consistent with a vasculitic process.

Cerebrospinal fluid studies revealed a pleocytosis of white cells and a positive IgG antibody titer to varicella-zoster virus. Serologic study results were unremarkable. The patient did not develop any focal neurologic signs or symptoms during the course of his follow-up.

Comment. On review of the literature, we found only 1 report of “nec-
rotizing herpetic retinitis” associated with cerebral vasculitis. Rousseau et al reported 3 patients with human immunodeficiency virus who developed bilateral retinal necrosis despite intravenous antiviral therapy. Given the highly immunocompromised state of these patients and the fulminating course despite therapy, it is more likely that the retinal necrosis in these patients represented progressive outer retinal necrosis rather than ARN. Additionally, each of these patients had a herpes zoster rash, which is important because herpes zoster ophthalmicus has often been associated with ipsilateral cerebral angiitis.4 The patients in the report by Rousseau et al did develop clinical manifestations and radiologic evidence of central nervous system strokes. Cerebral arteriography was performed in 1 of these patients with evidence of a focal vasculitis.

In contrast, our patient was healthy and had no prior zoster rash. Further, he had no clinical signs of a central nervous system stroke. Since this diffuse cerebral vasculitis was subclinical, it is plausible that other patients with ARN may have had similar central nervous system processes that were not discovered. This has clinical implications since parenteral corticosteroids have been used in addition to antivirals in treating central nervous system vasculitides, notably those caused by varicella-zoster virus.4

Although this may be the first report of cerebral vasculitis and ARN, the association of these entities might be expected given that ARN is, in part, a vasculitic process. Further, varicella-zoster virus has been linked to each separately. It is a known cause of ARN and as mentioned previously, herpes zoster ophthalmicus–related cerebral angiitis is well documented. Finally, animal models demonstrate that anterior chamber inoculation of herpesviruses results in spread to the brain.3

This case is unique in that clinical symptoms or an antecedent zoster rash did not herald the cerebral vasculitis. It demonstrates that cerebral vasculitis may occur when ARN is the only manifestation of a presumed herpesvirus infection, and screening may be warranted using high-resolution magnetic resonance imaging and magnetic resonance angiography, especially in cases where varicella-zoster virus is the implicated cause.

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REFERENCES


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