Optic Chiasm, Optic Nerve, and Retinal Involvement Secondary to Varicella-zoster Virus

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Immunocompromised patients are known to be at risk for varicella-zoster virus reactivation, often in atypical manners. We describe a 30-year-old man with simultaneous involvement of the retina, optic chiasm, and optic nerve with varicella-zoster virus who had a bitemporal visual field defect.

A 30-year-old man with chronic myelogenous leukemia who had undergone bone marrow transplantation developed blurred vision in both eyes, dizziness, and mild headaches. He had a bitemporal hemianopia that rapidly progressed to complete blindness. In his right eye a single focus of deep retinochoroiditis developed, which over time progressed and became more confluent. Three weeks later the patient died secondary to massive subarachnoid hemorrhage. Findings from the postmortem evaluation revealed viral intranuclear inclusions in the retina and optic nerve, and immunostaining of the optic nerve was positive for varicella-zoster virus (VZV).

REPORT OF A CASE

In April 1996, a 30-year-old white man received a diagnosis of chronic myelogenous leukemia that was Philadelphia chromosome positive. In September 1998 he had dizziness, bilateral blurred vision, and slight headaches of 5 days’ duration. He had undergone allogenic bone marrow transplantation in March 1998. His clinical course posttransplantation had been complicated by graft-vs-host disease, and he was being treated with cyclosporine and prednisone.

On evaluation September 20, 1998, his visual acuity was 20/400 OD and 20/200 OS. No afferent pupillary defect was present, and visual field testing showed a bitemporal defect (Figure 1). The slit-lamp examination findings were normal, and funduscopic examination of the right eye revealed a gray, deep retinochoroidal infiltrate nasal to the optic nerve (Figure 2 and Figure 3). The left retina and optic nerve were normal. Magnetic resonance imaging of the brain revealed an increased signal in the area of the posterior optic nerve and chiasm (Figure 4).

Within 1 week the patient had progressive deep and full-thickness areas of retinitis in the right eye, and he developed 2 small foci of retinitis in the left eye. In 2 days the patient had no light perception OU. Two weeks later he experienced a large subarachnoid hemorrhage secondary to pancytopenia and coagulopathy and died.

RESULTS

Findings from postmortem examination of the brain revealed diffuse multifocal subarachnoid hemorrhage covering most of the brain, brainstem, and cerebellum. Sections of the globes showed areas of full-thickness retinal necrosis with lymphocytic infiltration of the choroid (Figure 5A) and in some areas superficial retinitis with relative sparing of the deep retina (Figure 5B). Sections of the optic nerve and retina showed intranuclear inclusions suggestive of infection with a herpesvirus (Figure 5C). Focal areas of demyelination were present in the nerve with
infiltration of the tissue by macrophages. Immunostains for VZV were strongly positive (Figure 5D), while immunostains for herpes simplex virus and cytomegalovirus were negative. Polymerase chain reaction of the tissue was positive for VZV but negative for the other herpesviruses.

The ocular manifestations of VZV infections are protean and include dermatitis, keratitis, uveitis, retinitis, and optic nerve involvement.1 Pa-
tients who are immunocompromised are particularly susceptible to these complications, and they frequently occur without typical cutaneous dermatomal eruption. Simultaneous involvement of the retina and optic nerve from VZV in acute retinal necrosis syndrome is well documented.2 Optic disc edema with associated optic neuropathy and retrobulbar optic neuritis have been described as preceding the acute retinal necrosis syndrome in patients with the acquired immunodeficiency syndrome.3,4 Progressive outer retinal necrosis is another retinal manifestation of VZV that often occurs in patients with acquired immunodeficiency syndrome, presenting with multiple discrete areas of deep retinal opacification and progressing to confluent retinal involvement.5

The most common significant viral infections in bone marrow transplant recipients are due to members of the herpesvirus family.6 Infection with VZV is almost always due to a reactivation of a previous VZV infection. At 5 years, more than 50% of bone marrow transplant recipients have clinical infection with VZV.7 The greatest risk period for development of serious life-threatening VZV infections in patients undergoing bone marrow transplantation is 2 to 10 months posttransplantation, with a median occurrence at 5 to 7 months.7,8 The presence of graft-versus-host disease is a major risk factor for VZV reactivation.7

Our patient was severely immunocompromised by his underlying leukemia and graft-versus-host disease that required therapy with prednisone and cyclosporine. The atypical chiasmal involvement he had as evidenced by the bitemporal hemianopia appears to be unique. The diagnosis of VZV infection became more clinically apparent with the onset and progression of the retinitis in the right eye and was confirmed by autopsy findings. Varicella-zoster infection should be considered in immunocompromised patients with retrobulbar optic nerve or chiasmal lesions, even in the absence of cutaneous or retinal lesions.

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