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1. Hamade IH, Al Shamsi HN, Al Dhibi H, Chacra CB, Abu El-Asrar AM, Tabbara KF. Uveitis survey in children. *Br J Ophthalmol.* 2009;93(5):569-572.
2. Read RW, Holland GN, Rao NA, et al. Revised diagnostic criteria for Vogt-Koyanagi-Harada disease: report of an international committee on nomenclature. *Am J Ophthalmol.* 2001;131(5):647-652.
3. Iqniebi A, Gaafar A, Sheereen A, et al. HLA-DRB1 among patients with Vogt-Koyanagi-Harada disease in Saudi Arabia. *Mol Vis.* 2009;15:1876-1880.
4. Al-Halafi A, Dhibi HA, Hamade IH, Bou Chacra CT, Tabbara KF. The association of systemic disorders with Vogt-Koyanagi-Harada and sympathetic ophthalmia. *Graefes Arch Clin Exp Ophthalmol.* 2011;249(8):1229-1233.
5. Jabs DA, Nussenblatt RB, Rosenbaum JT; Standardization of Uveitis Nomenclature (SUN) Working Group. Standardization of uveitis nomenclature for reporting clinical data: results of the First International Workshop. *Am J Ophthalmol.* 2005;140(3):509-516.
6. Jabs DA. Improving the reporting of clinical case series. *Am J Ophthalmol.* 2005;139(5):900-905.

Use of Intravitreal Bevacizumab to Treat Macular Edema in West Nile Virus Chorioretinitis

West Nile virus (WNV) appeared in the United States in 1999; ocular sequela have been documented since 2003.¹⁻³ Associated chorioretinitis, ischemic vasculitis, retinal hemorrhages, and choroidal neovascularization have all been reported.³⁻⁵

Report of a Case. A 66-year-old man residing in a northern suburb of Chicago, Illinois, had increasing fatigue, severe headaches, confusion, and fevers for 4 days. Enzyme-linked immunosorbent assay of his cerebrospinal fluid was positive for WNV-specific IgM. The patient received supportive therapy for 2 weeks.

Two weeks after discharge, the patient had profound vision loss in his right eye. Visual acuity was counting fingers at 3 ft OD (no improvement with pinhole) and 20/20 OS with correction. Anterior segment examination showed a posterior subcapsular cataract in the right eye. Fundus examination showed absence of vitreous cells in each eye. In the right eye, vasculitis, intraretinal hemorrhages, and areas of nonperfusion were seen (**Figure 1A** and **B**). In the left eye, linear streaks of chorioretinal scarring were seen, with no vasculitis or retinal hemorrhages (**Figure 1D** and **E**). Fluorescein angiography confirmed areas of nonperfusion in the right macula (**Figure 1C**) and linear streaklike hyper-

fluorescent scars along the arcades of the left eye (**Figure 1F**). Spectral-domain optical coherence tomography (OCT) showed a central thickness of 415 μm OD (**Figure 2A**); findings from OCT of the left eye were unremarkable.

On routine eye examination for type 2 diabetes 5 months prior to hospitalization, visual acuity was 20/40 OD and 20/20 OS. There was no diabetic retinopathy, and diabetes was controlled with metformin hydrochloride (the hemoglobin A_{1c} level was 6.8% of total hemoglobin 6 months prior to hospitalization [to convert to proportion of total hemoglobin, multiply by 0.01]). Visual acuity of 20/40 OD was attributed to the mild cataract.

The macular edema was treated with topical nepafenac in the right eye, 4 times daily. The patient returned 2 weeks later; OCT showed worsening macular edema with a thickness of 502 μm centrally (**Figure 2B**). Bevacizumab (1.25 mg) was injected intravitreally into the right eye. Two weeks later, visual acuity had improved to 20/150 OD and the OCT central thickness was 346 μm . During the next 2 months, macular edema in the right eye remained stable on clinical examination and OCT; the patient underwent cataract extraction with a monofocal intraocular lens implant.

At the last follow-up, 7 months after the initial hospitalization, visual acuity was 20/80 OD and 20/20 OS. On dilated retinal examination, vascular sheathing was seen along the right superior arcade; macular edema and hemorrhage in the right eye had resolved. The left eye showed old, inactive pigmented streaks of chorioretinal scarring along the arcades. Spectral-domain OCT showed the right macula to have a normal thickness of 301 μm centrally (**Figure 2C**).

Comment. To our knowledge, this is the first case of WNV chorioretinitis with macular edema and the first use of intravitreal bevacizumab to treat it. Despite the patient's history of diabetes, the patient had no diabetic retinopathy on funduscopic examination 5 months prior to acquiring WNV. The course of WNV infection, onset of visual symptoms, and detection of macular edema strongly suggest that the macular edema was the result of increased vascular permeability from WNV chorioretinitis and retinal vasculitis. Diabetes likely predisposes patients with WNV to occlusive vasculitis in the brain and retina,³ and diabetes has been implicated as an independent risk factor in WNV-related death.⁶ While the macular edema may have regressed on its own, the prompt resolution of the macular edema and improvement in visual acuity that followed shortly after use of bevacizumab suggest a potential use for the drug in this condition that warrants further investigation.

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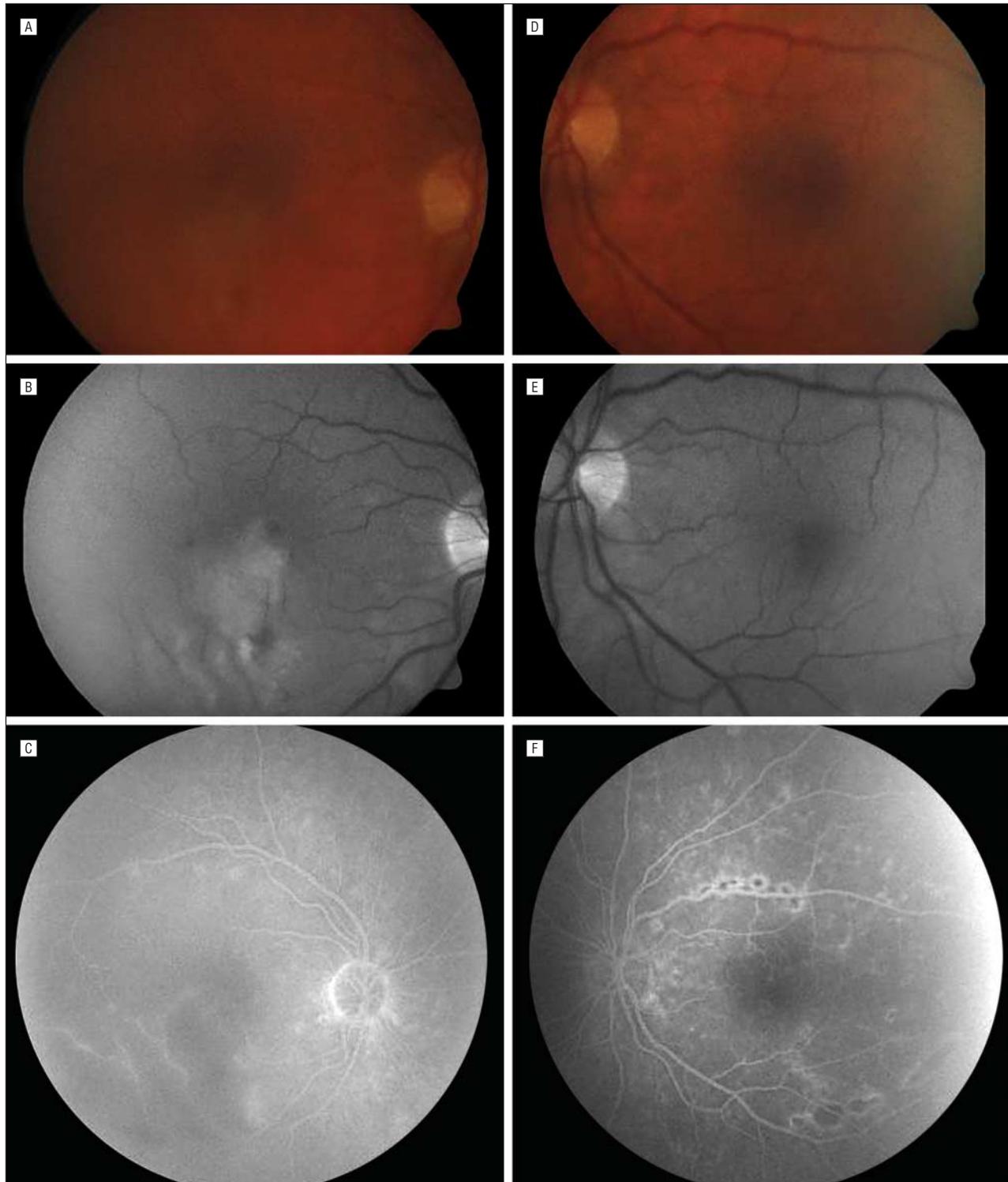


Figure 1. Fundus photograph (A), red-free photograph (B), and fluorescein angiogram (C) of the right eye and fundus photograph (D), red-free photograph (E), and fluorescein angiogram (F) of the left eye at the initial visit.

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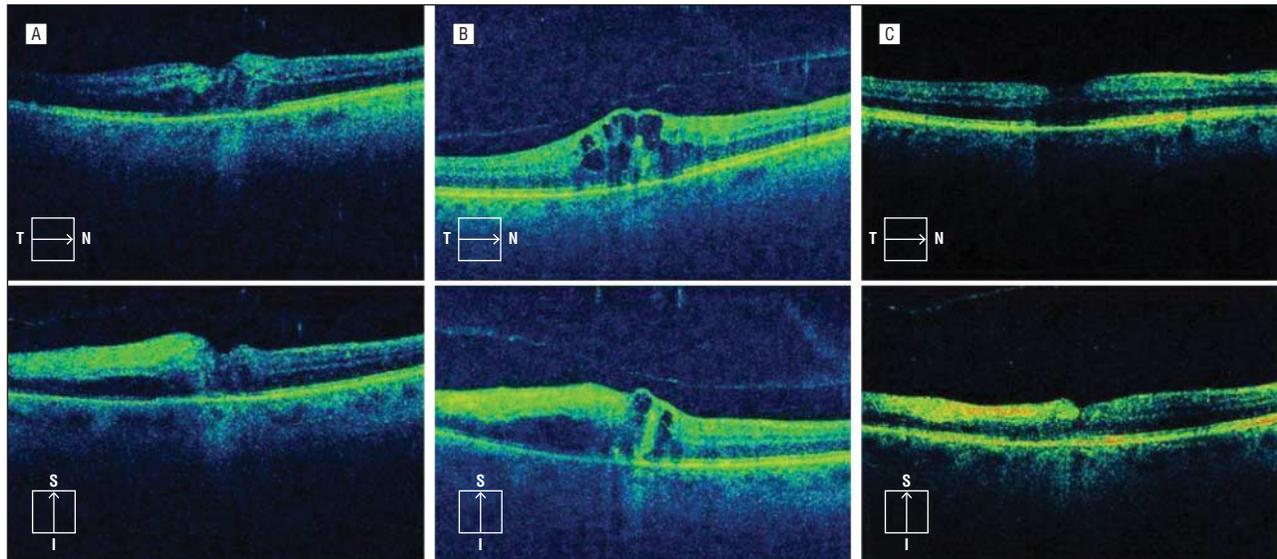


Figure 2. Spectral-domain optical coherence tomographic images of the right eye at the initial visit (A), 2 weeks after the initial visit (B), and 7 months after the initial visit (C). T indicates temporal; N, nasal; S, superior; and I, inferior.

1. Bains HS, Jampol LM, Caughron MC, Parnell JR. Vitritis and chorioretinitis in a patient with West Nile virus infection. *Arch Ophthalmol.* 2003;121(2):205-207.
2. Garg S, Jampol LM. Systemic and intraocular manifestations of West Nile virus infection. *Surv Ophthalmol.* 2005;50(1):3-13.
3. Garg S, Jampol LM, Wilson JF, Batlle IR, Buettner H. Ischemic and hemorrhagic retinal vasculitis associated with West Nile virus infection. *Retina.* 2006;26(3):365-367.
4. Chan CK, Limstrom SA, Tarasewicz DG, Lin SG. Ocular features of west nile virus infection in North America: a study of 14 eyes. *Ophthalmology.* 2006;113(9):1539-1546.
5. Seth RK, Stoessel KM, Adelman RA. Choroidal neovascularization associated with West Nile virus chorioretinitis. *Semin Ophthalmol.* 2007;22(2):81-84.
6. Nash D, Mostashari F, Fine A, et al; 1999 West Nile Outbreak Response Working Group. The outbreak of West Nile virus infection in the New York City area in 1999. *N Engl J Med.* 2001;344(24):1807-1814.

Peripheral Retinal Nonperfusion Associated With Optic Nerve Hypoplasia and Lissencephaly

Few entities exist in which optic nerve hypoplasia (ONH) is found in association with peripheral retinal nonperfusion. Among these are the congenital muscular dystrophies with abnormal glycosylation of α -dystroglycan, consisting of Walker-Warburg syndrome, muscle-eye-brain disease, and Fukuyama congenital muscular dystrophy, characterized by defective brain migration and ocular abnormalities. Posterior segment findings in these disorders have included ONH as well as retinal dysplasia.¹ Herein, we describe a full-term girl not only with ONH and bilateral peripheral retinal nonperfusion with resultant tractional retinal detachments but also with severe brain abnormalities including lissencephaly and hydrocephalus.

Report of a Case. A 3785-g girl born at 39 weeks' gestation was referred for bilateral retinal detachments. The prenatal course was uneventful, and there was no family history of eye or neurological abnormalities.

Shortly after birth, she developed seizures. Magnetic resonance imaging revealed massive dilatation of the lateral ventricles secondary to atrophy and hydrocephalus,

marked cortical dysplasia and lissencephaly, periventricular calcifications, and a thin corpus callosum (**Figure 1**). Although the calcifications were suspicious for cytomegalovirus infection, serologic and urine culture results were negative. The serum creatine kinase level, to evaluate for a muscular dystrophy, was normal.

Examination revealed normal anterior segments. Fundus photography of the right eye showed a large tractional retinal detachment involving the macula, obscuring both the macula and the optic nerve, with posterior retinal vessels dragged and distorted into a retinal fold (**Figure 2A**). Fluorescein angiography demonstrated massive leakage off the stalk and along the apex of the horseshoe-shaped retinal detachment (Figure 2B). The optic nerve of the left eye showed a double ring sign consistent with ONH as well as foveal hypoplasia (Figure 2C). The retinal vessels terminated posteriorly, especially temporally with extraretinal fibrovascular proliferation extending into the vitreous and tractional detachment inferonasally and superotemporally (Figure 2D and E).

Laser photocoagulation was applied to the avascular retinal zones. The patient subsequently underwent vitrectomy in both eyes. Three months later, the left retina was attached completely (Figure 2F) but the right retina remained detached.

Comment. There is a spectrum of disorders with ocular and neurological manifestations that overlap those of our patient. In the congenital muscular dystrophies, an underlying defect in glycosylation is thought to result in severe defects in neuronal migration, thus causing hypoplasia of various brain and eye structures.² Walker-Warburg syndrome is the most severe, with brain abnormalities including lissencephaly, hydrocephalus, cerebellar malformation, hypomyelination of the white matter, and agenesis of the corpus callosum.³ Ocular posterior segment abnormalities include retinal dysplasia as well as hypoplasia or atrophy of the optic nerve and macula. Lissencephaly can also be found in Fukuyama congenital