sent with the unusual fundus. However, because only 2 generations were analyzed, the pattern is also consistent with autosomal recessive, X-linked dominant, digenic, or mitochondrial inheritance. Because this is the first report of CGARPES in more than a single family member, we believe the appropriate name for this unique presentation is familial congenital grouped albinotic retinal pigment epithelial spots.

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Immune Choroiditis Following Contralateral Acute Retinal Necrosis

Acute retinal necrosis (ARN) syndrome is characterized by acute panuveitis and retinal arteritis that progresses to a diffuse necrotizing retinitis with late-onset rhegmatogenous retinal detachment.

The contralateral eye is involved in 10% of patients despite systemic antiviral treatment. It is thought to occur because of retrograde axonal transport between the suprachiasmatic nucleus of the hypothalamus and the contralateral retina in an animal model. Bilateral ARN is characterized by bilateral foci of retinal necrosis associated with arteritis and panuveitis. However, a noninfective choroiditis in the contralateral eye following ARN has never been described.

This article describes 2 cases of presumed reactive immune choroiditis following contralateral ARN.

Report of Cases. Case 1. A 54-year-old white woman was referred for treatment with a diagnosis of presumed ARN in her right eye. There was no other relevant medical history. Her best-corrected visual acuity (BCVA) was 20/200 OD, 20/20 OS. On examination, the right eye had panuveitis with retinal necrosis and associated arteritis. There was also a relative afferent pupillary defect in the right eye. The left eye was unaffected. Vitreous biopsy confirmed herpes simplex virus–associated ARN by polymerase chain reaction. She began taking 10 mg/kg of intravenous acyclovir 3 times per day for 7 days followed by 500 mg of oral valacyclovir 3 times per day for 3 months. Four months later, the right eye became phthisical owing to persistent inflammation despite topical corticosteroids. Ten months later, the right eye was no longer inflamed but she had increasingly blurred vision and mild discomfort in her left eye. Her BCVA was 20/40 OS. Anterior segment examination revealed a white eye with mild inflammation and a cataract. There was minimal vitritis, and funduscopy revealed disc swelling with multiple deep pale lesions throughout the fundus (Figure 1A). Fluorescein angiography showed early hyperfluorescence of the choroidal lesions with late staining accompanied by perivascular and optic disc leakage of dye (Figure 1B). A vitreous biopsy was performed and was negative for herpes simplex virus, Varicella zoster virus, cytomegalovirus, and Epstein-Barr virus. The sensitivity for detection of virus by polymerase chain reaction in ARN at our institution was 87.5%. With suspicion of atypical presentation of bilateral ARN, she started taking 500 mg of oral valacyclovir 3 times per day and 40 mg of prednisolone once per day. Her BCVA improved to 20/20 OS. During the next 12 months, her visual acuity diminished to 20/40, partly owing to a cataract that was removed with a peritomomorphic intravitreal triamcinolone.

Figure 4. The sister of case 1. A, In the right eye, note the smaller retinal flecks in the posterior pole. B, Lesions in the posterior pole of the left eye are shown. C, Lesions are present up to the superior quadrant periphery of the right eye. D, The superior temporal portion of the left eye contained the retinal flecks.
lone injection. This did not improve her vision, and her condition continued to deteriorate, with BCVA reducing to 20/200 OS owing to macular involvement (Figure 1C). The choroiditis eventually left pale white scars with associated optic neuropathy (Figure 1D). On the final follow-up, the left eye was phthisical and she had no light perception visual acuity.

Case 2. A 12-year-old white boy with a history of viral meningitis as an infant was referred with a diagnosis of ARN in the left eye. His BCVA was 20/20 OD, 20/40 OS. Examination revealed a herpetic lesion on his left upper eyelid and a moderate nongranulomatous anterior uveitis with associated vitritis in the left eye. The optic disc was swollen, with an area of retinal necrosis and associated arteritis. He started taking 10 mg/kg/d of intravenous acyclovir 3 times per day for 7 days followed by oral acyclovir for 3 months. Results of testing of vitreous biopsy samples were positive for Human herpesvirus 1 and negative for Human herpesvirus 2. Rhegmatogenous retinal detachment developed in the left eye but was deemed to have very poor visual potential and was therefore not operated on. The left eye eventually became phthisical. One year later, the right eye remained asymptomatic, with a BCVA of 20/20 OD. Anterior segment examination was unremarkable but fundoscopy revealed deep pale lesions in the retina located in the peripapillary area and along the vascular arcades (Figure 2A). Results of a Goldman visual field test were normal. Electrodiagnostics revealed mild dysfunction of both rod and cone systems. No treatment was prescribed. The lesions eventually left punched-out chorioretinal scars with good vision (Figure 2B). Two years later, he presented with blurred vision and BCVA of 20/80 OD. The right eye was quiet but fundoscopy revealed a peripapillary hemorrhage associated with a choroidal neovascular membrane (Figure 2C). This was managed conservatively and his visual acuity improved to 20/20 OD (Figure 2D).

Comment. These 2 cases describe a delayed immune response to previous ARN in the contralateral eye. The clinical presentation is totally different from bilateral ARN in that there is a lack of the typical features of ARN. However, the choroidal involvement could be explained by the granulomatous infiltration of the choriocapillaris and choroid in ARN, seen similarly in sympathetic ophthalmia and Vogt Koyanagi Harada syndrome. Both cases of ARN ultimately became phthisical but the difference in clinical presentation of the immune choroiditis is interesting. Case 1 had features reminiscent of sympathetic ophthalmia, whereas case 2 had features of multifocal choroiditis. Both cases could demonstrate a spectrum of a subthreshold presenta-
tion of ARN in which an inflammatory response is clinically visible but does not progress to ARN. These cases may also represent a spectrum of sympathetic ophthalmia. However, previous histological studies describe the absence of organisms in sympathetic ophthalmia. Therefore, if they were indeed a spectrum of sympathetic ophthalmia, the interesting factors would be the phthisis bulbi and previous vitreous biopsies in the ARN eye.

Because second eye involvement has been reported as late as 34 years after first diagnosis of eye involvement in ARN, it is important to differentiate between a purely inflammatory, reactive choroiditis and an infective etiology. The treatment for immune choroiditis, which involves immunosuppression, could potentially allow infection to progress unopposed if the diagnosis of ARN is not ruled out and appropriate antiviral treatment given to prevent reactivation of disease.

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Pemphigus Vulgaris Involving the Eyelids
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Figure 1. Photograph of the left eye of a 20-year-old women with pemphigus vulgaris, showing severe involvement of the skin of both the upper and lower eyelids. She also had multiple vesiculobullous lesions of pemphigus all over the body.

Figure 2. Direct immunofluorescence of a skin biopsy specimen showed IgG immune complex deposits in intercellular space consistent with the diagnosis of pemphigus vulgaris.