keratoconjunctivitis should be suspected in patients who have ocular surface symptoms and report a metallic taste in their mouth and who handle zoanthids, as occurred in case 1. After excluding the infectious causes, toxic keratoconjunctivitis might be treated with aggressive use of topical corticosteroids as well as topical cyclosporine and lubricants.

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Familial Congenital Grouped Albinotic Retinal Pigment Epithelial Spots

Congenital grouped albinotic retinal pigment epithelial spots (CGARPES), or polar bear tracks, is a rare anomaly characterized by multiple grouped, white, variably sized, albinotic spots. They generally involve the peripheral retina, similar to that of bear track grouped pigmentation.1 Usually the macula is spared, and the spots may occur in one or both eyes. The lesions seem to be stable, and visual acuity, visual fields, color examination, dark adaptation, and electrophysiologic findings are normal. There is only 1 report of a decrease in visual acuity in this entity.2(p614-615) Fluorescein angiography revealed a variable pattern related to the choroidal fluorescence seen through these lesions that was considered sporadic and rare in frequency. In this article, we describe 4 cases that occurred in a Brazilian family of Italian descent. This is the first reported instance of familial CGARPES.

Report of Cases. Case 1. A 19-year-old woman was referred for evaluation after her general ophthalmologist noted areas of retinal abnormality on ophtalmoscopy after a routine refraction. She had occasional headaches but no specific ocular symptoms, and she was in excellent general health. Visual acuity was 20/20 in both eyes with −0.50 cylinder correction in the left eye. Slitlamp examination of the anterior segments and vitreous cavity was unremarkable. Intraocular pressures were normal. Funduscopy of both eyes revealed multiple white and sometimes yellow flecks of variable size and configuration affecting all parts of the retina (periphery, equator, posterior pole, and fovea) (Figure 1). The size of the flecks was highly variable, from the diameter of one vessel to 4 times the diameter of the optic nerve but the flecks had a consistent color pattern: in the smaller lesions, the color was more homogeneous, and in the larger lesions, it was concentrated at the edge of the flecks (Figure 1). Fluorescein angiography showed a normal choroidal and retinal vascular perfusion. The arterial phase revealed a transmitted hyperfluorescent lesion (a window defect of the retinal pigment epithelium) that corresponded to the lesions seen during the ophthalmic examination. No intraretinal fluid was seen during the late phase of the angiogram (Figure 1).

Color vision tests with the Farnsworth dichotomous (D-15) test,
electrooculogram, electroretinogram, and visual field were normal. Follow-up 1 and 5 years later showed the same ocular fundus findings without progression of the flecks, and the headaches had disappeared. There was no history of consanguineous marriage. When other members of the family were examined, this unusual fundus was observed in her 2 sisters and her mother (Figure 2).

Case 2. A 39-year-old woman, the mother of the patient described in case 1, described no ocular or systemic symptoms. Her visual acuity was 20/20 in both eyes, with normal visual fields and no clinical night blindness. Results of anterior biomicroscopy examination, color testing (Farnsworth dichotomous D-15), electroretinogram, and electrooculogram were normal. Color fundus photographs showed deep lesions of the fundus, similar to those seen in case 1. The same color pattern was seen: more homogenous color in the smaller lesions, and color concentrated at the edges of the larger lesions (Figure 3). The ocular coherence tomographic findings of a small fleck showed no defect in the inner or outer retina.

Case 3. This 16-year-old girl is the sister of the patient described in case 1. Her corrected visual acuities were 20/20 in both eyes. Her visual fields were normal, and she had no night blindness. Retinal flecks were present in the posterior pole and all fundus quadrants up to the periphery. However, the lesions were smaller than those seen in either case 1 or case 2 (Figure 4).

Case 4. A 12-year-old girl, the youngest sister of cases 1 and 3, was also healthy, with 20/20 visual acuity and normal visual field without clinical night blindness. Both eyes were affected with fleck lesions, comparable in shape and distribution with the flecks seen in the eyes of her 2 sisters and mother. As in her 16-year-old sister (case 3), the fleck lesions were smaller but were distributed from the macula up to the periphery in all quadrants.

Comment. The retinal condition in the patients described in our article is compatible and consistent with a diagnosis of congenital grouped retinal pigment epithelial spots. The 4 cases were characterized by bilateral, multiple, deeply situated, white-yellow fleck lesions with a panretinal distribution (fovea, posterior pole, peripapillary, equator, and periphery). However, the lesions in original reports of CGAREPS tended to be white, spare the macula, and unicoar. On the other hand, our and more recent articles found those spots to be bilateral, affecting the macula and present with some yellow lesions, particularly in the posterior pole area. We hypothesize that yellow macular lesions could be explained by the increased amount of xanthophyll in these areas compared with the periphery, where the spots are white. Like in all reports of CGAREPS, the lesions vary in size and configuration; however, we believe there is a color pattern: larger lesions tend to have heterogeneous coloration between the edge and the central part; smaller lesions tend to be more homogeneous.

According to Gass, the spots may represent focal thickening of the retinal pigment epithelium that is filled with a white material that may be diffusely distributed or more concentrated in the periphery of the lesion. Histologic findings have not been reported so far to clarify the true composition of this material.

Fluorescein angiography showed hyperfluorescent spots with the initial phase of the angiogram (window defect) correlating exactly with the lesions observed in the ocular fundus. This finding is well documented in previous articles. The differential diagnosis of CGAREPS includes fundus flavimaculatus, fundus albipunctatus, and familial drusen. Gass recognized that CGAREPS is identical to the entity reported by Kandori and colleagues, sometimes referred to as the flecked retina of Kandori.

In contrast to previous studies that found this entity to be sporadic, we found an obvious familial component in our cases. The hereditary pattern in the presently described family is most likely an autosomal dominant form of the disease since only the mother and 3 daughters pre-
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 rheumatogenous 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 perioperative intravitreal triamcino-