only, with pale featureless optic discs. A computed tomodergraphic scan of the brain and orbits excluded mass lesions or intraorbital abnormalities, and magnetic resonance imaging revealed severe atrophy of the optic nerves, optic chiasm, and optic tract.

Genetic analysis revealed compound heterozygosity, with a GAA expansion on one allele and a G-to-T base substitution in exon 4 (Gly130Val) on the other. The mitochondrial point mutations associated with MELAS (mitochondrial myopathy, encephalopathy, lactic acidosis, and stroke-like episodes) or MIDD (maternally inherited diabetes and deafness), MERRF (myoclonic epilepsy with ragged-red fibers), NARP (neuropathy, ataxia, and retinitis pigmentosa), and Leber hereditary optic neuropathy were not identified.

Comment. This patient with FRDA had visual loss of a rapidity and severity not previously described in FRDA. She was also noted to have a pattern dystrophy but had normal electroretinographic results and pattern electroretinographic results, suggesting that it is the optic nerve or optic tract disease that is causing the visual loss rather than the pattern dystrophy. Macular abnormalities have been described in autosomal dominant spinocerebellar ataxia type 7 but not previously in FRDA; thus, this may be a coincidental finding in our patient. It is possible that the pattern dystrophy observed in this case could be associated with a mutation in peripherin/RDS gene, the ELOVL4 gene, or as-yet-unknown genes.

The cause of optic atrophy in FRDA is unknown, but it occurs more frequently in patients with larger GAA repeats and also more frequently in compound heterozygotes than homozygotes. Furthermore, optic neuropathy emerges late in the course of disease and may be more frequent in compound heterozygotes who tend to survive longer. Optic atrophy is a heterogeneous disorder often caused by inherited or acquired abnormalities of mitochondrial function. Further investigation of the molecular mechanisms causing FRDA in compound heterozygotes may provide important insights into the underlying pathogenesis that leads to optic atrophy.

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Autofluorescence Findings in Acute Exudative Polymorphous Vitelliform Maculopathy

Acute exudative polymorphous vitelliform maculopathy is a rare acute disorder characterized by multifocal, curvilinear or oval, yellow-white, posterior pole lesions with vision loss and preceding headaches. These retinal pigment epithelial (RPE) lesions are associated with bilateral serous detachments and evolve into deposits resembling those of Best disease. During subsequent weeks, patients experience gradual recovery of vision. Optical coherence tomography and indocyanine green angiography findings have been described. To date, 7 cases have been reported in the literature. The main aim of the current study was to ascertain the nature of the posterior pole lesions using autofluorescence imaging in 3 patients and to characterize macular, retinal, and RPE function using International Society for Clinical Electrophysiology of Vision–standard electrophysiology.

Report of Cases. Case 1. A healthy 49-year-old man noticed a gradual decrease in both distance and near vision, worse in his left eye, preceded by a flu-like illness. There was no other relevant medical, ophthalmic, or family history. On first examination several months later, his best-corrected visual acuity (VA) was 20/30 OD and 20/60 OS. Anterior segments, optic discs, and the peripheral retina were within normal limits. Posterior segment examination showed bilateral yellow-white confluent macular deposits (Figure 1 A and B). Autofluorescence imaging using a confocal scanning laser ophthalmoscope showed that the lesions were hyperfluorescent (Figure 1C and D), consistent with lipofuscin deposits. Pattern and full-field electroretinogram (ERG) and electro-oculogram results were normal, the latter suggesting normal generalized RPE function that would not be in keeping with Best disease. Six months later, the patient’s VA improved to 20/30 OU. The macular deposits had diminished (Figure 1E and F), with a corresponding reduction in autofluorescence (Figure 1G and H).

Case 2. A previously well 33-year-old woman had sudden onset of bilateral ring scotomas and distortion in the right eye following a flu-like illness and headaches. On first examination elsewhere, bilateral RPE detachments were noted. These settled spontaneously over a month. When referred to our service 9 months later, she had photophobia and difficulty adapting to dark. Her VA was 20/16 OU. A pale white curvilinear lesion appeared at each macula, with multiple yellow circular lesions around the posterior pole that were brightly autofluorescent (Figure 2A and B). The pattern ERG results were normal, in keeping with normal macular function. The full-field ERG and electro-oculogram results were normal bilaterally. There was partial resolution of the lesions on color photographs and confocal scanning laser ophthalmoscopy over 2 years (Figure 2C and D).
Case 3. A previously well 38-year-old man awoke one morning with a sudden decrease in central vision in both eyes and no recollected pro-
drome. His VA was 20/80 OD and 20/125 OS when first seen elsewhere. Some macular subretinal fluid was noted and confirmed by
optical coherence tomography. Fluorescein angiography (Figure 3A) showed early hyperfluorescence. Investigations were directed toward posterior scleritis, and results of a workup for systemic immune-mediated diseases were unremarkable other than a borderline-raised C-reactive protein level (11 mg/L). The patient was treated conservatively. On review in our service 15 months later, he reported fluctuating central vision, photophobia, and slow dark adaptation. His VA was 20/40 OD and 20/60 OS. Multilobular white-yellow lesions involving each macula appeared and extended beyond temporal arcades (Figure 4A and B). Fluorescein an-

Figure 1. From the first examination of case 1, fundus photographs of the right (A) and left (B) eyes and autofluorescence images of the right (C) and left (D) eyes. From 6 months later, fundus photographs of the right (E) and left (F) eyes showing reduction of the yellow-white deposits. Autofluorescence images of the right (G) and left (H) eyes show a corresponding reduction.

Figure 2. From the first examination of case 2, fundus photograph (A) and autofluorescence image (B) of the right eye. From 2 years later, fundus photograph (C) and autofluorescence image (D) of the right eye.
Giology (Figure 3B) showed complete resolution of all of the hyperfluorescent lesions. The yellow deposits were nonfluorescent. Four months later, his VA was 20/20 OU with gravitation of the lesions inferiorly (Figure 4C and D). All of the lesions were brightly autofluorescent (Figure 4E and F). Optical coherence tomography of the right eye showed anterior displacement of the RPE and photoreceptors overlying a hyporeflective space adjacent to the hyperreflective deposit (Figure 4G). Pattern and full-field ERG revealed no abnormality; the electrooculogram light rise was borderline on the right (170%) and normal on the left (185%).

Comment. The pathogenesis of acute exudative polymorphous vitelliform maculopathy is still unclear, but an inflammatory and immune-mediated cause has been proposed on the basis of associations with preceding viral illness and apparent steroid responsiveness. Dysfunction of the RPE initiated by different stimuli might explain the overload in lipofuscin. It has also been suggested that an inflammatory involvement of the inner choroid and RPE might result in leakage, causing the exudative detachments. Indocyanine green angiography findings in the acute phase have demonstrated choriocapillaris involvement and an affinity for the dye within the lesions thought to consist of inflammatory exudates. Hyporeflective spaces overlying the RPE, possibly corresponding to serous fluid beneath the yellow deposits, have been documented with optical coherence tomography. These spaces resolve in conjunction with diminution of the deposits and visual improvement correlating with the reestablishment of the normal RPE-photoreceptor complex anatomy.

Autofluorescence intensity has been found to parallel the amount and distribution of lipofuscin. The current data support previous suggestions that the yellow deposits in acute exudative polymorphous vitelliform maculopathy contain lipofuscin. Accumulation may result from impaired function of the RPE and its initial failure to clear the increased lipofuscin load or abnormally high

Figure 3. For case 3, early-phase fluorescein angiogram images of the right eye at the first examination (A) and 14 months later (B).

Figure 4. For case 3, fundus photographs taken at our institution 14 months after the first examination in the right (A) and left (B) eyes, fundus photographs taken 18 months later in the right (C) and left (D) eyes, and autofluorescence images of the right (E) and left (F) eyes and optical coherence tomographic images of the right (G) and left (H) eyes 16 months later.
outer-segment turnover. Spontaneous clinical resolution suggests active RPE phagocytosis. It is notable that previous articles\textsuperscript{1,4} have described more severely abnormal electro-oculogram results, possibly at a different or earlier stage of recovery; we have no other explanation for the apparent discrepancy. Previous studies have failed to show mutations in the VMD2 or RDS genes in affected patients that can be causative of inherited Best maculopathy and adult vitelliform macular dystrophy.\textsuperscript{2,3} This is in keeping with the absence of a family history in our patients and those previously described. In addition, the normal ERG results in case 3 precluded a diagnosis of Vogt-Koyanagi-Harada or paraneoplastic syndrome. Autofluorescence imaging is likely to be of help in the diagnosis and monitoring of acute exudative polymorphous vitelliform maculopathy and may further the understanding of its pathophysiology.

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