In this study, only 34.5% of the candidates used AREDS-related supplements, which is far lower than in the previous report in the United States (67%). This was in contrast to the high level of adherence among the candidates (48 of 78 participants [61.5%]; no significant difference according to age or sex). It should be noted that the instruction rate of the ophthalmologists was more critical than the adherence rate of the patients for taking supplements. Interestingly, AREDS plus lutein supplements were used more often than AREDS-like supplements in this study, in contrast to a US-based study. A recent survey in a Japanese cohort reporting the protective association of the serum levels of carotenoids with AMD and studies in animal models showing the tissue-protecting effects of lutein as a blue-light filter and antioxidant may have promoted the use of AREDS plus lutein supplements.

The ophthalmologists did not always recommend supplements in accordance with AREDS. Some ophthalmologists were not completely confident that the AREDS results are applicable to Japanese or other Asian patients with AMD because of the differences in the clinical features of Asian and white patients with AMD.

The increased incidence of AMD and the resulting vision loss are now serious issues. Preventive therapy is important for both patients’ personal health and their continued contributions to society. Reliable evidence based on a randomized controlled trial is still needed for ophthalmologists to appropriately instruct patients’ use of supplements to prevent the progression of AMD in Japan.

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Novel Clinical Manifestation of Congenital X-Linked Retinoschisis

Tradiitionally, congenital X-linked retinoschisis (CXLRS) has been defined as a juvenile macular dystrophy. It is characterized by foveal schisis accompanied by an abnormally reduced b-wave on electroretinography. The gene for CXLRS, RS1, has been mapped to the short arm of the X chromosome and encodes for the protein retinoschisin. Schisis spaces, visible on optical coherence tomography (OCT), have been reported in the fovea in virtually all cases. We report a case of CXLRS followed up for 6 years with no evidence of foveal schisis on OCT and excellently maintained central vision.

Report of a Case. The patient was referred to the Associated Retinal Consultants, Royal Oak, Michigan, in 2004 at age 9 years. At this time, he had a 2-year history of recurrent vitreous hemorrhages in the left eye with a recent spontaneous vitreous hemorrhage in the right eye. His visual acuity was 20/30 OU. Dilated fundus examination revealed bilateral nasal dragging of his discs, absent foveal reflexes, and bilateral peripheral schisis cavities (Figure 1). A diagnosis of probable CXLRS was given, and the patient was scheduled for electroretinography as well as an examination under anesthesia with OCT. His OCT findings confirmed the presence of bilateral peripheral schisis cavities, and electroretinography showed decreased a- and b-waves. A detailed family history revealed a maternal grandfather diagnosed as having retinitis pigmentosa, which likely represents a misdiagnosed case of CXLRS. Unfortunately, it was not possible to examine the patient’s grandfather to confirm or disprove this
The patient was followed up at regular 6-month intervals with serial clinical examinations and OCT scans. Vision and funduscopic examination findings remained stable. In 2010, his peripheral blood was screened for mutations in RS1. This test showed a positive missense mutation, R102Q (CGG to CAG), on exon 4 of RS1. At his most recent follow-up visit in October 2010, his visual acuity was 20/25−1 OD and 20/20−1 OS. Spectral-domain OCT showed redemonstration of peripheral schisis cavities but failed to show any detectable evidence of the foveal cystic changes that have been considered universal in CXLRS (Figure 2). Central foveal thickness was normal at 266 µm OD and 276 µm OS. A return visit was scheduled for 6 months later.

Comment. We report a case with the classic diagnostic criteria for CXLRS: abnormal electroretinographic findings, clinically evident peripheral schisis cavities with associated visual field abnormalities, and confirmed mutation in RS1. The R102Q mutation in RS1 has previously been reported in clinically diagnosed cases of CXLRS. This case is unique in that the patient has failed to develop foveal involvement during the 6 years in which he has been closely followed up. The novel clinical mani-
festation of this case underscores the need for a formal genetic evaluation in any child with suspected genetically controlled retinal pathology. In the updated classification system for CXLRS inclusive of OCT findings, all 4 subtypes demonstrate foveal cystic schisis.\(^1,2\) Although this patient has multiple features of CXLRS, his lack of this critical finding places him outside this previously described classification system. It has been observed that children with CXLRS may not have clinical foveal changes at birth, but they usually manifest by age 8 years in our experience. The clinical picture of this case is somewhat of an outlier. This case may represent a fifth type of CXLRS with peripheral schisis and no associated macular lamellar schisis or foveal cystic schisis.

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Idiopathic Retinal Vasculitis, Aneurysms, and Neuroretinitis in a Patient With Antiphospholipid Syndrome

We report a case of idiopathic retinal vasculitis, aneurysms, and neuroretinitis (IRVAN) in a patient with antiphospholipid syndrome.

Report of a Case. A 76-year-old white man had slightly blurred vision and visual acuity of 20/30 OU. Slitlamp examination showed no anterior chamber inflammation and rare cells in the anterior vitreous of both eyes. Funduscopy examination (Figure 1) showed hard exudates and intraretinal hemorrhages. Fluorescein angiography (Figure 2) emphasized the multiple arteriolar aneurysms, most prominent at bifurcations. Leakage from and staining of both arterial and venous vessels as well as the optic nerve is best appreciated in the right eye.

His history was significant for a seizure disorder treated with phenytoin sodium for 25 years and coronary artery disease treated with a stent 1 year prior to our examination. He has lived in Kenya, Uganda, and the West Bank and has been treated for malaria. He also had occipital-lobe stroke 16 years earlier, which was thought to be related to a patent foramen ovale. A patent foramen ovale was not noted on a transthoracic echocardiogram. He has been receiving warfarin sodium continuously since the stroke, with a goal international normalized ratio of 2.5 to 3.5.

A review of his previous ophthalmology records showed at least a 7-year history of vitritis, retinal vasculitis, and hard exudates more prominent in his right eye. During this time, no ocular treatment was performed.

Laboratory and imaging tests with results within normal limits included complete blood cell count, metabolic panel, liver enzymes, angiotensin-converting enzyme, erythrocyte sedimentation rate, hepatitis B and C antibodies, fluorescent treponemal antibody absorption, rapid plasma reagin, *Bartonella henselae* and *Bartonella quintana* IgG and IgM, *Toxoplasma gondii* IgM, an-

Figure 1. Fundus photographs of the right (A) and left (B) eyes showing vascular sheathing, hard exudates, intraretinal hemorrhages, and aneurysms.