We have shown that cone dystrophy frequently accompanies fundus albipunctatus in elderly patients with *RDH5* mutations.[3] In such cases, full-field photopic electroretinograms are severely reduced, a bull's-eye maculopathy is present, and visual acuity and fields are impaired.[3,4] We concluded that mutations of the *RDH5* lead to a progressive cone dystrophy resulting in severe loss of visual function with aging.[3]

To date, no fundus albipunctatus case with a maculopathy has been reported in a child. We describe a 9-year-old Japanese boy who presented with a 2-year history of night blindness with no family history of retinal diseases. Multiple discrete yellow-white dots were observed at the level of retinal pigment epithelium with scarring of the macula, in both eyes (Figure 1A). The eyes were otherwise normal.

Full-field electroretinograms elicited by Ganzfeld stimuli after 20 minutes of dark adaptation showed no scotopic electroretinograms and "negative-shaped" bright-flash electroretinograms. Normal scotopic b-wave and bright-flash electroretinograms were recorded after 3 hours of dark adaptation. Photopic electroretinograms were normal (Figure 2A).

Molecular genetic examination revealed a compound heterozygous mutation, a T to C mutation at nucleotide 841 (Tyr281His), and a C deletion at nucleotide 928 with insertion of GAAG (Leu310GluVal) in *RDH5*. His normal father showed a heterozygous mutation at nucleotide 841 and was normal at nucleotide 928. No such base substitutions were recognized in 100 alleles from normal individuals.

Unexpectedly for a young patient with fundus albipunctatus,[3] his fundi demonstrated symmetric atrophic lesions in the macula in both eyes (Figure 1A), and his corrected visual acuity was RE: 0.5 and LE: 0.3. Fluorescein angiography was normal in the macula (Figure 1B). Focal cone macular electroretinograms[5] (time constant, 0.03 seconds) demonstrated that the a- and b-waves were absent from 5 degrees and 10 degrees, and significantly reduced for 15° of the macula (Figure 2B). Because the function of his retina was deteriorated especially in the macula, he was found to have macular dystrophy and fundus albipunctatus.

This case is different from previously reported elderly patients[3] because the patient did not show a bull's eye but a foveal atrophy. He was not affected by general cone dystrophy because the full-field photopic electroretinograms were normal. We cannot determine whether the macular dystrophy is caused by a phenotypic variation of fundus albipunctatus or by a chance combination with fundus albipunctatus, however, we suggest that the macular dystrophy is caused by *RDH5* mutation because many elderly patients with *RDH5* mutations develop maculopathy.[3] Although the two mutations of Tyr281His and Leu310GluVal have been reported in other Japanese cases of fundus albipunctatus,[3] no case with this combination of a compound heterozygous mutation has been reported. It is not clear whether the phenotype in this case resulted from the genotype, and further data will be necessary to understand the clinical features and the genotype–phenotype correlation with the *RDH5* mutations.

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Spontaneous Reopening of a Spontaneously Closed Macular Hole

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PURPOSE: To report a case of spontaneous reopening after spontaneous closure of a full-thickness macular hole.
METHODS: Observational case report. Retrospective clinical practice case review.
RESULTS: A 57-year-old man with a full-thickness macular hole in his left eye developed spontaneous closure for 1.5 years with improved vision, followed by spontaneous reopening of the hole with loss of vision. Surgical repair resulted in repeat closure and recovery of 20/20 visual acuity.
CONCLUSION: Spontaneous reopening, which occasionally occurs after surgical closure of macular holes, can also occur after spontaneous closure of a macular hole.

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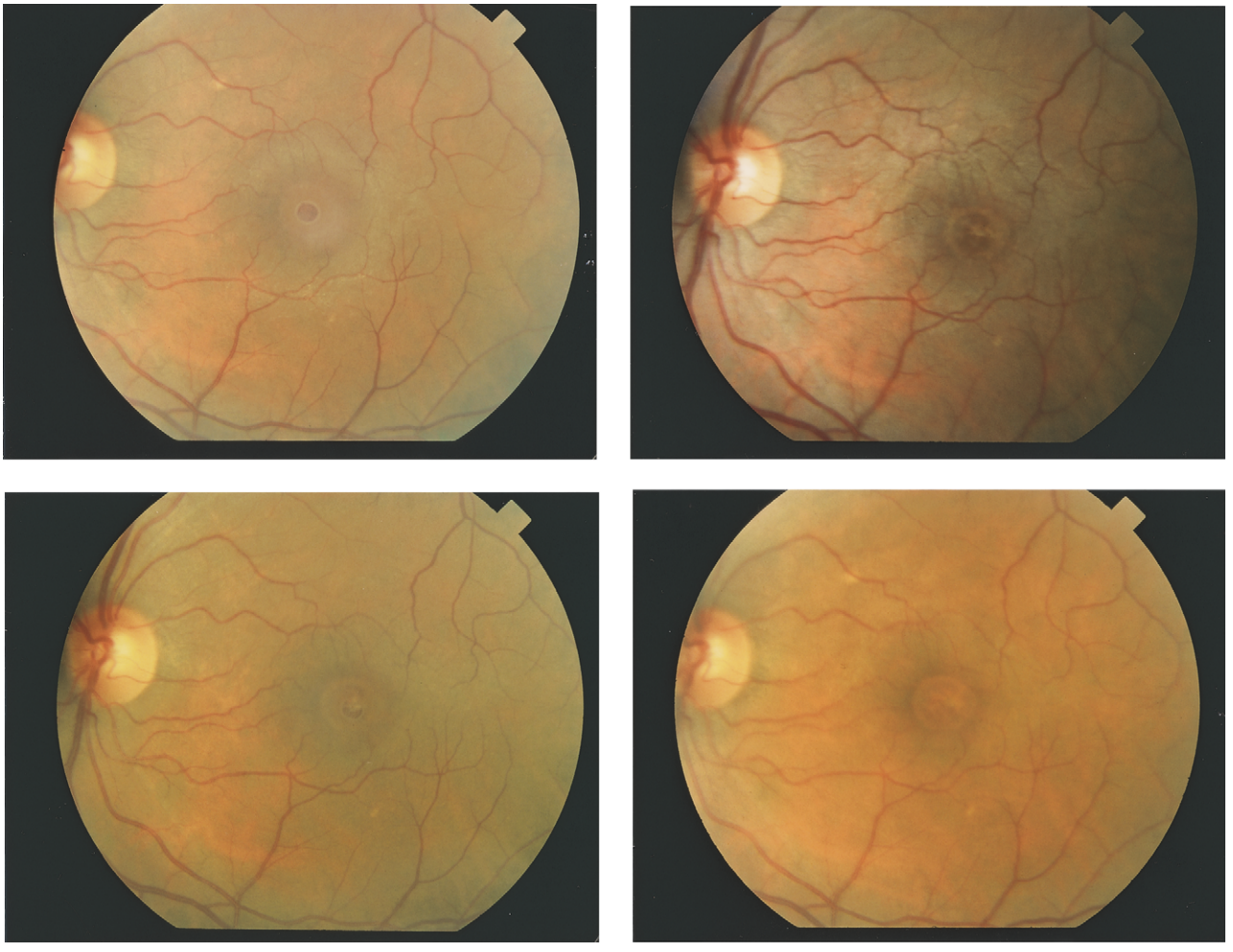

FIGURE 1. (Top left) Fundus photograph shows full-thickness macular hole and cuff of subretinal fluid. Best-corrected visual acuity is 20/70. (Top right) Fundus photograph shows spontaneous closure of macular hole with 20/40 vision. Note the glial plug and perifoveal pigmentary changes. (Bottom left) Fundus photograph shows spontaneous reopening of macular hole with cuff of subretinal fluid. Note the residual glial tissue superiorly within the macular hole. Visual acuity decreased to 20/60. (Bottom right) Fundus photograph shows closure of macular hole postoperatively. Visual acuity recovered to 20/20.

SPONTANEOUS REOPENING AFTER SURGICAL CLOSURE of full-thickness macular holes occurs in approximately 5% of successfully closed macular holes.[1–3] Spontaneous closure of a full-thickness macular hole occurs uncommonly, but it has been observed in 3% to 5% of macular hole series.[4] To our knowledge, this is the first reported case of a spontaneous reopening of a full-thickness macular hole that initially spontaneously closed.

• **CASE**: A 57-year-old man presented with a 6-month history of blurred vision in his left eye. His right eye was asymptomatic on initial presentation. On ocular examination, his best corrected visual acuity was 20/20, J1, in the right eye and 20/70 (+0.25 + 0.50 × 178), J7, in the left eye. Amsler grid revealed a central scotoma in the left eye. Contact lens biomicroscopy of the left eye showed a full-thickness 350-μm macular hole, 200-μm cuff of subretinal fluid, parafoveal cystic changes, and an overlying pseudo-operculum (Figure 1, top left). The patient noted a complete break in a slit-lamp beam projected across the hole (Watzke–Allen sign). The patient declined surgery in the left eye. The macular hole enlarged to 500 μm 1 year after presentation. Two years after initial presentation, the macular hole spontaneously closed with improvement in visual acuity to 20/40 (Figure 1, top right). One-and-a-half years after spontaneous closure, the patient's visual acuity decreased to 20/60. A recurrent, full-thickness 300-μm macular hole with surrounding subretinal fluid and epiretinal membrane was noted (Figure 1, bottom left). Pars plana vitrectomy, membrane peeling, and fluid–gas ex-

change with 18% perfluoropropane C_3F_8 were performed 4 months later with closure of the hole and recovery of stable 20/20 visual acuity for 4 years (Figure 1, bottom right).

The fellow right eye developed a stage Ib macular hole 7 months after initial presentation with decreased best-corrected visual acuity to 20/50. Pars plana vitrectomy, peeling of the posterior hyaloid, and fluid–gas exchange with sulfur hexafluoride (SF_6) was performed. The postoperative course was complicated by cataract, retinal detachment, and spontaneous reopening of the macular hole. After surgical repair, visual acuity has been stable at 20/20 for 7 years.

Spontaneous reopening occurs infrequently after successful macular hole closure after surgery.[1–3] The origin of spontaneous reopening is uncertain, but possible mechanisms include progressive epiretinal membrane formation resulting in tractional forces reopening the hole; residual cortical vitreous on the macula resulting in recurrent vitreofoveal traction; cystoid macular edema after subsequent cataract surgery; and internal retinal elasticity.[1–3] This case demonstrates that reopening, which occurs after surgical closure of macular holes, can also occur after a spontaneous macular hole closure.

Spontaneous closure of macular holes is uncommon.[4,5] Possible mechanisms for spontaneous closure[4,5] include spontaneous relief of vitreofoveal traction, bridging glial proliferation, and a protrusion of retinal tissue from the borders of the macular hole, as recently imaged with optical coherence tomography imaging.[5] In the above-reported case, glial proliferation was noted after spontaneous macular hole closure (Figure 1, top right). Possible mechanisms for reopening after this spontaneous closure include progressive glial proliferation resulting in tractional forces reopening the edges of the hole, recurrent vitreofoveal traction, or internal retinal elasticity overcoming the forces of glial proliferation closing the hole. Spontaneous reopening has also been reported after resolution of stage I or impending macular hole,[2] as occurred in the fellow eye of this case. Both eyes of this patient recovered excellent vision after surgery for reopened full-thickness macular holes.

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Intraoperative Choroidal Hemorrhage in the Osler–Rendu–Weber Syndrome

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PURPOSE: To describe a patient with Osler–Rendu–Weber syndrome who developed a nonsimultaneous intraoperative choroidal hemorrhage in each eye.

METHOD: Interventional case report. A 65-year-old Caucasian woman with Osler–Rendu–Weber syndrome developed a choroidal hemorrhage in the left eye during vitrectomy for a complicated retinal detachment with a poor visual outcome. Fifteen years later, she developed a macula on retinal detachment in the right eye, which also had a dense cataract. Immediately after uncomplicated phacoemulsification and intraocular lens implantation, under monitored anesthesia care and retrobulbar block, and without valsalva stress, ophthalmoscopy demonstrated a choroidal hemorrhage. A planned scleral buckle was replaced by pneumatic retinopexy.

RESULTS: Seven months postoperatively, the retina remained attached with resolution of the choroidal hemorrhage. Visual acuity was 20/30.

CONCLUSION: Choroidal hemorrhage may occur more commonly in individuals with Osler–Rendu–Weber syndrome. Recognition of this possible association and institution of appropriate intraoperative precautions may facilitate a good visual outcome. (Am J Ophthalmol 2002;133:282–284.

OSLER–RENDU–WEBER SYNDROME OR HEREDITARY hemorrhagic telangiectasia is a rare, autosomal dominant disease, characterized by widespread vascular anomalies comprising dilated thin-walled capillaries and venules.[1] Affected individuals may have recurrent epistaxis and spontaneous bleeding from different organs throughout the body.[1] Conjunctival and retinal telangiectasia have been described.[2–4] We are unaware of reports identifying choroidal hemorrhage associated with the Osler–Rendu–Weber syndrome and could find no reference to it in a computer MEDLINE search. We report a case of intraoperative choroidal hemorrhage in each eye of an individual with Osler–Rendu–Weber syndrome.

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