muscular dystrophy and muscle-eye-brain disease with associated ONH and retinal hypoplasia. A case of genetically proven muscle-eye-brain disease with ONH and peripheral retinal nonperfusion with secondary fibrovascular proliferation and retinal detachment was recently described. Our patient shares many similar features but had no evidence of a muscular dystrophy, with clinically absent hypotonia and a normal creatine kinase level. Additionally, a case of de Morsier syndrome with similar ocular findings of bilateral peripheral retinal nonperfusion and neovascularization with resultant retinal detachment was reported recently. We suggest that the abnormal neuronal development that led to lissencephaly resulted in optic nerve and retinal maldevelopment and subsequent associated retinal vascular maldevelopment. Tractional retinal detachment is an end stage of a process that starts with nonperfusion and progresses to ischemia and extraretinal fibrovascular proliferation in a variety of pediatric retinal diseases.

Although the cause of neural maldevelopment in our patient remains elusive, her case supports the idea that patients with severe neuronal migration deficits should be evaluated for ONH and abnormal retinal vasculature development, even without evidence of a muscular dystrophy.

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Subclinical Photoreceptor Disruption in Response to Severe Head Trauma

C ommotio retinae is a transient opacification of the retina due to outer retinal disruption occurring in a contrecoup fashion after blunt trauma. Histological studies in animals and humans after ocular blunt trauma have revealed that disruption occurs at the level of the photoreceptor outer segments and retinal pigment epithelium. Recent reports using optical coherence tomography (OCT) have shown detectable disruption at the level of the photoreceptor inner segment/outer segment junction and retinal pigment epithelium and that these changes may be reversible over time with restoration of normal outer retinal architecture. However, the resolution of existing OCT technology may not be sensitive enough to detect photoreceptor disruption. Adaptive optics (AO) imaging systems enable cellular-resolution imaging of the human retina, and there is a growing number of cases where deficits have been visible on AO images but not on OCT. Herein, we report a case of subclinical photoreceptor disruption after head trauma as seen by an AO scanning ophthalmoscope (AOSO) but not apparent clinically or on spectral-domain OCT (SD-OCT).

Report of a Case. A 43-year-old man described a 5-year history of a stable, crescent-shaped purple scotoma nasal to central fixation in his right eye that developed after an industrial accident in which he sustained significant head and body trauma. A complete ophthalmic examination revealed best-corrected visual acuity of 20/20 OU and no remarkable fundus findings or abnormalities. Fluorescein angiography and SD-OCT (Spectralis SD-OCT; Heidelberg Engineering) findings were unremarkable (Figure 1). Humphrey visual field 10-2 testing and microperimetry revealed a small nonspecific area of functional vision loss near fixation in the right eye. Images of the photoreceptor mosaic near the fovea were acquired using a newly developed AOSO. Images were processed and registered using custom Matlab software (MathWorks). While foveal cone density was normal, the AOSO images revealed a well-defined crescent-shaped area of photoreceptor disruption just temporal to the fovea (Figure 2A, large arrows). Other focal areas of photoreceptor irregularities were also seen superior, temporal, and inferotemporal to the fovea (Figure 2A, small arrows). Both cone and rod photoreceptors were visualized with this AOSO imaging, and both cell types appeared to be disrupted (Figure 2A and B).

Comment. The AOSO detected photoreceptor disruption resulting from head trauma and not apparent clinically or by other standard imaging modalities, including SD-OCT. Restoration of the outer retinal appearance in SD-OCT has been reported after commotio retinae, suggesting recovery of the outer retinal structure. Our data demonstrate that photoreceptor disruption may still exist. The SD-OCT axial resolution is likely not sensitive enough to reveal the full extent of photoreceptor disruption that may occur after ocular or head trauma. The
Figure 1. Clinical imaging of the right eye. A, Color fundus photograph shows no macular abnormalities. B, Late-frame fluorescein angiogram shows no window defect, staining, or leakage. White box indicates the area imaged by the adaptive optics scanning ophthalmoscope as seen in Figure 2. C, Spectralis spectral-domain optical coherence tomographic horizontal scan through the fovea shows no outer retinal abnormalities. Area between arrows indicates the region imaged by the adaptive optics scanning ophthalmoscope as seen in Figure 2.

Figure 2. Disrupted photoreceptor mosaic of the macula in the right eye. A, Adaptive optics scanning ophthalmoscope montage shows a large, crescent-shaped area of photoreceptor disruption (edges indicated by large arrows) temporal to the fovea. Other areas of photoreceptor disruption are also present (small arrows). The foveal center was not imaged (solid white rectangle). B, Magnified view of a patch of retina 1° temporal from the fovea, centered on an area of significant photoreceptor disruption. C, Magnified view of a patch of retina 1° nasal from the fovea, showing a regularly packed cone photoreceptor mosaic. D, Image from a healthy control subject, about 1° temporal from the fovea. Scale bars=50 µm.
AOSO imaging may prove useful in improved detection and understanding of photoreceptor involvement in ocular or head trauma. In addition, patients with traumatic brain injury often report visual symptoms. The AOSO may be of value to help differentiate retinal vs cortical contributions to vision loss in these patients.

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The postoperative complication rates stated by the authors are lower than those in other significant studies.2-4 The high number of patients lost to follow-up could easily mask a higher complication rate, and the results must be interpreted in this context.

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In reply

We thank Patel and colleagues for their interest in our study.1 In our article, we did not report the number of patients who had lost 2 or more Snellen lines of visual acuity as this will be examined as part of future work looking at surgical outcomes such as visual acuity and intraocular pressure. We...