Case report

Photoreceptor disruption and vision loss associated with central serous retinopathy

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Purpose: To present ophthalmic imaging findings in the case of a 40-year-old male with sustained visual loss after a single episode of acute central serous retinopathy (CSR).

Observations: A male subject presented with visual acuity decline to 20/50 OS and was diagnosed with acute CSR. The initial pigment epithelial detachment and subretinal fluid resolved within 6 weeks, but visual acuity remained impaired. Using directional optical coherence tomography (D-OCT) and confocal and split-detector adaptive optics scanning light ophthalmoscopy (AOSLO), we imaged pathologic alterations in the photoreceptor mosaic of the affected eye. A foveal region of intermittent missing cones, a temporal parafoveal region of confluent missing cones, and a nasal parafoveal region of misdirected cones were observed.

Conclusions and Importance: Pathologic alterations in photoreceptor microanatomy underlie residual visual acuity deficits in this case of acute CSR. Observations of missing cones correlated well across all imaging modalities in the fovea and the temporal parafoveal region of missing cones. However, in the nasal parafovea where cones were present but misdirected, D-OCT and AOSLO may be able to identify and image photoreceptors with greater fidelity as compared to non-directional SDOCT (spectral domain OCT). D-OCT may thus have a clinical role in rapidly assessing photoreceptor mosaic integrity in pathology.

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1. Introduction

Central serous retinopathy (CSR) is characterized by idiopathic pigment epithelial detachments (PED) and localized pockets of subretinal fluid (SRF). CSR typically resolves within three months, but rare cases of permanent vision loss may reflect photoreceptor mosaic changes visible with advanced ophthalmic imaging techniques. Confocal adaptive optics scanning light ophthalmoscopy (AOSLO) images cones with intact and aligned outer segments, previously uncovering both patchy and confluent areas of reduced or absent reflectivity in CSR. Split-detector AOSLO utilizes multiply-scattered light to image cone inner segments, unambiguously differentiating misdirected cones from missing cones. High-resolution spectral domain optical coherence tomography (SDOCT) can distinguish cone outer segment tips (COST) from rod outer segment tips (ROST). Additionally, directional OCT (D-OCT) utilizes multiple angles of illumination to infer photoreceptor orientation. Using these modalities in combination, we characterized photoreceptor disruption in a patient with sustained visual loss after acute CSR resolution.

2. Case report

A 40-year-old male presented with decreased vision in the left eye. Best-corrected visual acuity (BCVA) OS was 20/50 with examination and SDOCT imaging demonstrating moderate subretinal fluid (SRF) and a small pigment epithelial detachment (PED). OD was 20/20 with a normal exam and imaging. The patient was diagnosed with CSR. Six weeks later, the PED and SRF had spontaneously resolved; however, VA had not improved.

After informed consent was obtained, the patient was imaged with SDOCT, D-OCT, and confocal and split-detector AOSLO. The photoreceptor mosaic disruption was observed in all modalities.
Foveally (Fig. 1), confocal and split-detector AOSLO revealed a patchy pattern of both outer and inner cone segment loss closely correlating with COST dropout seen on the en face SDOCT, suggesting loss of individual photoreceptors. Parafoveally (Fig. 2), borders of the initial PED coincided with apparent COST dropout on SDOCT best seen en face. In the temporal parafovea, this border region corresponded to cone loss seen on AOSLO and is surrounded by a penumbra of present but...
misdirected cones. The nasal parafovea, however, revealed intermodal discrepancy between SDOCT and AOSLO imaging, which revealed an intact and reflective cone mosaic. D-OCT imaging of this region demonstrated off-axis COST reflectivity suggesting present, but misaligned, photoreceptors.

3. Discussion

Multimodal imaging uncovered significant photoreceptor mosaic defects underlying the patient’s sustained vision loss after the resolution of an acute episode of CSR. Foveal AOSLO showed patchy loss of both confocal reflectivity and split detector inner segments, suggesting absence rather than misalignment of photoreceptors. These gaps in the cone mosaic closely corresponded to COST dropout on SDOCT.

With the additional insight gained from AOSLO imaging in this patient, it is apparent that standard SDOCT images taken from a single pupil entry position would overestimate photoreceptor disruption in the parafovea. Unfortunately, the clinical utility of AOSLO is at present limited by financial, technical and time considerations. In cases with photoreceptor misalignment, D-OCT may provide a relatively rapid, simple, inexpensive, and clinically applicable assessment of photoreceptor integrity using existing clinical SDOCT instrumentation.

The relationship between photoreceptor integrity, orientation, and function was not assessed in this patient. While the anatomical presence of photoreceptors can be identified, it would be interesting to perform perimetric studies on the penumbral region of misdirected cones found outside the initial lesion in this patient.

Fig. 2. Directional OCT and AOSLO accurately characterizes parafoveal transition zones. Numbered arrows indicate the approximately corresponding areas, allowing for minor variations in eye fixation and rotation between imaging sessions. A: 3 × 3 mm en face Bioptigen spectral domain optical coherence tomography (SDOCT) of the cone outer segment tips (COST) band of a central serous retinopathy (CSR)-affected region. Fixation was deliberately offset during image acquisition, and the anatomic fovea is near Arrow 2. B: averaged 3 mm Bioptigen SDOCT line scan through the fovea. 1a–5a: confocal adaptive optics scanning light ophthalmoscopy (AOSLO) images of the photoreceptor layer; reflectivity is thought to originate from waveguiding cone outer segments. 1b–5b: split-detector AOSLO images of the photoreceptor layer; images are thought to result from light scattering from cone inner segments. Patchy gaps in the cone mosaic can be seen foveally (2a–b), while a confluent region of cone loss found at the temporal border of the CSR-affected region (3a–b). A penumbral region of misdirected cones is also seen (4a–b). In contrast, the temporal retina demonstrates a region of absent COST reflectivity independent of scan angle (C–F, Arrow 3). Scale bars: Panel A, 100 μm. Panel B, lateral 100 μm, axial 100 μm. Panels 1a–5b, 30 μm. Panels C–F, lateral 200 μm, axial 200 μm.
Future studies may elucidate whether these may represent cones with full visual potential.

4. Conclusions

Although most cases of acute CSR resolve without significant visual loss, permanent deficits have been noted in some cases, including the one presented in this report. Using advanced ophthalmic imaging techniques including D-OCT and confocal and split-detector AOSLO, we characterized pathologic alterations in photoreceptor microanatomy. Both missing and poorly reflective cones were identified, with the latter potentially representing misaligned cones. Observed changes correlated well across all imaging modalities in the fovea, while in the parafovea D-OCT and AOSLO might assess photoreceptor integrity with greater accuracy as compared to non-directional SD-OCT.

Patient consent

Research was conducted under an IRB-approved protocol, and written informed consent was obtained from the subject prior to the collection of any data. This report does not contain any patient identifiers.

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Conflicts of interest

L.W. Sun, None; J. Carroll, Co-inventor D-OCT; B.J. Lujan, Co-inventor D-OCT, UC Berkeley.

Authorship

All authors attest that they meet the current ICMJE criteria for Authorship.

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