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OCT Angiography Imaging in Serpiginous Choroidopathy

Ria Desai, MD, Peter Nesper, BA, Debra A. Goldstein, MD, Amani A. Fawzi, MD, Lee M. Jampol, MD, Manjot Gill, MD

Purpose: To report OCTA findings in 3 cases, 2 active and 1 inactive, of serpiginous choroidopathy (SC) and describe OCTA changes in response to treatment.

Design: Retrospective case series.

Participants: We studied 6 eyes of 3 patients with SC.

Methods: Retrospective case series of 3 patients with SC undergoing multimodal imaging, including OCTA. In 1 treated eye, both pre- and posttreatment images were compared.

Main Outcome Measures: Description of OCTA findings in patients with SC.

Results: In the active phase, OCTA images show an apparent absence of the choriocapillaris with variable outer retinal and retinal pigment epithelial thickening. After treatment, OCTA of previously active lesions demonstrates a partial reappearance of the choriocapillaris, especially at lesion margins. In inactive SC, the choriocapillaris, along with the retinal pigment epithelium and outer retina, is notably absent.

Conclusions: Optical coherence tomography angiography suggests absence of choriocapillaris in both active and inactive phases of SC with partial reestablishment following treatment. Although the exact pathogenesis of SC is not elucidated by these findings, OCTA images allow us to better evaluate choroidal involvement. *Ophthalmology Retina 2017;■:1–9 Published by Elsevier Inc. on behalf of the American Academy of Ophthalmology*

Serpiginous choroiditis (SC) is a well-known, but uncommon, usually bilateral disease process involving the retina, retinal pigment epithelium (RPE), and choroid.[1,2] In its active form, the disease presents with sharply demarcated, flat, gray-yellow lesions involving the outer retina and RPE.[1,3] Classically, these lesions are distributed in a peripapillary pattern.[1–3] A variant of SC, macular serpiginous, presents with similar lesions located in the central macula.[2–4] In both presentations, SC lesions transform into areas of chorioretinal atrophy within weeks.[1,2]

The natural history of SC is characterized by recurrence, typically adjacent to atrophic scars.[1,5,6] The presence of foveal involvement and the development of choroidal neovascularization (CNV), is associated with poor visual prognosis.[7]

The etiology of serpiginous choroiditis remains unknown. Late stages of SC demonstrate atrophy involving choriocapillaris, RPE, and photoreceptors,[1–3,5] yet debate regarding pathogenesis remains. Some argue for an inflammatory process at the level of the outer retina and RPE or choroid.[3,6,8–10] Others believe that the disease is primarily vaso-occlusive, resulting in choroidal nonperfusion[4] with secondary involvement of the outer retina and RPE.[3,9,11–13] Treatment is directed toward control of active inflammation via systemic or local immunosuppression.[1,6]

Various imaging modalities have been used in an attempt to identify the pathological changes associated with active SC. The most commonly employed are fluorescein angiography (FA) and indocyanine green angiography (ICGA). Both of these modalities involve the injection of IV agents, and neither modality allows visualization of the outer retina, RPE, and choriocapillaris simultaneously in 3 dimensions.

Optical coherence tomography angiography has been developed to simultaneously obtain structural images of the retina and assess retinal and choroidal vasculature without IV injection. In OCTA, decorrelation signals are generated by variable signal intensities and amplitudes obtained from sequential B scans at a single posterior segment location.[14,15] The variability of these signals results from the movement of intravascular blood cells contrasting with stationary adjacent tissues.[14,15] The generated signals can then be processed to create maps of the choroid and retina. B scans showing the retinal layers can then be viewed along with en face images. This imaging modality is ideal for SC, as it allows physicians to view outer retinal and choroidal pathology with a single technique.

Materials and Methods

Six eyes of 3 patients with SC were evaluated with ophthalmic examination, and multi-modal imaging including FA, ICGA, spectral-domain OCT (SD-OCT) and OCTA between August 13, 2015, and April 19, 2016. Of the 3 patients, 2 had clinically active disease and 1 had inactive disease. Disease activity was based on clinical examination and FA results.

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Optical coherence tomography angiography images were obtained using the RTVue-XR Avanti system (Optovue, Fremont, CA) with split-spectrum amplitude-decorrelation angiography (SSADA) software. This instrument has an A-scan rate of 70,000 scans per second and uses a light source centered on 840 nm with a bandwidth of 45 nm. A 3 × 3-mm scanning area centered on the lesion of interest on corresponding FA or SD-OCT was obtained in approximately 3 seconds.[15] Two consecutive B-scans (M-B frames), each containing 304 A-scans, were captured at each sampling location and SSADA was used to extract OCTA information. En face OCT angiograms were segmented using the built-in software algorithm to define the superficial and deep capillary plexuses, as well as the outer retina and choriocapillaris.

Areas of disease activity and inactivity were noted clinically and correlated with OCTA findings. These findings were then compared with those from other imaging modalities. OCTA images were obtained before and after treatment in 1 patient with active disease.

Institutional review board approval was obtained for imaging. The research adhered to the tenets of the Declaration of Helsinki. Data collection in this study was compliant with the Health Insurance Portability and Accountability Act of 1996.

Results

Case 1: Active SC

A 38-year-old male patient with a history of right eye "retinal atrophy" and myopia in both eyes presented with acute decreased vision in the right eye for 5 days. Initial best-corrected visual acuity (BCVA) was 20/400 (right eye) and 20/20 (left eye). Results of a slit-lamp examination of the right eye were unremarkable except for 1+ anterior vitreous cell. Fundus examination revealed 2 active creamy yellow-gray macular lesions contiguous with chorioretinal scarring extending in a peripapillary fashion (Fig 1). Spectral-domain OCT (Spectralis, Heidelberg Engineering, Heidelberg, Germany) images *showed* significant disruption and swelling of the outer retinal layers and RPE in the active lesion (Fig 1). In the left eye, results of a fundus examination and imaging were unremarkable.

Baseline FA and ICGA were obtained at presentation (Fig 2). FA, transit right eye, showed early hypofluorescence of the margins of the more temporal lesion and diffuse hypofluorescence of the larger central lesion. Both active lesions demonstrated late central hyperfluorescence consistent with staining. The areas of chorioretinal atrophy demonstrated late staining of the margins without any identifiable leakage. Early- and late-phase ICGA images showed marked hypofluorescence (hypocyanescence) corresponding with areas of clinically active disease.

OCTA images demonstrated a notable absence of choriocapillaris and large choroidal vessels in both active lesions. OCTA B scans corresponding with the en face images showed that areas of choriocapillaris and choroidal absence correlate with overlying areas of outer retinal and RPE swelling (Figs 3 and 4).

Results of QuantiFERON Gold (Qiagen, Hilden, Germany) and tuberculin skin testing were negative. The patient's right eye was treated with 4 mg of intravitreal Triesence (triamcinolone acetonide injectable suspension; Alcon, Fort Worth, TX). Five days after treatment, BCVA improved to 20/70 and fundus examination show increased pigmentation, with early atrophy, of both lesions. SD-OCT revealed decreased outer retinal and RPE swelling and disruption at the site of both previously active lesions (Fig 5).

The patient was followed with lengthening intervals and noted to have continued improvement in right eye BCVA. Fundus examination continued to show progressive atrophy. At the most recent follow-up, 7 months after presentation, BCVA was 20/20. SD-OCT showed no outer retinal or RPE swelling, with reorganization of the outer retinal layers, including partial reappearance of the subfoveal ellipsoid layer.

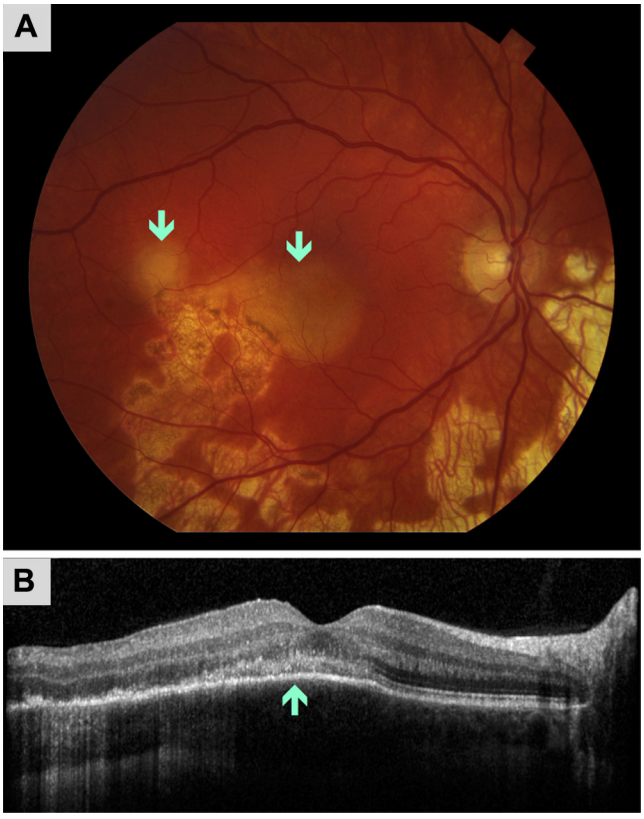

Figure 1. Case 1: A 38-year-old male patient at presentation **A**, Fundus photograph of the right eye with *arrows* pointing to 2 macular *yellow-gray* lesions, 1 central and the other temporal, extending from areas of chorioretinal atrophy. **B**, Spectral domain OCT imaging of the right eye demonstrating subfoveal outer retinal and retinal pigment epithelial thickening (*arrow*) through the active central lesion.

At follow-up visits, OCTA images of the 2 previously active lesions were obtained and showed progressive emergence of large choroidal vessels on en face angiograms corresponding with a decrease in outer retinal and RPE swelling on accompanying B scans (Figs 6 and 7). Of note, the large central lesion initially demonstrates the reappearance of large choroidal vessels and choriocapillaris at the lesion edges followed by the reappearance of these vessels within the lesion center (Fig 6).

In the temporal (more subacute) lesion, sequential OCTA images again show the progressive reappearance of both choriocapillaris and large choroidal vessels with a corresponding decrease in outer retinal and RPE edema. Unlike the larger central lesion, even at the most recent 7-month visit, the choriocapillaris reappearance here is limited to the margins of the lesion. Despite a demonstrated decrease in outer retinal and RPE thickening, there remains a notable absence of choriocapillaris in the center of the lesion.

Case 2: Active, Macular SC

A 59-year-old male patient with a history of macular choroiditis in the right eye, diagnosed 4 years previously, presented with a