Characterization of Birdshot Chorioretinopathy Using Extramacular Enhanced Depth Optical Coherence Tomography

Pearse A. Keane, MD, MRCOphth; Musarrat Allie, BSc; Stephen J. Turner, FRCOphth; H. Sue Southworth, BSc, MSc, RN; Srinivas R. Sadda, MD; Philip I. Murray, FRCOphth, PhD; Alastair K. Denniston, FRCOphth, PhD

Objective: To combine “extramacular” and “enhanced depth” optical coherence tomographic (OCT) scanning protocols to facilitate enhanced characterization of patients with birdshot chorioretinopathy.

Methods: Spectral-domain OCT images were prospectively collected from 24 eyes of 12 patients with birdshot chorioretinopathy. The images were acquired both from the macula and from 4 peripheral locations: superior and inferior to the temporal vascular arcades, nasal to the optic disc, and temporal to the macula. All images were obtained using enhanced depth scanning protocols. Qualitative and quantitative assessments were performed and compared with those from healthy, age-matched controls.

Results: Generalized loss of the photoreceptor inner segment/outer segment junction was seen more frequently on extramacular OCT image sets. Focal loss of the inner segment/outer segment junction was seen most commonly on inferior extramacular images. Generalized thinning and loss of retinal architecture, accompanied by outer retinal hyperreflective foci, were also commonly seen on extramacular scans. Assessment of choroidal morphology included thinning/absence of the Sattler layer, generalized thinning, discrete hyperreflective foci, focal depigmentation, and the presence of suprachoroidal hyporeflective space. The mean (SD) foveal choroidal thickness was significantly less for patients with birdshot chorioretinopathy (276 [101] μm) than for controls (337 [74] μm) (P=.04).

Conclusions: The OCT images obtained outside the macula often show significant retinal and choroidal changes in cases for which conventional OCT scans appear unremarkable. Use of extramacular scanning may thus allow improved phenotyping of uveitic disorders such as birdshot chorioretinopathy. Evaluation of the photoreceptor inner segment/outer segment junction, using this approach, may be of value for monitoring disease activity in clinical practice and as a surrogate end point in clinical trials.


Birdshot chorioretinopathy is an inflammatory disease that affects the posterior segment of the eye, that is characterized by multiple, hypopigmented choroidal lesions radiating from the optic nerve, and that has a strong association with the human leukocyte antigen (HLA)—A29 allele. Although advances have been made in our understanding of the immunogenetics of this disorder, much remains unclear about its morphology and pathogenesis. In particular, the nature of the “birdshot” lesions seen using biomicroscopy remains poorly understood. Fluorescein angiography and indocyanine green angiography have been widely used for the evaluation of this disease, such techniques allow excellent visualization of vascular architecture but provide minimal information about other anatomical features. Fortunately, recent advances in optical coherence tomographic (OCT) imaging, if applied to patients with birdshot chorioretinopathy, may be uniquely suited to facilitate enhanced characterization of this disease.

Optical coherence tomography has been widely adopted by uveitis specialists for use in clinical practice, principally for identifying and monitoring patients with cystoid macular edema. Initial efforts have also been made to use OCT for the morphologic characterization of uveitic disorders. For a variety of reasons, the full...
potential of OCT in this regard has yet to be realized. First, many uveitic disorders are both rare and pathologically heterogeneous. Second, many patients with uveitis have dense vitreous inflammation that precludes optimal OCT image acquisition. Third, many uveitic disorders predominantly involve the choroid, and, until recently, visualization of this layer was not possible using commercially available OCT systems. Finally, and perhaps most importantly, many uveitic disorders are not confined to, or avoid entirely, the macular region. Thus, OCT images focused on the macula may often appear unremarkable, even in the presence of vision-threatening posterior segment disease.

In recent years, many of these issues have been addressed. In 2008, the formulation of “enhanced depth” scanning protocols allowed for high-resolution visualization of choroidal anatomy using the current generation of OCT (Figure 1). In 2010, Kriechbaum et al described the acquisition of OCT image sets outside the macula/vascular arcades for the visualization of scatter laser photocoagulation scars in diabetic retinopathy. Taken together, the use of enhanced depth protocols and “extramacular” image acquisition may greatly facilitate characterization of posterior uveitic pathology, particularly for disorders not typically associated with severe viritis. In the present report, we use a standard protocol for extramacular, enhanced depth OCT image acquisition, to evaluate retinal and choroidal morphology in a cohort of patients with birdshot chorioretinopathy.

METHODS

DATA COLLECTION

For our study, 12 patients (24 eyes) with an established diagnosis of birdshot chorioretinopathy were prospectively recruited (from July 2010 to August 2011). In addition, images were acquired from 9 healthy volunteers (18 eyes). Exclusion criteria for these “normal” controls included (1) any history of ocular disease or previous ocular surgery and (2) the presence of high myopia or hypermetropia (ie, greater than −6 or 6 diopters of spherical equivalent refractive error). Ethics committee approval was obtained (reference 06/Q2702/63; Dudley Local Research Ethics Committee), and the research adhered to the tenets set forth in the Declaration of Helsinki. Information about age, sex, ethnicity, time since diagnosis, HLA-A29 status, and current treatments of patients was gathered before the clinical ophthalmic examination and OCT image acquisition (Table 1). Best-corrected visual acuity was measured using Snellen visual acuity charts.

OCT IMAGE ACQUISITION PROTOCOL

Optical coherence tomographic image sets were obtained by an experienced operator (M.A.), using a single Spectralis OCT device (Heidelberg Engineering) and our custom extramacular enhanced depth protocol (entitled “extramacular enhanced depth OCT” [EMEDOCT]). In each case, both “macular” (ie, image sets centered on the fovea) and “extramacular” image sets were obtained. For extramacular imaging, images were obtained from 4 locations: (1) nasal to the optic disc, (2) superior to the superotemporal vascular arcade, (3) inferior to the inferotemporal vascular arcade, and (4) temporal to the macula (Figure 2). To allow reproducible identification of extramacular image locations, areas closely adjacent to the upper and lower vascular arcades, and adjacent to the optic nerve, were chosen.

All image sets (both macular and extramacular) were obtained using the “enhanced depth imaging” protocols first described by Spaide et al in 2008. Specifically, the OCT device was placed close enough to the subject’s eye to obtain an inverted image, and 7 equally-spaced OCT B-scan sections, each composed of 100 averaged B-scans, were then obtained over a 5° × 20° rectangle.

QUALITATIVE ANALYSIS OF OCT IMAGES

Optical coherence tomographic scans were evaluated for a number of retinal and choroidal morphologic characteristics by a certified OCT grader (P.A.K.) at the Doheny Image Reading Center in Los Angeles, California, who was masked to associated clinical information at the time of grading (Table 2 and Table 3; Figures 3, 4, and 5). A standard form was then completed for each image set, with each tomographic feature recorded as “visible,” “questionable,” “not visible,” or “cannot grade.” At-
tempts were also made to correlate OCT findings with specific posterior pole features as seen on the near-infrared images obtained at the time of OCT acquisition in the Spectralis system.

QUANTITATIVE ANALYSIS OF OCT IMAGES

For the generation of retinal and choroidal thickness maps, raw OCT data were exported from the Heidelberg Spectralis OCT system and imported into validated custom grading software, entitled “OCTOR.” For each OCT image set, the inner and outer boundaries of both the retina and the choroid were manually segmented. For one patient with birdshot chorioretinopathy and one normal control subject, the inner and outer boundaries of the photoreceptor inner segment/outer segment (IS/OS) junction were also analyzed (Figure 6). All boundaries were drawn in accordance with a standard OCT grading protocol.

DATA ANALYSIS AND STATISTICAL METHODS

For macular OCT image sets, the mean and standard deviation of retinal and choroidal thickness were calculated at the Early Treatment Diabetic Retinopathy Study foveal central subfield. For extramacular OCT image sets, the mean and standard deviation of retinal and choroidal thickness were calculated at the center point of each volume scan image set.

Clinical and imaging data were analyzed with frequency and descriptive statistics. Snellen visual acuities were converted to logMAR visual acuities for the purposes of statistical analysis. For comparison of retinal and choroidal thickness measurements between groups, the t test (2-tailed distribution) was used in the case of normally distributed interval variables. For comparison of categorical variables, the χ² test or the Fisher exact test was used, depending on whether cells had an expected fre-
quency of 5 or more. *P* values of less than .05 were considered statistically significant. Statistical analysis was performed using commercially available software (Intercooled Stata for Windows, version 9; StataCorp).

## RESULTS

### BASELINE CHARACTERISTICS

Of the 12 patients (24 eyes) with birdshot chorioretinopathy included in our analysis, 5 (42%) were women, and 7 (58%) were men. The mean (SD) age of patients was 59 (12) years, and the median age was 61 years (range, 39-73 years). Although the presence of the HLA-A29 allele was not required for diagnosis, all 12 patients in the present study tested positive for this allele. Baseline characteristics are summarized in Table 1.

### Table 2. Assessment of Retinal Morphologic Parameters in Birdshot Chorioretinopathy Using Optical Coherence Tomography

<table>
<thead>
<tr>
<th>Retinal Morphologic Parameter</th>
<th>Participants, No. (%)</th>
<th>Extramacular Location</th>
</tr>
</thead>
<tbody>
<tr>
<td>Epiretinal membrane</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Yes</td>
<td>0 (0)</td>
<td>2 (8)</td>
</tr>
<tr>
<td>Questionable</td>
<td>10 (42)</td>
<td>0 (0)</td>
</tr>
<tr>
<td>Generalized thinning/loss of architecture</td>
<td>Yes</td>
<td>8 (33)</td>
</tr>
<tr>
<td>Questionable</td>
<td>6 (25)</td>
<td>5 (21)</td>
</tr>
<tr>
<td>Focal disruption of IS/OS junction</td>
<td>Yes</td>
<td>10 (42)</td>
</tr>
<tr>
<td>Questionable</td>
<td>6 (25)</td>
<td>3 (13)</td>
</tr>
<tr>
<td>Generalized loss of IS/OS junction</td>
<td>Yes</td>
<td>6 (25)</td>
</tr>
<tr>
<td>Questionable</td>
<td>0 (0)</td>
<td>0 (0)</td>
</tr>
<tr>
<td>Outer retinal hyperreflective foci</td>
<td>Yes</td>
<td>8 (33)</td>
</tr>
<tr>
<td>Questionable</td>
<td>1 (4)</td>
<td>4 (17)</td>
</tr>
<tr>
<td>Intraretinal cystoid space</td>
<td>Yes</td>
<td>6 (25)</td>
</tr>
<tr>
<td>Questionable</td>
<td>0 (0)</td>
<td>0 (0)</td>
</tr>
<tr>
<td>Subretinal fluid</td>
<td>Yes</td>
<td>1 (4)</td>
</tr>
<tr>
<td>Questionable</td>
<td>0 (0)</td>
<td>0 (0)</td>
</tr>
</tbody>
</table>

Abbreviation: IS/OS, inner segment/outer segment.

*A* A total of 12 patients with birdshot chorioretinopathy and 12 healthy age-matched controls.

### Table 3. Assessment of Choroidal Morphologic Parameters in Birdshot Chorioretinopathy Using Optical Coherence Tomography

<table>
<thead>
<tr>
<th>Choroidal Morphologic Parameter</th>
<th>Participants, No. (%)</th>
<th>Extramacular Location</th>
</tr>
</thead>
<tbody>
<tr>
<td>Thinning/absence of Sattler layer</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Yes</td>
<td>2 (8)</td>
<td>4 (17)</td>
</tr>
<tr>
<td>Questionable</td>
<td>5 (21)</td>
<td>4 (17)</td>
</tr>
<tr>
<td>Generalized thinning</td>
<td>3 (13)</td>
<td>4 (17)</td>
</tr>
<tr>
<td>Questionable</td>
<td>0 (0)</td>
<td>2 (8)</td>
</tr>
<tr>
<td>Discrete hyperreflective foci</td>
<td>7 (29)</td>
<td>4 (17)</td>
</tr>
<tr>
<td>Questionable</td>
<td>4 (17)</td>
<td>5 (21)</td>
</tr>
<tr>
<td>Focal depigmentation</td>
<td>3 (13)</td>
<td>6 (25)</td>
</tr>
<tr>
<td>Questionable</td>
<td>0 (0)</td>
<td>0 (0)</td>
</tr>
<tr>
<td>Suprachoroidal hyporeflective space</td>
<td>Yes</td>
<td>3 (13)</td>
</tr>
<tr>
<td>Questionable</td>
<td>4 (17)</td>
<td>1 (4)</td>
</tr>
</tbody>
</table>

*A* A total of 12 patients with birdshot chorioretinopathy and 12 healthy age-matched controls.
group did not differ significantly from the mean age of patients with birdshot chorioretinopathy \( (P = .17) \).

**RETINAL FEATURES**

The OCT-derived retinal morphologic characteristics for patients with birdshot chorioretinopathy are summarized in Table 2. Generalized loss of the photoreceptor IS/OS junction was seen to differ significantly by scanning location (including questionable grades: inferior, 75%; nasal, 54%; superior, 46%; temporal, 41%; and macular, 25% \( [P = .012] \). Focal loss of the IS/OS junction also differed significantly by scanning location (including questionable grades: inferior, 91%; nasal, 67%; macular, 67%; superior, 59%; and temporal, 55% \( [P = .04] \)).

For patients with birdshot chorioretinopathy, the mean (SD) retinal thickness at the foveal central subfield was 237 (54) \( \mu \)m. For normal controls, the mean (SD) retinal thickness at the foveal central subfield was not significantly different (234 [19] \( \mu \)m; \( P = .90 \) (Table 4). Retinal thickness measurements from the center point of each extramacular image set, for both patients with birdshot chorioretinopathy and controls, are also summarized in Table 4.

**CHOROIDAL FEATURES**

The OCT-derived choroidal morphologic characteristics for patients with birdshot chorioretinopathy are summarized in Table 3. Thinning/absence of the Sattler layer was seen to differ significantly by scanning location (including questionable grades: inferior, 58%; nasal, 50%; superior, 34%; macular, 29%; and temporal, 17% \( [P < .001] \). Suprachoroidal hyporeflective space was also

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Figure 3. Detection of novel retinal morphologic parameters using extramacular optical coherence tomographic (OCT) scanning protocols. A, Near-infrared fundus image and inferior extramacular OCT B-scan reveal patchy disruption of the photoreceptor inner segment/outer segment (IS/OS) junction. B, Near-infrared fundus image and inferior extramacular OCT B-scan reveal generalized thinning/loss of the retinal architecture, generalized loss of the IS/OS junction, and the presence of discrete outer retinal hyporeflective foci. C, Near-infrared fundus image and temporal extramacular OCT B-scan reveal the transition zone between a grossly normal and a diseased retina.
Figure 4. Detection of novel choroidal morphologic parameters using extramacular and enhanced depth optical coherence tomographic (OCT) scanning protocols. A, Near-infrared fundus image and inferior extramacular OCT B-scan reveal generalized choroidal thinning. B, Near-infrared fundus image and inferior extramacular OCT B-scan reveal focal choroidal hypopigmentation (boundaries of depigmented area highlighted on OCT B-scan and fundus image using arrowheads).

Figure 5. Detection of novel choroidal morphologic parameters using extramacular and enhanced depth optical coherence tomographic (OCT) scanning protocols. A, Near-infrared fundus image and nasal extramacular OCT B-scan reveal discrete choroidal hyperreflectivity. B, Near-infrared fundus image and macular extramacular OCT B-scan reveal probable suprachoroidal hyporeflective space.
found to differ significantly by scanning location (including questionable grades: temporal, 38%; macular, 30%; superior, 12%; inferior, 4%; and nasal, 0% \( P = .001 \)).

For patients with birdshot chorioretinopathy, the mean (SD) choroidal thickness at the foveal center was 276 (101) \( \mu \text{m} \). By comparison, for normal controls, the mean (SD) choroidal thickness at the foveal center was significantly greater (337 (74) \( \mu \text{m}; P = .04 \)) (Table 4). Choroidal thickness measurements from the center point of each extramacular image set, for both patients with birdshot chorioretinopathy and controls, are also summarized in Table 4. Choroidal thickness at each of the 4 extramacular locations was significantly reduced in patients with birdshot chorioretinopathy vs controls.

### CASE STUDY

Patient 8, a 60-year-old man with a diagnosis of birdshot chorioretinopathy made more than 4 years previously, underwent extramacular enhanced depth OCT imaging in the present study. At the time of imaging, there was evidence of active disease, with full-field electroretinography revealing reduced scotopic responses in both eyes and with a delay in the 30-Hz flicker implicit time of recent onset in the left eye. Extramacular OCT imaging revealed focal loss of the photoreceptor IS/OS junction in all locations, with the exception of the temporal region of the left eye (Figure 3A). Manual segmentation of the photoreceptor IS/OS junction was performed for all extramacular image sets to generate IS/OS intensity projection maps (Figure 6). The mean IS/OS intensities were 0.6021 (macula), 0.5508 (inferior), 0.5967 (superior), 0.5834 (nasal), and 0.6888 (temporal). By comparison, for a normal control subject, the mean IS/OS intensities were 0.7674 (macula), 0.6381 (inferior), 0.7081 (superior), 0.5764 (nasal), and 0.7211 (temporal).

### COMMENT

In this prospective observational study, we use custom OCT scanning protocols to allow enhanced structural characterization of the retina and choroid in patients with birdshot chorioretinopathy. Patients with birdshot chorioretinopathy often have significant visual impairment even in the absence of decreased central visual acuity.24-27 In particular, many patients experience symptoms attributable to changes in the peripheral retina, including photopsia, nyctalopia, and peripheral visual field...

![Figure 6. Generation of photoreceptor inner segment/outer segment (IS/OS) junction optical coherence tomographic (OCT) intensity maps. A, An inferior extramacular OCT B-scan, with superimposed IS/OS intensity map, was obtained from a healthy control subject. The mean IS/OS intensity is 0.6381. B, An inferior extramacular OCT B-scan, with superimposed IS/OS intensity map, was obtained from a patient with active birdshot chorioretinopathy. The mean IS/OS intensity is 0.5508.](https://archopht.jamanetwork.com/...)

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### Table 4. Optical Coherence Tomography–Derived Measurements of Chorioretinal Thickness (Macular and Extramacular)

<table>
<thead>
<tr>
<th>Parameter</th>
<th>Birdshot Chorioretinopathy Group ( (n = 24) )</th>
<th>Normal Control Group ( (n = 18) )</th>
<th>( P ) Value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Retinal thickness, ( \mu \text{m} )</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Foveal central subfield</td>
<td>237 (54)</td>
<td>234 (19)</td>
<td>.90</td>
</tr>
<tr>
<td>Extramacular location</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Superior</td>
<td>185 (58)</td>
<td>196 (14)</td>
<td>.53</td>
</tr>
<tr>
<td>Inferior</td>
<td>163 (51)</td>
<td>179 (11)</td>
<td>.48</td>
</tr>
<tr>
<td>Temporal</td>
<td>202 (39)</td>
<td>209 (9)</td>
<td>.92</td>
</tr>
<tr>
<td>Nasal</td>
<td>191 (41)</td>
<td>205 (11)</td>
<td>.33</td>
</tr>
<tr>
<td>Choroidal thickness, ( \mu \text{m} )</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Foveal central subfield</td>
<td>276 (101)</td>
<td>337 (74)</td>
<td>.04 ( a )</td>
</tr>
<tr>
<td>Extramacular location</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Superior</td>
<td>201 (76)</td>
<td>271 (68)</td>
<td>.007 ( b )</td>
</tr>
<tr>
<td>Inferior</td>
<td>148 (64)</td>
<td>203 (50)</td>
<td>.008 ( b )</td>
</tr>
<tr>
<td>Temporal</td>
<td>218 (104)</td>
<td>317 (74)</td>
<td>.001 ( b )</td>
</tr>
<tr>
<td>Nasal</td>
<td>169 (82)</td>
<td>225 (64)</td>
<td>.04 ( a )</td>
</tr>
</tbody>
</table>

\( a \) Correlation is significant at the \( \alpha = .05 \) level.
\( b \) Correlation is significant at the \( \alpha = .01 \) level.
In our report, the most striking abnormalities seen on extramacular scans involved disruption of the outer retinal substructures. For normal subjects, the layered structure of the retina appears well maintained outside the macula, with clear delineation of the external limiting membrane, photoreceptor IS/OS junction, and retinal pigment epithelium (Figure 2). However, for patients with birdshot chorioretinopathy, a spectrum of outer retinal changes were seen, ranging from patchy disruption of the photoreceptor IS/OS junction (in recent studies, this has also been referred to as the ellipsoid region of the photoreceptor inner segments) to generalized loss of this hyperreflective line (Figure 3A and 3B). In more severe cases, changes on OCT images extended throughout the retina, with generalized thinning and loss of the multilayered retinal architecture. In many such cases, clusters of small hyperreflective foci were then seen in the absence of the photoreceptor IS/OS junction (Figure 3B). These lesions may represent “clumping” of the photoreceptors in response to injury, or areas of intraretinal pigment migration (although their appearance is not typical of the pigmentary changes seen on OCT images in dry age-related macular degeneration).30

For a number of years, changes in the photoreceptor IS/OS junction have been recognized to be of prognostic significance in macular diseases such as retinal vein occlusion, neovascular age-related macular degeneration, and central serous chorioretinopathy. More recently, however, their importance has been appreciated in the context of inherited retinal degenerative disease. In particular, loss of the photoreceptor IS/OS junction has been shown to coincide with loss of visual field in patients with retinitis pigmentosa. Furthermore, even when the photoreceptor IS/OS junction appears relatively intact, its intensity has been shown to be reduced in patients with cone dystrophies and achromatopsia. Consequently, assessment of this OCT-derived parameter has been suggested as an end point for clinical trials for these disorders. Measurements of photoreceptor IS/OS junction thickness and intensity may also prove helpful for physicians in clinical practice, although, with experience, such abnormalities can be identified on visual inspection. In this report, we demonstrate the feasibility of quantitative IS/OS assessment in the context of birdshot chorioretinopathy (Figure 6). In future studies, more dense volume scanning of the peripheral retina should improve the accuracy of photoreceptor IS/OS junction intensity maps; such maps may prove useful for monitoring the subtle progression of this and other diseases.

In this report, we also used enhanced depth OCT protocols to allow for assessment of choroidal morphology in patients with birdshot chorioretinopathy. For many patients, the choroid appeared grossly abnormal, with thinning/absence of the Sattler layer, or had an appearance of generalized atrophy. Choroidal thickness measurements at the fovea, and at each of the 4 extramacular locations, were significantly reduced relative to normal controls. However, for a number of reasons, caution is required with respect to these interpretations. First, early experience with enhanced depth OCT imaging suggests that the choroid is highly variable in appearance. Normative studies indicate that subfoveal choroidal thickness is typically between 300 and 350 μm; however, these measurements appear to decrease with age (approximately 15 μm with every decade) and with increasing myopia (approximately 10 μm with each additional diopter of myopia). Furthermore, there is some evidence to suggest that choroidal thickness is greater in men (approximately 60 μm) than in women, and it is affected by systemic parameters such as blood pressure. Second, initial attempts at measuring choroidal thickness outside the macula suggest the presence of significant topographic variation, with marked reductions inferiorly. The quantitative findings from the present study are consistent with this finding of significant topographic variation.

In addition to the quantitative assessment of the choroid, we undertook a more descriptive, qualitative approach. In particular, we correlated the appearance of birdshot lesions on fundal imaging with their corresponding OCT findings. The appearance of these lesions on OCT images appears consistent with focal areas of choroidal demarcation, a hypothesis first presented by Gass in 1981. Specifically, increased scleral reflectivity was seen on OCT images in areas corresponding to birdshot lesions; of note, this suggestion of choroidal demarcation was inferred from the difference in OCT appearance compared with that seen in retinal pigment epithelial hypopigmentation, where increased choroidal light transmission is a prominent feature (eg, geographic atrophy in dry age-related macular degeneration). It has previously been suggested that birdshot lesions often appear to be bordered by medium- to large-sized choroidal vessels; the findings on enhanced depth OCT images provide further support for this (Figure 4B). In addition to the appearance of focal demarcation, discrete areas of choroidal hyperreflectivity were also noted in many cases (Figure 5A). These were often seen in the vicinity of birdshot lesions, and we speculate that they represent clusters of pigmented cells. Alternatively, they may represent clusters of inflammatory cells, such as the choroidal lymphocytic infiltrates that have previously been described in patients with birdshot chorioretinopathy.

Each patient enrolled in our study had an established diagnosis of birdshot chorioretinopathy, with an average time since diagnosis of approximately 6 years. Moreover, only 3 patients were defined as having “active” disease at the time of OCT image acquisition. Therefore, it is perhaps not surprising that intraretinal cystoid spaces and/or subretinal fluid were not a common finding in the current cohort. The clinical and angiographic appearance of birdshot lesions has also been suggested to differ in the acute vs chronic disease states. We also attempted to assess each eye for the presence of fluid in the suprachoroidal space (in eyes without pathology, this
exists only as a potential space). For the purposes of the present study, we defined this OCT appearance as a thin, linear hyporeflective space at the outer aspect of the choroid, present on 2 or more adjacent OCT B-scans (Figure 5B). We speculate that fluid in the suprachoroidal space may be indicative of ongoing choroidal inflammatory activity. In our patient cohort, suprachoroidal hyporeflective space was seen more commonly than cystoid macular edema or subretinal fluid, a finding consistent with previous work suggesting that retinal and choroidal inflammation may occur independently in patients with birdshot chorioretinopathy. Although no firm conclusions can be made regarding the accuracy of this designation, our findings may be useful for future studies.

Our study has a number of strengths, combining prospective, standardized data collected from a clinically well-phenotyped population and standardized qualitative and quantitative assessments of OCT images by a senior grader certified for image interpretation in clinical trials. At present, there is a paucity of research describing the OCT findings in birdshot chorioretinopathy, and, as such, our report presents hitherto undescribed morphologic features obtained using a novel scanning protocol. Of note, this protocol can easily be used with the current generation of commercially available spectral-domain OCT systems (ie, without need for specialized hardware) and may be of value for the assessment of other posterior segment diseases. Our study also has a number of limitations. In particular, it involves a small number of patients, with chronic disease, presenting to a single tertiary referral center. It also has a cross-sectional design and is specifically designed to be exploratory in nature. In the future, longitudinal studies involving larger patient cohorts will be required to clarify the clinical significance of our findings. In addition, future studies will likely benefit from more detailed structure-function correlations (in particular, prospective comparisons of electrophysiologic and psychophysical testing, with extramacular OCT scanning).

In conclusion, the use of novel scanning protocols, with commercially available OCT devices, allows for enhanced characterization of patients with birdshot chorioretinopathy. Evaluation of the photoreceptor IS/OS junction in peripheral locations using this approach may be of value for monitoring disease activity, even in situations where it appears grossly intact. Furthermore, assessment of the choroid for evidence of depigmentation, with accompanying discrete hyporeflective lesions, may be of diagnostic utility for this disease. Finally, the scanning protocols described in our study may be easily applied to the study of other posterior segment diseases and may be of particular use for phenotyping diffuse choroidal inflammatory diseases.

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Author Affiliations: National Institute for Health Research Biomedical Research Centre for Ophthalmology, Moorfields Eye Hospital National Health Service (NHS) Foundation Trust, London, and University College London Institute of Ophthalmology (Dr Keane), Birmingham and Midland Eye Centre, Sandwell and West Birmingham NHS Trust (Mr Allie, Dr Turner, and Ms Southworth), Academic Unit of Ophthalmology, University of Birmingham (Drs Turner, Murray, and Denniston and Ms Southworth), and Queen Elizabeth Hospital Birmingham, University Hospitals Birmingham NHS Foundation Trust (Dr Denniston), England; and Doheny Eye Institute and Department of Ophthalmology, Keck School of Medicine of the University of Southern California, Los Angeles (Dr Sadda).

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REFERENCES


