Spectral-domain Optical Coherence Tomography Characteristics of Chorioretinal Folds

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Abstract

Purpose: To describe the spectral-domain optical coherence tomography features of chorioretinal folds of various etiology.

Materials and methods: Cross-sectional observational case series of consecutive patients with chorioretinal folds. All patients underwent enhanced depth imaging optical coherence tomography over a two-month study period. Clinical variables and imaging features including subfoveal choroidal thickness and chorioretinal fold morphology were analysed.

Results: 11 of 628 patients evaluated presented with chorioretinal folds. Diagnoses included hyperopia, uveal effusion, and secondary to surgery or medications. 22 patients presented with lesions simulating chorioretinal folds on ophthalmoscopy but were found not to be true chorioretinal folds with optical coherence tomography. Subfoveal choroidal thickness was diffusely thick in hyperopia, uveal effusion, hypotony, and topiramate, and normal in cases following scleral buckle and trauma. Lesions simulating chorioretinal folds occurred in age-related choroidal atrophy, and demonstrated chorioretinal contour changes involving specific choroidal vessels in an overall thin choroid.

Conclusion: Chorioretinal folds occurred in the context of a diffusely thick choroid in high hyperopia and hypotony, and with a normal choroidal thickness following scleral buckle or trauma. Enhanced depth imaging OCT is helpful in differentiating chorioretinal folds from various etiologies and from simulating lesions.

Keywords: Choroid; Choroidal folds; Chorioretinal folds; Enhanced depth imaging; Optical coherence tomography

Introduction

Chorioretinal folds (CRF), or infoldings of the inner choroid, retinal pigment epithelium, and retina were originally described by Nettleship [1], and have been further characterized by Norton [2] and Newell [3] using ophthalmoscopy and fluorescein angiography in various posterior segment diseases. Reported associations include high hyperopia [4-7], age-related macular degeneration [2,4,8], hypotony [9], disc edema, idiopathic uveal effusion syndrome (IUES) [10], and external globe compression [11]. The term choroidal folds and chorioretinal folds have often been used interchangeably, however the former has occasionally been used to denote folds of the choroid and retinal pigment epithelium occurring without evidence of distortion of the overlying inner retinal tissue. Nevertheless, the two terms likely represent the same entity, and are recognized to have significant overlap by most authors [11,12].

Previous imaging reports have included angiography, fundus autofluorescence [13], and time-domain optical coherence tomography [12], but have lacked the resolution possible with current spectral-domain (SD-OCT) technology. Additionally, the pathogenesis of chorioretinal fold formation is incompletely understood. The authors hypothesized that enhanced depth imaging SD-OCT may reveal specific aspects of the choroid that may be helpful in understanding CRF pathogenesis. The purpose of the present report was to investigate the morphology of chorioretinal folds as imaged by high resolution SD-OCT, and to investigate aspects of choroidal thickness and morphology using enhanced depth imaging (EDI).

Materials and Methods

Consecutive patients were prospectively evaluated over a two-month period with EDI SD-OCT (Spectralis, Heidelberg Engineering, Heidelberg, Germany). All patients were seen and evaluated at a single vitreoretinal specialist office (Charles Retina Institute, Memphis, TN). Patients were evaluated with complete ophthalmic examination, EDI SD-OCT 12-line radial raster scan centered on the foveola with 20 scans averaged/raster. The treatment of all patients and data conformed with the tenets set forth in the Declaration of Helsinki, and was performed in accordance with the Health Insurance Portability and Accountability Act of 1996. The study was performed as part of an ongoing IRB approved study examining choroidal thickness and morphology.

Results

628 patient SD-OCT examinations were initially reviewed for the presence of CRF. CRF were present in 11 patients (2%). Lesions simulating CRF, but were not considered true CRF after OCT analysis were present in 22 patients. Overall mean age ± standard deviation in patients with CRF was 66 ± 10 years. Patients consisted of 5 males and 6 females, 9 Caucasians, and 2 African Americans. Chorioretinal folds were bilateral in 5 patients and unilateral in 6. Mean spherical equivalent refraction (phakic or prior to cataract extraction) was 0.82 ± 4.5. Chorioretinal folds consisted of distinct infoldings of the inner

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and two with chronic hypotony following pars plana vitrectomy or trabeculectomy. Cases 5 and 6 had previous scleral buckle without vitrectomy and reported metamorphopsia, each with vision of 20/40 in the affected eye. Case 5 had spherical equivalent of -2.00 diopters and subfoveal choroidal thickness of 275 µm which was identical to the fellow eye. Case 6 was a high myope with spherical equivalent of -8.25 diopters and subfoveal choroidal thickness of 82 µm, which was similar to the fellow eye. CRF in cases 5 and 6 appeared as distinct infoldings of the inner choroid, RPE, and outer retina (Figure 3) without a diffusely thick choroid.

Hyperopia

Cases 1-3 (5 eyes) presented with CRF and high hyperopia. Representative images are demonstrated in Figure 1. CRF were horizontal, asymptomatic, and unassociated with additional chorioretinal pathology in all cases. All patients were phakic and mean overall spherical equivalent was 7.25 ± 1.5 diopters. Mean subfoveal choroidal thickness was 493 ± 189 µm. Cases 1 and 2 were bilateral and associated with good vision (20/20-20/25). Case 3 was unilateral and associated with anisometropic amblyopia with BCVA of 20/70 and without any metamorphopsia or additional chorioretinal pathology.

Idiopathic uveal effusion

Cases 3 and 4 (4 eyes) presented with nanophthalmos and idiopathic uveal effusion syndrome (IUES). Both cases demonstrated marked nanophthalmos on high resolution computed tomography and magnetic resonance imaging of the orbits, with axial globe diameter between 14 and 16 mm, with posterior scleral thickness greater than 3 mm in both cases. Representative images are presented in Figure 2. Mean spherical equivalent was 10.5 ± 0.7 diopters. Mean subfoveal choroidal thickness was 647 ± 161 µm. Case 3 (Figure 2) presented with multiple serous pigment epithelial detachments (PED) in both eyes and subretinal fluid in her left eye with BCVA of 20/100 and 20/25 with metamorphopsia in her right eye. Case 4 lacked subretinal fluid but SD-OCT revealed serous PED and diffusely thick choroid.

Secondary

Cases 5-8 (4 eyes) presented with unilateral CRF following ocular surgery, two following encircling scleral buckle (Figure 3) and two with chronic hypotony following pars plana vitrectomy or trabeculectomy. Cases 5 and 6 had previous scleral buckle without vitrectomy, and reported metamorphopsia, each with vision of 20/40 in the affected eye. Case 5 had spherical equivalent of -2.00 diopters and subfoveal choroidal thickness of 275 µm which was identical to the fellow eye. Case 6 was a high myope with spherical equivalent of -8.25 and subfoveal choroidal thickness of 82 µm, which was similar to the fellow eye. CRF in cases 5 and 6 appeared as distinct infoldings of the inner choroid, RPE, and outer retina (Figure 3) without a diffusely thick choroid. Cases 7 had chronic hypotony with intraocular pressure of 3 mm Hg following trabeculectomy. EDI SD-OCT revealed typical CRF with a diffusely thick choroid of 428 µm and BCVA of 20/200. The fellow eye had choroidal thickness of 228 µm with BCVA of 20/20. Case 8 had a chronic retinal detachment and intraocular pressure of 4 mm Hg. Subfoveal choroidal thickness was 340 µm and 212 µm in the unaffected fellow eye. Cases 9 and 10 (4 eyes) had bilateral CRF with mean spherical equivalent of -1.25 and -0.75, respectively. Both patients had a diffusely thick choroid (mean 686 ± 74 µm) with normal anterior chamber angle. Both patients had BCVA of 20/20 in both eyes.

Figure 1: Imaging characteristics of chorioretinal folds in high hyperopia. (A) Fundus autofluorescence demonstrating horizontal chorioretinal folds (arrow) in this 54 year-old patient with a spherical equivalent of +7.50 diopters; (B) Scanning laser ophthalmoscopic image demonstrating scan position for SD-OCT in (C) Enhanced depth imaging of chorioretinal folds consist of distinct infoldings of the choriocapillaris, RPE, and outer retina; the subfoveal choroidal thickness is 463 µm.

Figure 2: Imaging characteristics of chorioretinal folds in idiopathic uveal effusion syndrome. (A) Fundus photography of the right eye of this 61 year-old female with a spherical equivalent of +9.75 diopters demonstrating horizontal chorioretinal folds (arrow) and a large serous pigment epithelial detachment (asterisk); (B) Left eye demonstrating horizontal chorioretinal folds and central macular subretinal fluid (arrowheads); (C and D) Scanning laser ophthalmoscopic images, green line denotes scan position for SD-OCT images in (E) demonstrating infolding of the choriocapillaris, RPE, and outer retina; subfoveal choroidal thickness is 731 µm and (F) Chorioretinal folds similar to (E), with central macular subretinal fluid, subfoveal choroidal thickness is 760 µm.
but reported metamorphopsia. CRF appeared identical to those seen in other patients with a diffusely thick choroid in the present series. Case 11 presented with unilateral CRF 2 years following blunt ocular trauma and choroidal rupture. Subfoveal choroidal thickness was 241 μm which was similar to the fellow eye.

Lesions simulating chorioretinal folds

22 patients (36 eyes) presented with an ophthalmoscopic appearance that was consistent with CRF, but SD-OCT revealed similar or simulating lesions that were not true chorioretinal folds. Mean age in this subgroup was 71 ± 12 years. In 17 patients, all with a diagnosis of atrophic age-related macular degeneration, EDI SD-OCT revealed a diffusely thin choroid, drusen, subretinal drusenoid deposits, and focal chorioretinal contour changes that corresponded to specific choroidal vessels (Figure 4). 5 additional patients had a similar SD-OCT appearance but lacked features of age-related macular degeneration, and thus represented age-related choroidal atrophy. The overall mean subfoveal choroidal thickness in this group was 88 ± 24 μm. One patient presented with metamorphopsia and a submacular choroidal varix (Figure 4) without additional chorioretinal pathology and demonstrated focal elevations of the choroid corresponding to a specific dilated choroidal vessel.

Discussion

The imaging results of the present series indicate that CRF consist of distinct infoldings of the inner choroid and outer retina, which are consistent with previous reports. With EDI SD-OCT, we have demonstrated a significantly thicker than average choroid in patients with CRF in the context of high hyperopia, uveal effusion, and hypotony. Patients with secondary CRF due to scleral buckle or trauma demonstrated choroidal thickness in the normal range, but with similar morphology of the inner choroid, RPE, and outer retina. Giuffre et al., in the only prior report of OCT analysis of chorioretinal folds, used time-domain OCT to evaluate 8 patients. In that series, Giuffre et al. demonstrated similar features to the present analysis, but lacked choroidal thickness data due to the low resolution and lack of enhanced depth imaging. They identified two variants of CRF, differentiated by the presence of retinal undulations in some patients (chorioretinal folds) and the lack of retinal involvement in others (choroidal folds). That report did stress, however, the significant overlap among the two entities. In the present series, all patients with true chorioretinal folds demonstrated infoldings of the inner choroid (choriocapillaris), RPE, and outer retina. We did not observe significant inner retinal contour changes in cases of true CRF. Interestingly, several patients in the present series presented with chorioretinal contour changes which appeared similar to CRF on ophthalmoscopy, but SD-OCT revealed a focal elevation involving the outer retina and retinal pigment epithelium (RPE) corresponding to a specific choroidal vessel (Figure 4). We have termed this entity focal choroidal elevation, which appears as a relatively common feature of patients with age-related choroidal atrophy and age-related macular degeneration, and is the subject of a forthcoming investigation. We do not feel this entity represents true CRF, but is rather the consequence of a relatively thin choroid resulting in the RPE being draped over the more rigid underlying choroidal vessels. This may represent a subset of previously reported CRF in age-related macular degeneration, and in fact has some features in common with the OCT appearance of Giuffre et al. [12] (Figures 1d and 2a). Gass [8] reported radial CRF in association with contracted choroidal neovascularization, which likely represents a subset of true CRF due to mechanical fibrovascular contraction of the RPE in exudative age-related macular degeneration.

The largest single group of patients with CRF in the present series represented bilateral high hyperopia and revealed horizontal chorioretinal folds in the absence of additional chorioretinal pathology that appeared asymptomatic. In contrast to patients with high hyperopia alone, patients with CRF due to uveal effusion or secondary causes reported metamorphopsia, and often had reduced vision either from CRF or subretinal fluid. Serous PED was a common feature in patients with uveal effusion, which was absent from patients with high hyperopia or hypotony. In patients with hypotony, the choroid appeared thickened secondarily, as evident by the asymmetric and normal
choroidal thickness in the fellow eye. This is similar to previous reports indicating that chronic sub-clinical CRF are often asymptomatic, but may lead to significant visual consequence when develop acutely or are secondary to additional underlying pathology [9,11].

The results of the present series may offer insight into the underlying pathogenesis of chorioretinal folds. As previously theorized [8,10,14], the diffusely thickened choroid in cases of high hyperopia, idiopathic uveal effusion, hypotony, and topiramate use is consistent with a spectrum of choroidal effusion. This may be secondary to anatomic variants, as in hyperopia or medication use, as in topiramate. We hypothesize that the development of CRF results from a diffusely thickened choroid which effectively decreases the internal circumference of the vitreous cavity, thereby leading to the RPE/inner choroid complex being thrown into redundant folds. We also observed cases in which the choroid demonstrated a normal thickness, such as in encircling scleral buckle and trauma. In the case of scleral buckle, the internal circumference of the vitreous cavity is reduced due to external compression, which likely represents a similar mechanical mechanism as in high hyperopia but without a primary choroidal process.

The present series is limited by small sample size, single-center study location, and lack of longitudinal analysis. We observed CRF in 2% of patients seen over a two-month study period that likely overestimates the true prevalence of the finding due to the sub-specialist referral center location. We conclude that CRF due to high hyperopia, IUES, topiramate, and hypotony involve an overall thick choroid and involve infoldings of the choriocapillaris, RPE, and outer retina. This is in contrast to CRF secondary to trauma or scleral buckle, which have an identical inner choroidal morphology but a normal choroidal thickness. EDI SD-OCT is helpful in differentiating true chorioretinal folds from additional simulating lesions, such as focal choroidal elevations, which may be more common than true chorioretinal folds.

Summary

We describe the morphologic characteristics of chorioretinal folds using spectral domain optical coherence tomography and identify a diffusely thickened choroid in cases associated with hyperopia and hypotony but a normal choroidal thickness in cases associated with previous surgery or trauma.

Conflict of Interest

The authors have no financial interest or conflict of interest regarding the material presented in this report.

References