The Expanded Spectrum of Focal Choroidal Excavation

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Objective: To describe the clinical and imaging findings in patients with focal choroidal excavation.

Methods: Retrospective observational case series. The medical records of 12 patients (13 eyes) with focal choroidal excavation were reviewed. Clinical histories and imaging findings (including color photography, fundus autofluorescence imaging, fluorescein angiography, indocyanine green angiography, spectral-domain optical coherence tomography, and enhanced depth imaging spectral-domain optical coherence tomography) were analyzed.

Results: The mean age of the patients was 45 years (range, 22-62 years). Four patients were Asian. Mean visual acuity was 20/31 (range, 20/20 to 20/100). Mean refractive error was −3.54 diopters (D) (range, 6.00 to −8.00 D). One patient had bilateral involvement. All patients manifested varying degrees of foveal pigmentary changes that were usually hypoautofluorescent on fundus autofluorescence images. Fluorescein angiographic findings varied with degree of retinal pigment epithelial alterations. Indocyanine green angiography revealed relative hypoautofluorescence. In 7 eyes, spectral-domain optical coherence tomography revealed outer retinal layers conforming to retinal pigment epithelial alterations within the excavation. In the other 6 eyes, spectral-domain optical coherence tomography revealed a separation between the outer retina and the retinal pigment epithelium within the excavation. In 7 eyes studied with enhanced depth imaging spectral-domain optical coherence tomography, there was no evidence of scleral ectasia. Mean choroidal thickness of the uninvolved choroid was thicker than normal at 319 µm (range, 244-439 µm). All lesions remained stable except for in 1 eye, which had findings of central serous chorioretinopathy and secondary type 2 (subretinal) neovascularization.

Conclusion: Focal choroidal excavation is a newly described idiopathic entity in eyes having 1 or more focal areas of choroidal excavation. In some patients, there may be an association with central serous chorioretinopathy. Although most lesions remain stable, secondary choroidal neovascularization may occur.


Video available online at www.archophthalmol.com

An unusual macular finding was reported by Jampol et al1 in which an excavation of the choroid was detected by optical coherence tomography (OCT) in an eye with good visual acuity and normal appearance of the overlying retina. The etiology of the lesion was unclear because it did not resemble a macular staphyloma and was not associated with prior insult to the choroid or sclera. The lesion appeared to enlarge slightly over a period of 1 year with thinning of the fovea seen on OCT. Analysis of these findings was limited by the inability of the time-domain OCT instrument to image beyond the choroid, making it impossible to determine whether the defect involved the choroid, sclera, or both. Three additional patients with similar findings were described by Wakabayashi et al2 using spectral-domain OCT (SD-OCT). Wakabayashi et al2 described 2 patterns of unilateral choroidal excavation without scleral involvement. In 2 patients, the outer retinal layers appeared to conform to the choroidal defect without disruption or an optically clear space between the photoreceptor tips and the hyperreflective retinal pigment epithelial band. A third eye showed a disorganization of the photoreceptor tips with a hyporeflective subretinal space. Another case was recently reported by Abe et al3 who noted a separation between the photoreceptors and the retinal pigment epithelium. In our report, we expand the spectrum of focal choroidal excavation (FCE) by describing the multimodal
imaging findings in 12 patients with FCE, including 1 patient with bilateral involvement.

METHODS

A retrospective review of 13 eyes was performed for 12 patients with 1 or more areas of FCE. Focal choroidal excavation was defined as an area of macular choroidal excavation detected on an SD-OCT scan without evidence of a posterior staphyloma or scleral ectasia, in a patient lacking a history of trauma, posterior uveitis, retinal or choroidal vascular disease, or prior retinal or choroidal infection. All patients had undergone complete ocular examinations, including slitlamp biomicroscopy, indirect ophthalmoscopy, and measures of best-corrected visual acuity, refractive error, and intraocular pressure. Ancillary testing was performed with color photography, fundus autofluorescence imaging, and SD-OCT. Enhanced depth imaging OCT (EDI-OCT) was used when available to image the entire choroid in cross section in order to rule out staphylomatous excavation of the sclerochoroidal junction.4 Angiographic studies were performed in only some of the outer border of the retinal pigment epithelium to the inner scleral border using EDI-OCT as previously described.4

RESULTS

Thirteen eyes of 12 patients were identified in which SD-OCT scans revealed 1 or more focal areas of choroidal excavation without evidence of a posterior staphyloma or scleral ectasia. Patients’ demographic and clinical characteristics are summarized in Table 1. The mean age of patients was 45 years (range, 22-62 years). Four patients were Asian. Mean visual acuity was 20/31 (range, 20/20 to 20/100). Ten eyes were myopic, and the mean refractive error was −3.54 diopters (D) (range, 6.00 to −8.00 D). None of the patients had a history of any medical illness or medication use that seemed relevant to the retinal findings. None of the patients reported a family history of retinal disease. Six patients reported a visual disturbance in the affected eye over a mean of 3 months prior to presentation, which included symptoms of metamorphopsia and blurred vision. The other 6 patients were asymptomatic. One patient had a history of central serous chorioretinopathy in the fellow eye not affected by excavation. A second patient had a history of serous retinal detachment in the involved eye consistent with central serous chorioretinopathy. This eye subsequently developed type 2 (subretinal) neovascularization, which was...
treated with intravitreal antivascular endothelial growth factor therapy with good visual and anatomic response.

In all eyes, the presence of choroidal excavation could not be determined on clinical examination or by color photography (Figures 1, 2, 3, 4, and 5). Clinical examination and color photography did show varying degrees of subfoveal and perifoveal pigmentary disturbances in all eyes (Table 2). Eleven eyes showed perifoveal ill-defined punctate areas of hyperpigmentation and hypopigmentation, and 2 of these eyes had a yellowish spot consistent with a small acquired vitelliform lesion. Larger, well-circumscribed lesions with more pronounced hypopigmentation were seen in 2 patients. For the most part, the pigmentary changes corresponded to areas of hypoautofluorescence on fundus autofluorescent images (Figure 4) in all but 2 eyes. The lesion in case 10 had both hypoautofluorescent and hyperautofluorescent components associated with subretinal fluid seen on SD-OCT scans. An autofluorescence image of case 11 revealed an oval area of hyperautofluorescence consistent with the patient’s history of resolved central serious chorioretinopathy.

The use of SD-OCT imaging for all patients was necessary to detect the presence of FCE (video, http://www.archophthalmol.com). Of the 13 eyes studied, 10 had a single lesion (cases 1-4 and cases 8-13), and 3 had 2 distinct areas of excavation (cases 5-7). In all eyes, SD-OCT scans revealed the retinal pigment epithelial band following the contour of the choroidal excavation. None of the eyes showed evidence of retinal pigment epithelial detachment on SD-OCT scans. In 7 eyes, there was no separation between the photoreceptor tips and the retinal pigment epithelium (conforming FCE) (Figures 1 and 2). In these cases, the outer nuclear layer appeared thickened compared with the retina in the areas not affected by excavation. In the remaining 6 eyes, the photoreceptor tips appeared to be detached from the underlying retinal pigment epithelium with an intervening hyporeflective space presumably representing subretinal fluid (nonconforming FCE) (Figure 3). In 2 cases of nonconforming FCE (cases 8 and 10), there was thinning of the hyperreflective retinal pigment epithelial band beneath the elevated retina. At attenuation or complete absence of the inner segment/outer segment junction was present in all nonconforming FCE eyes, with preservation of the outer nuclear layer and the external limiting membrane. In contrast, signals from the retinal pigment epithelium and the inner segment/outer segment junction appeared intact in eyes with conforming FCE. In 7 eyes studied using EDI-OCT, the mean thickness of

Figure 2. A 26-year-old woman with a conforming focal choroidal excavation. A, Color photograph of the left eye reveals retinal pigment epithelial alterations at the temporal edge of the fovea. B, Spectral-domain optical coherence tomographic scan through the fovea reveals an irregularly shaped conforming focal choroidal excavation. The outer retinal layers can be seen to conform to the retinal pigment epithelium within the excavation.

Figure 3. A 62-year-old woman with a nonconforming focal choroidal excavation. A, Color photograph of the left eye reveals atrophic retinal pigment epithelial changes beneath the fovea. B, Time-domain optical coherence tomographic scan from 2003 reveals an area of presumed hyporeflective subretinal fluid separating the photoreceptor outer segments from the underlying retinal pigment epithelium within the choroidal excavation. C, Spectral-domain optical coherence tomographic scan from 2009 reveals findings similar to the scan taken 6 years earlier (B).
the uninvolved choroid adjacent to the areas of FCE was 328 µm (range, 244-439 µm).

Fluorescein angiography was performed in 6 eyes that showed varying degrees of hyperfluorescence and hypofluorescence related to a range of retinal pigment epithelial changes. Five eyes with retinal pigment epithelial attenuation exhibited hyperfluorescence consistent with a transmission defect that suggested an intact underlying choriocapillaris (cases 2 and 8-11). In contrast, the lesion in case 13 was hypofluorescent centrally with circumferential hyperfluorescence. This angiographic pattern suggested some attenuation of the choriocapillaris in this eye, which had the largest excavation and a very thin layer of remaining choroid on SD-OCT scans. Varying degrees of hypofluorescence were seen in the 6 eyes studied using indocyanine green angiography. The degree of indocyanine green angiographic hypofluorescence appeared to correlate with the amount of choroidal thinning detected using SD-OCT.

The results of microperimetry were normal in case 3, even though the patient complained of mild distortion with visual acuity of 20/20. The results of microperimetry for case 13 showed decreased sensitivities within the excavation, and visual acuity was 20/100.

**COMMENT**

We report an expanded spectrum of a recently described entity in which 1 or more focal areas of macular choroidal excavation are detected in patients lacking a history of any ocular disease known to produce choroidal thinning. Because one of our cases had bilateral involvement (Figure 5), we suggest that a more appropriate name for this entity would be *focal choroidal excavation* rather than *unilateral choroidal excavation*, as was recently proposed by Wakabayashi et al.²

Most of our patients were diagnosed with FCE in their fourth or fifth decades, and most were moderately myopic (mean, −3.54 D). In our series, there was no clear sex or race predilection. In the report by Wakabayashi et al,² all 3 patients were Asian women in their fourth decade. The patient reported by Abe et al³ was a 29-year-old Asian man.

Patients were either asymptomatic or reported no history of central visual disturbances until shortly prior to presentation. Clinically, the lesions appeared as either pigmentary disturbances or yellowish spots. The lesions were mostly hypoautofluorescent and typically showed transmission defects with retinal pigment epithelial attenuation on fluorescein angiography. We attributed hyperautofluorescence in 2 cases to current (case 10) or resolved (case 11) subretinal fluid.
All lesions involved the foveal or perifoveal region. Two patterns of excavation were detected using SD-OCT. In conforming FCE, there was no separation between the photoreceptor tips and the retinal pigment epithelium, with the outer nuclear layer appearing thicker than it does in areas not affected by excavation. In this type of excava-
tion, the inner segment/outer segment junction and reti-

nal pigment epithelium were undisturbed. In noncon-
forming FCE, the photoreceptor tips appeared to be
detached from the underlying retinal pigment epithel-

um, with the intervening hyporeflective space presum-
ably representing subretinal fluid. In these eyes, the sig-

nals corresponding to the retinal pigment epithelium
and the inner segment/outer segment junction were often

disrupted. Hyperreflective material, presumed to repre-

sent shed outer segment debris, was often present below
an intact external limiting membrane.

We hypothesize that FCE may be a congenital pos-
terior segment malformation. Initially, the elasticity of
the retina would allow the photoreceptors to remain
attached to the retinal pigment epithelium. In time,
eyes with conforming FCE could progress to noncon-
forming lesions as stress on the outer retina results
in separation of the photoreceptor tips from the apical
surface of the retinal pigment epithelium, leading to
subsequent accumulation of varying amounts of the
shed outer segment material in the subretinal space.
At this point, photoreceptor death may accelerate as seen
in other conditions with chronic serous detachments.

Also, because the normal choroid has been shown to
become thinner with age, it is possible that the choro-
dal excavation may enlarge with time, resulting in fur-
ther ischemia to the overlying retina, atrophic changes,
and visual disturbances. Although we did not observe
conforming FCE eyes convert to nonconforming
lesions, our mean follow-up of the conforming FCE
eyes was less than 1 year. Long-term follow-up of these
lesions may help elucidate their natural history.

It is interesting that 2 patients had a concurrent di-
agnosis of central serous chorioretinopathy, one in the
involved eye and one in the fellow eye. Although EDI-
OCT was not performed for all patients, a review of scans
in 6 eyes demonstrated thicker choroidal measure-
ments than would be expected for myopic eyes, which
typically have a thin choroid. Similarly, the choroid in
eyes with central serous chorioretinopathy has been
shown to be significantly thicker than in normal eyes.

The patients reported by Wakabayashi et al also had cho-
roids that appeared thicker than normal. In fact, in case
1 and case 3 of that report, the outer choroidal border
could not be discerned on the SD-OCT scans. Although
the significance of this observation is unknown, it is plau-
sible that a thick choroid may predispose to FCE or, al-
ternatively, that FCE is difficult to detect in eyes with thinner-than-average choroidal thickness, which is more
typical of myopic eyes.

The etiology of FCE is unclear. All patients were young
and healthy, and there was no evidence of any systemic
or ocular condition that may have caused the choroidal

disturbance. Ten of the 13 eyes were myopic, 5 of which
had 6 D of myopia or more, so it is possible that these
lesions represent a “microstaphyloma.” However, the ex-
cavation appears to affect only the choroid because the
junction between the choroid and the sclera appears
smooth and undisturbed on EDI-OCT scans. Although
we hypothesize that FCE is a congenital lesion, an alter-
native possibility is a congenital or acquired choroiditis
that leaves a legacy of focal choroidal atrophy. How-
ever, FCE lesions have an intact outer retina and cho-
riocapillaris, whereas these layers are typically absent in
chorioretinal scars. Cases 12 and 13 had a discrete area
of pigmentary and fundus autofluorescence changes that
resemble torpedo maculopathy. Unlike torpedo macu-
lopathy, FCE is not usually oval in shape, typically in-
volves the fovea, and is associated with less significant
outer retinal thinning and degeneration.

Our study has limitations inherent to a small retro-
spective analysis. Focal choroidal excavation has only
recently been described using OCT imaging techniques and,
thus far, has no histopathologic confirmation. Owing to
the small number of cases identified and the short follow-
up, our understanding of this condition and our inter-

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### Table 2. Clinical Findings in 13 Eyes with Focal Choroidal Excavation

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Fundus Appearance</th>
<th>FAF Finding</th>
<th>SD-OCT Finding</th>
<th>Lesion(s), No.</th>
<th>Choroidal Thickness, µm</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Punctate pigmentary changes</td>
<td>Hypoautofluorescent</td>
<td>Conforming</td>
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<td>260</td>
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<td>2</td>
<td>Punctate pigmentary changes</td>
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<td>Conforming</td>
<td>1</td>
<td>321</td>
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<tr>
<td>3</td>
<td>Punctate pigmentary changes</td>
<td>NA</td>
<td>Conforming</td>
<td>1</td>
<td>NA</td>
</tr>
<tr>
<td>4</td>
<td>Punctate pigmentary changes</td>
<td>NA</td>
<td>Conforming</td>
<td>1</td>
<td>NA</td>
</tr>
<tr>
<td>5</td>
<td>Punctate pigmentary changes</td>
<td>Hypoautofluorescent</td>
<td>Conforming</td>
<td>2</td>
<td>333</td>
</tr>
<tr>
<td>6a</td>
<td>Punctate pigmentary changes</td>
<td>Hypoautofluorescent</td>
<td>Conforming</td>
<td>2</td>
<td>NA</td>
</tr>
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<td>7a</td>
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<td>Conforming</td>
<td>2</td>
<td>NA</td>
</tr>
<tr>
<td>8</td>
<td>Yellow spot</td>
<td>Hypoautofluorescent</td>
<td>Nonconforming</td>
<td>1</td>
<td>330</td>
</tr>
<tr>
<td>9</td>
<td>Yellow spot</td>
<td>NA</td>
<td>Nonconforming</td>
<td>1</td>
<td>244</td>
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<tr>
<td>10</td>
<td>Punctate pigmentary changes</td>
<td>Hyperautofluorescent and hypoautofluorescent</td>
<td>Nonconforming</td>
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<td>Punctate pigmentary changes</td>
<td>Hyperautofluorescent</td>
<td>Nonconforming</td>
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<tr>
<td>12</td>
<td>Discrete pigmentary changes</td>
<td>Hypoautofluorescent</td>
<td>Nonconforming</td>
<td>1</td>
<td>328</td>
</tr>
<tr>
<td>13</td>
<td>Discrete pigmentary changes</td>
<td>Hypoautofluorescent</td>
<td>Nonconforming</td>
<td>1</td>
<td>439</td>
</tr>
</tbody>
</table>

Abbreviations: FAF, fundus autofluorescent; NA, not available; SD-OCT, spectral-domain optical coherence tomographic. 

A bilateral involvement.
pretation of the images are currently limited. Further prospective study of eyes with an FCE is necessary to elucidate its etiology, clinical course, and visual prognosis.

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REFERENCES


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