

## The Frequency Of Focal Choroidal Excavation Detected By Optical Coherence Tomography

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**Conflict-of-interest statement:** The author(s) declare(s) that there is no conflict of interest regarding the publication of this paper.

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Received: March 1, 2017  
Revised: March 21, 2017  
Accepted: March 22, 2017  
Published online: March 25, 2017

### ABSTRACT

**AIM:** To determine the frequency and the causes of focal choroidal excavation (FCE) detected by spectral domain optical coherence tomography (SDOCT), and to comment on its clinical importance.

**MATERIALS AND METHODS:** In this study, 632 eyes of the patients with untreated wet age-related macular degeneration (w-AMD,  $n = 368$ ), central serous chorioretinopathy (CSCR,  $n = 214$ ), Vogt-Koyanagi-Harada Disease (VKHD,  $n = 3$ ), adult vitelliform macular dystrophy (AVMD,  $n = 32$ ), Best vitelliform macular dystrophy (BVMD,  $n = 2$ ) and other macular dystrophies and specific maculopathies ( $n = 13$ ) were retrospectively reviewed for the presence of the FCE. Additionally, 228 unaffected fellow eyes without obvious macular pathology of the cases with unilateral

involvement were also evaluated as control cases in this study. The medical records of ten patients with FCEs detected by SDOCT were reviewed. The causes, frequency, associated diseases, types, size and shape, associated SDOCT pathologies, SDOCT findings of the FCEs in ten cases were evaluated and commented on.

**RESULTS:** The FCE was detected in ten eyes of ten cases. Of the patients with FCE, 4 (40%) were women and 6 (60%) were men. The ages of patients were ranged between 63 to 76 years (median 66.1 years). Bilateral FCE was not detected. The FCE was detected in one eye with w-AMD (1/368 patients), in two eyes with active CSCR (2/214), in one with AVMD (1/32 patients) and in 5 eyes of the control (idiopathic FCE; 5/228).

**CONCLUSIONS:** FCE may be detected in various macular chorioretinal diseases. However, it should be considered that FCE might be idiopathic.

**Key words:** Focal Choroidal Excavation; Optical Coherence Tomography; Causes; Idiopathic

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Turgut B. The Frequency Of Focal Choroidal Excavation Detected By Optical Coherence Tomography. *International Journal of Ophthalmic Research* 2017; 3(1): 220-225 Available from: URL: <http://www.ghrnet.org/index.php/ijor/article/view/2018>

### INTRODUCTION

Choroidal excavation is a new optical coherence tomography (OCT) finding firstly described by Jampol *et al*<sup>[1]</sup>. Later, it was called focal choroidal excavation (FCE) by Margolis *et al*<sup>[2]</sup>. FCE is characterized by focal concavity at the RPE-choriocapillaris line in the choroid in an OCT scan, with a good visual acuity, and normal appearance of the overlying retina. In the FCE cases, there is no history of trauma and current or prior retinal or choroidal vascular or infectious disease<sup>[3-5]</sup>. It is considered that this lesion often remains stable<sup>[2-5]</sup>. However, some lesions may be related to the development of CNV<sup>[6-8]</sup>.

It has been reported that FCE may occur in various macular diseases such as age-related macular degeneration (AMD), central serous chorioretinopathy (CSCR), epiretinal membrane

(ERM), choroidal neovascularization (CNV), polypoidal choroidal vasculopathy (PCV), Best vitelliform macular dystrophy (BVMD), Vogt-Koyanagi-Harada Disease (VKHD), multifocal choroiditis and punctate inner choroidopathy, focal retinochoroiditis, foveoschisis, multiple evanescent white dot syndrome, combined hamartoma of the retina and retinal pigment epithelium<sup>[9,23]</sup>.

The FCE is not clearly visible in mono/polychromatic fundus photography, fundus angiography or ophthalmoscopy and it might be only detected using OCT<sup>[2-5]</sup>. Reports on the FCE has been increasing in recent years and the authors emphasize its clinical importance. To our knowledge and only Pubmed search, there are almost 40 publications on the FCE in the literature since firstly described 2006 and the majority of these have been published in the last five years.

In our study, we aimed to determine the frequency and the causes of FCE detected by in spectral-domain (SD) OCT and, to comment on its clinical importance.

## MATERIALS AND METHODS

### Study Design

This study was designed as an institutional, retrospective trial and followed the tenets of the Declaration of Helsinki. The patients underwent complete ophthalmic examination, fundus photography, fundus autofluorescence (FAF) and fundus fluorescein angiography (FFA) (Zeiss FF450 plus IR; Carl Zeiss Meditec, Jena, Germany) and SDOCT. The files were retrieved in the databases of the SDOCT machines at our university hospital that was taken between January 2007 and February 2017.

### Inclusion Criteria

In this study, 632 eyes of the patients with untreated wet AMD (w-AMD,  $n = 368$ ), central serous chorioretinopathy (CSCR,  $n = 214$ ), Vogt-Koyanagi-Harada Disease (VKHD,  $n = 3$ ), adult vitelliform macular dystrophy (AVMD,  $n=32$ ), Best vitelliform macular dystrophy (BVMD,  $n = 2$ ) and other macular dystrophies and specific maculopathies ( $n = 13$ ) were retrospectively reviewed for the presence of the FCE. 228 fellow (unaffected) eyes without obvious macular pathology of the cases which have no bilateral involvement were also reviewed as control cases in this study. OCT scanings of these were not different than healthy cases with no retinal pathology. Ten eyes of ten patients having FCE were included in this study. The medical records of ten eyes of these patients with FCEs detected by SDOCT were reviewed. The causes, frequency, associated diseases, types, shape, associated SDOCT pathologies, SDOCT findings of the FCEs in ten cases were evaluated and commented on.

### Exclusion Criteria

The patients with pathologic myopia, angioid streaks, intraocular inflammation, history of previous ocular trauma, intraocular surgery and macular laser photocoagulation and the patients without clear OCT image were excluded from the study. Additionally, the patients having bilateral macular involvement were also excluded because the unaffected fellow eyes of only the patients with unilateral involvement were used as controls.

### OCT imaging and the measurement

In our study, SDOCT scans obtained from two different trades of SDOCT were retrospectively reviewed for the presence of the FCE: OPKO OCT (OTI/OPKO Inc, Toronto, Canada) and Zeiss Cirrus HD-OCT (Cirrus HD-OCT 5000, Carl Zeiss Meditec Inc, Dublin, CA, USA). OPKO OCT has transverse and axial resolutions of 20 and 5–6  $\mu\text{m}$ , respectively. Cirrus HD-OCT has transverse and axial

resolutions of 15 and 5  $\mu\text{m}$ , respectively. The acquisition speeds of both devices were set at 28000 A-scan/sec. Every B-scan of both devices was composed of 512 A-scans. The measurement mode of the Macular Cube in Cirrus D-5000 SDOCT device has acquisition of 128 B-scans each composed of 512 A-scans.

*In vivo* OCT images with high-resolution were obtained and analyzed for the presence of FCE. Additionally, the SDOCT characteristics and types of the FCE were also evaluated in all SDOCT scans obtained. The FCE was defined as the presence of a focal intrachoroidal concavity or downward deflection of the Bruch's membrane-retinal pigment epithelium (RPE)-choriocapillaris complex line in the macula detected in SD-OCT scans analyzed by a single retina specialist (BT), without the evidence of a posterior staphyloma or scleral ectasia<sup>[2-5]</sup>.

The FCEs were categorized into two types described by Wakabayashi *et al* as "nonconforming and conforming" depending on the separation between the neurosensory retina and RPE, and three types described by Shinjima *et al* as "cone-shaped type, bowl-shaped type and the mixed type"<sup>[24,25]</sup>.

## RESULTS

The FCE was detected in ten eyes of ten cases. The associations and frequencies of the FCE were given in Table 1. Of the patients with the FCE, 4 (40%) were women and 6 (60%) were men. The ages of patients were ranged between 63 to 76 years (median 66.1 years). Bilateral FCE was not detected. In fellow eyes, FCE was not detected in any case. The FCE was detected in one eye with w-AMD (1/368 patients, 0.0027%), in two eyes with active CSCR (2/214, 0.0093%), in one with AVMD (1/32 patients, 0.03125%), in one eye with torpedo maculopathy (1/1) and in 5 eyes of the control (idiopathic FCE; 5/228, 0.02193%).

The SDOCT scans of ten patients with FCE were given in Figure 1 (A-J). The sample OCT images from two FCE types were demonstrated in Figure 2 (A and B).

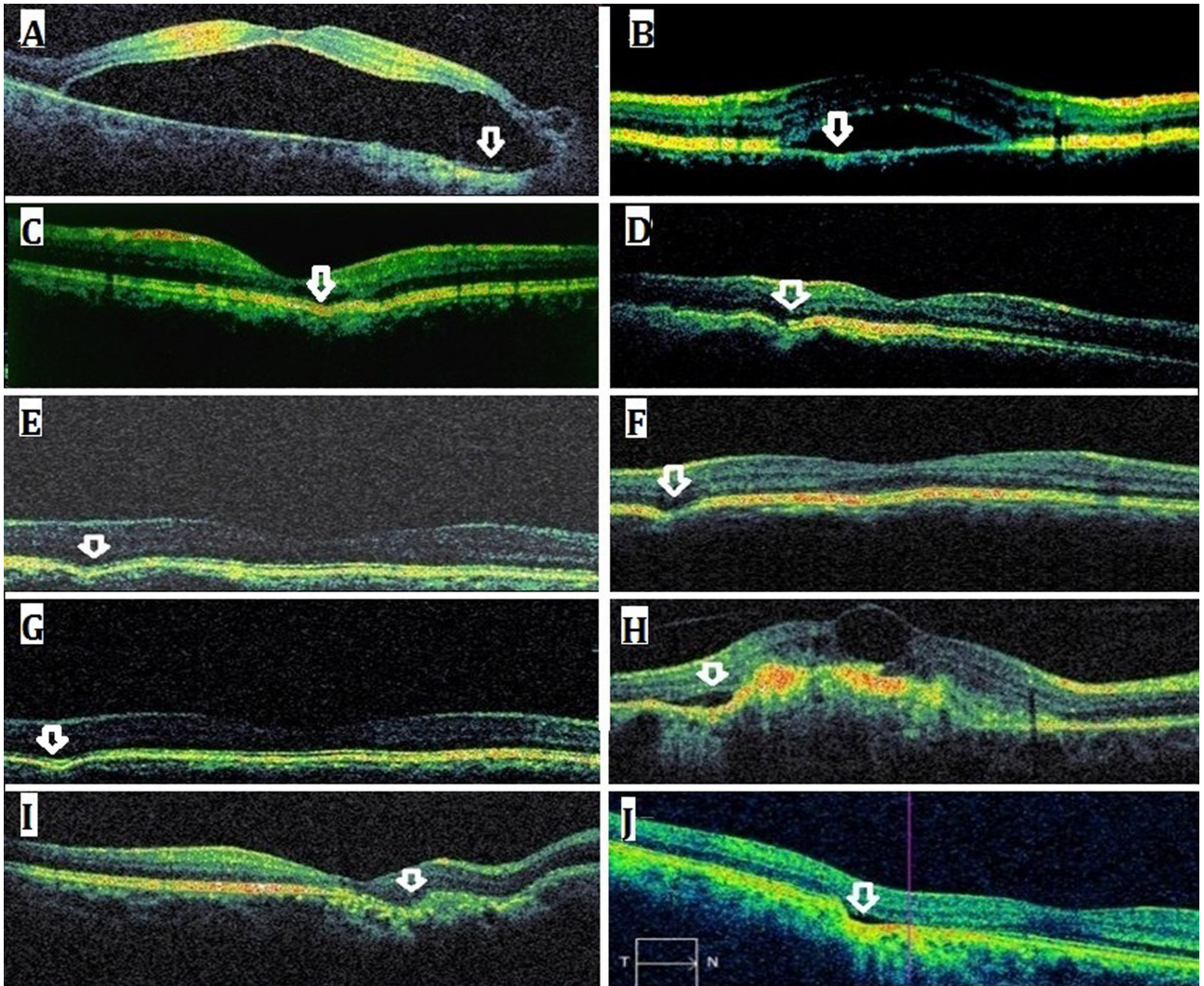
In one of two cases with active CSCR, FCE was located at nasally next to the optic disc. The overlying retina was thinned. Its type was nonconforming (Figure 1A). In the other one had FCE located in the fovea and beneath serous macular detachment. In that case, the overlying retina had a normal appearance. Its type was also nonconforming (Figure 1B).

In the case with w-AMD, FCE was located extrafoveally next to the neovascular scar tissue and its type was nonconforming (Figure 1H).

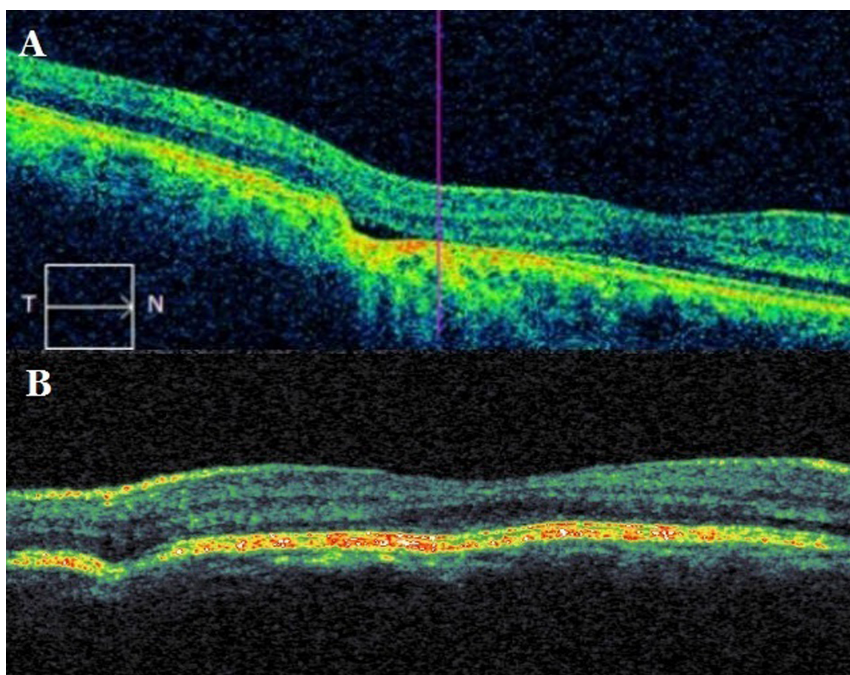
In the case with torpedo maculopathy, FCE was located extrafoveally in the temporal portion of the lesion and its type was nonconforming (Figure 1I, 2A and 3).

**Table 1** The associations, frequencies, and types of FCE. FCE; focal choroidal excavation; w-AMD, wet-type age-related macular degeneration; CSCR, central serous chorioretinopathy; AVMD, adult vitelliform macular dystrophy.

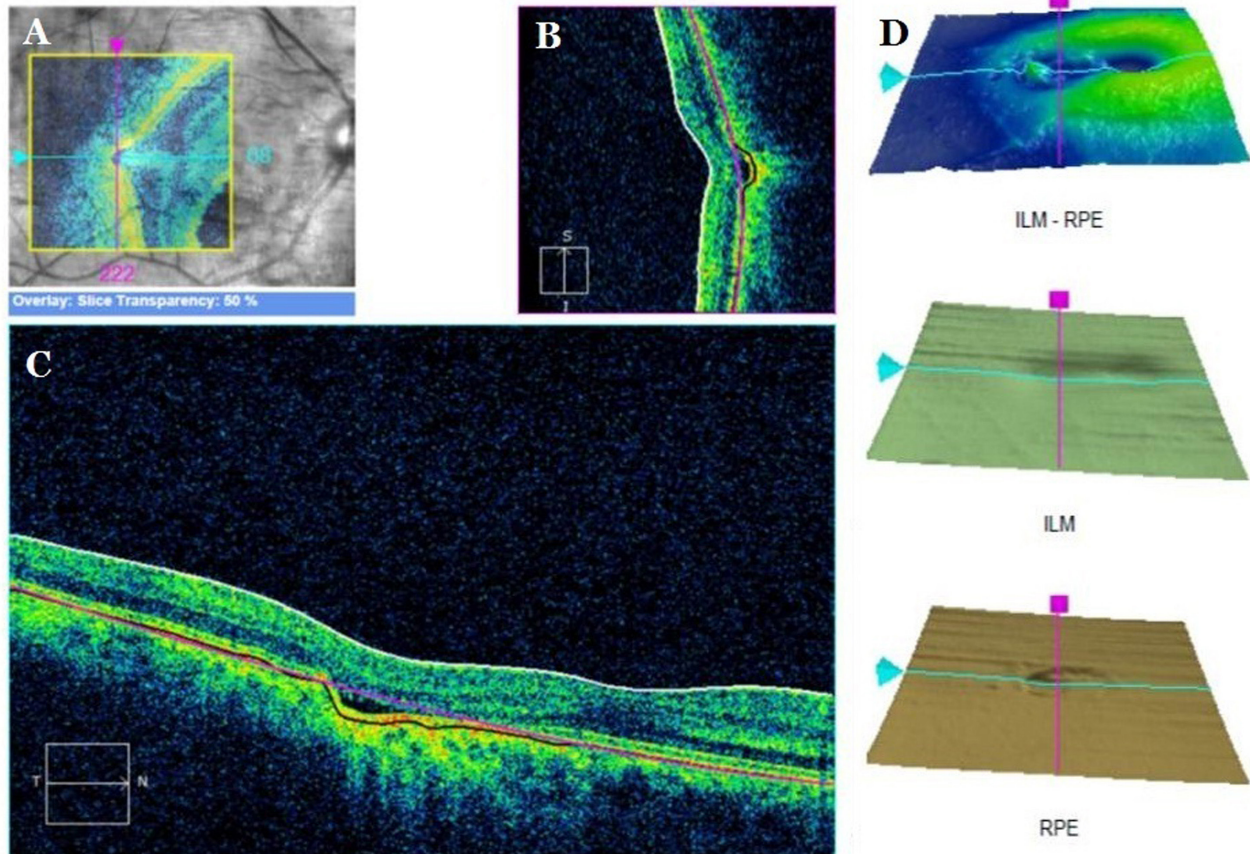
Association of FCE	Numbers (%)	Frequency (%) in cohort	Type of FCE based on two classification system
CSCR	2 eyes (20%)	2/214 patients (0.0093%)	Nonconforming; Bowl-shaped
w-AMD	1 eye (10%)	1/368 patients (0.0027%)	Nonconforming; Bowl-shaped
AVMD	1 eye (10%)	1/32 patients (0.03125%)	Conforming; Cone-shaped
Idiopathic	5 eyes (50%)	5/228 controls (0.02193%)	Conforming; Cone-shaped
Torpedo maculopathy	1 eye (10%)	1/1 patient	Nonconforming; Bowl-shaped



**Figure 1** SDOCT scans of ten patients with FCE: CSCR (A, B), idiopathic (C-G), AMD (H), AVMD (I) and torpedo maculopathy (J). The arrows indicate FCE. SDOCT; spectral domain optical coherence tomography, FCE; focal choroidal excavation, CSCR; central serous chorioretinopathy, AMD; age-related macular degeneration, AVMD; adult vitelliform maculopathy.



**Figure 2** Types of FCE in OCT scans: Nonconforming FCE (A) and conforming FCE. FCE; focal choroidal excavation; SDOCT; spectral domain optical coherence tomography.



**Figure 3** The en face imaging (A), vertical (B) and horizontal (C) scans, and sections (D) at different layers of FCE in SDOCT. FCE: focal choroidal excavation; SDOCT: spectral domain optical coherence tomography.

In the case with AVMD, it was located in the foveal region with the normal appearance of overlying retinal tissue. It was conforming FCE (Figure 1I).

Of 5 eyes with normal appearance of overlying retinal tissue and without obvious maculopathy, in 4, FCEs were located extrafoveally while one was in the fovea. In cases with idiopathic FCE, their type was the conforming type (Figure 1C-G).

We observed that the choroidal tissue beneath the FCE in eyes with w-AMD, CSCR, AVMD, torpedo maculopathy and 2 eyes with idiopathic FCE has with internal hyper-reflectivity while as the choroidal reflectivity in 3 of the eyes without obvious macular pathology was normal or nearly normal.

According to the classification described by Shinjima *et al*, the FCEs in the cases without macular pathology (idiopathic cases) and that in the case with AVMD were cone-shaped type while as the remaining cases had bowl-shape type FCE.

We could not measure the choroidal thickness and imagine the external limiting membrane because of poor imaging quality. Thus, we failed to evaluate the choroidal or scleral thickness. However, we observed that the junction between inner segment and outer segment of photoreceptors was healthy in the case with CSCR and in the cases without macular pathology.

## DISCUSSION

Focal choroidal excavation is a new choroidal entity, which can only be detected with OCT. It was firstly described by Jampol *et al* using OCT in 2006<sup>[1]</sup>. In 2011, this tomographical entity was called as FCE by Margolis *et al*<sup>[2]</sup>. FCE is defined as a focal area of

intrachoroidal concavity in the macula detected in OCT in an eye with preserved visual acuity and normal fundus without a posterior staphyloma or scleral ectasia, a history of trauma, inflammatory/infectious or vascular disease in retina or choroid<sup>[2-5, 26-33]</sup>. Although the FCE is a relatively stationary lesion, it can cause the CNV<sup>[6-8]</sup>. FCE affects Bruch's membrane-RPE-choriocapillaris complex line and photoreceptors<sup>[11, 26-33]</sup>.

The pathogenesis of the FCE is not well defined. However, it is considered that FCE could be an embryonic developmental disorder or an acquired pathology. Some authors hypothesize that FCE is a congenital malformation while as others theorize that it might due to the failure of chorioretinal development in the embryonic stage, micro staphyloma, congenital focal choroidal dysplasia, focal choroidal atrophy caused by congenital or acquired choroiditis<sup>[2, 3, 11, 26-33]</sup>.

Focal choroidal excavation often does not appear in mono/polychromatic fundus photography, fundus angiography or ophthalmoscopy and it might be only detected using OCT. According to OCT findings, FCE has been firstly classified by Wakabayashi *et al* into two types as nonconforming and conforming depending on the separation between the neurosensory retina and retinal pigment epithelium (RPE)<sup>[24]</sup>. The nonconforming type exists a separation between the outer retina (photoreceptor tips) and the RPE within the excavation while as in the conforming type, outer retina (photoreceptor tips) are attached to the apical surface of the RPE. The nonconforming type is frequently associated with CSCR having localized neurosensory detachment.

In a recent study, Shinjima *et al*<sup>[25]</sup> morphologically classified FCEs according to its appearance in OCT. They described three types of the FCE including cone-shaped type having a tethered appearance;

## Turgut B. The focal choroidal excavation

bowl-shaped type with RPE irregularity in whole FCE and the mixed type having findings of both cone-shaped and bowl-shaped types<sup>[25]</sup>.

In recent studies on the frequency and associations with FCE, it was found that FCE might be idiopathic or might be associated with especially CSCR, AMD and PCV<sup>[5, 9-25]</sup>. Obata *et al* demonstrated in a study which 21 eyes with the FCE were retrospectively reviewed that the cause of the FCE was one case with CSCR and one case with CNV<sup>[5]</sup>. In another study, Shinjima *et al* studied FCE with 29 eyes of 26 patients and they reported that the associated diagnoses were AMD in 5 eyes, CSCR in 6 eyes, idiopathic CNV in one eye, idiopathic FCE in 8 eyes, and PCV in 9 eyes in their cohort<sup>[25]</sup>. In our study, we observed that of 10 cases, five were idiopathic FCE.

In a recent study, including the largest case number, 37 eyes with the FCE of 32 patients, Liu *et al* reported that FCE associated with CSCR was found in four cases<sup>[26]</sup>.

In another study including 13 eyes with FCE, Margolis *et al* demonstrated that FCE associated with CSCR was observed in only one case<sup>[2]</sup>. In a study, Luk *et al* reported that FCE was found at 6.0% in patients with CSCR<sup>[11]</sup>.

In our study, we found the frequency of FCE in the CSCR patients as 0.0093% (2/214). This lower frequency of FCE than that which we expected may be related to racial differences. In our study, we detected FCE in only one case with wet AMD. In that case, FCE was located at next to vascular RPE detachment and it was Type 1 FCE.

The several limitations of our study are the small number and the lack of follow-up of the cases of FCE, the retrospective nature of the study and single ethnicity of the cases.

Although the FCE is an interesting and popular OCT finding, we think that it is a not common entity and that it occurs sporadically in CSC and wet AMD. Currently, the importance of this OCT entity in retinal and choroidal diseases has not been fully identified. The most common OCT findings affecting vision in most macular diseases are subretinal/intraretinal fluid or hemorrhage, hyper-reflective dots, confluent and soft drusen and atrophy involving fovea. When the FCE was compared with these and considering its infrequency, we consider that the FCE might be lesser important cause for visual loss unless it has large dimensions and foveal location. However, in a minority of the cases, FCE may be related to the development of CNV. Thus, we consider that it should be monitored for the development of CNV in the case of its detection in OCT scan.

In conclusion, it seems that FCE is a very rare entity. OCT is essential to detect the FCEs. FCE may be detected in various macular chorioretinal diseases. However, it should also be considered that FCE might be idiopathic. Further studies are needed to understand the importance of the presence as an SDOCT finding, the pathogenesis and natural clinical progress and long-term prognosis of FCE.

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Peer reviewer: Fei Yuan