Focal choroidal excavation: Cause or effect?

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We report the imaging characteristics of focal choroidal excavation (FCE) and associated choroidal neovascular membrane (CNVM) and interpret the probable etiopathogenesis of FCE through findings in four patients detected by spectral-domain optical coherence tomography (SD-OCT). FCE was found as an acquired entity in two of our cases subsequent to treatment of CNVM, whereas in the two other cases FCE was pre-existing. Furthermore, association of FCE with pachychoroid spectrum is reaffirmed through this case series.

Key words: Choroidal neovascular membrane, focal choroidal excavation, pachychoroid

Focal choroidal excavation (FCE) is a concavity in the choroid of unknown etiology, occurring without any adjacent scleral abnormality or ectasia, and characterized by good visual acuity and shows minimal changes over time. It was first described with time-domain optical coherence tomography (OCT) by Jampol et al. in 2006.[1]

FCE is believed by most authors to be a congenital condition, though its etiopathogenesis is yet unclear. FCE is detected primarily as an OCT finding. It is frequently associated with pachychoroid diseases, including central serous chorioretinopathy (CSCR), choroidal neovascular membrane (CNV) and polypoidal choroidal vasculopathy (PCV). Although FCE is classically thought to be congenital, acquired FCE forms possibly exist.[2]

To our knowledge, the known reports of acquired FCE following CNVM was by Lee et al.,[3] where a small choroidal excavation developed during CNV scarring changes in age-related macular degeneration, whereas another single case report by Hashimoto et al.[3] was secondary to multifocal choroiditis.

Here, we report four cases of FCE associated with CNVM, including two of acquired FCE post treatment of CNVM, one with pre-existing FCE, which later increased in size over time after treatment of CNVM, and one where the FCE lesion remained stable over the follow-up period.

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Case Reports

Case 1
A 17-year-old female presented with metamorphopsia and blurred vision in right eye (OD) since 1 week. Her best corrected visual acuity (BCVA) was 6/9;N8 in OD and 6/6;N6 in left eye (OS). Clinical examination revealed findings suggestive of choroidal neovascular membrane (CNVM). Her spectral-domain optical coherence tomography (SD-OCT) showed an active Type 2 CNVM associated with a FCE, which was confirmed by fundus fluorescein angiography (FFA). She was treated with multiple intravitreal antiVEGF injections. At her follow-up visits there was significant CNVM resolution, but FCE was progressively increasing in depth, as measured from a horizontal line drawn at the level of RPE to the inner margin of FCE, from 121 µm to 256 µm over the period of 2 years, possibly due to the contraction of the underlying scar [Fig. 1].

Figure 1: (a) Color fundus photograph showing serous macular detachment with grayish membrane and overlying hemorrhage. (b, c) Fundus fluorescein angiography early and late phase showing classic CNVM leak. (d) Enhanced depth imaging showing thick choroid (480 µm). (e, f) SD-OCT of FCE with CNVM at presentation and posttreatment with antiVEGF therapy.

Case 2
A 21-year-old male presented with distortion of vision in OS noted accidentally since 8 days with BCVA of 6/6;N6 in both eyes. Fundus examination revealed an extrafoveal grayish membrane with overlying hemorrhages. Enhanced depth imaging showed pachychoroid with thickness of 443 µm. SD-OCT revealed CNVM associated with FCE. AntiVEGF monotherapy was given with complete resolution of CNVM, but the FCE remained stable in size and configuration during subsequent follow-ups. Final BCVA was maintaining to 6/6; N6 in both the eyes [Fig. 2].

Case 3
A 48-year-old female presented to us with blurring and distortion of vision in OS since 2 weeks. Her BCVA was 6/6; N6 in OD and 6/24;N10 in OS at presentation. Fundus examination showed an extrafoveal CNVM. Based on multimodal imaging including SD-OCT and FFA, patient was diagnosed to have

Figure 2: (a) Color fundus photograph showing subretinal grayish membrane with hemorrhage suggestive of active CNVM. (b, c) FFA early and late phase showing staining of occult CNVM. (d) Enhanced depth imaging showing thick choroid (443 µm). (e, f) SD-OCT images show CNVM at the site of excavation with the size of FCE (269 µm) remaining almost constant during follow-up.

Figure 3: (a) Color fundus photograph showing grayish membrane surrounded by hemorrhage. (b) Fundus fluorescein angiography showing hyperfluorescence. (c) SD-OCT at presentation showing active CNVM features of subretinal fluid and hyper-reflectivity without FCE. (d, e, f) Serial SD-OCT images showing development of acquired FCE during the treatment course.

Figure 4: (a) Color fundus photograph showing orangish mass (choroidal osteoma) with hemorrhages. (b) FFA showing staining of osteoma and blocked fluorescence. (c) Enhanced depth imaging of the osteoma. (d) B-scan image showing highly reflective choroidal mass. (e) SD-OCT showing an active CNVM. (f) Development of subfoveal FCE (arrow) during the treatment course.
type-2 CNVM with subfoveal choroidal thickness of 505 µm. She was also treated with antiVEGF monotherapy on pro re nata (PRN) basis and had multiple recurrences over 5 years. Interestingly, she developed FCE at the site of resolved CNVM scar, during 23 months of treatment, which progressively increased from 85 µm to 154 µm in depth over the subsequent follow-ups. We report this as a case of acquired FCE secondary to CNVM [Fig. 3].

Case 4
A 46-year-old male patient presented to us with blurring of vision OS since 2 months. His BCVA was 6/6, N6 in OD and 6/12, N8 in OS at presentation. Fundus examination of OS showed an irregular elevated, yellowish-white choroidal lesion with well-defined geographic borders involving the inferior macula with areas of subretinal hemorrhage at and above the fovea suggestive associated choroidal neovascular membrane. SD-OCT showed a dense hyper-reflective choroidal mass with significant widening of the choroidal layers. B-scan ultrasonography revealed high-spike echoes through the lesion suggesting choroidal osteoma. FFA in that area showed staining conforming to the diagnosis of choroidal osteoma with a secondary CNVM. Patient underwent treatment with multiple antiVEGF injections and standard fluence photodynamic therapy (PDT). The patient developed an acquired FCE after 17 months of treatment [Fig. 4].

Discussion
Several authors have postulated that FCE lesions are congenital choroidal abnormalities related to developmental defects or some other type of focal structural defect within the choroid.[1,2,4,5]

Ellabban et al.[4] and Hashimoto et al.[3] suggested: the OCT finding of an “unusual hyper-reflective tissue” beneath some FCE lesions could represent scarring of choroidal connective tissue from a previous inflammatory process. Presumably, scar contraction could draw the choroid and RPE toward the sclera producing FCE. Our cases support the above alternative etiopathogenesis leading to an acquired subtype of FCE in addition to the known congenital form of FCE complicated secondarily by CNVM/CSCR.

The occurrence of FCE in eyes with increased choroidal thickness has been noted by several investigators.[6,7] Our cases also had increased choroidal thickness with pachyvessel/s adjacent to the excavation, reaffirming the fact that FCE is associated with the pachychoroid spectrum.

Conclusion
To conclude, FCE is an OCT-based diagnosis. FCE is known to be associated with pachychoroid spectrum of diseases, with known associations being CSCR and CNVM. Origin of FCE can be of two types: congenital and acquired. The significance of the acquired variety remains unknown until more such cases are reported. Periodic monitoring of such cases is of importance to detect and treat any of the known associations.

Declaration of patient consent
The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest
There are no conflicts of interest.

References