Focal choroidal excavation and giant choroidal cavern in an eye with pachychoroid

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Abstract:
Focal choroidal excavation (FCE) is a localized excavation of the choroid, which can be diagnosed by enhanced depth optical coherence tomography (OCT). Choroidal caverns are focal cavitation areas in the choroid which appear hyporeflective on OCT. These are angular or round, empty spaces with posterior tail of hypertransmission. A 47-year-old female presented to us for a routine eye check-up. The best-corrected visual acuity was 20/20 in both the eyes. On fundus examination, a localized pigmented lesion was seen in the right eye inferior to the optic disc, while the fundus of the left eye was normal. Spectral-domain OCT scan through the lesion showed an FCE and a giant cavern with a posterior tail of hypertransmission. The diameter of the cavern was 977 µ ×264 µ, with a subfoveal choroidal thickness (SFCT) of 360 µ. The SFCT of the other eye was 300 µ. Coexistence of FCE and cavern in an eye with thickened choroid is not yet reported in the literature, and their coexistence provides possible insight into the formation of FCE and caverns in the setting of thickened choroid.

Keywords:
Cavern, choroidal cavern, focal choroidal excavation, pachychoroid disease spectrum

Introduction
Focal choroidal excavation (FCE) is an excavation of the choroid seen mostly in patients with good visual acuity and a normal-appearing overlying retina. FCE can be easily identified on a routine optical coherence tomography (OCT). The use of multimodal imaging in routine clinical practice has helped clinicians to identify and diagnose these subtle retinal and choroidal pathologies. It is mostly congenital but can be acquired. Choroidal caverns are focal cavitation areas in the choroid which appear hyporeflective on OCT. There are angular or round, empty spaces with punctate or linear internal hyperreflectivity present in the Sattler and Haller layers. It is presumed to form as a result of ischemic or inflammatory pathology. Both FCE and choroidal caverns have been described separately in the literature with pachychoroid eyes.

Case Report
A 47-year-old female presented to us with complaints of gradual diminution of vision. The best-corrected visual acuity was 6/7.5 in both the eyes. She had a history of diabetes mellitus, heart disease, and systemic hypertension for the last 6 years. Anterior segment was within normal limits in both the eyes. Intraocular pressure by Goldmann applanation tonometry was 14 mmHg in both the eyes. On routine clinical fundus examination, a localized lesion was seen in the right eye inferior to the optic disc, while the fundus of the left eye was within normal limits for the age. Routine investigation was done for the right eye. Clinical fundus photograph
of the right eye showed a localized lesion inferior to the optic disc surrounded by a hyperpigmented border [Figure 1a (and c magnified)]. Multicolor imaging highlighted the lesion as light orangish surrounded by a dark border, which could be well delineated in the red-free image [Figures 1b (and d magnified) and 2a]. The lesion appeared hyporeflectant in the infrared image [Figure 2c], while in the blue autofluorescence, it appeared hypofluorescent [Figure 2b]. The infrared autofluorescence image was insignificant [Figure 2d]. Spectral-domain OCT (SDOCT) scan through the lesion showed an FCE [Figure 3b] and a giant cavern with a posterior tail of hypertransmission and hyporeflective core [Figure 3a] besides it. On SDOCT, Heidelberg, Germany inbuilt caliper software, the diameter of the cavern was 977 μ × 264 μ, with a subfoveal choroidal thickness (SFCT) of 360 μ. The SFCT of the other eye was 276 μ.

**Discussion**

FCE was first described by Jampol et al. in 2006. In 2010, Wakabayashi et al. described three cases of “unilateral choroidal excavation,” which was later on termed as “FCE” by Margolis et al. Two types of FCE have been described; conforming and nonconforming. “Conforming FCE” has a direct contact between the photoreceptors and retinal pigment epithelium (RPE). They show preservation of the ellipsoid zone and RPE within the lesion, while in “nonconforming FCE,” the photoreceptor tips are detached from the underlying RPE. The intervening space is thought to represent the subretinal fluid with varying amounts of shed outer segment debris or inflammatory material. These lesions are congenital choroidal abnormalities, which are a result of some developmental defects or focal structural defect within the choroid. FCE seen in adults is thought to be mostly acquired. Pachychoroid features have been described in eyes with FCE such as increased SFCT and choroidal vascular hyperpermeability in indocyanine green angiography (ICGA). It has been found to be associated with many diseases, such as central serous chorioretinopathy, choroidal neovascularization, and nonneovascular age-related macular degeneration.

Choroidal caverns were first seen in eyes having geographic atrophy in patients having age-related macular degeneration. They have also been reported in best vitelliform dystrophy. They appear as hyporeflective cavities in areas with no choroidal vessels and are presumed to arise from nonperfused ghost vessels and persistence of stromal pillars, where the vessels were originally situated. Sakurada et al. found choroidal caverns in the areas traversed by pachyvessels. This was detected in 52% of eyes with pachychoroid disease. They hypothesized that it might indicate a loss of normal choroidal architecture with dilated Haller layer veins and increased choroidal thickness. It is possible that trauma leading to choroidal rupture may cause focal areas of choroidal thinning akin to FCE. Using multimodal imaging, Dolz-Marco et al. defined the characteristics of caverns, which are: (a) nonreflective spherical to polyhedral structures visible on en face and cross-sectional OCT; (b) posterior tail of hypertransmission; (c) in cases of RPE loss, frequently hyperreflective on near-infrared and rarely reflective on color photographs or hyperfluorescent on ICGA; (d) not visible on fluorescein angiography or
fundus autofluorescence; and (e) no evidence of flow signal on en face or cross-sectional OCT angiography (OCTA).[11]

The formation of FCE may be caused by the formation of a giant cavern due to an ischemic or inflammatory insult in thick or pachychoroids having dilated pachyvessels, with subsequent collapse of the overlying structures. The presence of a giant choroidal cavern and FCE together in a pachychoroid eye in our case report indicates a relationship between all these three entities. A further look into a larger pool of patients in the future studies would be useful to bring out even more features of this novel association. The limitation of the present report is a lack of OCTA.

**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.

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