

Case report

Development of focal choroidal excavation in non-neovascular age related macular degeneration with pachy-choroid features

Touka Banaee^{a,b}, Lance J. Lyons^a, Jaafar El-Annan^{a,c,d,*}

^a University of Texas Medical Branch, Galveston, TX, USA

^b Mashhad University of Medical Sciences, Mashhad, Iran

^c University of Texas at MD Anderson, Houston, TX, USA

^d Blanton Eye Institute, Houston Methodist Hospital, Houston, TX, USA

Received 22 June 2019; revised 10 August 2019; accepted 16 August 2019

Available online 17 September 2019

Abstract

Purpose: To present a documented case of development of focal choroidal excavation (FCE) in non-neovascular age related macular degeneration (AMD).

Methods: An 86-year-old female with pachy-choroid was followed clinically for non-neovascular AMD. Successive optical coherence tomography (OCT) scans were reviewed with tracking software.

Results: Over the course of follow-up development of a FCE adjacent to a pachy-vessel along with disappearance of the pachy-vessel was documented in OCT.

Conclusions: This case is a documented development of FCE in an eye with pachy-choroid features. The possible mechanism in this scenario may be thrombosis of pachy-vessels.

Copyright © 2019, Iranian Society of Ophthalmology. Production and hosting by Elsevier B.V. This is an open access article under the CC BY-NC-ND license (<http://creativecommons.org/licenses/by-nc-nd/4.0/>).

Keywords: Focal choroidal excavation; Choroid; Optical coherence tomography; Pachy-choroid; Age related macular degeneration

Introduction

Focal choroidal excavation (FCE), first described by Jampol in 2006,¹ is an outward bowing of Bruch's membrane and retinal pigment epithelium (RPE) usually with a normal overlying retina. The pathogenesis of FCEs is still unclear. Once believed to be congenital in nature, association of FCEs with different categories of retinal and choroidal diseases, such as retino-choroidal inflammations,^{2,3} hereditary retinal dystrophies,⁴ and the pachy-choroid spectrum, has called this

theory into question.⁵ In this report, we present optical coherence tomography (OCT) images of a patient with non-neovascular age related macular degeneration (AMD) showing development of FCE during follow-up, and providing clues to the pathogenesis.

Case report

The patient is an 86-year-old female followed for intermediate AMD with subretinal drusenoid deposits visible on the central OCT B scan (Fig. 1). Her notable past medical and surgical histories are hyperlipidemia, hypothyroidism, diverticulitis, and colon cancer treated only by resection.

At her most recent visit, both eyes (OU) had visual acuity of 20/20, normal intraocular pressures, and anterior segments with pseudophakia. There were no signs of inflammation in the anterior chamber or vitreous cavity. Fundoscopy revealed

The authors have not received any grants or funding for this case report, and have no proprietary interest in the instruments or medications presented in this article.

* Corresponding author. 301 University Blvd., Galveston, TX, 77555, USA.
E-mail address: jaelanna@utmb.edu (J. El-Annan).

Peer review under responsibility of the Iranian Society of Ophthalmology.

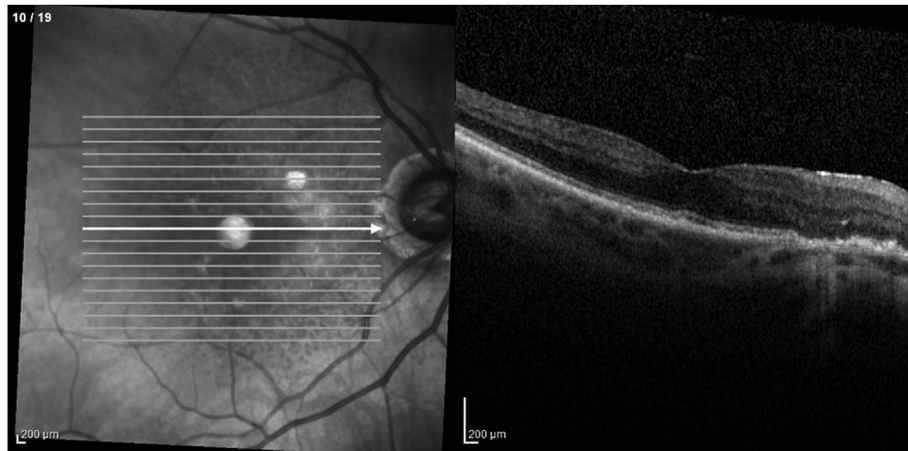


Fig. 1. Central optical coherence tomography (OCT) B scan shows the presence of subretinal drusenoid deposits along with a choroid which is of normal thickness.

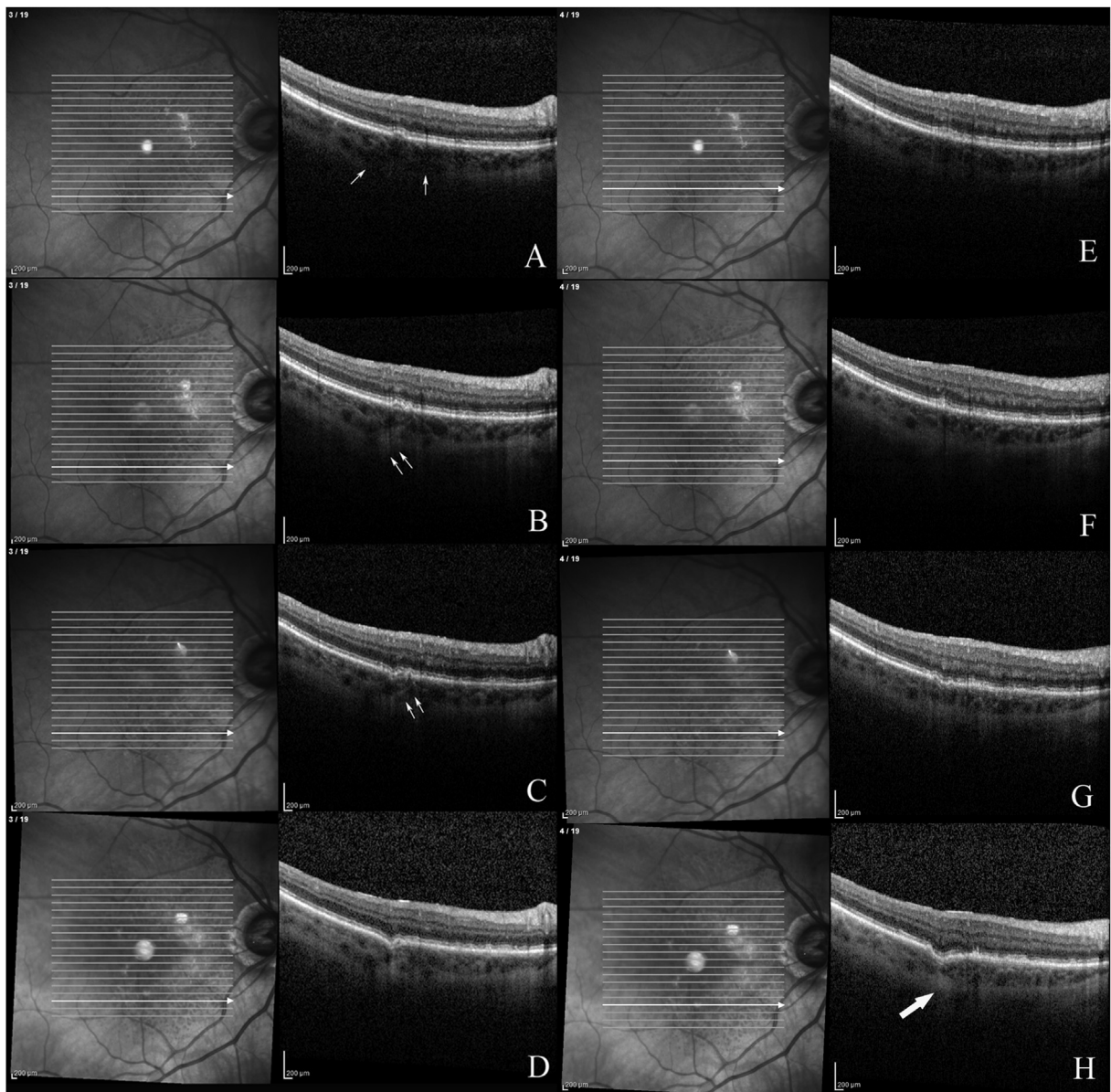


Fig. 2. Successive optical coherence tomography (OCT) B scans through two adjacent locations (A-D and E-H) taken in 2015 (A and E), 2016 (B and F), 2017 (C and G) and 2018 (D and H). There is pigment epithelial thickening and derangement over pachy-vessels in 2015 (A-arrows), replacement of the pachy-vessels by a hyper-reflective tissue in 2016 (B- double arrows) and 2017 (C- double arrows) and development of a focal choroidal excavation (FCE) in 2018 (D and H). The hyper-reflective tissue underneath the FCE has a round shape (H- arrow).

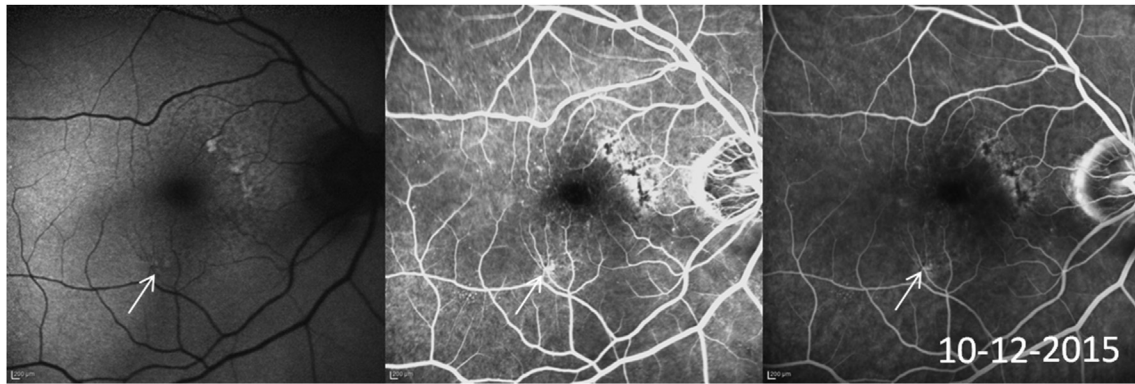


Fig. 3. Auto fluorescence and fluorescein angiogram taken in 2015 show hyper-auto fluorescent spot and window defect at the site that later developed focal choroidal excavation (FCE) (arrows).

subtle pigmentary changes, medium sized drusen, and peripapillary atrophy OU.

There was a FCE in the OCT (Spectralis, Heidelberg engineering, Germany) of the right eye, which was traced back on the same site with Heidelberg Explorer software (Heidelberg engineering, Germany). Three years earlier, the same spot had pigment epithelial changes over pachy-vessels (Fig. 2-A), was hyper-auto fluorescent on auto fluorescence imaging and showed window defect on fluorescein angiography (Fig. 3).

Later OCTs at the same location clearly show development of a hyper-reflective tissue in the outer choroid and disappearance of the pachy-vessels (Fig. 2B, double arrows) culminating in development of a FCE (Fig. 2-D and Fig. 2-H).

Discussion

Although there are reports of documented development of FCE in eyes under treatment for neovascular AMD^{6,7} and after inflammatory choroidal diseases,^{2,3} we are not aware of any documented development of FCE in an eye with non-neovascular AMD.

Pseudodrusen are visible in the infrared images and OCTs of this patient. Although pseudodrusen are typically associated with thin choroid, submacular choroidal thickness in this patient is normal to thick for her age (Fig. 1), so in spite of the presence of pseudodrusen, we believe that this patient had pachy-choroid pigment epitheliopathy before development of FCE. FCEs are associated with pachy-choroid diseases,^{5,8} and pachy-vessels are usually visible in the vicinity of FCEs, but not under the FCE. Underlying FCE, a dense hyper-reflective tissue is usually visible in the choroid.⁵ In the current case, large pachy-vessels are visible in the first OCT B scans (Fig. 2-A, arrows) at the site of future FCE. Development of the hyper-reflective tissue in the outer choroid is associated with disappearance of the pachy-vessels; its rounded shape in the final scans (Fig. 2-H, single arrow) may be inferred as a fibrosed vessel lumen. There is a progressive contraction of the hyper-reflective choroidal tissue and outward bowing of the Bruch's membrane-RPE complex, with resultant FCE. Based on these findings, we suggest thrombosis of large choroidal

vessels as the underlying process in the pathogenesis of this FCE.

Although metastasis from previous colon cancer is a possible differential diagnosis, shrinkage of the hyper-reflective tissue during two years of follow-up, absence of any clinical correlates and the good general health of the patient make this diagnosis a remote possibility.

FCEs in the context of pachy-choroid diseases are often seen at a site of choroidal hyperpermeability.^{5,9} The exact cause of choroidal hyper permeability is still unknown, but leakage has been proposed to stem from the choriocapillaris layer.¹⁰ Hypothetically, both inflammation and ischemia of the choroid can lead to hyper permeability, and both may be associated with thrombosis; in the former the inflammation is the cause, and in the latter the ischemia is the effect of thrombosis. Unfortunately due to the retrospective nature of this case, we do not have fluorescein and indocyanine green angiograms from the time of development of the hyper-reflective tissue to be confident that the process had been thrombotic/ischemic in origin, but disappearance of the pachy-vessels and the round hyper-reflective tissue in the outer choroid are in favor of this etiology. Optical coherence tomography angiography (OCTA) was not available to us, but we believe that OCTA would not be helpful in evaluation of the deeper choroidal vessels involved in this case.

In summary, this case clearly demonstrates the acquired nature of this FCE, and proposes vascular thrombosis in pachy-vessels as a pathophysiologic mechanism in development of FCE in pachy-choroid spectrum of diseases.

References

1. Jampol LM, Shankle J, Schroeder R, Tornambe P, Spaide RF, Hee MR. Diagnostic and therapeutic challenges. *Retina*. 2006;26(9):1072–1076.
2. Hashida N, Fok A, Nishida K. Choroidal excavation in vogt-koyanagi-harada disease. *Case Rep Ophthalmol*. 2014;5(2):222–225.
3. Jabbarpoor Bonyadi MH, Hassanpour K, Soheilian M. Recurrent focal choroidal excavation following multiple evanescent white dot syndrome (MEWDS) associated with acute idiopathic blind spot enlargement. *Int Ophthalmol*. 2018;38(2):815–821.

4. Battaglia Parodi M, Casalino G, Iacono P, Introini U, Adamyan T, Bandello F. The expanding clinical spectrum of choroidal excavation in macular dystrophies. *Retina*. 2018;38(10):2030–2034.
5. Chung H, Byeon SH, Freund KB. Focal choroidal excavation and its association with pachychoroid spectrum disorders: a review of the literature and multimodal imaging findings. *Retina*. 2017;37(2):199–221.
6. Lee CS, Woo SJ, Kim YK, et al. Clinical and spectral-domain optical coherence tomography findings in patients with focal choroidal excavation. *Ophthalmology*. 2014;121(5):1029–1035.
7. Shinojima A, Kawamura A, Mori R, Yuzawa M. Morphologic features of focal choroidal excavation on spectral domain optical coherence tomography with simultaneous angiography. *Retina*. 2014;34(7):1407–1414.
8. Cheung CMG, Lee WK, Koizumi H, Dansingani K, Lai TYY, Freund KB. Pachychoroid disease. *Eye (Lond)*. 2019;33(1):14–33.
9. Lim FP, Wong CW, Loh BK, et al. Prevalence and clinical correlates of focal choroidal excavation in eyes with age-related macular degeneration, polypoidal choroidal vasculopathy and central serous chorioretinopathy. *Br J Ophthalmol*. 2016;100(7):918–923.
10. Dansingani KK, Balaratnasingam C, Naysan J, Freund KB. En face imaging of pachychoroid spectrum disorders with swept-source optical coherence tomography. *Retina*. 2016;36(3):499–516.