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Pachychoroid, an update from the new finding to the usual investigation in selected diseases

Atualização em espessamento de coroide

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ABSTRACT

The authors make an update of pachychoroid in a group of the choroidal-retinal diseases that choroidal thickening is an usualenhanced depth image - optic coherence tomography (EDI-OCT) finding like as central serous chorioretinopathy, pachychoroid neovasculopathy, polipoidal choroidal vasculopathy and pachychoroid pigment epitheliopathy.

Keywords:*Choroid/pathology; Choroidal neovascularization/pathology; Retinal pigment epithelium/pathology; Tomography, optical coherence*

RESUMO

Os autores fazem uma atualização da presença do espessamento de coroide, um achado de tomografia de coerência óptica com imagem de profundidade melhorada (EDI-OCT) empatologias retino-coroidianas como coroido-retinopatia central serosa, espessamento de coroide com neovascularização, vasculopatiacoroidianapolipoidal e espessamento de coróide com epiteliopatia.

Descritores: Coróide/patologia; Neovascularização de coroide/patologia; Epitélio pigmentado da retina/patologia; Tomografia de coerência óptica

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Introduction

Pachychoroid is defined as choroidal thickening and it was related at the first time by Freund in a description of a new disease called pachychoroid pigment epiteliopathy[1] and posteriorly of another disease pachychoroid neovasculopathy[2]. Spaide[3] was the first to visualize choroid in spectral – domain ocular coherence tomography (SD-OCT) using-enhanced depth imaging (EDI) and Immamura et al.[4] related the choroidal findings in cases of central serous chorioretinopathy.

According Freund[1] pachychoroid pigment epitheliopathy (PPE) is a clinical entity characterized by a range of retinal pigment epithelium (RPE) abnormalitiesoverlying the areas of choroidal thickening.

The other related disease by Freund was pachychoroid neovasculopathy[2](PN)a Type 1 neovascularization associated with choroidal thickening . In this entity, patientswith no evidence of AMD, myopic degeneration, orother causes of degeneration develop Type 1 neovascular tissue overlying focal areas of choroidal thickeningand choroidal hyperpermeability Fung et al.[5] showed a series of patients with long-standing CSC who developed Type 1 neovascularization, 36% of which went on to develop PCVPPE and CSC sharea similar pathophysiologic profile, it may be that eyes with long-standing "silent" PPE develop Type 1 neovascularization in the absence of an overt CSC manifestation including submacular exudative detachment or gravitational tracts of chronic SRF.In addition to Type 1 neovascularization, CSC hasalso been shown to be associated with polypoidal choroidal vasculopathy(PCV)[2,5].Originally describe as a primary choroidal pathology, PCV is increasing thought to be a manifestation of long-standing Type neovascularization in AMD and CSC, as well as a varietof other diseases[2]. The strikingly similar characteristicsshared between PCV and CSC, including choroidal hyperpermeability as seen with indocyanine green angiography(ICGA) and increased choroidal thickness as demonstrated with EDI-OCT and histopathology showing dilated thin-walled choroidal vessels in PCV,support the theory that CSC and PCV may be part of pachychoroid-driven disease spectrum in which CSC may develop into Type 1 neovascularization and, ultimately PCV[2].

A new paper comparing pachychorod neovasculopathy and AMD with choroidal thickening was published[6] and the conclusionwas that pachychoroid neovasculopathy was different from neovascular AMD not only phenotypically but also genetically.

Pachychoroid neovasculopathy may represent up to one quarter of diagnosed neovascular AMD cases[6]. Although pachychoroid neovasculopathy often masquerades as neovascular AMD, their etiology is likely to be different because pachychoroid neovasculopathy not shows lack of drusen and the genotype distribution of AMD susceptibility SNPs differed significantly between the two conditions [6]. Pachychoroid neovasculopathy should be distinguished from neovascular AMD in future epidemiological and genetic studies[6].

In the original description of three patients with pachychoroid neovasculopathy[2](PN)using EDI-OCT to measurements the choroidal thickness, the mean subfoveal choroidal thickness in the affected eyes was 310 μm(range 244–407 μm). This was in contrast to the unaffected fellow eyes, in which the mean

Subfoveal choroidal thickness was 172 μm(range, 150–210 μm).We[7] show a case of PNof a 58-year-old white male patient was seen three years ago for impaired visual acuity in his left eye. The visual acuity was 20/25 on the OD and 20/200 in OS. In the ocular fundus examination we noticed reduced fundus tessellation in the OD . A semitranslucent epiretinal membrane created macular distortion. Membrane contraction had pulled the paramacular vessels toward the horizontal raphe in the OS . Fluorescein angiography (FA) of the OD showed window defects at any points. The OS displayed staining in the vascular area in the epiretinal membrane. Spectral domain (SD)-OCT revealed retinal pigment epithelium abnormalities on the OD and an epiretinal membrane with wrinkling of the inner retina on the OS.

Two years after epiretinal membrane surgery on the left eye, the patient presented with a loss of visual acuity in his right eye. His vision in the right eye had decreased to 20/40; however, the visual acuity in the left eye had improved to 20/100. A multimodal evaluation was performed. Color photographs of the OD showed progress in terms of reduced fundus tessellation . In contrast to the first visit showing paramacular vessels pulled toward the horizontal raphe, FA of the OD indicated poorly demarcated leakage, and regularity of the retinal capillaries in the OS (Figure 1). SD-OCT of the OD revealed small pigment epithelial detachments and subretinal fluid (Figure 2). In the OS, the epiretinal membrane and wrinkling of the inner retina persisted. By EDI-OCT, the subchoroidal thickness in the affected eye was 247 μm and 165 μm in the OS (Figure 3).

Fundus autofluorescence was used to detect any hipoautofluorescence points on the OD. No abnormalities were observed in the OS. The results of an indocyanine green angiographic analysis of the area of hyperfluorescence in the OD were consistent with leakage from a Type 1 occult choroidal neovascularization (Figure 4).

We show cases of central serous chorioretinopathy (CSCR) in figures 5-11.

The occurrence of PCV in a initial PN case was related in two cases of Freund original paper[2]. New images of pachychoroid cases with Em Face ,swept souce[8]and optical coherence tomography angiography of shallow irregular pigment epithelial detachments[9] were published.

The treatment of CNV is with anti-VEGF drugs , when the evolution is for PCV the PDT treatment can be effective associated or not with anti-angiogenics drugs.

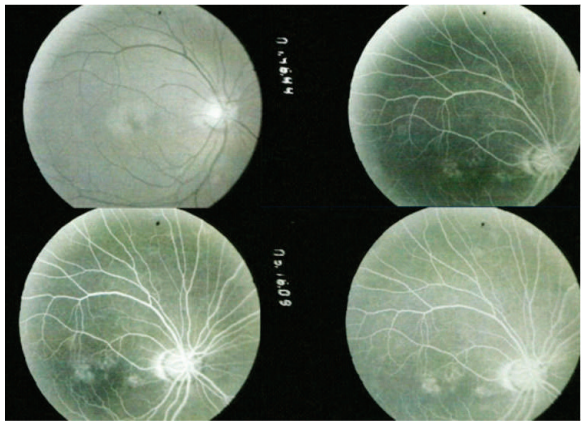

Figure 1: FA demonstrating poorly demarcated leakage in the right eye

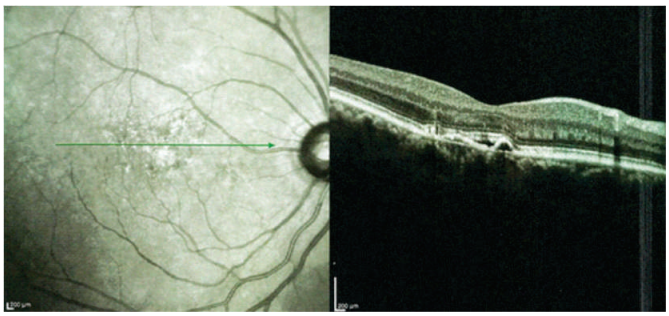

Figure 2: SD-OCT images demonstrating a small PED and subretinal fluid in the right eye

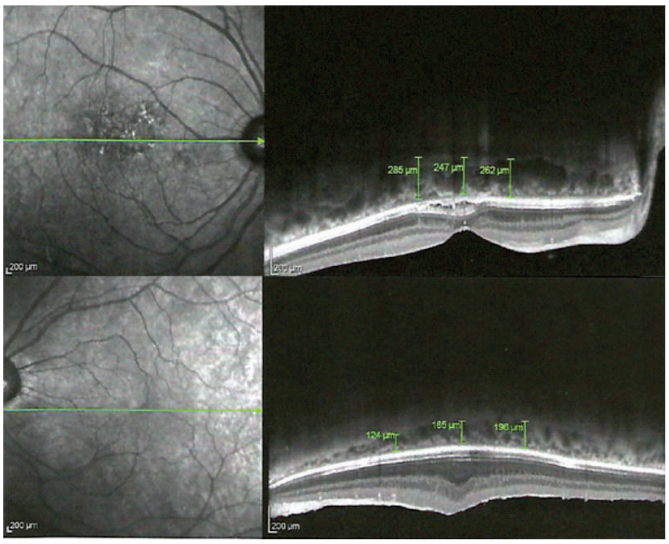

Figure 3: Choroidal thickness in the affected right eye and left eye

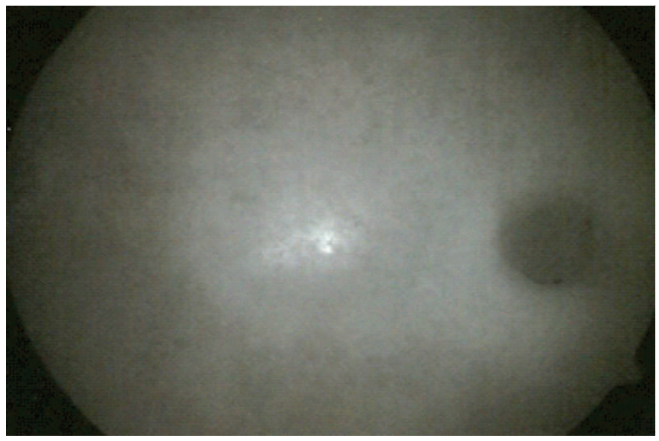

Figure 4: Indocyanine green angiography revealed a region of hyperfluorescence consistent with leakage from a type 1 occult CNV.

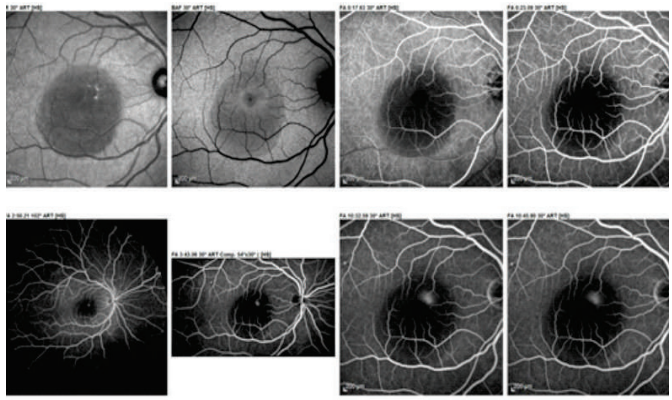

Figure 5: FA in CentralSerousChorioretinopathy (CSCR)

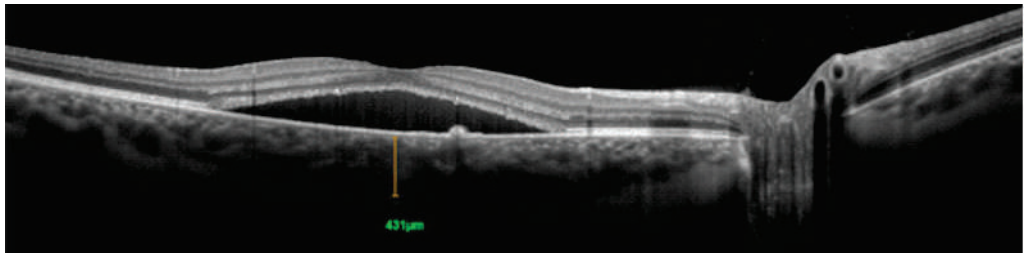

Figure 6: CSCR case's. EDI-OCT measurement of subfoveal choroid thickness and subretinal fluid

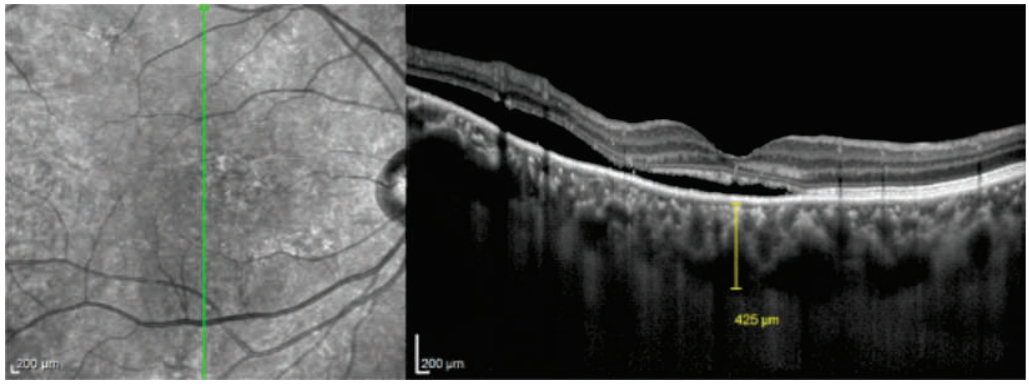

Figure 7: EDI-OCT measurement of subfoveal choroid thicknessandsubretinalfluid from OD of other CSCR case's

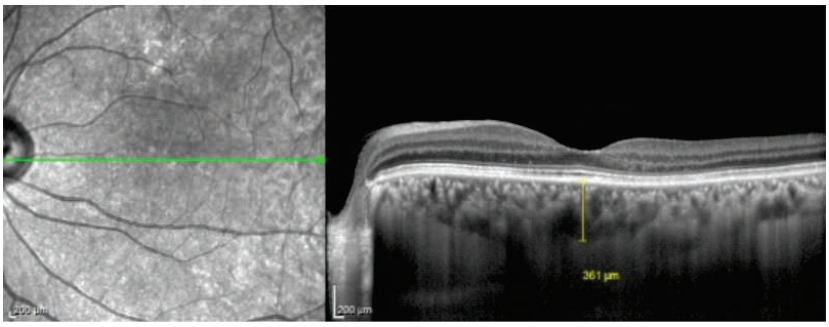

Figure 8: EDI-OCT EDI-OCT measurement of subfoveal choroid thicknessof the OS

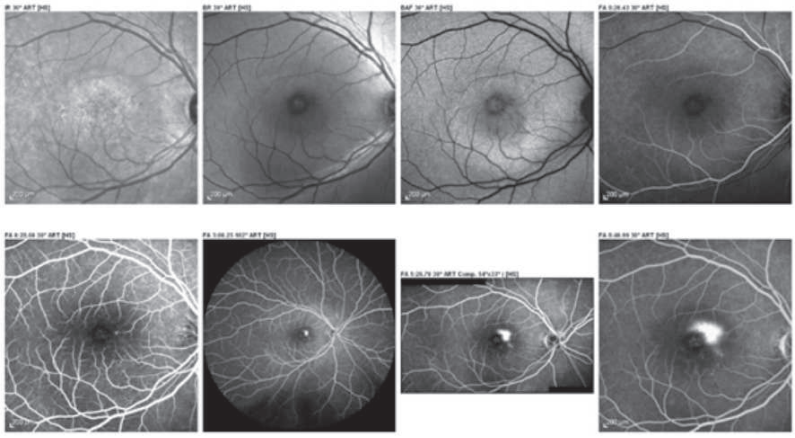

Figure 9: FA of typical CSCR case's

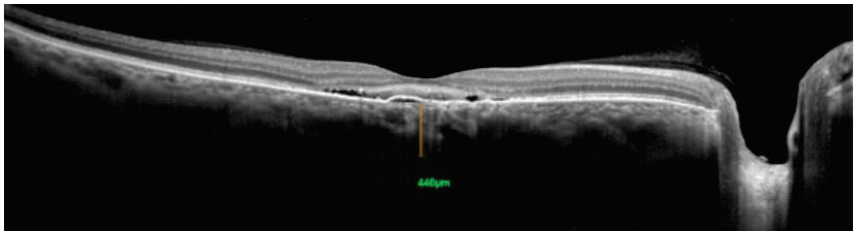

Figure 10: EDI-OCT EDI-OCT measurement of subfoveal choroid thicknesswith subretinal fluid and PED

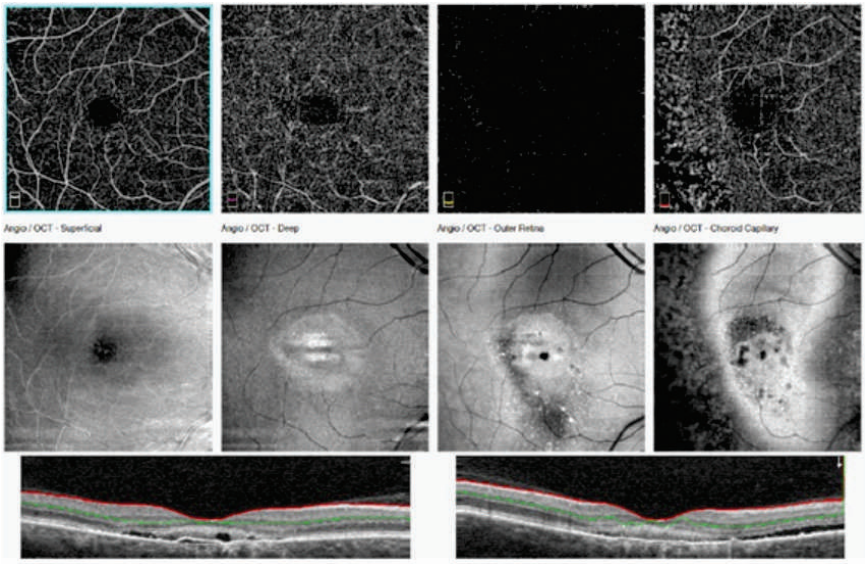

Figure 11: Angio-OCT (OCTA) of the patient; No choroidal neovascularization was detected

References

1. Warrow DJ, Hoang QV, Freund K.B. Pachychoroid pigment epitheliopathy. Retina. 2013;33(8):1659–72.
2. Pang CE, Freund KB. Pachychoroidneovasculopathy. Retina. 2015;35(1):1–9.
3. Spaide RF, Koizumi H,Pozzoni MC. Enhanced depth imaging spectral-domain optical coherence tomography. Am JOphthalmol. 2008;146(4):496–500.
4. Imamura Y, Fujiwara T, Margolis R, Spaide RF. Enhanced depth imaging optical coherence tomography of the choroid in central serous chorioretinopathy. Retina. 2009;29(10):1469–73.
5. Fung AT, Yannuzzi LA,Freund KB. Type 1 (sub-retinal pigment epithelial) neovascularization in central serouschorioretinopathy masquerading as neovascular age-related macular degeneration. Retina. 2012;32(9):1829–37.
6. Miyake M, Ooto S, Yamashiro K , Takahashi A,Yoshikawa M, Akagi-Kurashige Y, et al. Pachychoroidneovasculopathy and age-related macular degeneration.Sci Rep. 2015; 5:16204. doi: 10.1038/srep16204.2015
7. Amaro MH, Belfort Jr, R. Pachychoroidneovasculopathy in a male patient: a case report.ArqBrasOftalmol. 2015; 78(6):385-7.
8. DansiganiKK, Balaratnasigan C, Naysan J, Freund KB. Emface imaging of pachychoroid spectrum disorders with swept source optical cohence tomography.Retina. 2016;36(3):499-516.
9. Dansigani KK,Balaratnasigan C, Klufas MA, SarrafD, Freund BK. Optical coherence tomography angiography of sallow irregular pigment epithelial detachments in pachychoroid spectrum diseases. Am J Ophthalmol. 2015;160(6):1243-54.

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