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 usual enhanced depth image - optic coherence tomography (EDI-OCT) finding like as central serous chorioretinopathy, pachychoroid neovascularopathy, polypoidal 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) em patologias 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 epitheliopathy and posteriorly of another disease pachychoroid neovasculopathy. Spaire was the first to visualize choroid in spectral domain optical coherence tomography (SD-OCT) using enhanced depth imaging (EDI) and Imamura et al. related the choroidal findings in cases of central serous chorioretinopathy.

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

The other related disease by Freund was pachychoroid neovasculopathy (PN) a Type 1 neovascularization associated with choroidal thickening. In this entity, patients with no evidence of AMD, myopic degeneration, or other causes of degenerative development Type 1 neovascular tissue overlying focal areas of choroidal thickening and choroidal hyperpermeability Fung et al. showed a series of patients with long-standing CSC who developed Type 1 neovascularization, 36% of which went on to develop PCV and CSC show a similar pathophysiological 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 has also been shown to be associated with polypoidal choroidal vasculopathy (PCV). Originally describe as a primary choroidal pathology, PCV is increasing thought to be a manifestation of long-standing Type 1 neovascularization in AMD and CSC, as well as a variety of other diseases. The strikingly similar characteristics shared 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.

A new paper comparing pachychoroid neovasculopathy and AMD with choroidal thickening was published and the conclusion was 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. 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. Pachychoroid neovasculopathy should be distinguished from neovascular AMD in future epidemiological and genetic studies.

In the original description of three patients with pachychoroid neovasculopathy (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 show a case of PN of a 58-year-old male patient who developed Type 1 neovascularization and, ultimately, PCV. 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. Fundus autofluorescence was used to detect any hippoautofluorescence 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 OS indicated poorly demarcated leakage, and regularity of the retinal capillaries in the OS. 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.

Fundus autofluorescence was used to detect any hippoautofluorescence 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. 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. New images of pachychoroid cases with Em Face, swept source and optical coherence tomography angiography of shallow irregular pigment epithelial detachments 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 showing 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 Central Serous Chorioretinopathy (CSCR).

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

Figure 7: EDI-OCT measurement of subfoveal choroid thickness and subretinal fluid from OD of other CSCR case's.

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

**Figure 9:** FA of typical CSCR case's

**Figure 10:** EDI-OCT measurement of subfoveal choroid thickness with subretinal fluid and PED

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


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