Outer retinal defects in pachychoroid pigment epitheliopathy

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Disclosures

• Allergan: Advisory board
• Alimera: Advisory board
• Regeneron: Advisory board
Summary

This is a case series of 11 eyes of 8 patients with outer retinal defects in the setting of pachychoroid pigment epitheliopathy. Patients demonstrated focal outer retinal defects (EZ +/- IZ), with an intact RPE. Patients maintained good visual acuity and the defects were stable in patients with long-term follow-up.

At presentation
20/20

9 months later
20/20
• **Purpose:** To describe unique outer retinal defects in patients with pachychoroid pigment epitheliopathy.
Methods

• **Study design:** prospective, observational case series of patients examined between October 2017 and January 2020 at a single center (New York University)

• **Evaluation performed:** comprehensive ophthalmologic examination and imaging: enhanced depth imaging-OCT (Heidelberg Spectralis), fluorescein angiography, OCT-angiography (Zeiss)
Methods

- **Inclusion criteria**: presence of pachychoroid vessels or choroidal thickness ≥390 μm on EDI-OCT, outer retinal disruption of the EZ and/or IZ on SD-OCT, pigmentary changes on fundus examination, history of or concurrent IRF or SRF on OCT.

- **Exclusion criteria**: history of or concurrent IRF or SRF or serous PED on OCT, choroidal neovascularization, typical AMD and macular telangiectasia Type 2.
Results

- 11 eyes of 8 patients
- Gender: 3 female and 5 male
- Mean age: 61.38 years (range 48 – 71)
- Co-morbid conditions
  - Diabetes mellitus type 2: 4 patients
    - 3 eyes with diabetic retinopathy
    - 3 eyes without diabetic retinopathy
  - Hypertension: 4
  - Obstructive sleep apnea: 3
- Mean follow-up: 12.75 months*

*in patients with long-term follow-up (4)
Results

• Mean subfoveal choroidal thickness: 481 ±104 μm (range 320 – 699)
• Mean BCVA: LogMAR 0.14 (approximately Snellen 20/30)
• Foveal involvement: 6 eyes
• Dilated choroidal vessels immediately underlying focal disruption: 4 eyes
• OCT findings:
  • EZ/IZ disruption: 10 eyes
  • IZ disruption only: 1
  • Pachyvessel underlying defect: 4
  • Overlying ELM hyper-reflectivity: 7
  • Transmission defect: 5
Results

EZ/IZ disruption
Results

IZ disruption only
Transmission defect
Results

Pachyvessel underlying defect
Results

ELM hyper-reflectivity
Results

- SD-OCT demonstrates focal defects in the EZ and IZ with preservation of the RPE. The focal defects demonstrate stability over 9 months.

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Results

- OCT-A demonstrates focal defects in the sub-RPE and choriocapillaris layers only, corresponding to the focal defects in the EZ/IZ.
Conclusions

• Patients with pachychoroid pigment epitheliopathy may develop focal outer retinal defects in the EZ and/or IZ.

• Patients have a stable disease course and retain visual acuity.

• There may be an increased risk of these defects in patients with diabetes.
Thank you!

Susan Elner, MD