PERIPAPILLARY PACHYCHOROID SYNDROME (PPS): A CASE REPORT OF A RARE ENTITY

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Purpose: Peripapillary Pachychoroid Syndrome (PPS) is considered a distinct variant of central serous chorioretinopathy, within the pachychoroid disease spectrum. It is characterized by pachychoroid features associated with intraretinal and/or subretinal fluid in the nasal macula surrounding the optic disc. The aim of our case report is to present the challenges in diagnosing a patient with PPS.

Methods: A 72-year old white male presented to our hospital complaining of painless, gradual visual loss. His visual acuity (VA) was 4/10 in the right eye and 5/10 in the left. Examination of the anterior segment showed cataract formation in the right eye. Fundoscopy, optical coherence tomography (OCT), and indocyanine green angiography (ICGA) were also performed.

Results: Fundoscopy and OCT revealed cystoid macular edema (CME) in the left eye with no obvious pathology present. The patient was treated with a combination of dexamethasone and nepafenac eye drops with no clinical improvement. Intravitreal anti-VEGF treatment was also administered with similarly disappointing results. The patient received a combination of dorzolamide eye drops and acetazolamide tablets. CME persisted and at this point the visual acuity in the left eye was counting fingers. The ICGA performed revealed optic disc edema, peripapillary choroidal thickening and diffuse choroidal leakage in the posterior pole, findings supportive of PPS in the left eye. Photodynamic therapy was applied peripherally at the leakage sites. The CME finally resolved and the patient’s vision significantly improved (VA=7/10).

Conclusions: It is important for the ophthalmologists to be able to recognize and correctly treat this challenging, novel clinical entity.

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