Peripapillary pachychoroid syndrome successfully treated with topical prednisolone acetate 1% drops

Lojain A. Azizalrahman[1], Abdullah N. Almousa[1], Fadwa Al Adel[2], Abdulrahman F. AlBloushi[1]

Access this article online

Quick Response Code:

Website: www.saudijophthalmol.org

DOI: 10.4103/sjopt.sjopt_187_22

Abstract:

Peripapillary pachychoroid syndrome (PPS) is a rare disease characterized by choroidal thickening around the optic disc. Visual acuity might be impaired secondary to the associated peripapillary intraretinal and/or subretinal fluids. We reported a case of a 70-year-old male patient who presented with a gradual bilateral decrease in vision. His best-corrected visual acuity was 20/60 in the right eye and 20/25 in the left eye. Dilated fundus examination showed yellowish peripapillary lesions and intraretinal fluid (IRF) surrounding the optic disc in both eyes. Optical coherence tomography showed that the outer retinal layers were disrupted, and IRF affected the nasal macula. A bilateral increase in the thickness of the choroid around the optic discs was found. The patient was successfully treated with a tapering dose of topical prednisolone acetate (1%). Three months after treatment, his vision improved to 20/25 in the right eye and 20/20 in the left eye. Topical steroid drops might be administered to treat PPS.

Keywords:

Pachychoroid, peripapillary pachychoroid syndrome, prednisolone acetate

Introduction

Pachychoroid disease spectrum (PDS) includes a group of chorioretinal disorders that have common characteristics, including choroidal thickening due to the dilation and congestion of choroidal vessels or "pachyvessels."[1] Several conditions are included in PDS, such as pachychoroid pigment epitheliopathy, central serous chorioretinopathy, pachychoroid neovasculopathy, and polypoidal choroidal vasculopathy.[2,3]

Peripapillary pachychoroid syndrome (PPS) is a novel variant of the pachychoroid spectrum characterized by choroidal thickening localized to the peripapillary area and associated with peripapillary intraretinal and/or subretinal fluid (SRF), choroidal folds, and occasionally, optic disc edema.[2] The disease is typically bilateral and predominantly occurs in elderly males and hypermetropes.[2,3]

PPS is benign, and most patients experience a decrease in retinal edema and choroidal thickening.[4] However, some patients might experience a visual decline with persistent retinal edema. Therefore, several treatment options were investigated for treating PPS, including photodynamic therapy, topical steroid drops, and others.[4-6] Topical steroid drops were previously administered to a few patients and showed promising results.[5,6] Here, we described a peculiar case of PPS, which was successfully treated by administering topical prednisolone acetate (1%) (Pred Forte™ [PF]).

Case Report

A 70-year-old male patient presented with a gradual loss of vision in both eyes. His ocular history revealed uneventful cataract extraction in both eyes 6 years ago. On examination, his best-corrected visual acuity (BCVA) was 20/60 in the right eye and 20/25 in the left eye. His intraocular pressure (IOP) was 19 mmHg in the right eye and 17 mmHg in the left eye. A slit-lamp examination revealed a clear cornea with pseudophakia in both eyes. Dilated fundus examination showed yellowish peripapillary lesions and chorioretinal folds. Fundus autofluorescence showed bilateral

[1]Department of Ophthalmology, College of Medicine, King Saud University, [2]Department of Ophthalmology, College of Medicine, Princess Nourah Bint Abdulrahman University, Riyadh, Saudi Arabia

Address for correspondence:
Dr. Abdulrahman F. AlBloushi, Department of Ophthalmology, College of Medicine, King Saud University, Riyadh 11411, Saudi Arabia.
E-mail: aalbloushi1@ksu.edu.sa

Submitted: 30-Oct-2022
Revised: 16-Jan-2023
Accepted: 29-Jan-2023
Published: 08-Jul-2023

How to cite this article: Azizalrahman LA, Almousa AN, Al Adel F, AlBloushi AF. Peripapillary pachychoroid syndrome successfully treated with topical prednisolone acetate 1% drops. Saudi J Ophthalmol 2024;38:89-91.

hyperautofluorescent lesions corresponding to the yellowish lesions. Optical coherence tomography (OCT) showed disruption of the outer retinal layers, and intraretinal fluid (IRF) affected the nasal macula (right > left). A significant increase in the thickness of the choroid around the optic disc was found through enhanced depth imaging [Figure 1]. The axial length measurement was 22.3 mm in the right eye and 22.5 mm in the left eye.

The patient was diagnosed with PPS and was initially observed for 3 months. Since his visual acuity and the clinical picture remained unchanged, he was administered topical prednisolone acetate (1%) (PF; 10 mg/mL). The drops were prescribed three times daily for 1 month, then reduced to twice daily for 1 month, once daily, continued for 1 month, every other day for 1 month, and then maintained every 2 days for 6 months. Three months after starting the treatment with PF drops, the BCVA of the patient improved to 20/25 in the right eye and 20/20 in the left eye. His IOP was 20 mmHg in the right eye and 16 mmHg in the left eye. At the last follow-up, i.e. 9 months after starting the treatment, his condition was stable without the presence of recurring intraretinal fluids; however, the yellowish subretinal deposits were still present in both eyes.

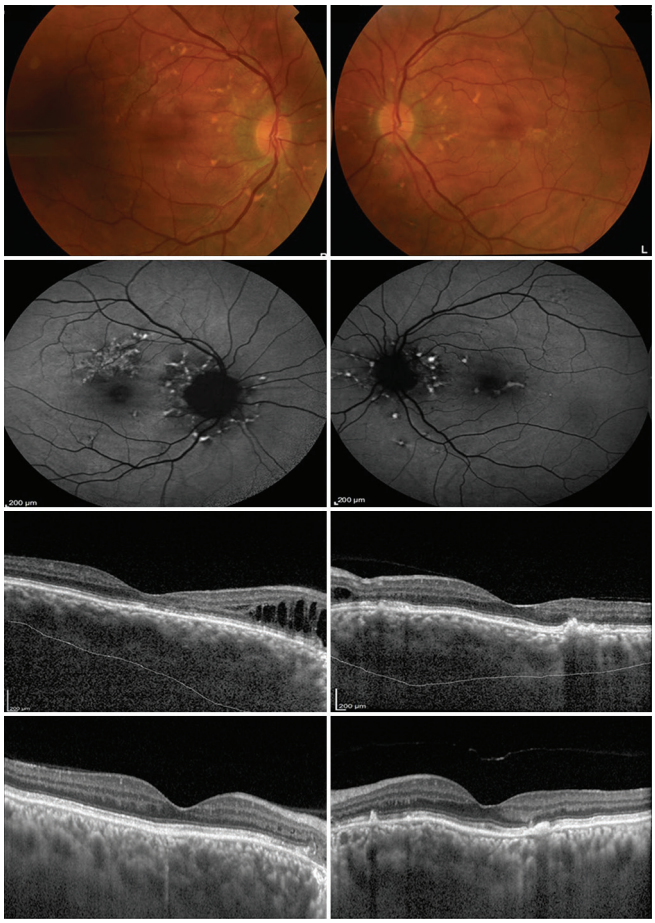

Figure 1: (First row) At presentation, fundus photographs of both eyes showed yellowish peripapillary lesions and chorioretinal folds. (Second row) Fundus autofluorescence revealed bilateral hyperautofluorescent lesions corresponding to the yellowish lesions. (Third row) Enhanced depth imaging-optical coherence tomography showed disruption of the outer retinal layers. Intraretinal fluid affected the nasal macula (right > left), and a significant increase in the choroid thickness around the optic disc (white dotted line) was observed. (Fourth line) Three months after treatment with topical prednisolone acetate (1%). Note the complete resolution of intraretinal fluids in both eyes

An informed consent was obtained from the patient for the anonymous use of data and photos.

Discussion

The underlying pathophysiology of PPS is not well understood; however, few studies have suggested that the disease occurs secondary to abnormal anastomotic choroidal vessels located in the peripapillary area.[2] The presumed high hydrostatic pressure in these anastomotic choroidal vessels might lead to congestion and fluid leakage into the retina in PPS.[2,7] The degeneration of the peripapillary outer retinal layers might facilitate the leakage of fluid into the retina from the hyperpermeable choroid.[2]

Isolated posterior uveal effusion syndrome (PUES) was previously described in a few cases that share similar findings to PPS, including thick posterior choroid with serous macular detachment, nasal cystoid macular edema, choroidal folds, and short axial length. However, unlike PUES, patients with PPS usually displayed serous retinal pigment epithelium detachment and alteration with IRF and/or SRF predominantly located around the optic disc. Moreover, patients with PPS showed similar indocyanine green angiography (ICGA) features seen in PDS, including dilated large choroidal veins (pachyvessels) and choroidal hyperpermeability best seen in the mid-phase of the study, which fades gradually at the late phase. However, in PUES, the ICGA usually shows a diffusely granular hyperfluorescence in the very early phase, which increases with time and persists until the late phase.[2]

Intraretinal and SRFs in PPS might exhibit significant fluctuations and even complete spontaneous resolution during the follow-up. Hence, observations might be a viable initial option in this disease if there is no significant change in vision.[3,4] No clear guidelines are available for managing PPS; however, several treatment options have been investigated for treating PPS. These include the administration of intravitreal anti–vascular endothelial growth factor,[2,8] photodynamic therapy,[9,10] topical or systemic carbonic anhydrase inhibitors,[2,5] and mineralocorticoid-receptor antagonists;[5] however, the outcomes are variable.

Topical steroids were administered to effectively treat IRF associated with PPS in a few cases.[5,6] It has been suggested that choroidal hyperpermeability resulting in PPS might be secondary to an underlying inflammatory etiology.[5,6] In this study, we found that the usage of topical prednisolone acetate drops could effectively treat IRF 3 months after starting the administration of PF drops; moreover, no recurrence was observed until the last follow-up. In another study, all

participants with PPS who received PF drops showed a reduction in the IRF after 4 weeks of treatment.[6] However, IRF might reaccumulate in some patients following the discontinuation or a reduction in the dosage of PF.[5,6] Thus, although topical prednisolone is effective, recurrence of the disease is possible. Therefore, it is recommended to continue this topical treatment over extended periods to prevent relapses.

Another finding in our study, which was also reported in other studies among some patients with PPS, was the presence of acquired vitelliform lesions (AVL).[11] In OCT, hyperreflective deposits are found lying over the compressing pachyvessels. This association between AVL and PPS necessitates distinguishing these lesions from macular neovascularization. This is because PPS can present IRF and/or SRF.

To summarize, for PPS cases complicated by IRF and/or SRF, topical steroids, either dexamethasone or PF, might be a simple and effective treatment, especially when other options, such as PDT, are unavailable. Prospective studies with more participants need to be conducted to prove the safety and efficacy of using topical steroid drops in patients with PPS.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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