Int Ophthalmol (2019) 39:2661–2665
https://doi.org/10.1007/s10792-019-01086-9

CASE REPORT

Purtscher's retinopathy case report: short posterior ciliary arteries contribution to radial peripapillary capillary system observed with optical coherence tomography angiography

Elena Ávila-Marrón · John P. Liscombe-Sepúlveda · Laura Manfreda-Dominguez · Prudencia Rochina-Pérez · Antonio Duch-Samper

Received: 8 January 2018 / Accepted: 23 February 2019 / Published online: 9 March 2019

Abstract
Purpose The authors report a case of Purtscher's retinopathy whose optical coherence tomography angiography (OCT-A) analysis allows us to better understand the vascular network of the optic disk.
Methods Case report.
Results A 75-year-old man presented with vision loss in his left eye (LE) after chest compression. The best-corrected visual acuity (BCVA) in the LE was 20/200, and the anterior segment showed a relative afferent pupillary defect. Dilated fundus examination revealed white peripapillary retinal patches and macular hemorrhage. The OCT scan showed edema and hyper-reflectivity of the inner retinal layers at macular level. In turn, OCTA evidenced capillary dropout in both macular retinal plexus, though with preservation of the radial peripapillary capillaries (RPC) and choriocapillary layer. At 9 months, BCVA was 20/20 associated with persistence macular ischemia but unaffected RPC.
Conclusion In conclusion, this case suggests that the RPC does not depend exclusively on retinal capillaries as there was a reversible damage after a microvascular retinal disorder such as Purtscher's retinopathy. Possibly, the contribution from short posterior ciliary arteries ensures proper vascularization as choriocapillary layer also remained unaffected. Furthermore, OCTA is considered a useful tool that affords better assessment of RPC than FA.

Keywords Optical coherence tomography angiography · Purtscher's retinopathy · Radial peripapillary capillaries · Case report

E. Ávila-Marrón · J. P. Liscombe-Sepúlveda · L. Manfreda-Dominguez · P. Rochina-Pérez · A. Duch-Samper
Department of Ophthalmology, Valencia University Clinic Hospital, Avenida Blasco Ibáñez, 17, 46010 Valencia, Spain

A. Duch-Samper
Department of Ophthalmology, Faculty of Medicine, University of Valencia, Valencia, Spain

E. Ávila-Marrón (✉)
Cardenal Cienfuegos 2, Esc 3-8C, 33007 Oviedo, Spain
e-mail: eleavilamarron@gmail.com

Introduction

Purtscher's retinopathy (PR) is an occlusive and hemorrhagic microvascular disorder mainly reported after thoracic compression, head injuries and other non-traumatic causes. The diagnosis is primarily based on the clinical findings and funduscopic signs.

Fluorescein angiography (FA) and indocyanine green angiography (IGA) have been the gold standard methods in clinical practice for evaluating retinal and choroidal vascular changes, though they provide little in-depth information due to the two-dimensional nature of the images. Optical coherence tomography angiography (OCTA) is a recently introduced noninvasive technique capable of detecting capillary dropout in the superficial and deep vascular plexus of the retina, as well as alterations in the choriocapillaris and choroidal layers [1]. There is a specific capillary network referred to as the radial peripapillary capillary (RPC) system that is limited in its distribution to the posterior pole. This vascular layer was thought to be dependent upon the central retinal artery. However, the introduction of OCTA allows to evaluate its vascular supply thoroughly [2].

The present case report analyzes the structural and vascular abnormalities present in the macula and RPC network evaluated by OCTA and OCT *B*-scans and describes the patient course over 9 months of follow-up.

Case report

A 75-year-old male presented with sudden vision loss in his left eye (LE) 48 h after chest compression injury. He reported no other systemic conditions.

At first ocular examination, visual acuity (VA) in the right eye was 20/20, with no significant findings. The LE presented a relative afferent pupillary defect, and the BCVA was 20/200. The anterior segment showed nasal subconjunctival hemorrhage, and the anterior chamber proved normal. However, indirect ophthalmoscopy examination under mydriasis showed multiple peripapillary white patches with periarteriolar sparing and cotton wool spots, as well as important pre-retinal macular hemorrhage (Fig. 1). Intraocular pressure (IOP), measured with a Goldmann applanation tonometer, was 14 mm Hg in both eyes. Based on the clinical findings, the tentative diagnosis was Purtscher's retinopathy, and no treatment was prescribed.

Fundus color pictures were obtained, and optical coherence tomography *B*-scans (OCT, Triton, Topcon, Tokyo, Japan) and OCT angiography (OCTA, Triton, Topcon, Tokyo, Japan) were performed. It provides segmentation of en face automated slabs guided by OCT *B*-scans within cubes (6 × 6 and 9 × 9 mm) via a software algorithm (IMAGEnet). The macular OCT *B*-scan showed hemorrhage under the internal limiting membrane (ILM) associated with a neurosensory retinal detachment, edema and hyper-reflectivity of the retinal layers (Fig. 2). OCTA of the optic disk (6 × 6 mm) demonstrated severe edema of the nerve fiber layer (NFL) that hid the vasculature. At this time, macular OCTA and RPC could not be evaluated.

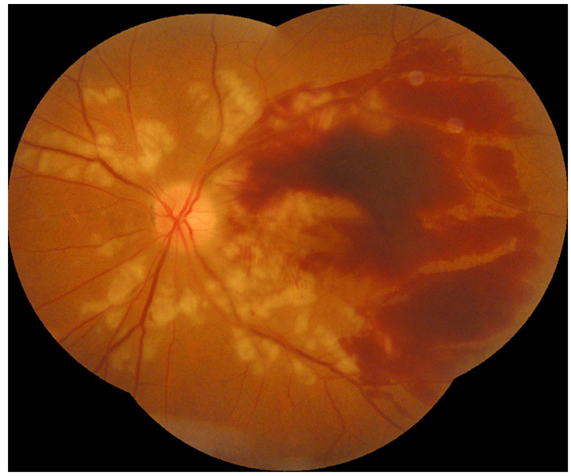

Fig. 1 Retinography of the left eye: pre-retinal macular hemorrhage and multiple peripapillary white lesions (Purtscher flecken and cotton wool spots)

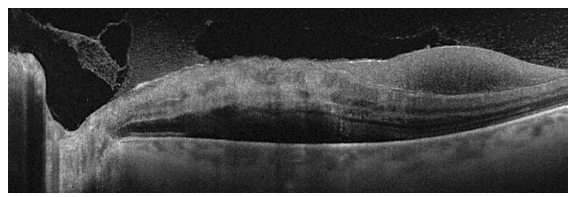

Fig. 2 OCT *B*-scan of the macula: pre-retinal hemorrhage, disorganization and hyper-reflectivity of the retinal layers and detachment of the neurosensory retina

One month later, retinography revealed incomplete resolution of the peripapillary white lesions and macular hemorrhage at the posterior pole. After 4 months, the BCVA of the affected eye was 20/20 eccentric, and fundus examination showed resolution of the retinal lesion and a normal optic nerve (Fig. 3). Despite the clear fundus, macular OCTA (6 × 6 mm) showed capillary loss in the superficial (including 2.6 µm under the ILM, the ganglion cell layer and 15.6 µm under the internal plexiform layer) and deep plexus (54.6 µm) of the nasal macula, though the choriocapillary layer remained unaffected (Fig. 4). The OCT *B*-scan of the macula demonstrated important atrophy of the inner retinal layers (Fig. 5).

With regard to the optic nerve, OCTA (6 × 6 mm) showed preservation of the RPC network, with mild

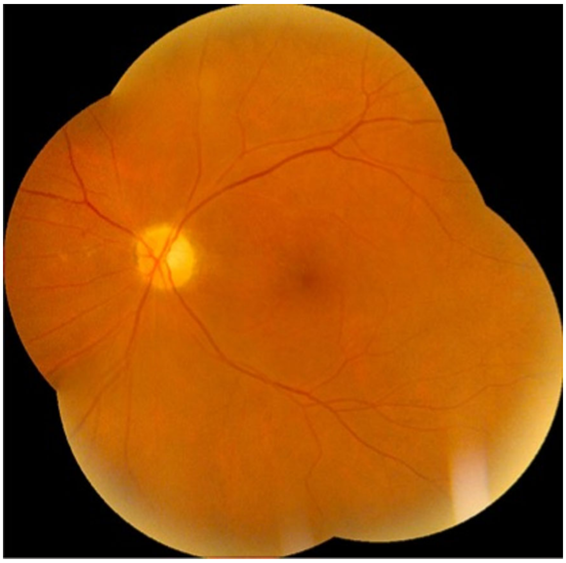

Fig. 3 Retinography of the left eye: resolution of macular hemorrhage and peripapillary white lesions, arteriolar attenuation

involvement in the superior-temporal quadrant (Fig. 6). The RPC was visualized in 6 × 6 scans centered on the optic nerve head, including a slab thickness sufficient to contain the full NFL (70.2 µm). The disk boundary was manually delineated into an ellipse base on the choriocapillary OCTA layer, and the peripapillary area was defined as a 700-µm-wide elliptical annulus external to the optic disk margin [3].

After 9 months of follow-up, the retinographic findings of the LE remained unaltered. However, macular OCTA (6 × 6 mm) revealed a persistence of ischemic areas (Fig. 7), and the OCT *B*-scan showed an increase in retinal atrophy. Nevertheless, the peripapillary capillary dropout visible at the start in the RPC was resolved, exhibiting normal vascular density and good perfusion of the area proximal to the optic nerve (Fig. 8).

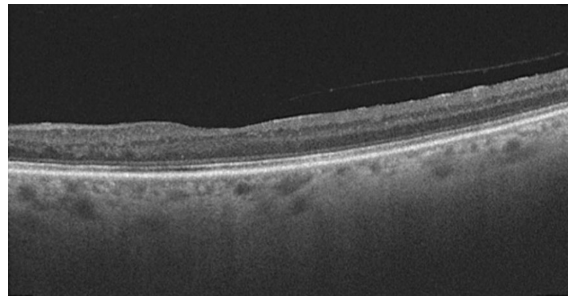

Fig. 5 Macular OCT *B*-scan: atrophy of the inner retinal layers. Preservation of the outer retina and choriocapillary layer

Conclusion

Several studies have described vascular changes in the PR using FA and IGA. However, in 2017 Vinod Kumar reported vascular abnormalities using OCTA focused on the macula. Unlike the macula, few studies have been published involving OCTA centered on radial peripapillary capillary (RCP) changes. The RPC system is considered to be a vascular network localized within the retinal nerve fiber layer (RNFL) around the optic disk. Dissimilar to the other capillaries, this area has fewer arteriolar anastomoses, so restriction of the lesions in PR to the nasal macula and peripapillary area may be related to their unique blood supply, vulnerable to retinal vascular occlusions [4].

In our case, apart from the clinical findings, OCTA was used to complement the study, as it is a novel noninvasive method offering high-resolution images of the retinal, choroidal and optic nerve microvasculature down to the capillary level. At macular level, the

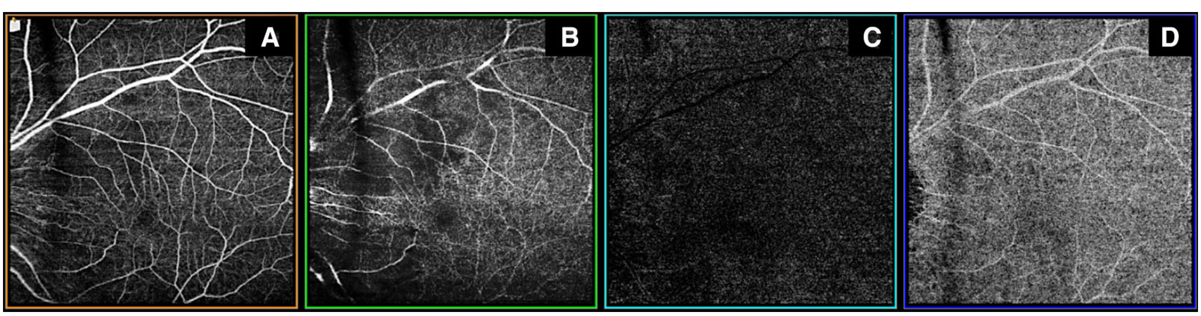

Fig. 4 Macular OCTA (6 × 6 mm): superficial vascular plexus (**a**), deep vascular plexus (**b**), avascular layer (**c**), choriocapillary layer (**d**): dropout of capillary perfusion in nasal macula in **a** and **b**. No vascular abnormalities in **d**

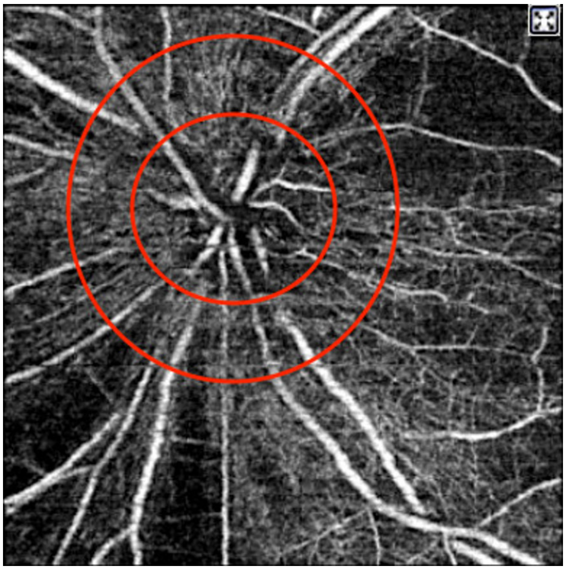

Fig. 6 Optic nerve OCTA (6 × 6 mm): delineation of optic nerve boundary (inner red ellipse) and radial peripapillary capillaries (outer red ellipse). Preservation of radial peripapillary capillaries with mild involvement in the superior-temporal quadrant

OCTA scan demonstrated a dropout of capillary perfusion in both retinal plexus that persisted for 9 months. This was related to an important thinning of the inner retinal layers observed by the OCT *B*-scan in the chronic phase. Although involvement of the choroidal vasculature has been reported in some cases, there were no such alterations in our patient. This demonstrated that the short ciliary arterioles were intact and established good perfusion of the outer retina.

With regard to the RPC, we found the capillary loss observed in the acute phase around the optic disk to have been resolved. In contrast to the purposed vulnerability of this region in retinal vascular diseases, we demonstrated that the unaffected choriocapillaris layer might make an important contribution to the perfusion of the optic nerve head, due to numerous anastomoses between the arterioles of the central retinal artery and the short posterior ciliary arteries [5].

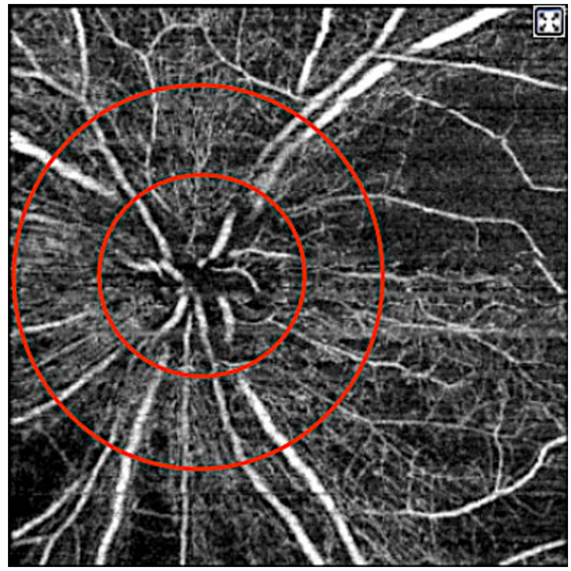

Fig. 8 Optic nerve OCTA (6 × 6 mm): preservation of radial peripapillary capillary perfusion

Thus, peripapillary capillary loss was located external to this area, where there is no contribution from the double vascularization.

To conclude, our results suggest that the perfusion of the RPC does not depend exclusively on the central retinal artery. Probably, an important contribution is made by the short posterior ciliary arteries that ensure proper vascularization in the context of ischemic events. Furthermore, OCTA is considered to be a

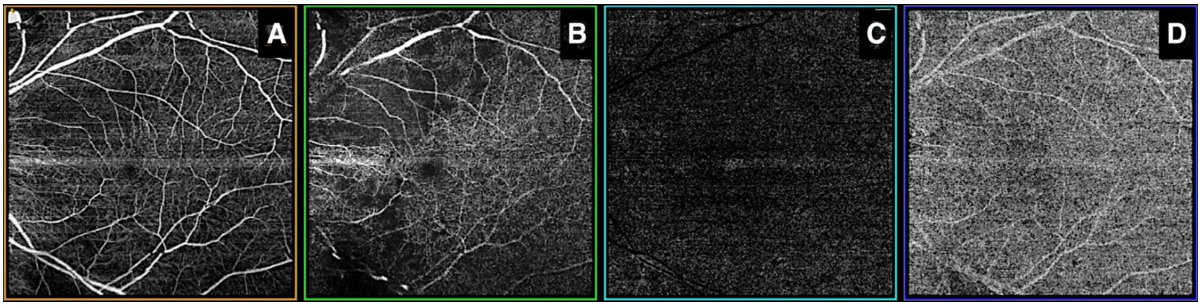

Fig. 7 Macular OCTA (6 × 6 mm): superficial vascular plexus (**a**), deep vascular plexus (**b**), avascular layer (**c**), choriocapillary layer (**d**): persistence of capillary dropout in nasal macula greater in **b**

useful tool for assessing retinal vascular alterations and extends FA findings.

Funding No funding was received for this work from any of the following organizations: National Institutes of Health (NIH); Wellcome Trust; Howard Hughes Medical Institute (HHMI); and other(s).

Compliance with ethical standards

Conflict of interest None of the authors have a proprietary interest. The authors declare that they have no conflicts of interest regarding the publication of this paper.

Ethical approval All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki Declaration and its later amendments or comparable ethical standards.

Informed consent Informed consent was obtained from all individuals participants included in the study.

References

1. Gao SS, Jia Y, Zhang M et al (2016) Optical coherence tomography angiography. Invest Ophthalmol Vis Sci 57:27–36
2. Mansoori T, Sivaswamy J, Gamalapati JS et al (2017) Measurement of radial peripapillary capillary density in the normal human retina using optical coherence tomography angiography. J Glaucoma 26(3):241–246
3. Pechauer Alex D, Jia Yali, Liu Liang et al (2015) Optical coherence tomography angiography of peripapillary retinal blodd flow response to hyperoxia. Invest Ophthalmol Vis Sci 56:3287–3291
4. Mo Shelley, Phillips Eirka, Krawitz Brian D et al (2017) Visualization of radial peripapillary capillaries using optical coherence tomography angiography: the effect of image averaging. PLoS ONE 12(1):2–4
5. Cerdá-Ibáñez M, Duch-Samper A, Clemente-Tomás R et al (2017) Correlation between ischemic retinal accidents and radial peripapillary capillaries in the optic nerve using optical coherence tomographic angiography: observations in 6 patients. Ophthalmol Eye Dis 9:1–3