Purtscher-like retinopathy presented a honeycomb-like pattern in optical coherence topography angiography

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Case report

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Abstract

Background To report a case of Purtscher-like retinopathy (PUR) and his optical coherence tomography (OCT) and OCT-angiography (OCT-A) findings before and after treatment. Case presentation A 65-year-old male presented with acute onset of vision loss for 2 weeks. Fundus examination revealed cotton-wool spots, retinal hemorrhage, and purtscher flecken spreading around the optic disc in the right eye. He was diagnosed with Purtscher-like retinopathy because of the absent reports of any traumatic medical history. OCT presented some band-like hyperreflective lesions at the layer of inner nuclear layer known as paracentral acute middle maculopathy (PAMM). OCT-A revealed apparent reduction of blood flow signal at the layers of deep retina and choriocapillaris with a pattern of honeycomb-like hypointense signal. After 3-month follow-up, OCT scan showed a resolution of retinal edema, but the PAMM lesions were still visible. OCT-A presented the honeycomb-like pattern turned into a homogeneous reduction of blood flow with small patches of hypointense signal areas in choriocapillaris. Conclusion This case presented a new OCT-A sign in Purtscher-like retinopathy with a honeycomb-like hypointense signal at the layer of choriocapillaris indicates the involvement and the ischemia condition of the choroid in its pathological process.

Background

Purtscher's retinopathy was first described in 1910 by Otmar Purtscher in a middle-aged man who fell off a tree and suffered cranial trauma [1]. When there are non-traumatic etiologies, the correct definition is Purtscher-like retinopathy (PUR). The exact mechanism of PUR remains unknown. The most accepted theory for its pathogenesis attributes to an embolic phenomenon resulting in occlusion of the precapillary arterioles[2]. We reported a case of PUR with some notable signs in optical coherence tomography (OCT) and OCT-angiography (OCT-A), which may provide more information about the pathological features.

Case Presentation

A 65-year-old male presented to our clinical center with a chief complaint of acute onset of vision loss in the right eye for 2 weeks without clear inducement before the onset. He reported neither trauma nor special systemic medical history. Ophthalmic inspections showed the best-corrected visual acuity (BCVA) was hand movements in the right eye and 20/20 in the left eye. The affected right eye presented a positive result of relative afferent pupillary defect (RAPD). Other signs of anterior segment and intraocular pressure were unremarkable. Dilated fundus examination revealed retinal hemorrhage, cotton-wool spots and Purtscher fleckens confined to peripapillary area and posterior pole in the right eye. The contralateral eye was almost normal except for mild arteriosclerosis. Fluorescence angiography (FA) indicated slight delayed arteriovenous circulation time (14s) and mottled hypoflorescence corresponding to cotton-wool spots and Purtscher fleckens. Optical coherence topography (OCT) presented retinal thickening and edema especially the inner layer. In addition, it presented lesions of hyperreflective band at inner nuclear layer (INL) beside the fovea corresponding to the perifovea wedge-shaped white-grey lesions known as paracentral acute middle maculopathy (PAMM). OCT angiography (OCT-A) presented a reduced blood

flow in both inner and deep retinal vascular plexus and a honeycomb-like pattern of hypointense signal at the layer of choriocapillaris. (Figure 1)

He was diagnosed with Purtscher-like retinopathy (PUR) despite of the lack of trauma or other specific systemic diseases. We ordered laboratory testing to exclude common etiologies like systemic lupus erythematosus (SLE) and thrombotic thrombocytopenic purpura (TTP). The results including antinuclear antibodies (ANA), antineutrophil cytoplasmic antibody (ANCA), erythrocyte sedimentation rate (ESR), C-reactive protein (CRP) and blood routine examination were all within normal limits. He was also referred to internal medical department for further investigation. Only obsolete lacunar infarctions were detected after a thorough review of the whole body. *Alprostadil* 10U intravenous injection (Q.D. for 10 days) was prescribed as vascular dilation therapy to improve retinal blood supplement and prevent further damage. After 3-month rehabilitation, his visual acuity recovered to 20/400. The retinal hemorrhage, cotton wood spots and Purtscher fleckens were mostly resolved. OCT noted a remission of retinal edema, but the PAMM lesions were still visible. OCT-A presented the previous honeycomb-like pattern in choriocapillaris turned into homogeneously reduction of blood flow with some small patches of hypointense signal areas. (Figure 2)

Discussion And Conclusions

According to a recent systemic review of PUR, the most frequent etiologies are trauma and acute pancreatitis[3]. A previous report presented a patient developing PUR after a myocardial infarction with a concomitant transient ischemic attack[4]. Although the exact mechanism of PUR remains unknown, currently the most accepted theory for its pathogenesis attributes to an embolic phenomenon resulting in occlusion of the precapillary arterioles[2]. Air, fat, platelets, fibrin, leukocyte aggregates as well as exogenous particles are all potential embolus. In this case, the present of lacunar infarction brought up a hint that the retinal disorders share the same etiology as the vasculature of retina has similar embryologic origins to the cerebral vasculature.

The diagnosis of PUR is mostly clinical depending on specific medical histories including sudden vision loss after trauma or other special systemic disease and typical funduscopic signs like retinal hemorrhage, cotton wool spots and Purtscher fleckens. Multimode-image inspections including OCT, OCT-A and FA may provide more information for diagnosis and follow-up. Previous researches reported common signs of FA findings in PURs including areas of non-perfusion, retinal ischemia and slower filling of vessels [2]. OCT-A may reveal extensive nonperfusion in the macular area in both inner and deep capillary plexus [5, 6]. This patient also presented a hypoperfusion condition in retina layer. But we found its presentation in choriocapillaris is even characteristic with a notable sign of honeycomb-like hypointense signal pattern indicating the ischemic involvement of the choroid.

Many patients of PUR could regain their visual acuity to normal level spontaneously after the etiology is resolved, while some patients remain poor prognosis despite of various treatments. There is still no exact prognostic factor for PUR. To this case, some special circumstances should be taken into consideration

to predict the visual outcomes. Owing to his delayed consultation for 2 weeks, the exact process of the disease was unknowable and the best opportunity for intervention might have lost. And the present of PAMM lesions surrounding the fovea indicated a poor prognosis as this sign represented the ischemia of both inner and deep capillary plexus at the fovea [7]. Furthermore, OCT-A presented a honeycomb-like hypointense signal pattern in choriocapillaris just corresponds with anatomic structures of choroidal lobules. This provided a sign of choroidal lobular infarction as well and explained the restricted recoverability of this patient. At present, there is still no consensus on the treatment of PUR. Most viewpoints consider observation and treatment of the underlying etiology may be the most reasonable therapeutic option without risk of adverse drug effects.

In conclusion, PURs were reported mostly related to trauma or other special systemic conditions. It's an embolic occlusion disease substantially. This case provides us another perspective to detect for potential embolic etiologies, which makes sense especially for patients in 60s if no trauma or other specific systemic diseases reported. FA and OCT-A provide more information for diagnosis and follow-up visit. The present of PAMM lesion in OCT scan and a honeycomb-like pattern of hypointense signal pattern in OCT-A at the layer of choriocapillaris may indicate a poor visual prognosis because of the ischemia involvement of macula fovea and choroid.

Abbreviations

PUR	Purtscher's / Purtscher-like retinopathy
FA	fluorescein angiography
OCT	optical coherence topography
OCT-A	optical coherence topography angiography
PAMM	paracentral acute middle maculopathy
INL	inner nuclear layer
SLE	systemic lupus erythematosus
TTP	thrombotic thrombocytopenic purpura
ANA	antinuclear antibodies
ANCA	antineutrophil cytoplasmic antibody
ESR	erythrocyte sedimentation rate
CRP	C-reactive protein

Declarations

Ethics approval and Consent for participate

This study was approved by the Review Board of Peking Union Medical College Hospital. Written informed consent was obtained from the patient for publication of the case report and accompanying images.

Consent for publication

Written informed consent for publication was obtained from the patient. A copy of the written consent is available for review by the editor of this journal.

Availability of data and material

Some datasets generated and/or analyzed during the current study are not publicly available due to the article word limit, but are available from the corresponding author on reasonable request.

Competing interests

One of the authors, Youxin Chen, is a member of the editorial board of BMC ophthalmology. No other competing interests exist.

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Authors' contributions

BL contributed to the collection of the medical record and clinical data of the case and the writing of the manuscript. YC contributed to the whole management and medical care of the patient's diagnosis, ophthalmic and laboratory examination, and follow-up clinic. HL contributed to the acquirement of ophthalmic images including fundus, FFA, OCT images. (*E-mail: lidh@pumch.cn*)

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Figures

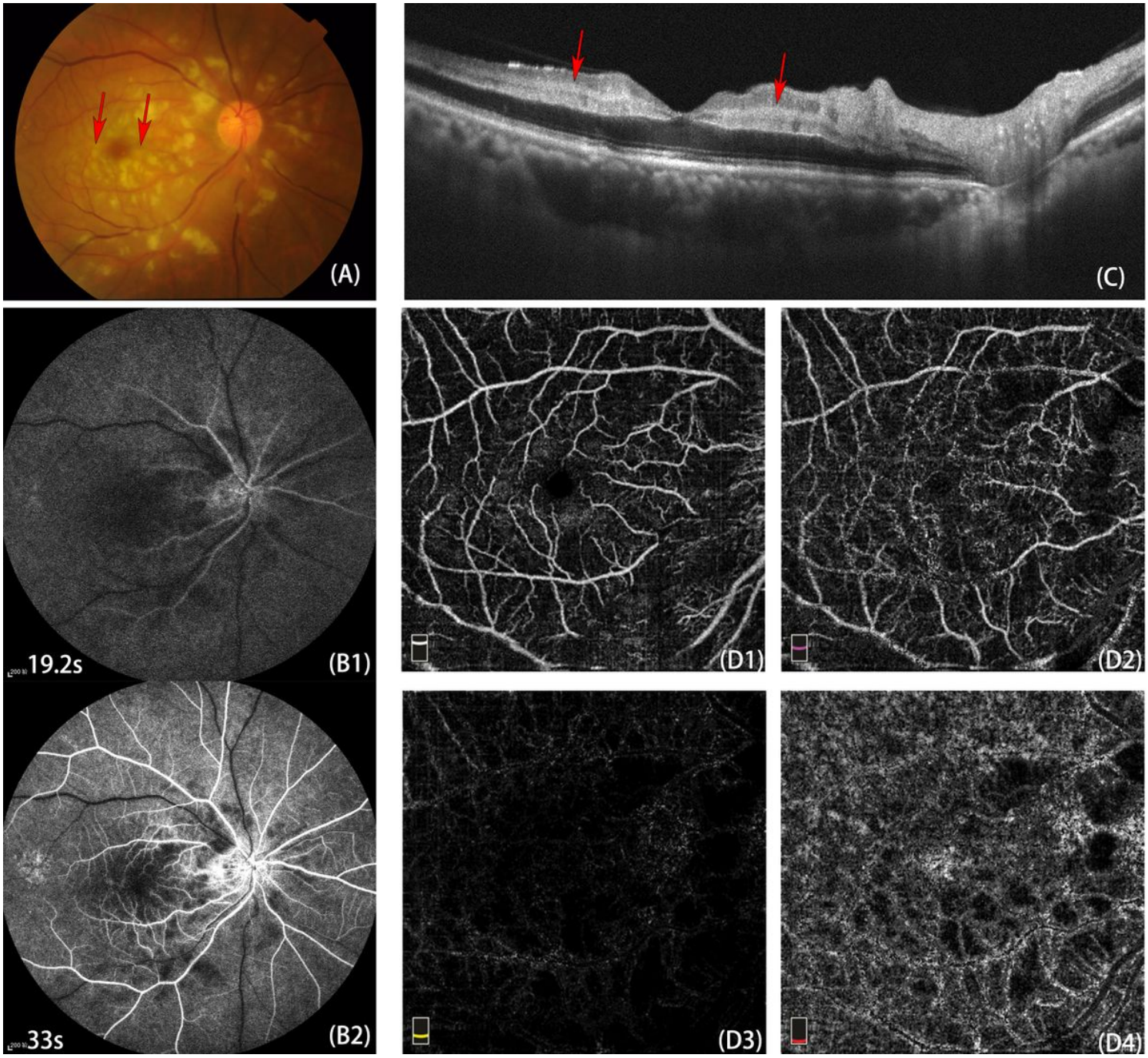

Figure 1

Multimodal imaging of the affected right eye. Fundus photography showed retinal hemorrhage, cotton-wool spots and Purtscher fleckens confined to peripapillary area and posterior pole (A). FA presented delayed arteriovenous circulation time (B1, B2). OCT B-scan of the fovea noted retinal edema and PAMM lesion in INL (red arrows)(C). OCT-A of the fovea (6×6mm) indicates the condition of blood supplement in superficial retina (D1), outer retina (D2), deep retina (D3) and choriocapillaris (D4). Blood flow signal decreased apparently in inner and deep retinal capillary layers. The honeycomb-like pattern of hypointense signal at the layer of choriocapillaris is the most remarkable sign.

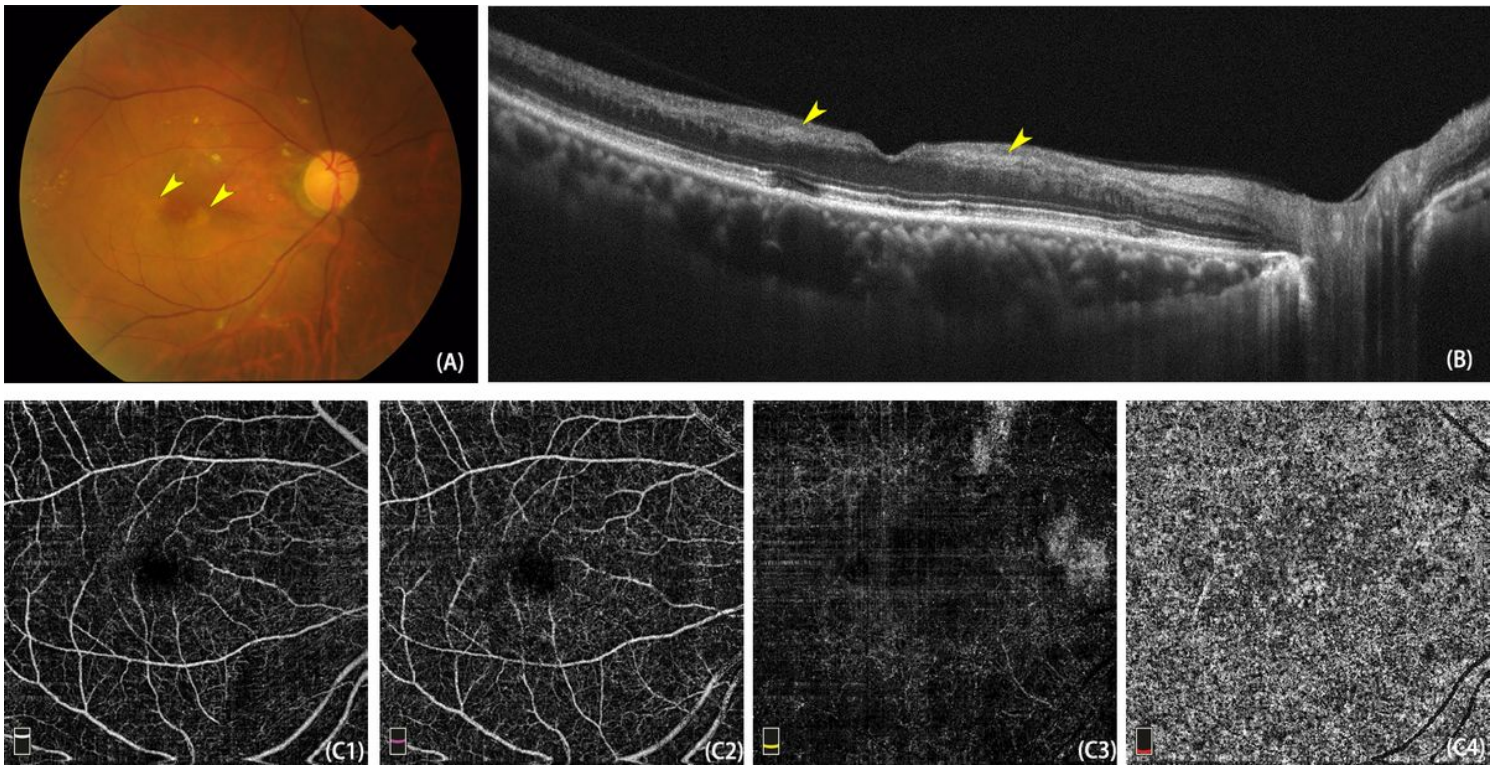

Figure 2

After 3 month follow-up, the retinal lesions of his right eye were mostly absolved (A), OCT noted PAMM lesions corresponding to perifovea wedge-shaped white-grey lesions (yellow arrowhead). OCT-A showed blood flow of different layers in retina and choroid: superficial retina (C1), outer retina (C2), deep retina (C3) and choriocapillaris (C4). OCT-A presented a hypoperfusion condition in both inner and outer retina with some small patches of hypointense signal areas.

Supplementary Files

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- supplement1.pdf