Optical Coherence Tomography Angiography Findings in Purtscher and Purtscher-like Retinopathy

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INTRODUCTION

The Purtscher retinopathy was first defined as changes in fundus following head injury by Othmar Purtscher in 1910; currently, it is defined as microangiopathic retinopathy developed following indirect trauma.¹

The Purtscher-like retinopathy is defined similar clinical picture developed secondary to causes other than trauma. In the literature, cases related to acute pancreatitis, pancreas adenocarcinoma, chronic renal failure, cryoglobulinemia, retrobulbar injection, fat/air/amnion fluid embolism, preeclampsia, connective tissue diseases, HEELP syndrome, chemotherapy, hemolytic uremic syndrome, synthetic cannabinoid use and malignant hypertension were reported.²⁻⁷ In a study from UK, its incidence was reported as 0.24: 1,000,000/years.⁸ Loss of vision is generally acute and severe. Visual acuity is generally decreased as low as ≤1/10.² In general, Purtscher retinopathy is bilateral but unilateral involvement was reported up to 40% of cases.⁹⁻¹¹

Clinically, it is an occlusive retinopathy characterized by acute loss of vision together with white retinal spots, soft exudate and retinal hemorrhages.¹²

The infarcts developed are generally seen as white retinal spots at posterior pole and nasal to disk with polygonal localization. This appearance is termed as Purtscher flecks. Central macular involvement by infarct areas can cause pseudo-cherry red spot in macula. In addition, soft exudates and retinal hemorrhages can be seen in fundus.

In this study, we discussed multimodal imaging and OCT-A findings in Purtscher retinopathy and Purtscher-like retinopathy.

Case 1

A 47-years old woman, a medical doctor, was hospitalized with head injury together with fractures at mandible, zygomatic bone, nasal bone, and rib following traffic accident. On day 1 after admission, the patient was consulted to ophthalmology department due o blurred vision and central loss of vision in right eye. In ophthalmological examination, best-corrected visual acuity (BCVA) was found as 1/10 in right eye and 10/10 in left eye. Biomicroscopic examination and intraocular pressure measurement were normal in both eyes. Fundus examination was normal in left eye while multiple retinal white spots which begin from superior temporal...
margin of optic disc and extend to macula along superior temporal arcuate and retinal hemorrhage at temporal margin of optic disc were detected in right eye (Picture 1A). In fundus auto-fluorescent (Spectralis; Heidelberg Engineering, Heidelberg, Germany) imaging, there was hypo-auto-fluorescence image corresponding to lesion area (Picture 1B). In optical coherence tomography (OCT, Spectralis; Heidelberg Engineering, Heidelberg, Germany), subretinal fluid and hyper-reflective image at inner retinal layer were observed on macular section (Picture 1C). On lesional section, hyper-reflective image and edematous appearance were seen (Picture 1D). On OCT angiography (Topcon DRI OCT Triton, Topcon, Japan), capillary ischemic areas at superficial (Picture 2A) and deep (Picture 2B) capillary plexuses were detected at same region. Black areas that may be compatible with ischemia were observed at choriocapillaris (Picture 2C). Prednisolone (40 mg; PO) was prescribed and tapered gradually (5 mg in every 3 days). In final control visit at month 7 after accident, BCV A was 6/10 in right eye. It was also seen that fundus findings were markedly improved as fundus autofluorescence finding and wedge nerve fiber defect was detected at lesion area (Picture 3A and B). It was observed that edema at macula (Picture 3C) and lesion area (Picture 3D) was regressed on OCT. In addition, it was seen that ischemic area was resolved but there was wedge nerve fiber defect on OCT-A (Picture 2D, E and F).

Case 2

A 47-years old man was referred with vision disorder over one week and potential treatment with intravitreal injection. The patient with known hypertension and diabetes mellitus had complaints of blurred vision and impaired balance over one week. In ophthalmological examination, best-corrected visual acuity (BCVA) was found as 1/10 in right eye and 2/10 in left eye. Biomicroscopic examination and intraocular pressure measurement were normal in both eyes. In fundus examination, it was seen that diffuse retinal white spots, retinal hemorrhages and macular edema at posterior pole in both eyes (Picture 4A and B). In fundus autofluorescence imaging, there was hypo-auto-fluorescence image corresponding to lesion area (Picture 4C and F). On fundus fluorescence angiography, hypo-fluorescence areas due to retinal white spots and optic disk leakage at late phases were observed (Picture 4D, E, G, H). There was serous elevation, hyper-reflective dots and exudates in both eyes on macular OCT (Picture 4I and J). On OCT-A, there was ischemic areas at superficial and deep capillary plexus corresponding to retinal white spots (Picture 5A, B and Picture 6A, B). Black areas that may be compatible with ischemia were observed at choriocapillaris (Picture 5C and 6C). The cranial diffusion-weighted MR imaging was found to be normal. In systemic examination, blood pressure was found as 240/150 mmHg; thus, patient was admitted to nephrology department. A mass lesion was detected at adrenal gland and the patient was diagnosed as Conn syndrome. The blood pressure control was achieved by carvedilol, spironolactone, valsartan plus hydrochlorothiazide and amlodipine. The patient was diagnosed as Purtscher-like retinopathy related to malignant hypertension. Prednisolone (40 mg; PO) was prescribed and tapered gradually (5 mg in every 3 days). In final control visit at month 3, BCVA was 6/10 in both eyes and it was seen that retinal white spots at fundus were markedly regressed (Picture 7A and B). It was also seen that hypo-auto-fluorescence secondary to lesion was decreased fundus autofluorescence imaging (Picture 7C and D).
OCT, it was observed that serous elevation was resolved with intraretinal hyper-reflective spots (Picture 7E and F). On OCT-A, it was seen that ischemia at superficial and deep venous plexuses and choriocapillaris was partially resolved when compared to baseline (Picture 5D, E, F and 6D, E, F).

Picture 2. Case 1; Optical coherence tomography angiography image at level of superficial vascular plexus: capillary ischemia adjacent to optic disc (A). Similar ischemic appearance in sections at level of deep vascular plexus (B) and choriocapillaris are present (C). It is seen that lesions were regressed at sections of superficial vascular plexus (D), deep vascular plexus (E) and choriocapillaris (F) with an image compatible to nerve fiber defect in same area on OCT-A obtained on month 7.

Picture 3. Case 1; it is seen that lesions were resolved in color fundus image (A), autofluorescence imaging (B), macular optical coherence tomography images (C) and lesional optical coherence tomography images (D) on month 7.
Case 2: diffuse white spots, retinal hemorrhages and macular exudation are seen in bilateral fundus at presentation; right eye (A) and left eye (B). There are diffuse hypo-autofluorescence areas in both eyes on fundus autofluorescence imaging; right eye (C) and left eye (F). In areas corresponding to retinal white spots, hypo-fluorescence due to blockade and optic disc leakage are seen in both eyes on fundus fluorescein angiography (D, E, G and H). There is subretinal serous fluid and hyper-reflective dots outer retinal layers in both eyes on optical coherence tomography; right eye (I) and left eye (J).
Picture 5. Case 2: diffuse capillary ischemia areas in superficial vascular plexus (A) of right eye on optical coherence tomography angiography obtained at presentation. Similar ischemic images are also seen at deep vascular plexus (B) and choriocapillaris sections (C). In control visit on month 3, it is seen that ischemic areas at superficial vascular plexus (D), deep vascular plexus (E) and choriocapillaris sections (F) were partially resolved.

Picture 6. Case 2: diffuse capillary ischemia areas in superficial vascular plexus (A) of left eye on optical coherence tomography angiography obtained at presentation. Similar ischemic images are also seen at deep vascular plexus (B) and choriocapillaris sections (C). In control visit on month 3, it is seen that ischemic areas at superficial vascular plexus (D), deep vascular plexus (E) and choriocapillaris sections (F) were partially resolved.
Although pathophysiology of Purtscher retinopathy hasn't been fully elucidated, it is though that Purtscher retinopathy develops due to pre-capillary occlusion and nerve fiber layer infarct secondary to micro-embolization. In a systematic review by Miguel et al., it was suggested that at least 3 of 5 following criteria should have to be present for diagnosis of Purtscher and Purtscher-like retinopathy.12

1) Purtscher fleck
2) Retinal hemorrhages (mild-to-moderate)
3) Soft exudates limited to posterior pole
4) Potential etiology
5) Ancillary test compatible with diagnosis

On fundus fluorescence angiography, blockade of choroid fluorescence in early phases, occlusion and leakage in retinal arterioles, capillary non-perfusion areas, delayed perivenous staining and optic disc edema can be observed.\(^\text{13}\). Although retinal findings are resolved over time, central scotoma and loss of vision are generally persistent. Final visual acuity was found as ≤1/10. Poor prognostic criteria include edema at optic disc head, choroid hypo-perfusion, outer retinal layer involvement and history of previous Purtscher retinopathy.\(^\text{2}\)

Although there is no standardized or proven treatment modality, there are case reports suggesting that steroid therapy is useful or ineffective.\(^\text{15, 16}\) It is thought that steroid therapy prevents leukocyte aggregation and reduces free radical formation. A recent review by Xia et al. included 76 studies and 139 eyes of 88 patients were reviewed regarding treatment.\(^\text{17}\) It was found steroids are most common agents used in the treatment (63.29%). However, no significant difference was detected in visual acuity between patients treated and those left untreated.

Optic coherence tomography angiography is a novel imaging technology that allows imaging retinal vascularity in a rapid and non-invasive manner. Hamoudi et al. followed a 16-years old patient with unilateral Purtscher retinopathy following traffic accident and found that there was ischemia at superficial and deep vascular plexus were detected on OCT-A. Xiao et al. reported a 65-years old patient with bilateral Purtscher retinopathy following traffic accident and found that there was ischemia at superficial and deep capillary plexuses and related dilatation in foveal avascular zone on OCT-A. In addition, multifocal filling defects and dilatation of foveal avascular zone were reported in florescence angiography.

In conclusion, OCT-A evaluation is valuable in Purtscher retinopathy since it can demonstrate capillary ischemia early in a rapid and non-invasive manner.

**REFERENCES**